Histological Typing of Kidney Tumours

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Histological Typing of Kidney Tumours. Mostofi/Davis (1998)

Histological Typing of Kidney Tumours

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In Collaboration with L. H. Sobin
and Pathologists in 6 Countries

Second Edition

With 145 Colour Figures

Springer

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General Preface to the Series

Among the prerequisites for comparative studies of cancer are international agreement on histological criteria for the definition and classification of cancer types and a standardized nomenclature. An internationally agreed classification of tumours, acceptable alike to physicians, surgeons, radiologists, pathologists and statisticians, would enable cancer workers in all parts of the world to compare their findings and would facilitate collaboration among them.

In a report published in 1952[1], a subcommittee of the World Health Organization (WHO) Expert Committee on Health Statistics discussed the general principles that should govern the statistical classification of tumours and agreed that, to ensure the necessary flexibility and ease of coding, three separate classifications were needed according to (1) anatomical site, (2) histological type, and (3) degree of malignancy. A classification according to anatomical site is available in the International Classification of Diseases[2].

In 1956, the WHO Executive Board passed a resolution[3] requesting the Director-General to explore the possibility that WHO might organize centres in various parts of the world and arrange for the collection of human tissues and their histological classification. The main purpose of such centres would be to develop histological definitions of cancer types and to facilitate the wide adoption of a uniform nomenclature. The resolution was endorsed by the Tenth World Health Assembly in May 1957[4].

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[1] WHO (1952) WHO Technical Report Series, no. 53. WHO, Geneva, p 45

[2] WHO (1977) Manual of the international statistical classification of diseases, injuries, and causes of death, 1975 version. WHO, Geneva

[3] WHO (1956) WHO Official Records, no. 68, p 14 (resolution EB 17.R40)

[4] WHO (1957) WHO Official Records, no. 79, p 467(resolution WHA 10.18)

Since 1958, WHO has established a number of centres concerned with this subject. The result of this endeavour has been the International Histological Classification of Tumours, a multivolumed series whose first edition was published between 1967 and 1981. The present revised second edition aims to update the classification, reflecting progress in diagnosis and the relevance of tumour types to clinical and epidemiological features.

Preface to Histological Typing of Kidney Tumours – Second Edition

The first edition of the *International Histological Classification of Kidney Tumours* was published in 1981[1]. Since then, a number of new entities have been recognised, necessitating a revision of the classification. For this publication, a draft of the proposed revision was circulated among pathologists from seven countries.

It will be appreciated, of course, that the classification reflects the existing state of knowledge, and modifications are almost certain to be needed as experience accumulates. Although the present classification has been adopted and recommended by the members of the group, it represents a view from which some pathologists may wish to dissent. It is hoped, nevertheless, that in the interests of international cooperation, all pathologists will try to use the classification as proposed. Criticisms and suggestions for its improvement will be welcomed. These should be sent to the World Health Organization, 1211 Geneva 27, Switzerland.

The histological classification which appears on pages 3–5 contains the corresponding morphology code numbers of the *International Classification of Diseases for Oncology* (ICD-O)[2] for tumours, and of the *Systematized Nomenclature of Medicine* (SNOMED)[3] for tumour-like lesions.

The publications in the series, *International Histological Classification of Tumours* are not intended to serve as textbooks,

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[1] Mostofi FK (1981) Histological typing of kidney tumours. Geneva, World Health Organization (International Histological Classification of Tumours, No. 25)

[2] World Health Organization (1990). *International classification of diseases for oncology* (ICD-O). Geneva

[3] College of American Pathologists (1982). *Systematized nomenclature of medicine* (SNOMED). Chicago, IL

but rather to promote the adoption of a uniform terminology of tumours that will facilitate and improve communication among cancer workers. For this reason, we have intentionally cited only two literature references. The readers should, therefore, refer to standard works on the subject for bibliographies.

Contents

Introduction

This classification is based primarily on the microscopic characteristics of tumours and, therefore, is concerned with morphologically identifiable cell types and histological patterns, as seen with conventional light microscopy.

The term *tumours* is used synonymously with *neoplasm.* The phrase *tumour-like* is applied to lesions which resemble neoplasms, clinically or morphologically, but do not behave biologically in a neoplastic manner. They are included in this classification because they give rise to problems in differential diagnosis and because of the unclear borderline between neoplasms and certain non-neoplastic lesions. Synonyms are listed only if they have been used widely, or if they are considered to be helpful to the understanding of the lesion. In such cases, the preferred term is given first, followed by the synonym.

Although the emphasis of this classification is on histological typing, in the examination of kidney tumours, consideration should be given to the degree of cellular anaplasia, the extent of local spread, vascular and lymphatic invasion, and the occurrence of metastasis.

The scheme of histological grading suggested here is as follows: *Grade I* applies to the tumours that have the least degree of cellular anaplasia compatible with a diagnosis of malignancy; *grade III* applies to tumours with the most severe degrees of cellular anaplasia; and *grade II* applies to those tumours in between. This scheme is applicable to the carcinomas of the renal parenchyma and pelvis.

In addition to histological assessment and growth pattern, the clinical and histopathological staging of the extent of the tumour should be taken into account for the purposes of treatment and prognosis. Such a system of staging (TNM) has been developed by the *International Union Against Cancer* (see p. 35).

Histological Classification of Kidney Tumours

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[1] Morphology code of the International Classification of Diseases for Oncology (ICD-O) and the Systematized Nomenclature of Medicine (SNOMED).

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Definitions and Explanatory Notes

1 Epithelial Tumours of Renal Parenchyma

1.1 Benign – adenoma (Figs. 1–8)

A benign epithelial tumour of renal parenchyma.

1.1.1 Papillary/tubulopapillary adenoma (Figs. 1–3)

An adenoma composed of small cells with little cytoplasm; small, uniform, dark-staining nuclei; and a tubular and/or papillary growth pattern that may be compact or cystic.

The tumours are circumscribed but not encapsulated and merge imperceptibly with adjacent nephrons. Mitotic figures are rare. Collections of foamy histocytes are often present.

The tumours are usually small, asymptomatic and discovered incidentally at autopsy, in end-stage kidneys or kidneys removed surgically for other reasons. They are benign, but most adenomas larger than 1 or 2 cm will usually show areas with cellular characteristics of renal cell carcinoma. Irrespective of its size, any tumor that has clear cell areas must be considered as malignant.

1.1.2 Oncocytic adenoma (oncocytoma) (Figs. 4–7)

An adenoma composed of cells with intensely granular eosinophilic cytoplasm; the granules representing mitochondria.

The nuclei are usually small, uniform, round and vesicular, but may be large with nucleoli. In focal areas, the cells may have larger, bizarre, dark-staining nuclei, but the cytoplasm re-

mains as described above. The cells occur in nests, ovoid aggregates, tubules or microcysts.

In most cases, the cell aggregates are tightly packed at the periphery with little or no stroma. Towards the centre, they become progressively spaced, separated by oedematous or acellular hyalinised stroma, constituting the central scar seen in imaging studies. A negative vimentin might help differentiate this lesion from granular cell carcinoma. The tumour lacks the endocrine vascularity of renal cell carcinoma.

The larger tumours often expand into perinephric fat. The tumour is sharply demarcated, but a capsule is rarely present.

1.1.3 Metanephric adenoma (Fig. 8)

An adenoma composed of cells with very little pink cytoplasm in an acellular stroma.

The nuclei are only slightly larger than lymphocytes and are irregularly rounded or ovoid with delicate chromatin, sometimes with a central fold. Nucleoli are not a prominent feature, and if present, they are small and inconspicuous. The nuclei are closely spaced and often overlapping.

The cells form very small acini, separated by an acellular stroma, and are often circumscribed by a basement membrane. These acini may consist of many cells, two cells, and sometimes even a single cell. The fields are relatively uniform except for the degree of acinar crowding. Solid areas or tubular structures may be present, but the cells are identical to acinar cells. The tumours may have a papillary component, consisting of tubular or microcystic structures with invagination of the tumour into the lumen to produce a glomeruloid appearance. These may be calcified with the formation of one or more psammoma bodies. Mitotic figures are rare.

The stroma consists of oedematous tissue, a smoothly hyalinised matrix, or both, with very few cells. Occasionally, much of the tumour may be replaced by hyalinised scar, containing deposits of calcification or dystrophic ossification. None of the tumours show aggressive behaviour.

1.2 Malignant – carcinoma

1.2.1 Renal cell carcinoma (Figs. 9–29)

A malignant epithelial tumour of the renal parenchyma with variable morphologic features.

The tumours are classified according to cell type, grade and growth pattern. For each category, the tumours should be graded as grade I, grade II, or grade III, depending on the degree of anaplasia and the nuclear size compared with those in the adjacent normal renal tubules (nuclear grade I, smaller; grade II, same size; grade III, larger, sometimes pleomorphic and bizarre). Renal cell carcinomas are generally positive for cytokeratin and vimentin.

These tumours are characterised by loss of genetic material in 3 *p*; 50 % show somatic mutations in the von Hippel-Lindau (VHL) gene and an additional 10 %–20 % of clear cell renal carcinomas show inactivation of the VHL gene by epigenetic changes (hypermethylation). Sarcomatoid change occurs in approximately 5 % of these carcinomas.

Not infrequently, the tumour may show more than one cell type. The predominant cell determines the category but the cell types should be mentioned.

1.2.1.1 Clear cell carcinoma (Figs. 9–15)

A renal cell carcinoma composed of cells with clear cytoplasm.

This tumour has large, cuboidal, columnar, polygonal or wedge-like cells. Cell borders are distinct and the cytoplasm is water clear. The nucleus varies from small or large to bizarre, depending upon the grade. It may be round or ovoid with irregular, angular forms. Mitotic figures are rare.

The cytoplasm usually contains a large amount of glycogen. Fat stains reveal a considerable amount of lipid. No epithelial mucin is seen in any of the tumour cells.

In some areas, the nuclei may be round and uniform with fine, granular, evenly distributed chromatin. Nucleoli may either be absent or large and prominent, depending on the tumour grade.

The cells may show various structural patterns. They may form alveoli, cords, or acini surrounded by a network of delicate, thin-walled capillaries or sinusoids resembling an endocrine-

type of vascularity. The alveolar pattern may show little or no lumina, or a central rounded space filled with slightly acidophilic serous fluid. The tumour may form solid sheets with foci of haemorrhage and necrosis.

The stroma often contains many xanthoma cells. Haemorrhage and necrosis are often present resulting in cholesterol clefts.

Of patients with VHL disease 10 %–35 % have clear cell carcinoma. In such cases, the tumours usually occur in younger patients. They are multiple, often bilateral and increase with age.

1.2.1.2 Granular cell carcinoma (Figs. 13–17)

A renal cell carcinoma in which the cytoplasm is eosinophilic and granular.

The amount of glycogen is much less than in the clear cell type. The nuclei are similar to those seen in clear cell carcinoma, with their appearance varying with the grade. Foci of haemorrhage and necrosis are common. The cells form small sheets or closely packed trabecular structures.

The distinction between well-differentiated granular cell carcinoma and oncocytoma may sometimes be difficult, but in granular cell carcinoma the cells are less intensely granular, the cells are more cohesive and endocrine-like vascularity is more prominent.

1.2.1.3 Chromophobe cell carcinoma (Figs. 18–20)

A renal cell carcinoma with two cell types: eosinophilic and very granular due to the presence of many mitochondria and larger, nearly transparent cells with a ballooned appearance.

The two cells may be admixed, although the distended cells are often oriented along the vascular channels. The tumour cells usually occur in broad sheets that are subdivided by a prominent vascular bed. The nuclei of both cell types are similar. They are slightly pleomorphic with coarse granular chromatin and occasional prominent nucleoli. There is a perinuclear transparent zone in many cells. The periodic acid-Schiff reaction shows a fine, granular, positive reaction in some of the cells, which disappears after diastase treatment. Colloidal iron stains are positive. The tumour cells react positively with antibodies to cytokeratins, epithelial membrane antigen and soybean agglutinins. They do not react with vimentin. Numerous cytoplasmic microvesicles

are demonstrated ultrastructurally. Chromophobe renal cell carcinoma is characterised by monosomy of multiple chromosomes (1, 2, 6, 10, 13, 17 and 21) and hypodiploidy.

1.2.1.4 Spindle cell carcinoma (Figs. 21, 22)

A renal cell carcinoma containing a predominance of interlacing or whorled bands of spindle cells.

In most cases, nests of differentiated clear or granular cells are present, either separate from the spindle cell component or admixed with it. Sometimes, a distinct transition may be seen between the two cell types. Immunocytologically, the cells show considerable positivity for vimentin but heterogeneity for epithelial membrane antigen. In some tumours, both cell types react with antibodies to cytokeratins. The larger cells may react with epithelial membrane antigen. In most cases, only the differentiated areas react with either. In addition to clear cell and granular cell carcinomas, collecting-duct carcinomas may also show a spindle cell component. This is less common with chromophobe cell carcinomas and is rare with papillary carcinomas.

Synonym: sarcomatoid carcinoma.

1.2.1.5 Cyst-associated renal cell carcinoma

Two categories are identified: renal cell carcinoma originating in a renal cyst and cystic renal cell carcinoma.

1.2.1.5.1 Renal cell carcinoma originating in a cyst (Fig. 23)

A renal cell carcinoma whose cyst wall is lined by clear cells or has islands of clear cells in the fibrous wall of the cyst.

Immunohistology markers show that cytokeratins are positive and that macrophage markers are absent.

1.2.1.5.2 Cystic renal cell carcinoma (Fig. 24)

A renal cell carcinoma containing multiple cysts of varied size and number.

Some of the cysts may be lined by cells, typical of renal cell carcinoma, and in most cases these are clear cells. Similar cells may occur as aggregates within septae that usually resemble scar tissue. This category is often misdiagnosed as a multilocular cyst. In the latter, the cysts are lined by a single layer of eosinophilic cells, which may be flattened or have a teardrop or hob-

nail appearance. The stroma consists of well-differentiated spindle cells. Cystic carcinomas should be distinguished from cystic degeneration of solid tumours. Cystic lesions resulting from haemorrhage and/or necrosis and resorption of the debris show residual carcinomas buried beneath the organised exudate (Fig. 25).

1.2.1.6 Papillary renal cell carcinoma (Figs. 26, 27)

A renal cell carcinoma with rows of cuboidal or low columnar cells that are arranged around fibrovascular fronds.

The cytoplasm is usually eosinophilic, but uncommonly clear cells may be present. Nuclei are generally small and uniform indicating a low-grade neoplasm but exceptions occur when nuclei are larger and may have prominent nucleoli. Foamy macrophages often enlarge the papillary fronds, and tumour cells occasionally contain an abundance of haemosiderin. Psammoma bodies may also be numerous. If a papillary tumour is associated with tubular structures, this should be recorded (Figs. 28, 29). At least 50 % of the tumour must have a papillary structure to be included in this category.

1.2.2 Collecting-duct carcinoma (Figs. 30–33)

A malignant epithelial tumor of the kidney simulating collecting ducts of Bellini.

The tumour cells may be clear or granular, similar to renal cell carcinomas, except that cell borders are generally less distinct. They usually exhibit a moderate or severe degree of nuclear anaplasia (grade II or III). Tubular or duct-like structures are present, usually associated with a papillary element, and both show some degree of stratification of the lining cells. The papillary element frequently projects into the pelvis. These tumours react with broad-spectrum cytokeratin and with peanut lectin but neither is specific for this neoplasm.

The tumour generally recapitulates the morphology of the ducts of Bellini but, in areas, it may resemble renal cell carcinoma. The adjacent ductal epithelium may show atypical changes.

2 Epithelial Tumours of Renal Pelvis

The classification, definitions and terminology of tumours of the renal pelvis correspond to the WHO classification of urinary bladder tumours.

2.1 Benign – papilloma

2.1.1 Transitional cell papilloma

A benign papillary tumour with a delicate fibrovascular stroma covered by transitional epithelium indistinguishable from that of the normal renal pelvis and not more than six layers thick.

The individual cells are slender and lie parallel to each other at right angles to the basement membrane. Mitotic figures are rare, usually absent. If present, they are located in the basal region. The nuclei are of uniform size and show finely distributed chromatin.

Transitional cell papilloma, as defined, is very rare and most papillary tumours of the renal pelvis are carcinomas.

2.1.2 Inverted papilloma

A benign tumour of transitional cells with an endophytic growth pattern.

The tumour is similar to that observed in the bladder.

2.2 Malignant – carcinoma

2.2.1 Transitional cell carcinoma (Figs. 34–37)

A tumour of transitional epithelium showing anaplasia or invasion.

The criteria for anaplasia are: increased cellularity; nuclear crowding; disturbances of cellular polarity; failure of differentiation from the base to the surface; irregularity in size and shape of cells; variations of shape and chromatin pattern of the nuclei; displaced or abnormal mitotic figures; and giant cells. It must be

emphasised that some of these features may be present in certain inflammatory, reactive or regenerative conditions, without signifying malignancy. However, if a lesion consists of distinctive papillary fronds, with delicate, fibrovascular stalks, it is unlikely to represent a reactive process and is probably a papilloma or papillary carcinoma, depending on the presence or absence of cellular anaplasia.

The degree of cellular anaplasia forms the basis of histological grading of these tumours (see Introduction).

The growth patterns of transitional cell carcinomas of the renal pelvis are similar to those of the bladder, namely: papillary, papillary and infiltrating, infiltrating, and nonpapillary-noninfiltrating (carcinoma in situ). It is important not to confuse those cases of transitional cell carcinoma, which involve the ducts as well as the surface of the papilla, with collecting-duct carcinomas (Fig. 37). Renal pelvic transitional cell carcinomas often occur as one of multifocal lesions of the urinary tract; however, tumours at this site are particularly associated with analgesic ingestion, smoking and Balkan endemic nephropathy.

As in the bladder, transitional cell carcinoma may contain foci of squamous and/or glandular elements.

2.2.2 **Squamous cell carcinoma** (Fig. 38)

A malignant tumour of squamous epithelium.

This type of tumour must be devoid of a transitional cell elements.

2.2.3 **Adenocarcinoma of renal pelvis** (Fig. 39)

A malignant tumour with cells forming mucous glands.

This tumour resembles colonic adenocarcinoma and must be distinguished from pyelitis glandularis, although both are rare in the renal pelvis.

2.2.4 **Renal medullary carcinoma** (Figs. 40–44)

A rapidly growing tumour of the calyceal epithelium or renal papillae associated with sickle-cell trait.

These tumors typically have a reticular growth pattern, prominent desmoplasia, basophilic cytoplasm, clear nuclei and prominent nucleoli. They may occasionally have a squamous appearance, sometimes with intercellular bridges and occasional mucin droplets. Keratinisation has not been observed. The cells may have a rhabdoid appearance but do not resemble transitional cells. The tumours are centered in the medulla and associated usually with satellites in the renal cortex and peripelvic tissue.

Stromal desmoplasia is a prominent feature and constitutes a substantial bulk of the tumour. The stroma usually has a myxoid, mucous, or oedematous appearance and tends to be hypocellular. In focal areas, it may be more densely collagenous but rarely entirely collagenous.

A reticular pattern of growth is the most common morphology. The tumour-cell aggregates form spaces of varied size, reminiscent of testicular yolk-sac tumour. Transitions are observed to be of a more compact adenoid cystic appearance or, less often, the cell aggregates exhibit a microcystic pattern with micropapillations. Most cases also contain poorly differentiated areas in the form of solid sheets of cells. Occasionally, spindling of the tumour cells may be seen. Other growth patterns consist of tubular, trabecular or discrete glandular patterns.

The tumours often show haemorrhagic areas and extensive areas of necrosis which may be ischaemic, geographic in configuration, comedo or suppurative. A prominent inflammatory element is often present with polymorphonuclear leucocytes in the tumour and lymphocytes around the advancing margins. Vascular and lymphatic invasion are usually present. Invasion into renal vein, perinephric tissue and adrenals is commonly present.

These tumours occur, almost exclusively, in patients with sickle-cell trait, and sickling will usually be found focally, although some searching may be required. None exhibits the massive sickling characteristic of sickle-cell disease and none has clinical features that would suggest that diagnosis.

These tumours have been seen in younger patients (11–40 years), with a male to female ratio of two to one. The tumour has an amazing rate of growth with metastases usually present at the time of initial diagnosis. The mean duration of life after diagnosis is 15 weeks, but it may be as short as 12 weeks and rarely as long as 52 weeks.

The rapidly destructive nature of the tumour prevents definite identification of the site of origin. In some, it appears to ari-

se from calyceal epithelium, while in others it seems to arise in or near the renal papillae.

2.2.5 Undifferentiated carcinoma of renal pelvis

A malignant tumour of epithelial structure that is too poorly differentiated to be placed in any of the other groups of pelvic carcinoma.

The term *undifferentiated* is used in a histological sense and is not employed here as a synonym for high-grade cellular anaplasia. When an undifferentiated pelvic tumour invades the kidney parenchyma, there may be difficulty in distinguishing it from a primary tumour of the parenchyma or a secondary tumour from elsewhere. Identification of an in-situ renal pelvic carcinoma is the most reliable method of making this distinction.

2.2.6 Carcinosarcoma (Fig. 45)

A malignant tumour consisting of malignant epithelial and stromal components.

These tumours are usually of renal pelvic origin. The epithelial component is commonly transitional or squamous cell carcinoma. To distinguish the lesion from a carcinoma with spindling, the sarcomatous component should consist of differentiated elements which will usually be chondrosarcoma, osteosarcoma and, less frequently, rhabdomyosarcoma. Some osteosarcomas have been associated with renal cell carcinoma and likely represent carcinosarcomas.

3 Nephroblastic Lesions

The entities described in this section consist of nephroblastoma, nephrogenic rests, mesoblastic nephroma, and cystic nephroma.

3.1 Nephroblastoma (Wilms tumour) (Figs. 46–56)

A malignant tumour of primitive nephroblastic tissues which forms structures resembling those of embryonic kidney.

This type of tumour occurs primarily in children but may rarely also be encountered in adults. In particular it is found in horseshoe kidneys, Beckwith-Wiedemann syndrome, hemihypertrophy, pseudo-hermaphroditism and aniridia. Most tumours contain varying quantities of blastemal, epithelial and mesenchymal elements but many will be essentially monophasic, usually blastemal or epithelial. Each is described separately, as the tumour may consist predominantly of any element.

Blastemal elements consist of small, slightly ovoid cells with indistinct cell borders, scanty cytoplasm and round or slightly ovoid nuclei. The cells are usually closely packed, forming nodules or trabeculae. At the periphery of the nodules, the cells appear to merge with stromal elements. The nuclear chromatin tends to be slightly coarse but evenly distributed. The nucleoli are usually small. Three patterns are encountered: serpentine, nodular and basaloid. The serpentine pattern is characterised by undulating cords of blastemal cells set in a loose myxoid or fibromyxoid stroma. The cords are often relatively even, of the same size and usually sharply demarcated but there may be frequent anastomosis and the cords may not be circumscribed. The nodular pattern differs from the serpentine pattern in that the blastemal aggregates are rounded and spherical. The stroma is similar to that of the serpentine pattern. The basaloid pattern presents either a serpentine or nodular pattern, but is surrounded by a layer of epithelial cells.

The *mesenchymal* elements may present an extremely varied picture. They most often consist of spindled and myxoid cells reminiscent of embryonic mesenchyme. Skeletal muscle elements are the most common heterologous mesenchymal tissue type, ranging from primitive myoblasts to mature skeletal muscle with cross striations. Mature fibroblasts, smooth muscle, adipose tissue, cartilage, bone, osteoid, neuroglial tissue and mature ganglion cells and foci suggesting rosettes may be seen.

The *epithelial* elements also show a wide range of differentiation. The cells may consist of low columnar epithelium with a primitive appearance. They may be cuboidal or flattened. Less commonly, the epithelium may include squamous, mucous or ciliated elements. The epithelial cells may be arranged in columns

but, more often, they form primitive tubular structures resembling developing nephrons of foetal kidneys. They may present as tufts, suggesting glomeruli, usually without a capillary component. At times, the tubules are dilated, forming cystic structures; at other times they are papillary.

The presence of these three components, blastemal, mesenchymal, and epithelial, as defined, irrespective of the predominance of any one, is generally accepted as diagnostic of nephroblastoma. Occasionally, the variety and maturation of the varied elements have led to a diagnosis of teratoma. Such tumours should be sampled extensively for the presence of other components and, if found, the tumour should be labelled as nephroblastoma.

Irrespective of cellular heterogeneity, the majority of nephroblastomas are considered within the favourable histology group, defined as those which respond to treatment. A small percentage is associated with an unfavourable prognosis. This latter category, the "unfavourable" histology group, is identified by the presence of cellular anaplasia: cells with multipolar mitotic figures and marked nuclear enlargement, with major dimensions of affected nuclei being at least three times that of apparently non-anaplastic nuclei in other areas of the tumour. Anaplasia correlates with a poor prognosis when it is diffuse. *The National Wilms Tumour Study Group*[1] has redefined focal and diffuse anaplasia as follows:

- *Focal anaplasia:* Anaplastic nuclear changes are confined to one or more clearly defined loci within the primary tumour, without evidence of anaplasia or prominent nuclear atypia in extratumoural or extrarenal sites. A tumour with more than one anaplastic focus is acceptable if each is small enough to be contained on a single microscopic section, provided that the other criteria for focal anaplasia are met.
- *Diffuse anaplasia:* Anaplastic changes are present but do not meet the revised criteria for focal anaplasia for one or more of the following reasons:
  1. Anaplastic changes in the primary tumour are less confined than required by the definition of focal anaplasia.

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[1] Faria P, Beckwith B, Mishra K, Zuppan C, Weeks DA, Breslow N, Green DM. (1996) Focal versus diffuse anaplasia in Wilms tumor – new definitions with prognostic significance – a report from the National Wilms Tumor Study Group. Am J Surg Pathol 20: 909–920

2. Tumour cells with anaplastic nuclear changes are present in intrarenal or extrarenal vessels, renal sinus, extracapsular invasive sites, or metastatic deposits.
3. Anaplasia is focal but nuclear atypia, approaching the criteria for anaplasia, are present elsewhere in the tumour.
4. Anaplasia is present in a biopsy or other incomplete tumour sample.
5. Nondemarcated anaplastic nuclear changes extend to the edge of more than one section, and the report does not document that the involved sections were from the same tumour locus.

If multicentric, each tumour should be sampled similarly. Rarely, nephroblastoma may have areas of renal cell carcinoma. If of high grade, it indicates an unfavourable prognosis.

Anaplastic nephroblastoma constitutes about 5 % of the tumours. It is rare in infants, but increases to 10 % by age six and remains at this level throughout childhood and adolescence. The rate of anaplasia is two or three times greater in African-Americans than in whites.

3.2 Nephrogenic rests (Figs. 57–65)

A focus of abnormally persistent embryonic cells which have the potential of developing into nephroblastoma.

This term supplants the designation *persistent nodular blastema* since the lesions are not always nodular and often contain no blastemal cells. Nephrogenic rests are classified as perilobar or intralobar, depending on whether they are located in the periphery of the renal lobe or deeper within it. The *perilobar* type is typically multiple or diffuse and has sharp peripheral margins, while the *intralobar* type is usually single with irregular margins. They may be found in 1 % of infant (less than 3 months of age) kidneys and in up to 40 % of kidneys with nephroblastoma.

Both types of rests may be subdivided, depending on their development and morphology. Incipient rests appear as blastemal nodules in newborns and young infants; dormant rests refer to identical lesions when observed in older infants and children; involuting (sclerosing) rests are the most commonly observed lesions and consist of multiple tubules, lined by a single cell layer of dark-staining epithelium within a dense collagenous stroma.

The endstage of the latter constitutes the obsolescent rest and consists entirely of collagen. In the absence of other rests, this is not clearly distinguishable from other types of focal renal scars. Hyperplastic rests are those that have undergone proliferation and have enlarged but retained their original shape. Neoplastic rests occur as rounded, expanding nodules within any of the other types of rests. The latter is often compressed by the nodule which exhibits an increased density of embryonal-type epithelium with mitotic activity identical to nephroblastoma. Otherwise similar nodules composed of mature epithelium, less-densely packed, constitute adenomatous rests.

3.2.1 Nephroblastomatosis

The presence of diffuse or multifocal nephrogenic rests.

This should be designated as perilobar or intralobar in type. Combined nephroblastomatosis refers to the presence of both types.

3.3 Mesoblastic nephroma (Figs. 66–70)

A benign tumour consisting of interlacing fascicles of spindle-shaped cells interdigitating with groups of nephrons.

The *classic* type is characterised by interlacing fascicles of spindle cells, resembling fibroblasts or myofibroblasts, interspersed with scanty collagen. The cells are fairly uniform with little or no pleomorphism. The blood vessels are thin walled and usually sinusoidal. Haemorrhages, necrosis and cysts are common. The characteristic distinguishing feature is the peculiar growth pattern, in which the spindle-cell bundles interdigitate with groups of nephrons. The latter often have an embryonic appearance and papillary hyperplasia. The tumour margins are irregular, with radiating bands of spindle cells often extending into the perirenal or hilar region. Nodules of hyaline cartilage are frequent and in infants haematopoiesis is common.

The *cellular* or atypical variant is more common than the classic type, described above, and differs from it chiefly due to the fact that it exhibits increased cellular density and a loss of fascicular morphology. The cellular type usually consists of plump cells with large vesicular nuclei and moderate amounts

of cytoplasm. Some nuclear pleomorphism may also be present. It also tends to be more sharply circumscribed than the classic type, with little or no interdigitation with parenchymal elements. However, it is not uncommon to see both classic and cellular types within the same tumour.

3.4 Cystic nephroma (Figs. 71–77)

3.4.1 Benign cystic nephroma (Figs. 71–74)

A cystic encapsulated tumour lined by eosinophilic cuboidal or hobnail epithelium, with a supporting stroma which is myxoid or collagenous with no functioning nephrons in the septae.

In older patients, the stroma is reminiscent of ovarian stroma but no follicles are seen. Cystic nephroma, in children, occurs between 2 months and 4 years of age, with equal sex incidence. In middle-aged patients, females predominate and the stroma has a more mature fibroblastic appearance. Cystic nephroma is not to be confused with the nonencapsulated maldeveloped lesion, the multicystic kidney.

Synonyms: benign multilocular cystic nephroma, multilocular cyst.

3.4.2 Cystic, partially differentiated nephroblastoma (Fig. 75)

A lesion similar to benign cystic nephroma but with blastemal and/or epithelial elements of nephroblastoma within the septal stroma.

If such nephroblastomatous tissue should impart a nodular appearance to the gross specimen, the lesion should be classified as cystic nephroblastoma.

3.4.3 Malignant cystic nephroma (Figs. 76, 77)

Cystic nephroma with sarcomatous stroma.

This is a rare lesion, occurring entirely in adult patients, mostly in males. The epithelial lining of the cysts is identical to that of benign lesions and it is the stromal component that is anaplastic and the component that metastasizes. The proliferat-

ing stromal element often compresses the cysts into slit-like spaces and also infiltrates through the tumour capsule into the renal parenchyma.

Synonym: malignant multilocular cystic nephroma.

4 Other Paediatric Tumours

4.1 Clear cell sarcoma (Figs. 78–90)

A sarcoma composed of cords of undifferentiated cells, usually with vacuolated cytoplasm within a network of vascular channels.

The cells are polygonal and monomorphous and usually lack distinct cell borders. The nuclei have fine, granular chromatin, and nucleoli are either absent or inconspicuous. The cytoplasm is eosinophilic and often contains the clear vacuoles which impart the "clear cell" appearance of the tumour. These cells are segregated into aggregates or cords by an evenly distributed network of capillary-sized vessels supported by a variable number of spindle cells (septal cells). The histological picture just described constitutes the "classical" pattern of clear cell sarcoma of the kidney and comprises the bulk of most tumours. However, it may be present only focally, with much of the lesion exhibiting *variant* patterns which may cause confusion with a wide variety of other neoplasms. Such variant patterns generally result from variations in the morphology of the "cord cells" and/or the "septal cells". Bone metastasis is a characteristic feature of this type of sarcoma.

Variant patterns of clear cell sarcoma:

1. The *epithelioid* pattern. The tumour cells are condensed into nodular cell aggregates or ribbons simulating epithelial lesions, particularly nephroblastoma.
2. *Spindled* pattern. This consists of a proliferation of the spindle cells associated with the vessels (septal cells). These may obscure the cords to the extent of resembling storiform fibrohistocytic tumours. Spindling of cord cells may also occur, with or without septal-cell spindling.
3. *Sclerosing* pattern. The tumour often undergoes hyalinisation with dense collagen replacing the cord pattern. Extensive sclerosis should suggest a diagnosis of clear cell sarcoma of the

kidney, since this is most unusual in other renal tumours with the exception of previously treated nephroblastomas.
4. *Myxoid* pattern. Extensive accumulation of mucopolysaccharide may replace large areas of recognisable tumour morphology and be confused with myxoid areas of nephroblastoma.
5. *Cystic* pattern. This is very common and results from coalescence of mucoid ground substance into cysts of variable size. Dilatation of entrapped renal tubules may also occur, resulting in epithelial-lined cysts.
6. *Pericytomatous* pattern. Dilatation of the vascular element has sometimes been a prominent feature, resulting in some resemblance to haemangiopericytoma.
7. *Palisading* pattern. Nuclear palisading, resembling neurilemomas, is observed focally in about 15 % of cases.
8. *Pleomorphic* pattern. Although rarely seen, this occurs as foci of bizarre, pleomorphic cells and may include abnormal mitoses. Such areas might be confused with a wide variety of sarcomas and other malignancies.

4.2 Rhabdoid tumour (Figs. 91–97)

A malignant tumour with a monomorphic population of noncohesive cells with large vesicular nuclei, prominent nucleoli, and scattered cytoplastic inclusions.

This tumour occurs in sheets of large, loosely cohesive cells with eosinophilic granular cytoplasm and distinct cell borders. The distinctive cytoplasmic inclusions which impart the "rhabdoid" appearance tend to occur only in focal areas. Cross striations have not been demonstrated. The stroma tends to be collagenous and accumulations of basophilic ground substance may suggest chondroid differentiation. Numerous variant patterns occur:

1. *Epithelioid pattern:* trabecular, mucoid, alveolar, pseudoglandular
2. *Spindled cell pattern:* broad fascicular, myxoid, pericytomatous, storiform or palisading
3. *Sclerosing pattern:* fibrotic, osteosarcomatoid or chondroid
4. *Lymphoid pattern:* solid or histiocytoid

In nearly all cases, the characteristic cellular morphology, with large central nucleoli, is present. The proportion of cells with in-

clusions is variable and their identification may occasionally require extended searching. Immunostains have given variable results but the most consistently positive results have been with cytokeratin and vimentin.

The median age of patients with rhabdoid tumours is 11 months and more than 90 % are under 3 years of age. The tumour predominates in males. Renal rhabdoid tumour is one of the most lethal tumours of childhood. Eighty percent die of their disease, and most deaths occur within 1 year of diagnosis.

4.3 Neuroblastoma (Fig. 98)

This neurogenic tumour, like that seen in the adrenals, can also occur in the kidney. It is composed of sheets of small rounded cells which are divided into small aggregates by a delicate fibrovascular stroma. The cells have scant cytoplasm. The nuclei are round or polygonal and deeply staining. In better-differentiated tumours, the cells have attenuated cytoplasmic processes (neurites) which are polarised to form rosettes.

Renal involvement is an occasional feature of adrenal neuroblastoma and ganglioneuroma. In some instances, the kidney has been reported to be the primary site. Neuroblastoma-like areas with rosettes and ganglion cells may occur in nephroblastoma.

5 Non-epithelial Tumours

5.1 Benign

5.1.1 Angiomyolipoma (Figs. 99–103)

A hamartoma composed of three elements: adipose tissue, blood vessels, and smooth muscle cells.

These tumours are usually large and solitary. Multifocal lesions are sometimes present, particularly with the tuberous sclerosis complex. They are benign tumours (hamartomas). The adipose tissue cells vary in size and the smooth muscle cells may show considerable variation in size, shape and staining qualities. Their cytoplasm typically has a moth-eaten appearance. Occasionally, the cells may be darker staining and bizarre in shape.

There may be mono- or multinucleated giant cells and rare mitotic figures. The arrangement and organisation of the smooth muscle cells vary from small groups between adipose-tissue cells and perivascular collars, to large sheets similar to leiomyoma. Rarely, the tumour may have areas resembling juxtaglomerular cells.

The vascular component displays great variation in type, size, arrangement and structure. The major vascular component consists of large, arterial-type vessels which are extremely tortuous and reminiscent of cirsoid aneurysms. The lumina show much variation in diameter and, frequently, are eccentric. The subendothelial connective tissue and the media are frequently of variable thickness and structure, consisting of smooth muscle, collagen or both. Most vessels have little or no elastic tissue. Smaller vessels include immature-appearing smooth muscle cells arranged circumferentially. The large size of many of the tumours, the absence of encapsulation, frequent multicentricity, frequent haemorrhage and necrosis, extension outside the kidney, pleomorphism of muscle cells and occasional extension into regional lymph nodes simulate a malignant neoplasm, but are regarded as benign tumours, although they have recurred when incompletely removed. It should be emphasised that some angiomyolipomas contain very little adipose tissue, while in others, the smooth muscle cells are sparse and inconspicuous. Immunohistology using antibodies to HMB 45 (a melanoma-related antigen) is positive and can be helpful in difficult cases.

5.1.2 **Leiomyoma** (Figs. 104–106)

Renal leiomyomas are usually capsular but may be cortical. They are usually an incidental finding. Clinically evident lesions are generally seen in adults but have also been reported in neonates and children. These have been regarded as hamartomas and are closely related to the angiomyolipoma.

Synonym: capsuloma.

5.1.3 Lipoma

True intrarenal lipomas are extremely rare and it is questionable if they occur. Most reported cases have appeared to occur in or adjacent to the renal capsule, making it all the more likely that these represent predominantly lipomatous angiomyolipomas or myolipomas.

5.1.4 Renomedullary interstitial cell tumour (Fig. 107)

A benign medullary tumour composed of spindle cells with indistinct cell borders and vesicular nuclei.

The stroma is loose, oedematous and either eosinophilic or basophilic. Varying amounts of collagen are seen and there are occasional entrapped tubules. Calcification is sometimes present. The cells resemble renal medullary interstitial cells and contain neutral lipids, phospholipids and mucopolysaccharides. They have no demonstrated significance.

Synonym: renal medullary fibroma.

5.1.5 Haemangioma (Fig. 108)

This tumour occurs most commonly in the wall of the renal pelvis, adjacent to the pyramids. It may be difficult to find in resected kidneys because of collapsed vessels.

5.1.6 Lymphangioma

The tumour consists of flattened endothelial cells lining spaces and separated by fibrous tissue which may contain smooth muscle cells. The spaces generally contain lymphocytes. This type of tumour is less frequent than haemangioma and usually occurs in young adults, in the peripelvic region, rarely in the kidney.

5.1.7 Juxtaglomerular cell tumour (Figs. 109–111)

A tumour of juxtaglomerular cells

This tumour is composed of a uniform population of polyhedral, ovoid or round cells with granular acidophilic cytoplasm and nu-

clei that are round or oval. The cells may be arranged in an organoid or trabecular pattern. The tumour may contain tubular or papillary structures. The cytoplasmic granules react positively with periodic acid – Schiff and Bowie reagents. Mitotic figures are rare. The tumour occurs mostly in young adults with an age range of 8–53 years. All patients have hypertension (due to renin-production by the tumour), often for many years prior to detection of the tumours. The elevated blood pressure is due to secretion of renin. Hypertension disappears after removal of the tumours. The tumours are usually small and unilateral.

5.2 Malignant soft tissue tumours (Figs. 112–117)

For these rare tumours, the *World Health Organization Histological Typing of Soft Tissue Tumours is used*[1]. The most common intrarenal sarcoma is the leiomyosarcoma. It most likely arises from the large vessels and renal pelvic muscularis and does not differ histologically from those seen elsewhere. Haemangiopericytoma, angiosarcoma, and rhabdomyosarcoma rarely occur as primary renal tumours. The same is true of malignant fibrous histiocytoma and fibrosarcoma, although it is difficult to distinguish these from spindled (sarcomatoid) renal cell carcinomas. Intrarenal liposarcomas have been reported but angiomyolipoma must always be ruled out before this diagnosis is accepted. Osteosarcomas are occasionally seen.

6 Miscellaneous Tumours

6.1 Carcinoid tumour (Fig. 118)

Most of the cells are argyrophilic. About one third of cases with typical histology have metastasised, chiefly to bone. More poorly differentiated variants (atypical carcinoids/neuroendocrine carcinomas) metastasise more frequently.

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[1] Weiss SW (1994) World Health Organization. Histological Typing of Soft Tissue Tumours. Second Edition. Springer-Verlag, Heidelberg

6.2 Small cell carcinoma

Small cell carcinomas, like those of the lung, occur rarely in the renal parenchyma and some have developed as an element of renal pelvic carcinoma. Their histological features, immunohistochemical reactions and aggressive behaviour do not differ from their pulmonary counterparts.

6.3 Primitive neuroectodermal tumour (Figs. 119, 120)

This small round cell tumour (SRCT), closely related to Ewing sarcoma, is of neural origin and occurs in the kidney. The tumour is composed of sheets of loosely cohesive neoplastic cells separated by scattered bands of collagen. Poorly formed rosette-like structures are occasionally found. The cells are small, uniform and have little cytoplasm with high nuclear-cytoplasmic ratios. Nuclei have slightly irregular contours with coarse granular chromatin and are about three times the size of lymphocytes. Adjacent tissue is invaded in a broad-front rather than an infiltrative pattern. The monoclonal antibodies HBA71 or 013 are useful for distinguishing these from other SRCT. Also, they often react with neuron-specific enolase. Synaptophysm and chromogranin may be focally positive but cytokeratin AE1/AE3 is negative. These are highly aggressive tumours usually seen in young adults.

Synonym: peripheral neuroepithelioma.

6.4 Ossifying renal tumour (Figs. 121, 122)

A benign tumour composed of proliferating spindle cells admixed with partially calcified, osteoid matrix.

This type of tumor occurs mostly in infants less than 1 year old. It consists of a calcified mass in the pelvis attached at the papilla and simulates a calculus.

6.5 Renal hamartoma

6.5.1 Renal cortical hamartoma (Figs. 123, 124)

A hamartoma of the cortex composed of epithelial and mesenchymal elements.

Most often the epithelial elements are distributed sparsely around the periphery of the lesion and resemble the common cortical tubulopapillary adenoma, while the mesenchymal elements are, most commonly, mature smooth muscle. This type of lesion has also been interpreted as mesoblastic nephroma of adults although the complete histological spectrum indicates a more complex lesion. The epithelial element may be diffuse and, at times, the stroma may include adipose tissue, collagen and/or angiomatous elements. The cortical hamartoma is benign and occurs chiefly in adult females. Descriptively, the majority of these tumours are essentially adenoleiomyomatous in appearance.

6.5.2 Renal pelvic hamartoma (Figs. 125–127)

A hamartoma of the pelvis with a diverse histological appearance containing epithelial and mesenchymal constituents. The latter is typically undifferentiated.

In this type of tumour, the epithelium may resemble cortical adenoma although the tubules may be more diffuse. Microcysts may be present, lined by cuboidal, columnar, or hobnail type cells. The stroma usually contains mature smooth muscle fascicles alternating with sheets of small mesenchymal cells. Numerous vessels are usually present. This lesion occurs as a polyp attached to the juxtapelvic cortical or medullary parenchyma and projects into the pelvic cavity. It is benign and mostly occurs in adult females.

6.6 Nephrogenic adenofibroma (Figs. 128, 129)

A benign tumour of epithelial and stromal elements involving a segment of the kidney.

This is an unencapsulated lesion which involves a segment of the kidney. In the cortical aspect, it consists of an epithelial element similar to the metanephric adenoma, while the remainder

of the segment is composed of a cellular, collagenous stroma. In some cases, there is, in addition, a papillary lesion projecting into the pelvic cavity which resembles collecting duct carcinoma.

6.7 Intrarenal teratoma

Renal teratoma is a very rare lesion and most reported cases have been nephroblastomas with teratoid features. A true teratoma in the kidney should include non-renal elements such as intestinal tract, skin adnexal structures, brain, etc.

6.8 Malignant lymphoma (Figs. 130, 131)

Although the kidneys are commonly involved in the late stages of systemic lymphoma, primary renal lymphoma is rare. The latter lesion is usually localized to the hilar region, but may occur as one or more discrete nodules in the parenchyma. Histologically, these exhibit an interstitial growth pattern with entrapment of glomeruli, nephrons and ducts. Most are of the diffuse, large B-cell type. Chronic lymphocytic leukaemia should be suspected when the infiltrate is diffuse rather than nodular, particularly if bilateral. Malignant lymphoma also arises in the kidney or elsewhere in renal allograft recipients. Some examples of posttransplantation lymphoproliferative disease contain Epstein-Barr virus and the deposits may regress when immunosuppressive treatment is withdrawn.

6.9 Malignant melanoma (Fig. 132)

This may be found in the kidney without an apparent primary tumour elsewhere.

7 Secondary Tumours

The tumours in this category may be metastatic, direct extensions or the kidneys may be involved by a systemic neoplastic process such as leukaemia. The common primary sites are lung,

breast, melanoma, tumours of gastrointestinal tract, ovary, testes or contralateral kidney.

8 Tumour-like Lesions

A number of non-neoplastic lesions can resemble renal tumours clinically or histologically. Developmental abnormalities may result in a mixture of primitive renal elements which may be mistaken for nephroblastic tumours.

8.1 Renal dysgenesis (Figs. 133, 134)

A malformed kidney consisting of cysts and tubules surrounded by mesenchymal stroma and admixed with varying numbers of nephrons.

In renal dysgenesis, the presence of cartilage is not infrequent and the lesion may involve the entire kidney or a segment of the kidney. In adults the stroma is more mature.

Synonyms: renal dysplasia or multicystic kidney.

8.2 Vascular malformations (Fig. 135)

Certain vascular malformations and anastomoses which are not true angiomas may simulate tumours.

8.3 Cysts

Cysts may be lined by a single layer of epithelial cells, ranging from flattened to low cuboidal or columnar cells. Cysts can pose as neoplasms, clinically. Carcinoma may be found in association with cysts or arising in a cyst wall, as noted in 1.2.1.5.

8.4 Renal tubular hyperplasia

Focal tubular hyperplasia, e. g. adjacent to foci of pyelonephritis, may be indistinguishable from a small adenoma.

8.5 Xanthogranulomatous pyelonephritis (Fig. 136)

An inflammatory condition characterised by the accumulation of large histiocytes with foamy cytoplasm.

The lesion is characterised by a zone of necrotic debris or granulation tissue which outlines the contour of the destroyed calyceal epithelium and adjacent medulla. Deep to this zone is a band of histiocytes (or foamy macrophages) with a variable number of multinucleated giant cells. These are associated with numerous plasma cells and lymphocytes. Abscesses may also be present. The cortex shows much scarring. This zonal configuration produces a typical gross appearance since the lipid-filled macrophages appear as a wavy, yellow band outlining the lumen of the calyces. The zonal morphology also distinguishes this from clear cell carcinoma as does the absence of tubules, alveoli or other organoid structures.

8.6 Malakoplakia (Figs. 137, 138)

An inflammatory process in which histiocytes have a granular eosinophilic cytoplasm containing Michaelis-Gutmann bodies.

The lesion in malakoplakia primarily involves the pelvis. A variant of this lesion, located primarily in the cortex and containing poorly defined Michaelis-Guttman bodies, is referred to as megalocytic interstitial nephritis or as cortical malakoplakia.

8.7 Cholesteatoma (Fig. 139)

Squamous metaplasia, particularly in hydronephrosis and in pyelonephritis with stones, may lead to entrapment of squamous epithelium within the renal parenchyma, which may mimic squamous cell carcinoma. Large collections of keratin may result in the formation of a cholesteatoma.

8.8 Inflammatory pseudotumour (Figs. 140–142)

A rare lesion of the kidney which presents a rounded, tumour-like configuration composed of dense collagen and admixed with variable numbers of plasma cells and lymphocytes. It may also in-

clude a spindle cell myofibroblastic element mimicking the fibrous histiocytoma.

Distinction of these pseudotumours from burned-out neoplasms may be difficult, but the latter generally exhibit much calcification in addition to cystic areas resulting from resorption of remote haemorrhage or necrosis. These lesions may resemble plasma cell granulomas and myofibroblastic pseudotumours as seen in other organs.

Pseudotumour has been used in the radiological literature for a congenital hyperplasia of renal tissue which may present as a tumour-like mass. This is rare but has led to nephrectomy.

8.9 Adrenal rests (Fig. 143)

Adrenal rests occur within the superficial renal cortex, usually as a flat plaque or disc-like structure of adrenal cortex, and are found in 4–6 % of the population. On rare occasions, the various adrenal cortical neoplasms have been reported to involve this tissue.

Synonym: adrenal-renal fusion.

8.10 Nephrogenic adenoma (Figs. 144, 145)

Cuboidal metaplasia of transitional epithelium of renal pelvis.

Following ulceration or other forms of chronic irritation, the transitional epithelium becomes cuboidal and lines the surface, often with papillations, and forms tubular or microcystic structures in the adjacent stroma. Hob-nail epithelium often lines the microcysts and tubules and some may have prominent basement membranes. The stroma usually shows oedema and chronic inflammation. The lesion is benign.

8.11 Others

A fibroepithelial polyp, as described for the bladder, occurs occasionally in the renal pelvis.

TNM Classification of Tumours of the Kidney[1]

Rules for Classification

This classification applies only to renal cell carcinoma. There should be histological confirmation of the disease.

The following are the procedures for assessing T, N and M categories:

T categories	Physical examination and imaging
N categories	Physical examination and imaging
M categories	Physical examination and imaging

Regional Lymph Nodes

The regional lymph nodes are the hilar, abdominal para-aortic and paracaval nodes. Laterality does not affect the N categories.

TNM Clinical Classification

T – Primary Tumour

TX Primary tumour cannot be assessed
T0 No evidence of primary tumour
T1 Tumour 7.0 cm or less in greatest dimension, limited to the kidney
T2 Tumour more than 7.0 cm in greatest dimension, limited to the kidney
T3 Tumour extends into major veins or invades adrenal gland or perinephric tissues but not beyond Gerota fascia
  T3a Tumour invades adrenal gland or perinephric tissues but not beyond Gerota fascia
  T3b Tumour grossly extends into renal vein(s) or vena cava below diaphragm

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[1] Sobin LH, Wittekind Ch (eds) (1997) TNM classification of malignant tumours, 5th edition. Wiley, New York.

T3c Tumour grossly extends into vena cava above diaphragm
T4 Tumour invades beyond Gerota fascia

N – Regional Lymph Nodes

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single regional lymph node
N2 Metastasis in more than one regional lymph node

M – Distant Metastasis

MX Distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

pTNM Pathological Classification

The pT, pN and pM categories correspond to the T, N and M categories.

Stage Grouping

Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1	N1	M0
	T2	N1	M0
	T3	N0, N1	M0
Stage IV	T4	N0, N1	M0
	Any T	N2	M0
	Any T	Any N	M1

TNM Classification of Tumours of the Renal Pelvis and Ureter[1]

Rules for Classification

This classification applies only to carcinomas. Papilloma is excluded. There should be histological or cytological confirmation of the disease.

The following are the procedures for assessing T, N and M categories:

T categories	Physical examination, imaging and endoscopy
N categories	Physical examination and imaging
M categories	Physical examination and imaging

Anatomical Sites

1. Renal pelvis (C65.9)
2. Ureter (C66.9)

Regional Lymph Nodes

The regional lymph nodes are the hilar, abdominal para-aortic and paracaval nodes and, for the ureter, the intrapelvic nodes. Laterality does not affect the N classification.

TNM Clinical Classification

T – Primary Tumour

TX	Primary tumour cannot be assessed
T0	No evidence of primary tumour
Ta	Noninvasive papillary carcinoma
Tis	Carcinoma in situ
T1	Tumour invades subepithelial connective tissue

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[1] Sobin LH, Wittekind Ch, (Eds) (1997) TNM classification of malignant tumours, 5th edition. Wiley, New York.

T2 Tumour invades muscularis
T3 (Renal pelvis) Tumour invades beyond muscularis into peripelvic fat or renal parenchyma
(Ureter) Tumour invades beyond muscularis into perireteric fat
T4 Tumour invades adjacent organs or through the kidney into perinephric fat

N – Regional Lymph Nodes

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single regional lymph node 2 cm or less in greatest dimension
N2 Metastasis in a single lymph node more than 2 cm but not more than 5 cm in greatest dimension or multiple lymph nodes, none more than 5 cm in greatest dimension
N3 Metastasis in a lymph node more than 5 cm in greatest dimension

M – Distant Metastasis

MX Distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

pTNM Pathological Classification

The pT, pN and pM categories correspond to the T, N and M categories.

Stage Grouping

Stage 0a	Ta	N0	M0
Stage 0is	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T3	N0	M0
Stage IV	T4	N0	M0
	Any T	N1, N2, N3	M0
	Any T	any N	M1

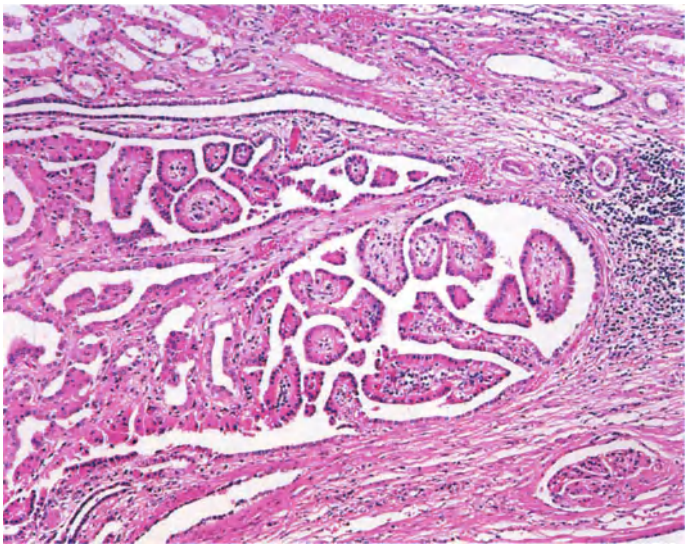

Fig. 1. *Papillary adenoma.* Fibrovascular stalks lined by a single layer of uniform cells

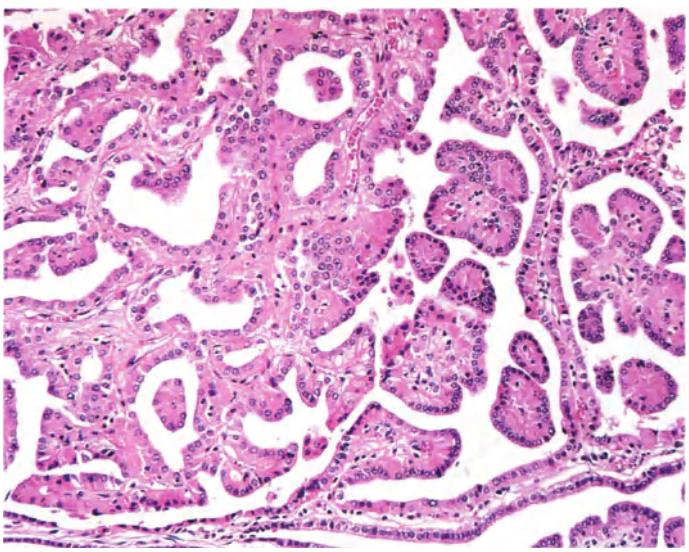

Fig. 2. *Tubulopapillary adenoma.* Epithelial cells have scant eosinophilic cytoplasm and uniform, small nuclei

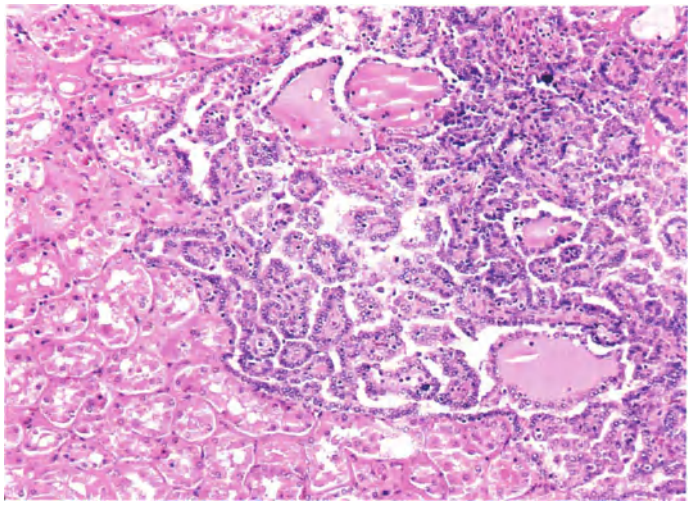

Fig. 3. *Papillary adenoma.* This example includes acini but the cells are small and uniform

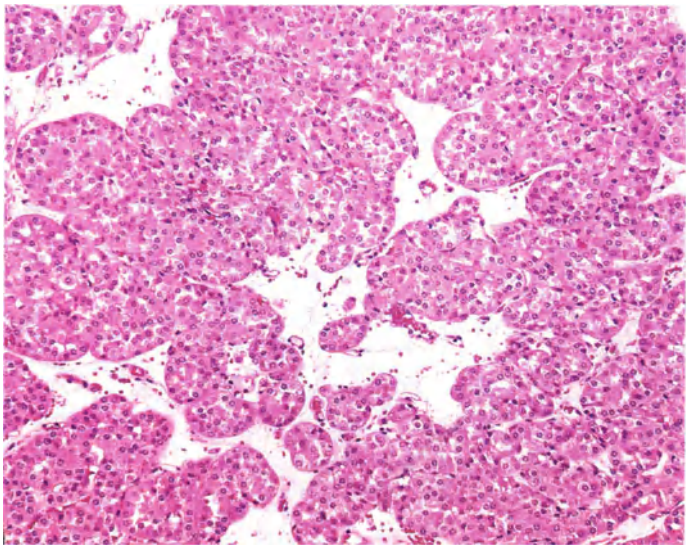

Fig. 4. *Oncocytic adenoma (Oncocytoma).* This is the common morphology, with eosinophilic cells in rounded aggregates

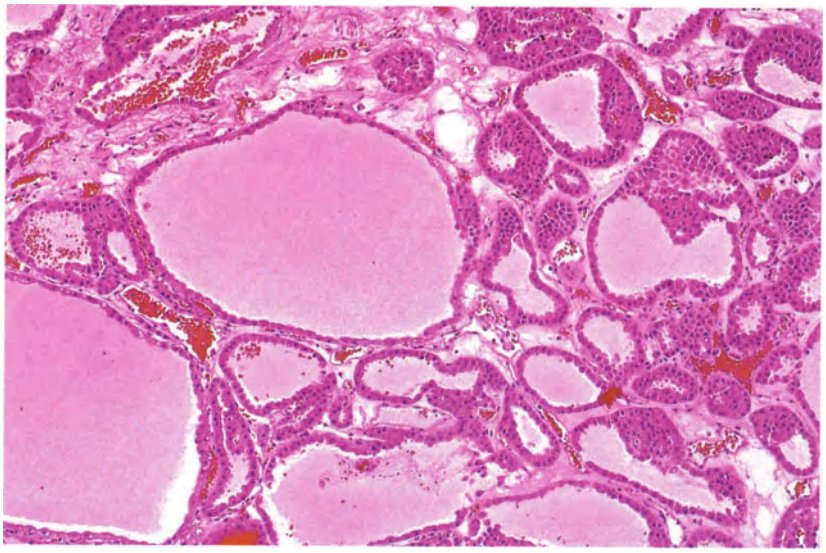

Fig. 5. *Oncocytic adenoma, microcystic pattern*

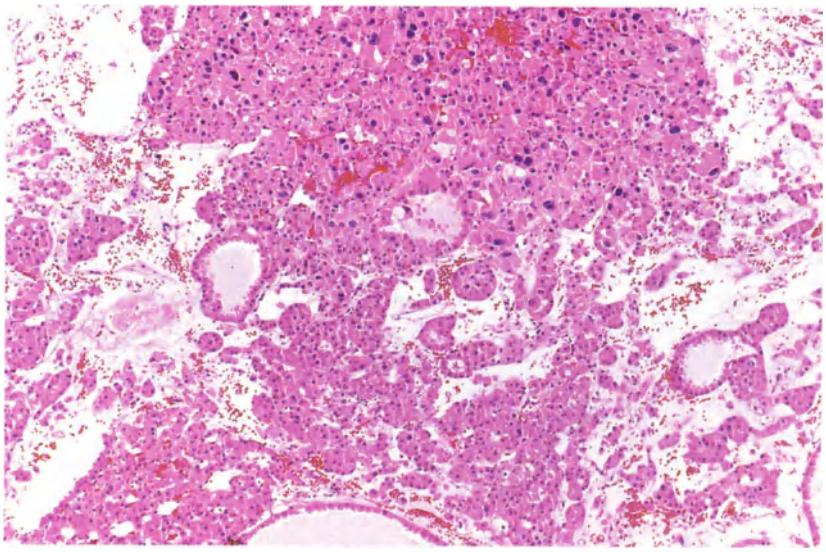

Fig. 6. *Oncocytic adenoma with focal pleomorphism*

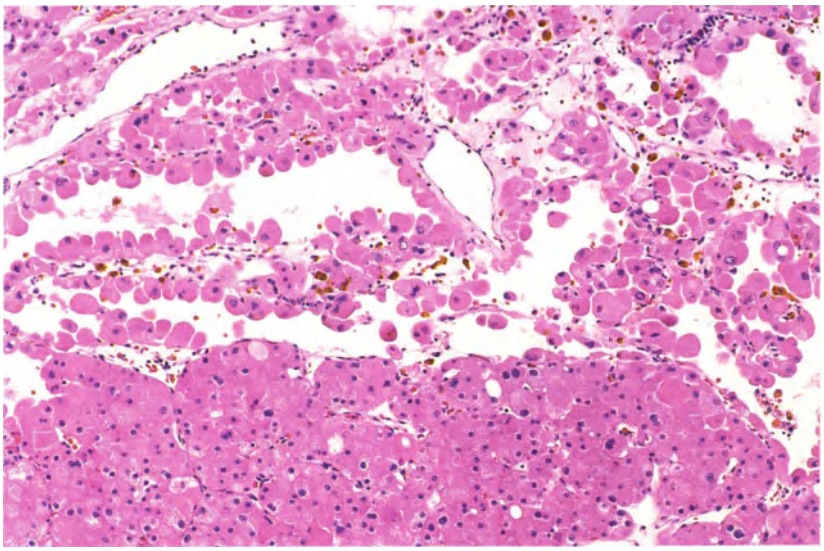

Fig. 7. *Oncocytic adenoma.* Lack of intercellular cohesion and, in this example, much larger cells

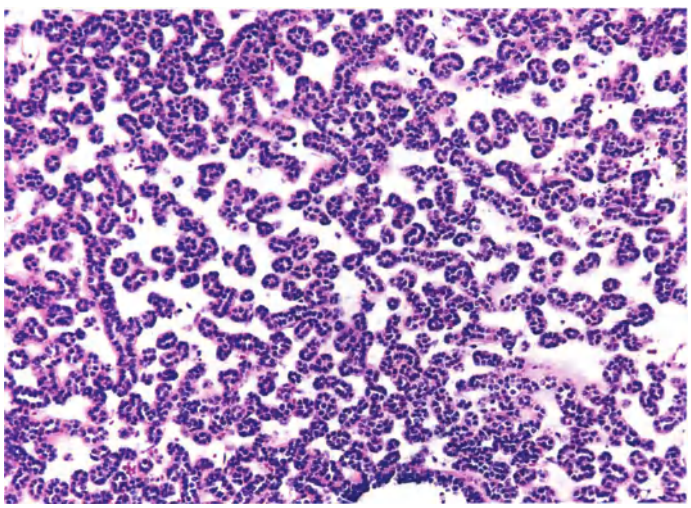

Fig. 8. *Metanephric adenoma.* Small cells, small tubules and acellular stroma

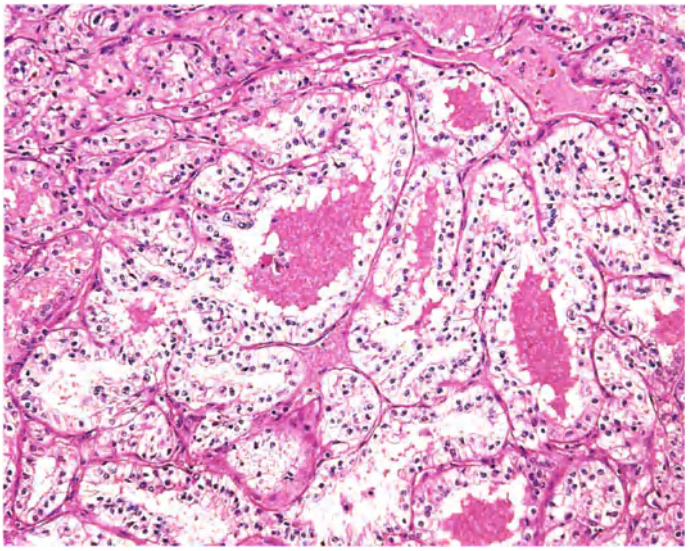

Fig. 9. *Renal cell carcinoma, clear-cell type.* Acinar pattern with eosinophilic secretion

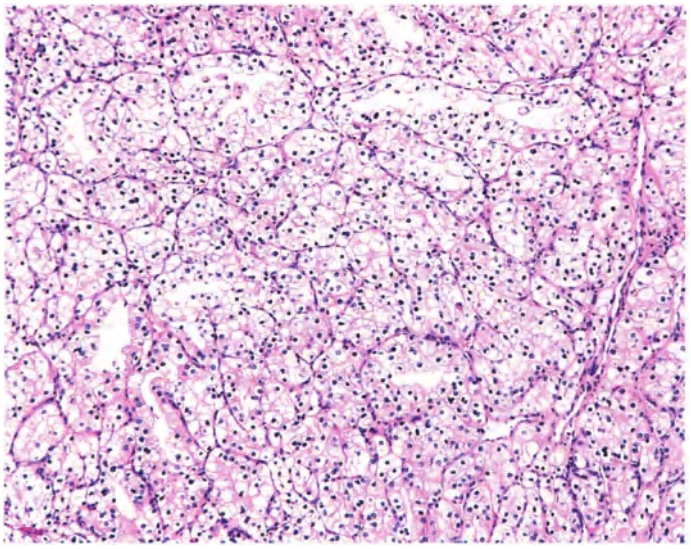

Fig. 10. *Renal cell carcinoma, clear-cell type.* Solid, acinar and tubular features

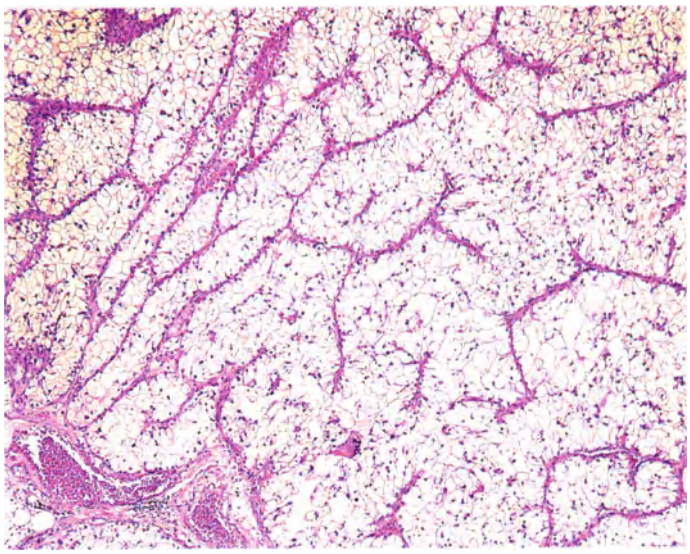

Fig. 11. *Renal cell carcinoma, clear-cell type*

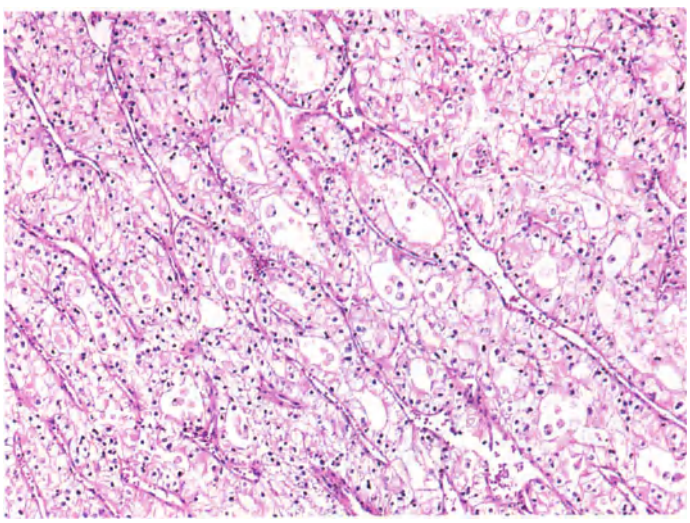

Fig. 12. *Renal cell carcinoma, clear-cell type.* Tubular pattern

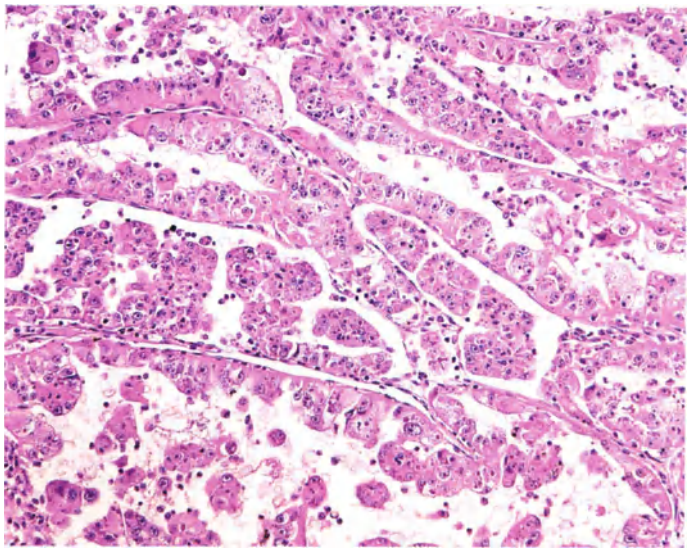

Fig. 13. *Renal cell carcinoma, clear-cell and granular-cell types*

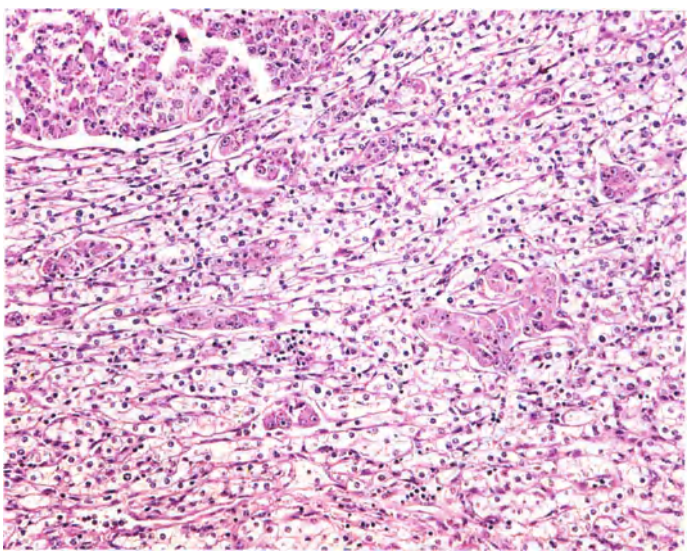

Fig. 14. *Renal cell carcinoma, clear-cell and, focally, granular-cell type*

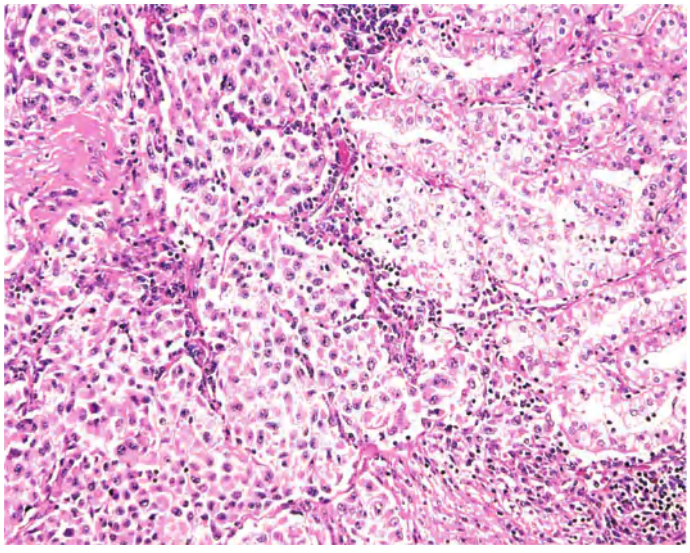

Fig. 15. *Renal cell carcinoma, clear-cell and granular-cell types.* Granular-cell elements often show a greater degree of nuclear anaplasia than do the clear cell elements

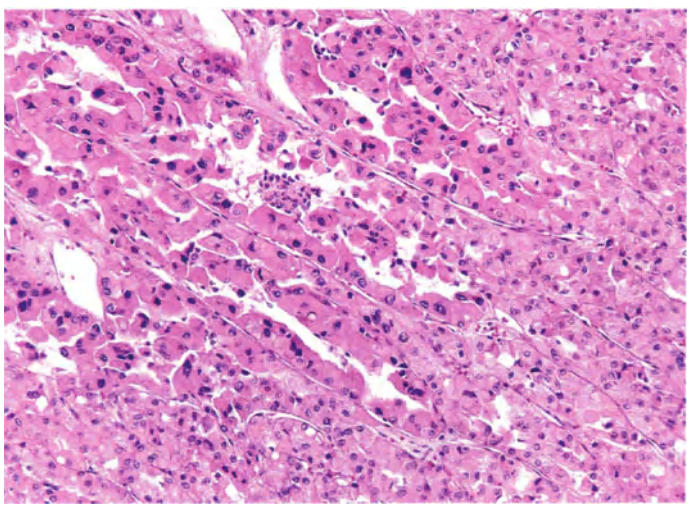

Fig. 16. *Renal cell carcinoma, granular-cell type*

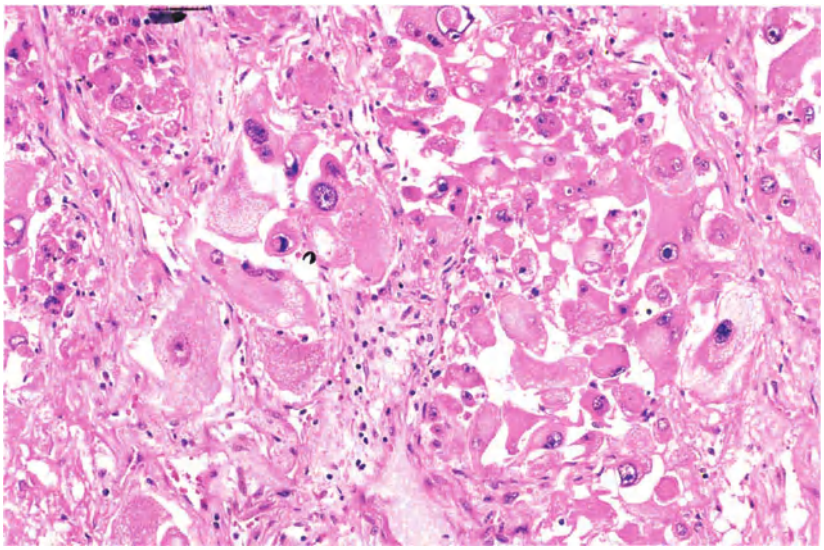

Fig. 17. *Renal cell carcinoma, granular-cell type.* Marked cellular anaplasia indicates a high grade tumour

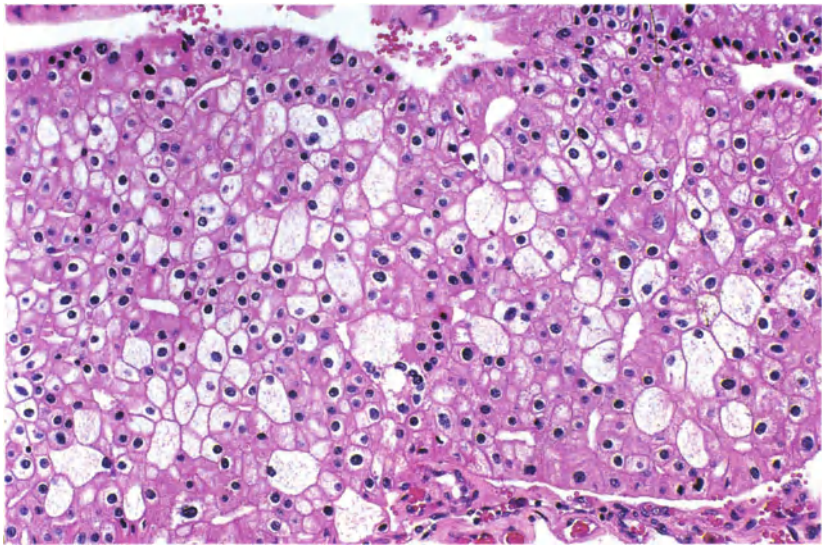

Fig. 18. *Renal cell carcinoma, chromophobe-cell type.* Large pale cells with a ballooned appearance and the smaller, eosinophilic cells

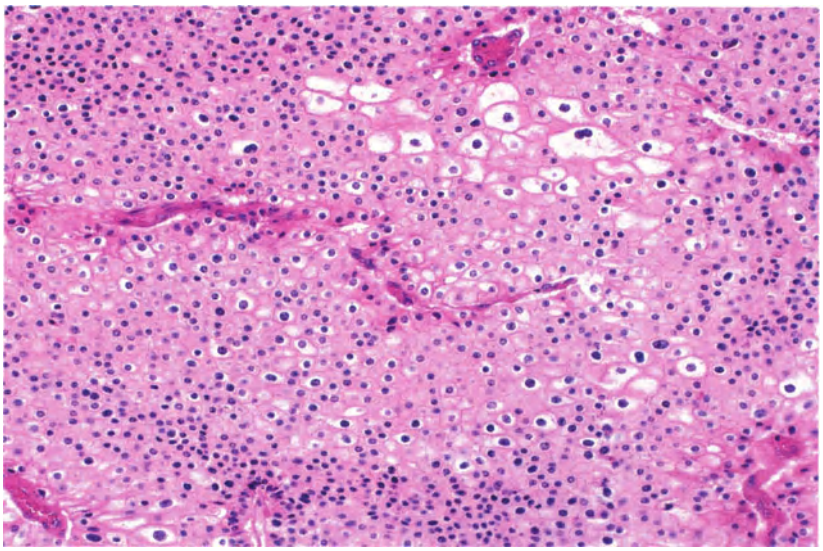

Fig. 19. *Renal cell carcinoma, chromophobe-cell type.* Most cells in this field are the smaller eosinophilic type and these often have a prominent perinuclear halo

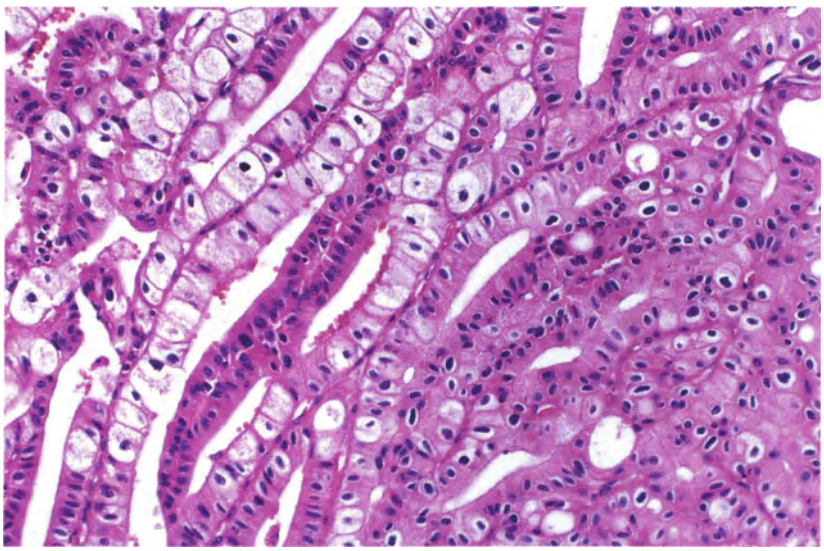

Fig. 20. *Renal cell carcinoma, chromophobe-cell type.* Most of these tumours have a solid growth pattern but this one is tubular

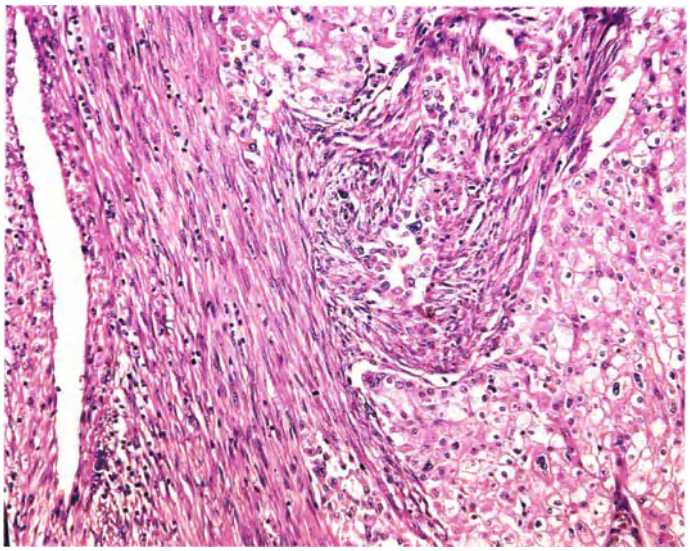

Fig. 21. *Renal cell carcinoma, spindle-cell type.* Differentiated carcinoma is on the right

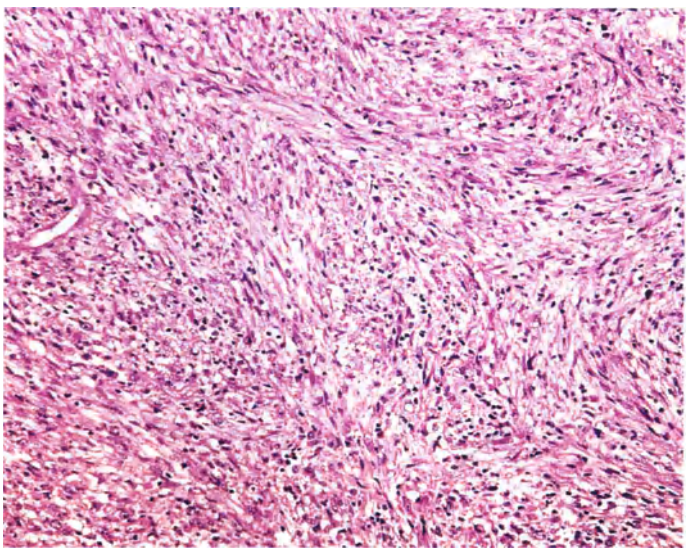

Fig. 22. *Renal cell carcinoma, spindle-cell type*

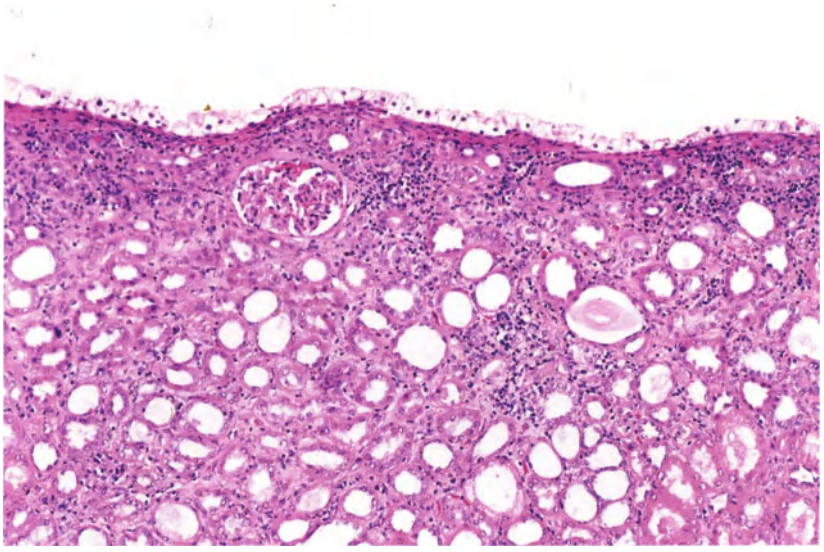

Fig. 23. *Renal cell carcinoma* arising in a solitary cyst. The cyst is lined by a row of clear cells typical of renal-cell carcinoma

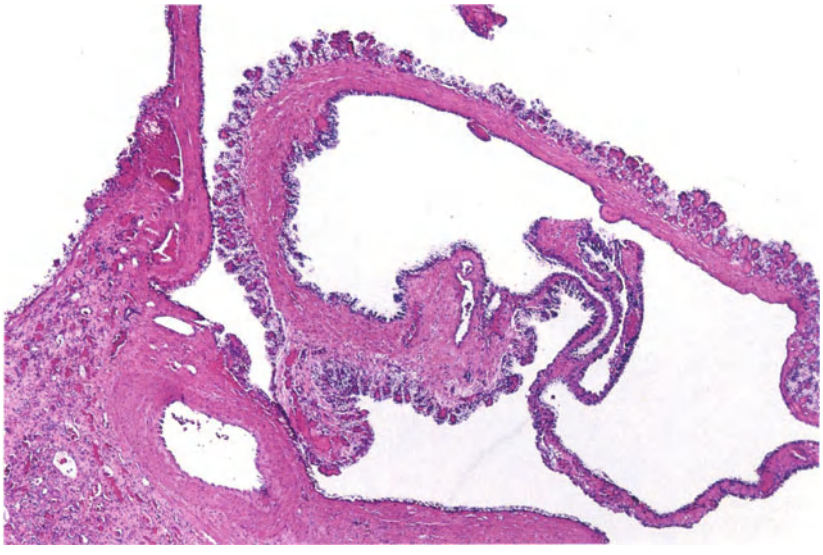

Fig. 24. *Cystic renal cell carcinoma.* The cysts that comprise the tumour are lined by clear cells

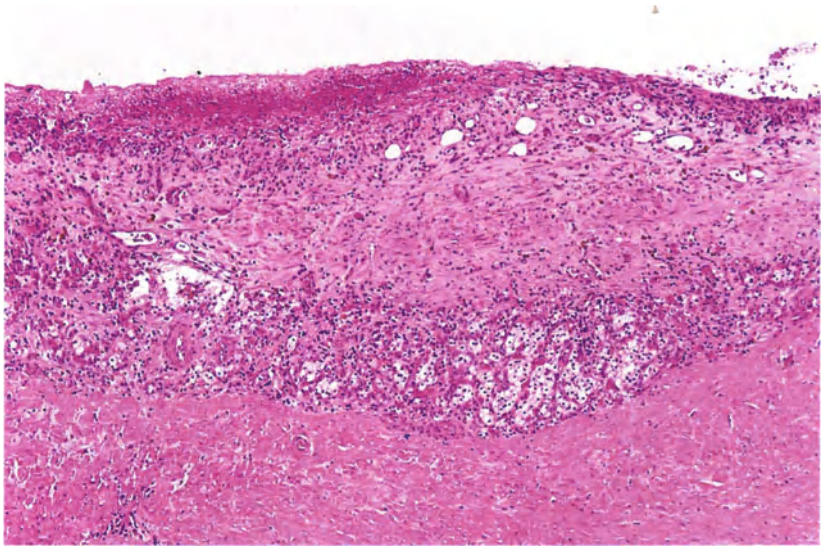

Fig. 25. *Renal cell carcinoma with cystic degeneration.* The lumen of the cyst is at the top and the residual carcinoma is located beneath the organised exudate

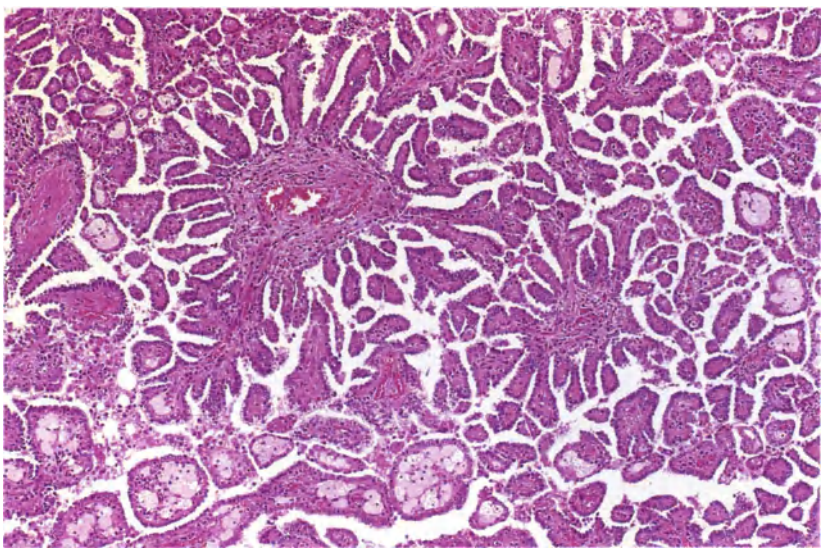

Fig. 26. *Renal cell carcinoma, papillary.* Some of the fronds are distended by foamy macrophages (bottom)

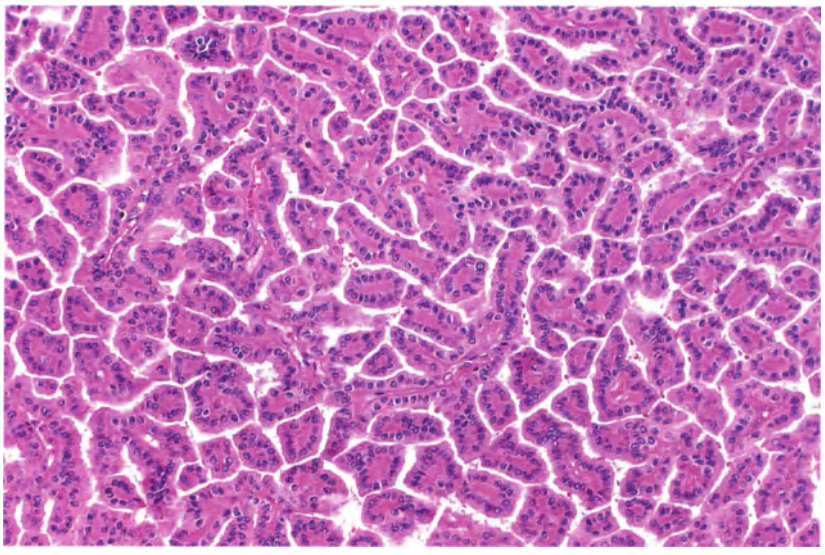

Fig. 27. *Renal cell carcinoma, papillary*

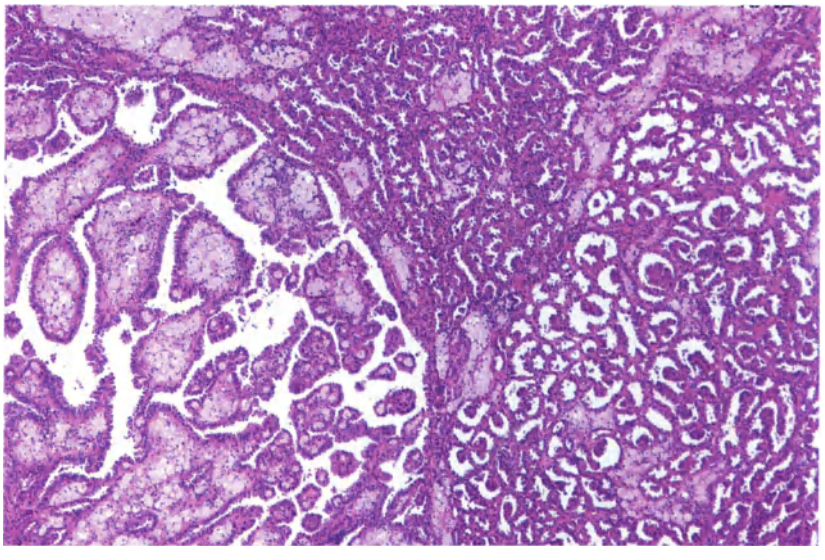

Fig. 28. *Renal cell carcinoma, tubulopapillary*

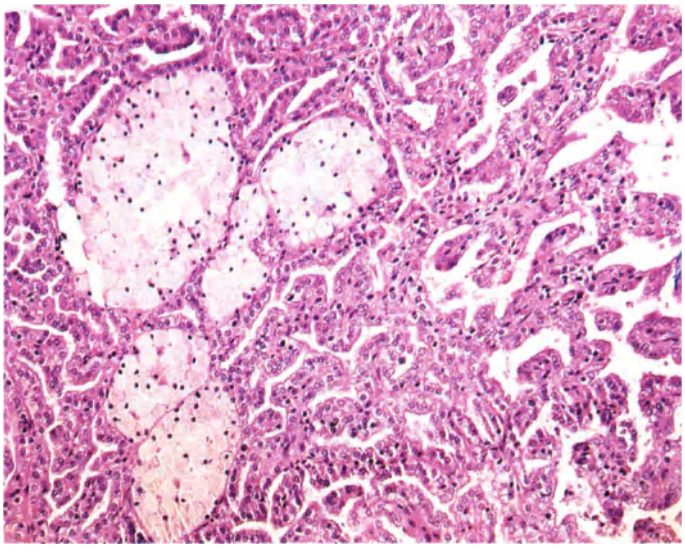

Fig. 29. *Renal cell carcinoma, tubulopapillary.* Accumulations of foamy macrophages

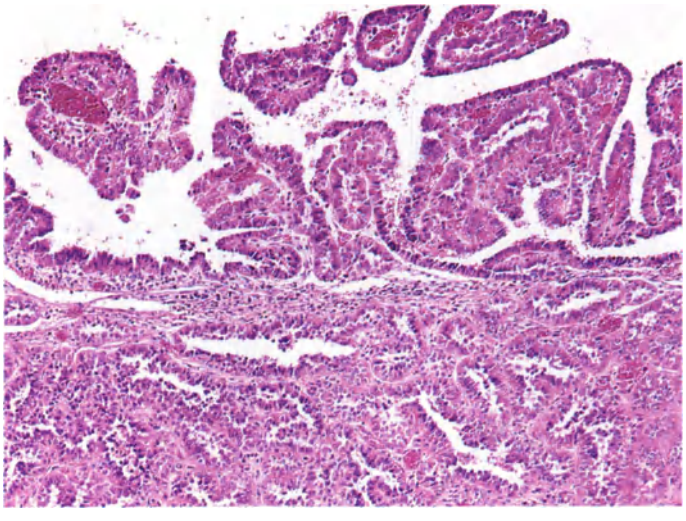

Fig. 30. *Collecting-duct carcinoma.* Tumour of medulla projecting into the renal pelvis

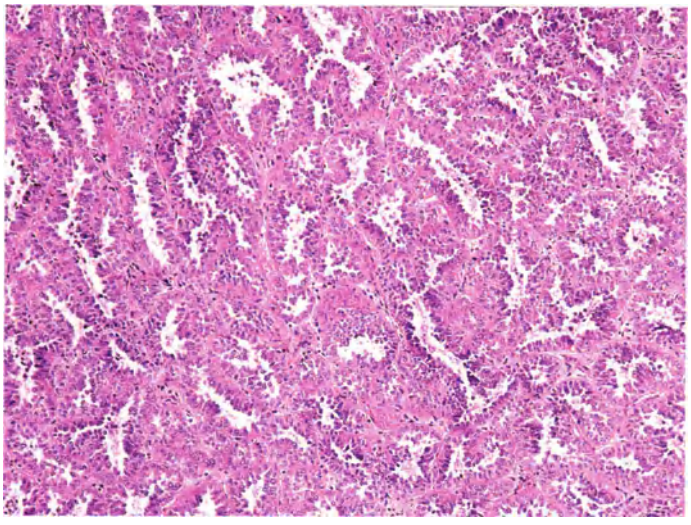

Fig. 31. *Collecting-duct carcinoma.* Tumour cells tend to be stratified and have indistinct cell borders

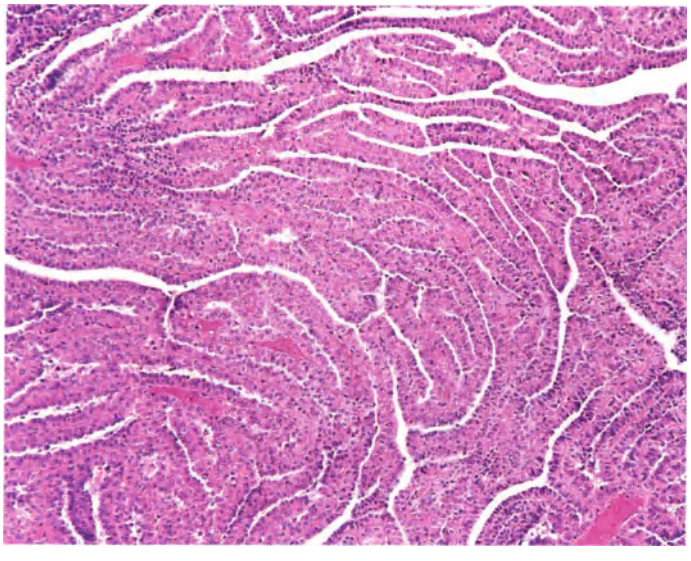

Fig. 32. *Collecting-duct carcinoma.* Tubulopapillary features

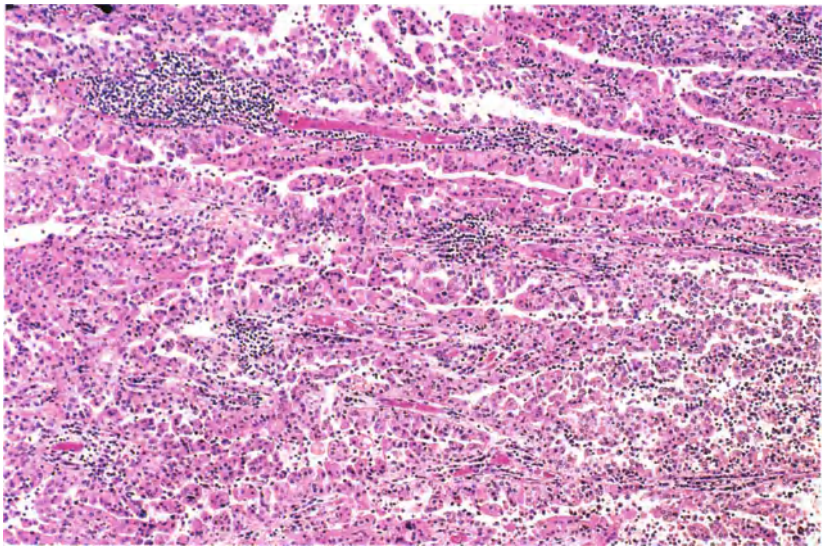

Fig. 33. *Collecting-duct carcinoma.* The tumour recapitulates the medullary ducts of origin

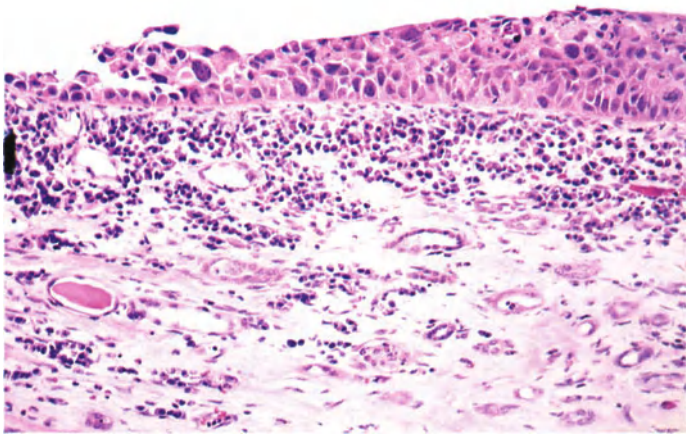

Fig. 34. *Transitional cell carcinoma in situ, renal pelvis*

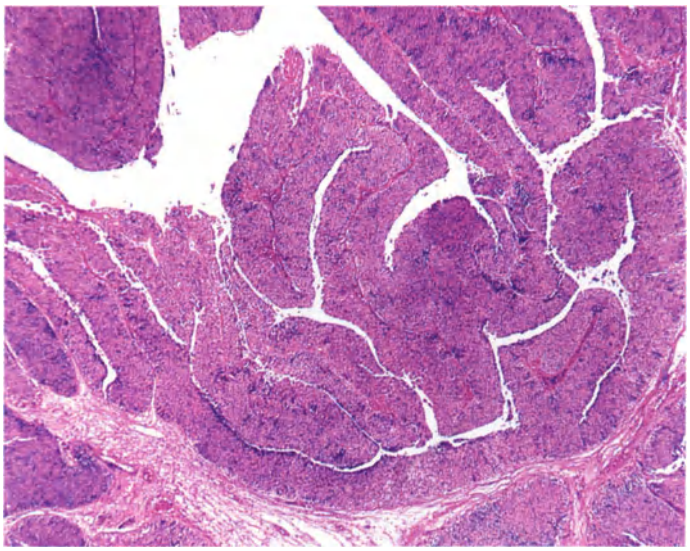

Fig. 35. *Transitional cell carcinoma, papillary, renal pelvis*

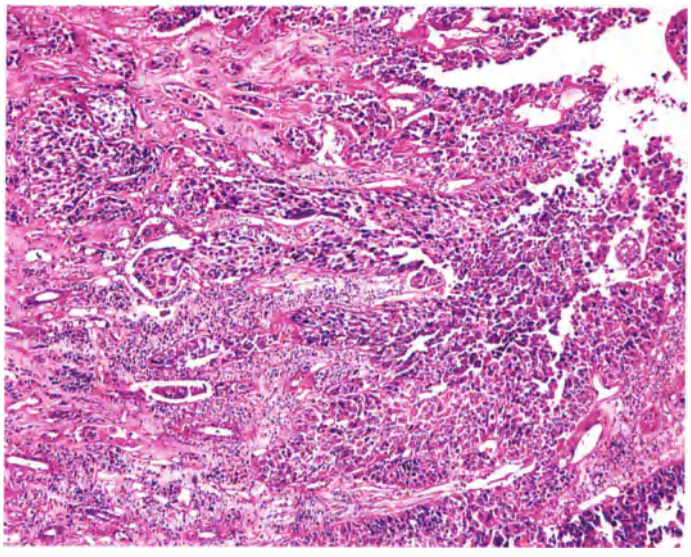

Fig. 36. *Transitional cell carcinoma, renal pelvis.* Infiltrating kidney

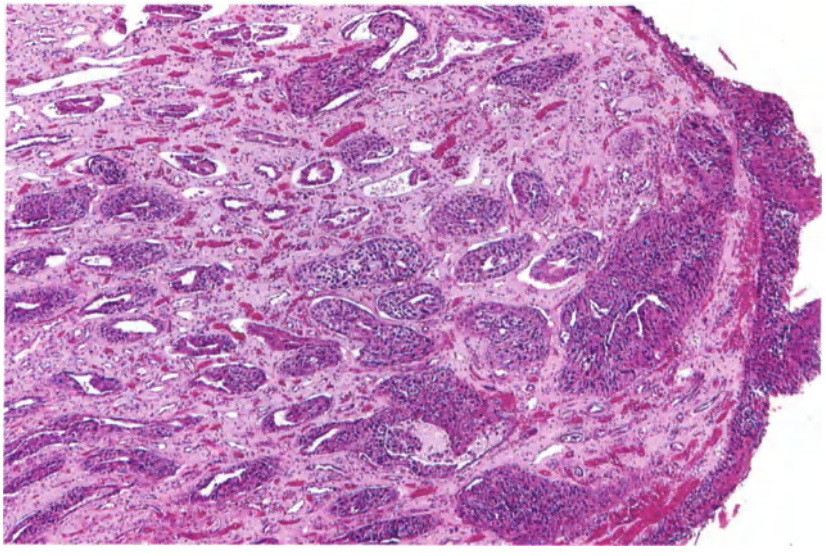

Fig. 37. *Transitional cell carcinoma, renal pelvis.* Infiltrating collecting ducts

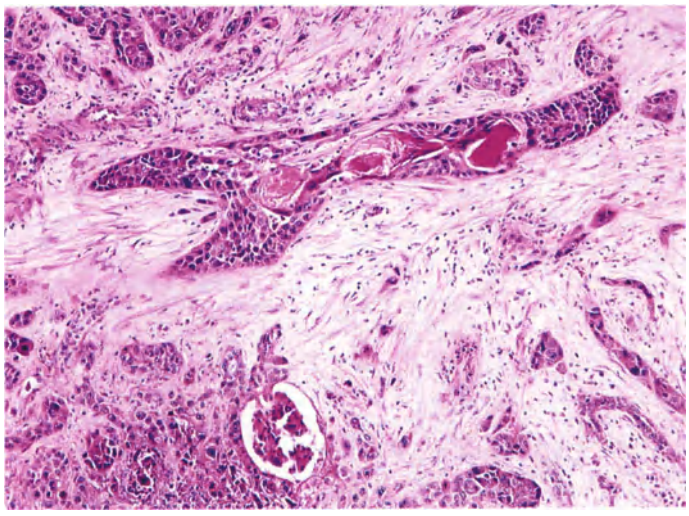

Fig. 38. *Squamous cell carcinoma, renal pelvis.* Infiltrating renal cortex

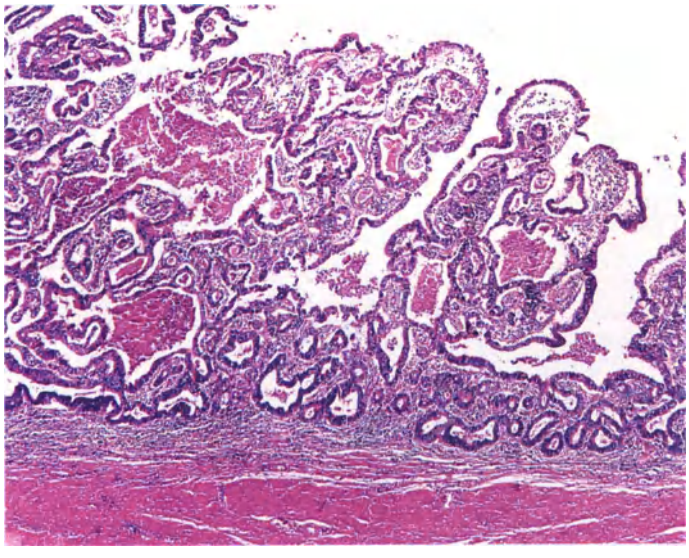

Fig. 39. *Adenocarcinoma, renal pelvis.* There is superficial stromal invasion

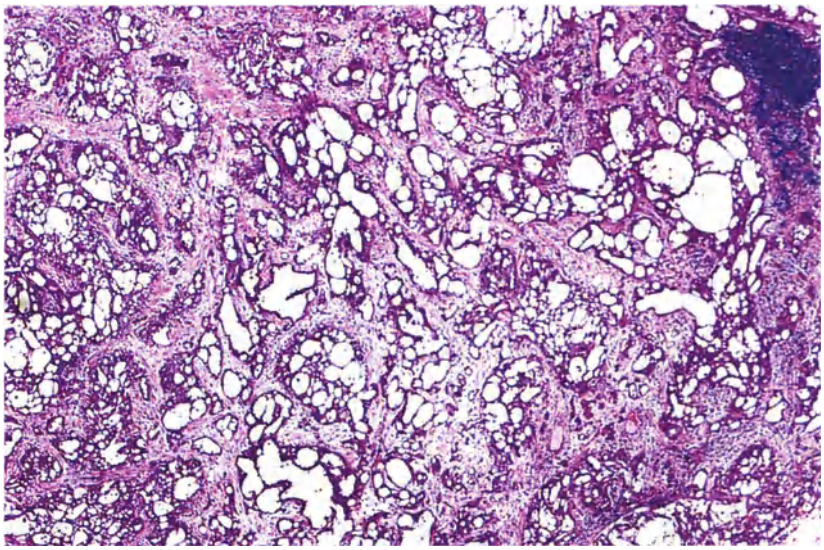

Fig. 40. *Renal medullary carcinoma.* This shows the more common yolk sac-like and adenoid cystic morphology

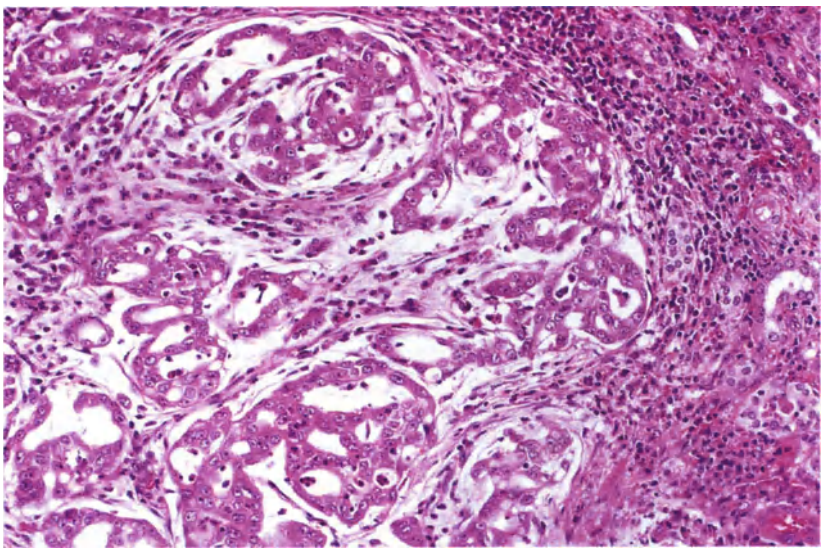

Fig. 41. *Renal medullary carcinoma.* Polymorphonuclear leucocytes infiltrate through the tumour

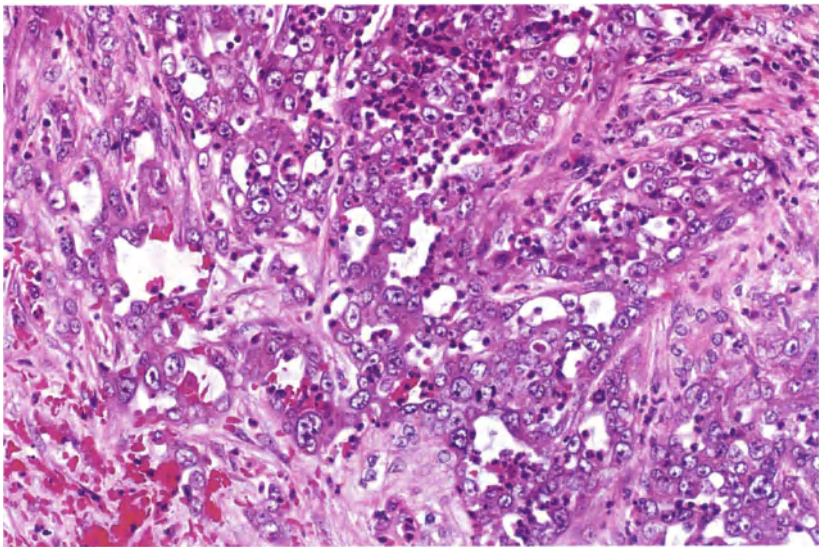

Fig. 42. *Renal medullary carcinoma.* The tumour cells are generally dark, have large nucleoli and frequently are infiltrated by inflammatory cells

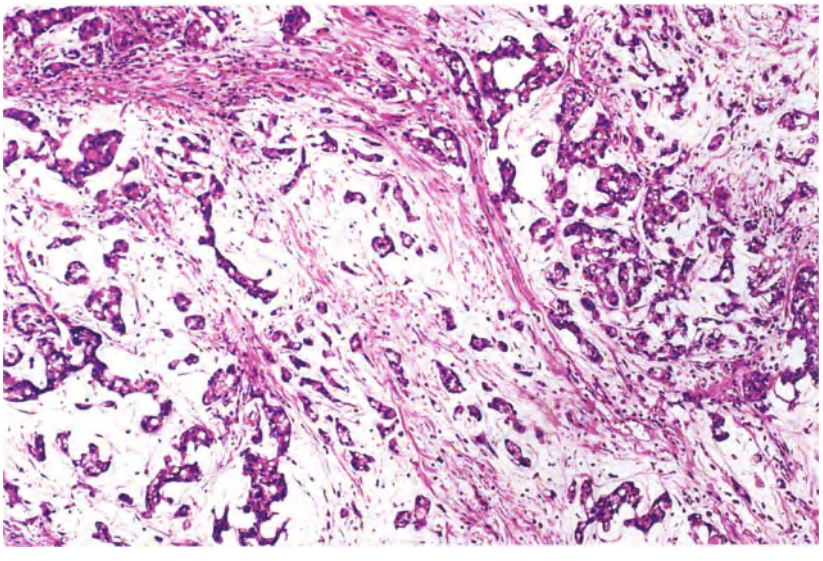

Fig. 43. *Renal medullary carcinoma.* Marked degree of stromal oedema

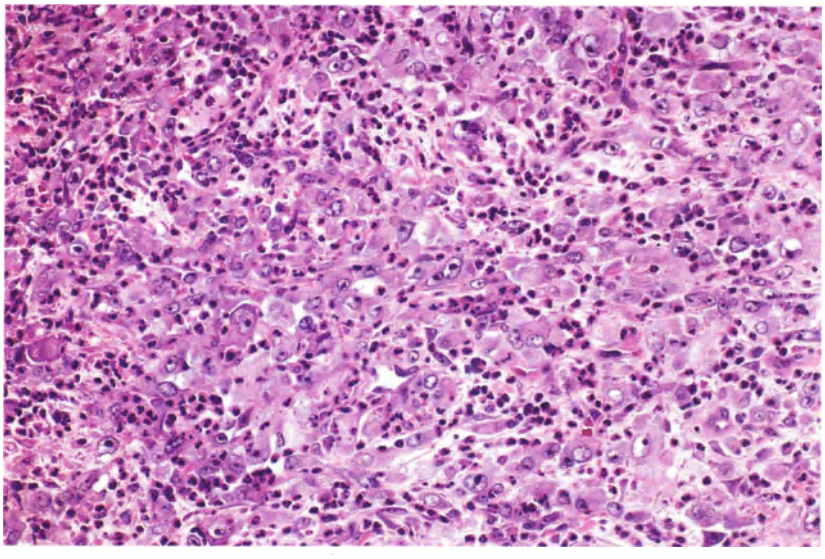

Fig. 44. *Renal medullary carcinoma.* A poorly differentiated area admixed with polymorphonuclear leucocytes

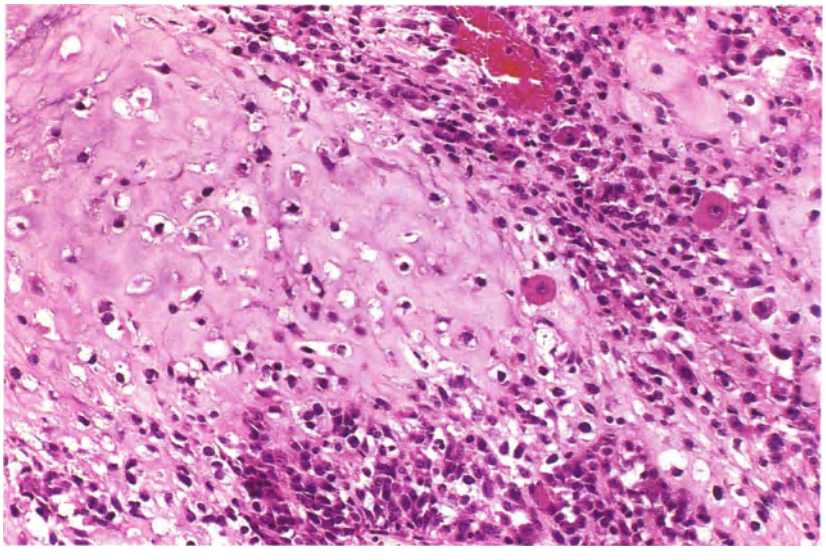

Fig. 45. *Carcinosarcoma, renal pelvis.* This field shows chondrosarcoma and rhabdomyosarcoma, including several rhabdomyoblasts

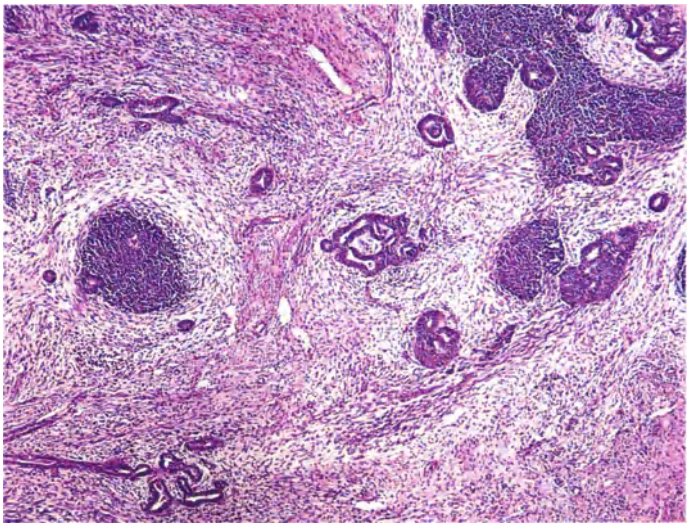

Fig. 46. *Nephroblastoma.* Triphasic pattern

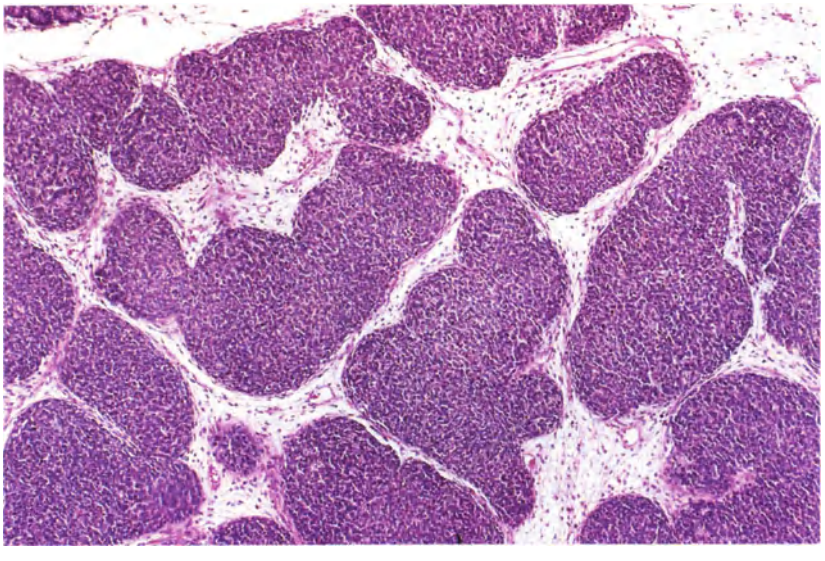

Fig. 47. *Nephroblastoma.* Nodular blastemal pattern

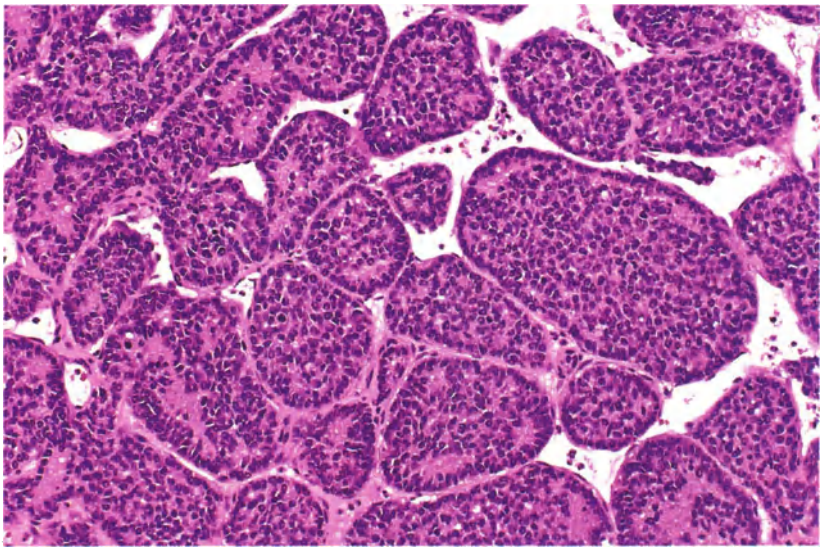

Fig. 48. *Nephroblastoma.* Basaloid blastemal pattern

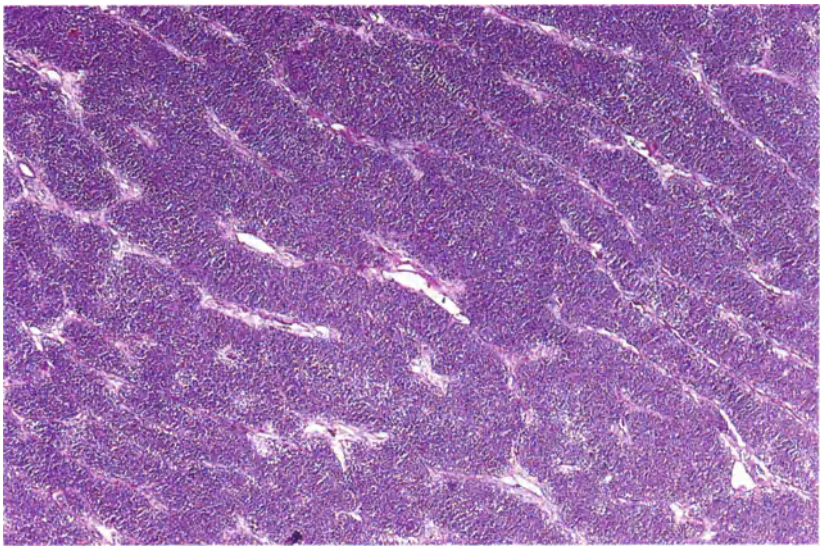

Fig. 49. *Nephroblastoma.* Serpentine blastemal pattern

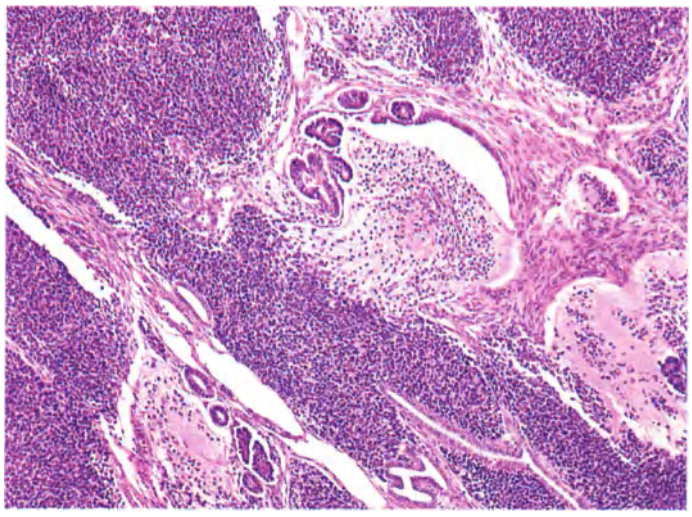

Fig. 50. *Nephroblastoma.* Glial tissue in addition to blastemal and epithelial elements

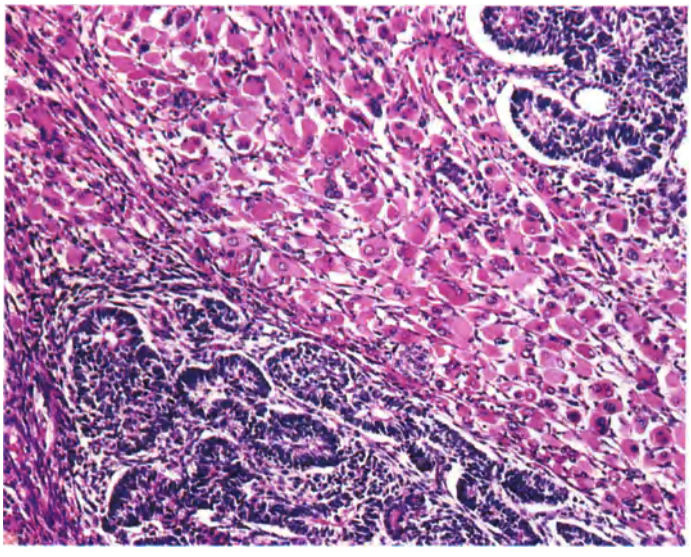

Fig. 51. *Nephroblastoma.* The mesenchymal element is striated muscle

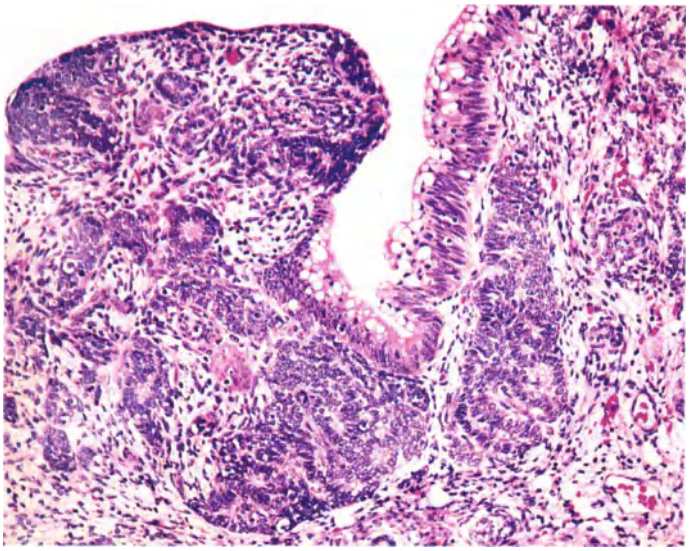

Fig. 52. *Nephroblastoma.* Wall of this cystic tumour is partly lined by differentiated mucinous epithelium

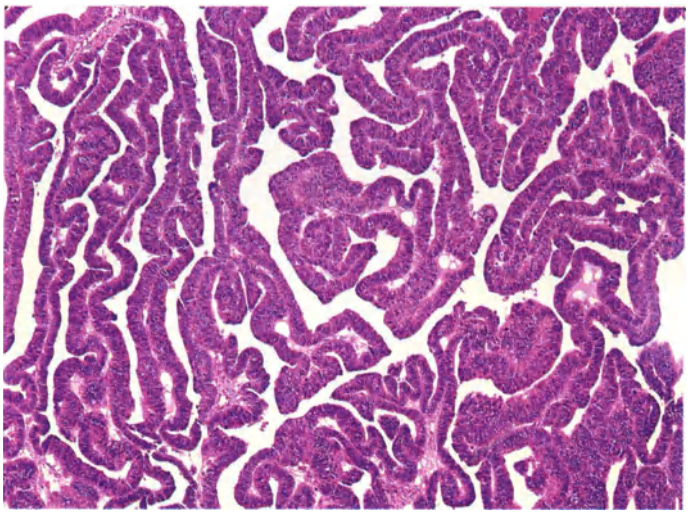

Fig. 53. *Nephroblastoma.* Epithelial pattern

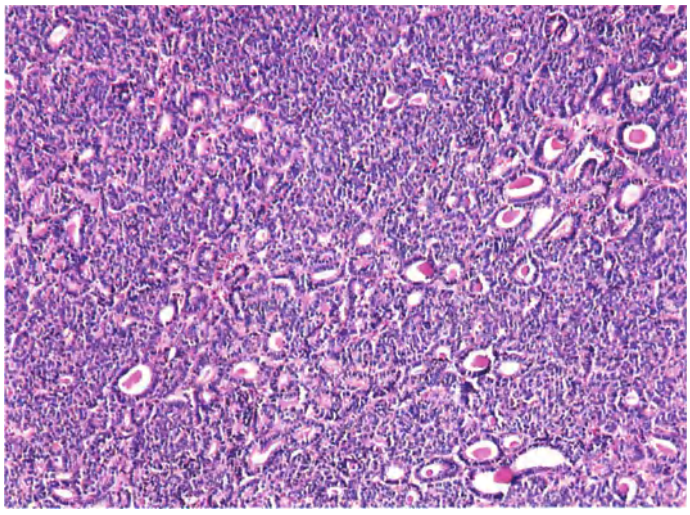

Fig. 54. *Nephroblastoma.* Epithelial and blastemal elements

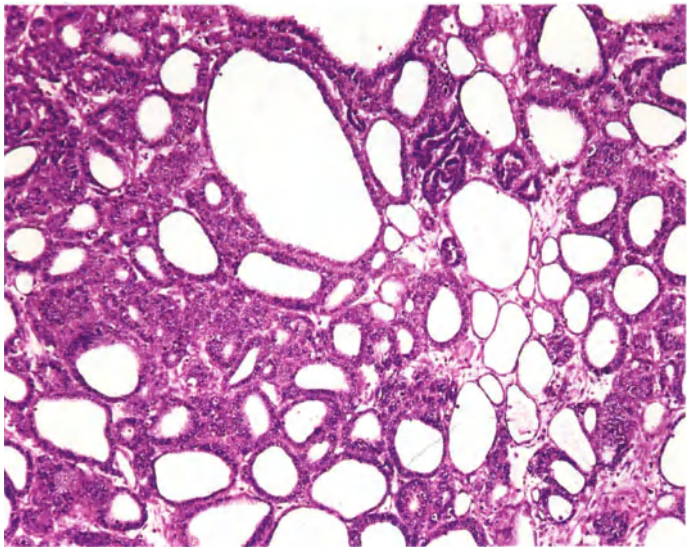

Fig. 55. *Nephroblastoma.* Numerous cysts in a predominantly epithelial field

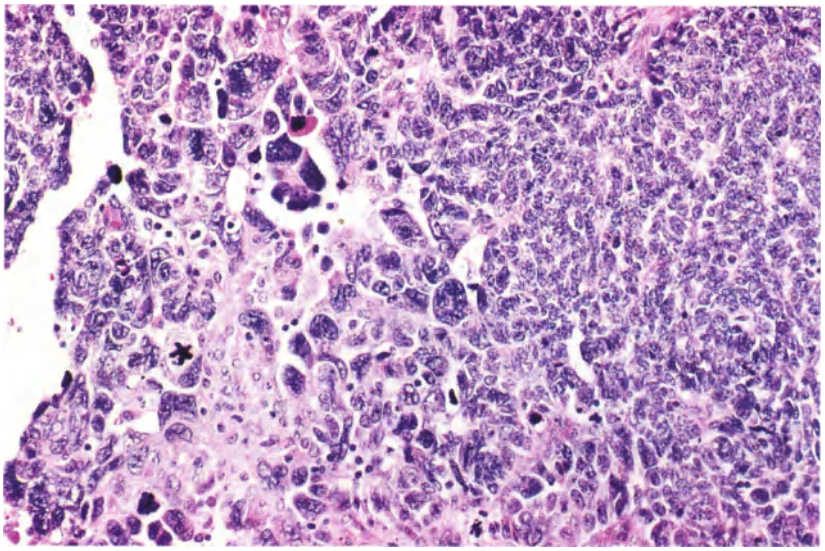

Fig. 56. *Nephroblastoma.* Anaplasia

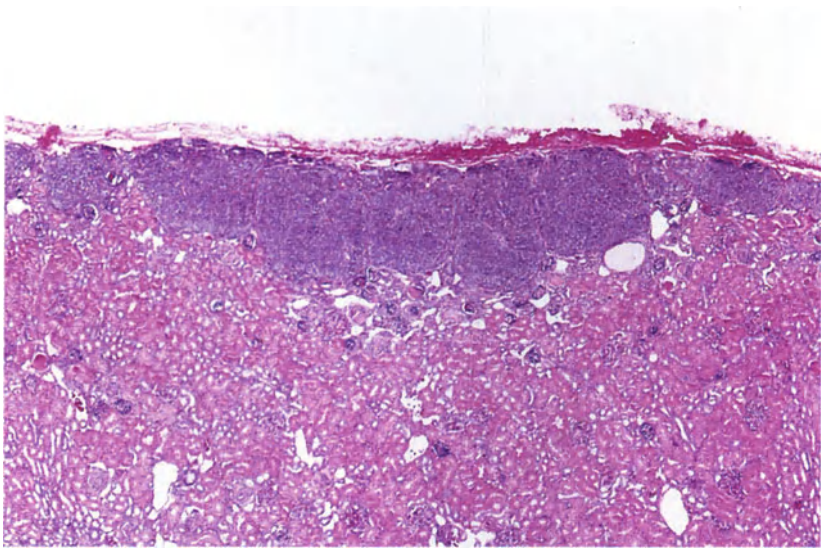

Fig. 57. *Perilobar nephrogenic rest.* Blastemal type. This field shows diffuse hyperplasia

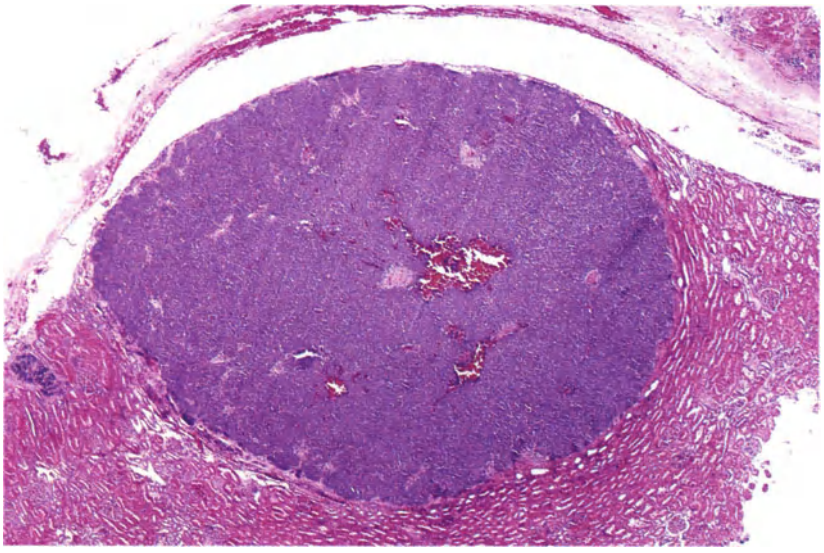

Fig. 58. *Perilobar nephrogenic rest.* Hyperplastic

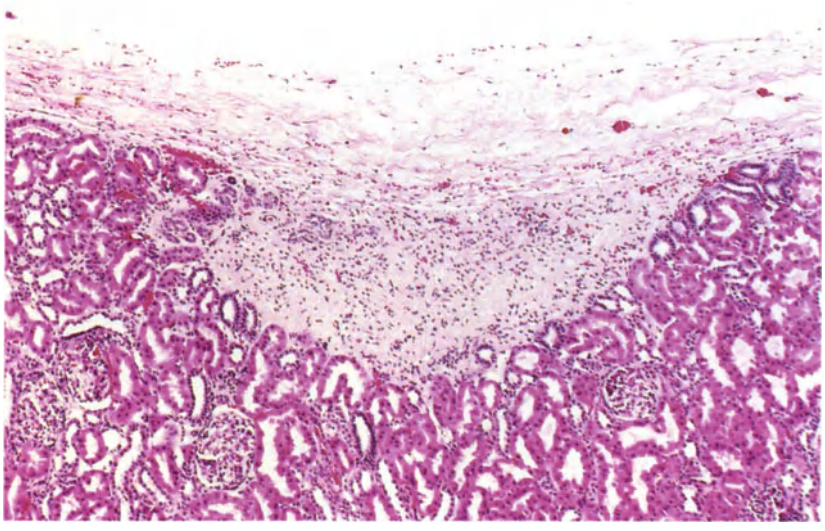

Fig. 59. *Perilobar nephrogenic rest.* Obsolescent

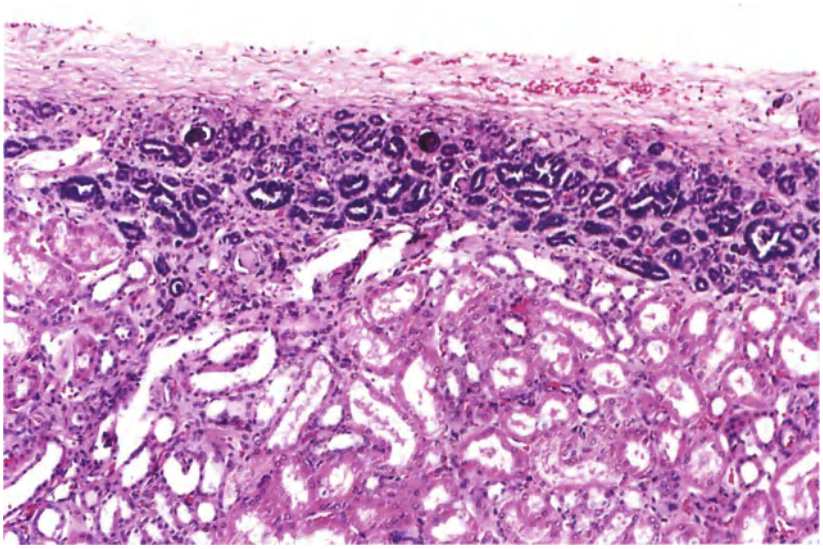

Fig. 60. *Perilobar nephrogenic rest.* Sclerosing type

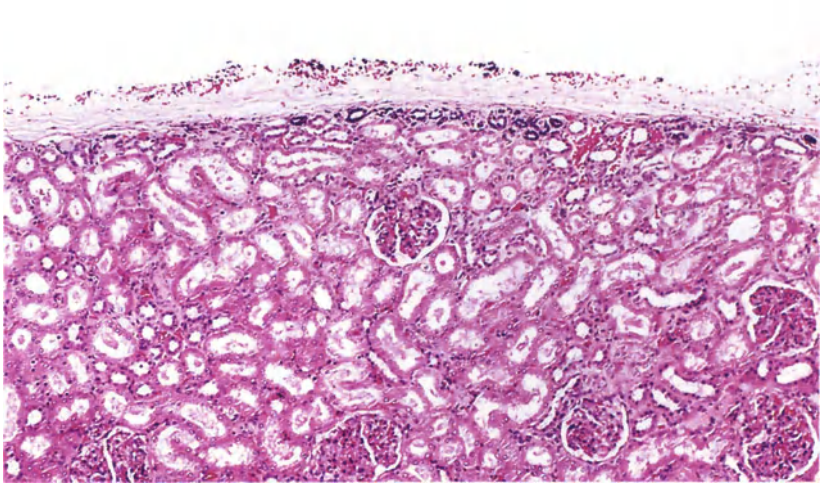

Fig. 61. *Perilobar nephrogenic rest.* This is a subtle example of the sclerosing type

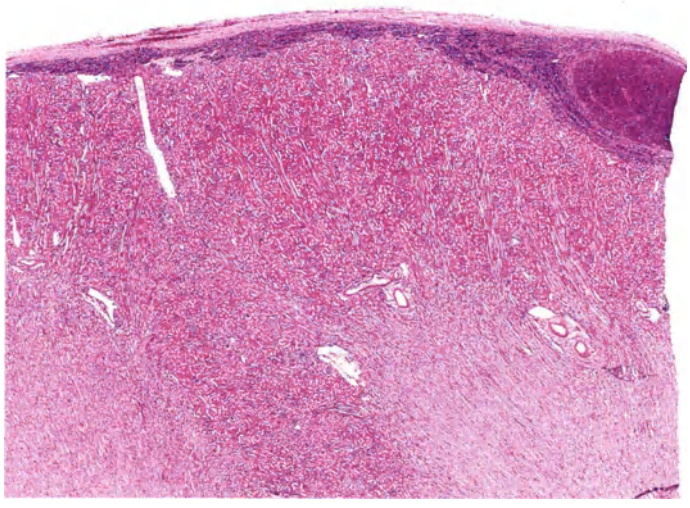

Fig. 62. *Perilobar nephrogenic rest.* Blastemal and diffuse sclerosing types

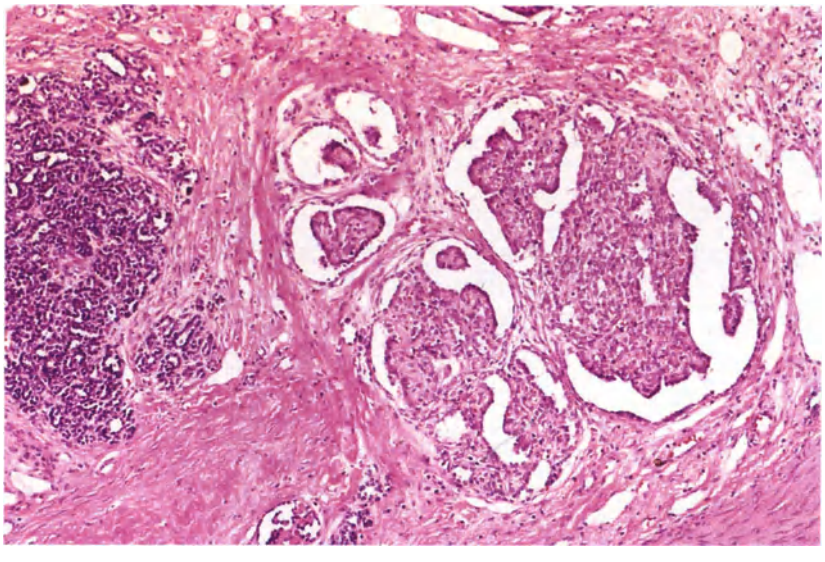

Fig. 63. *Perilobar nephrogenic rest.* Sclerosing and adenomatous types

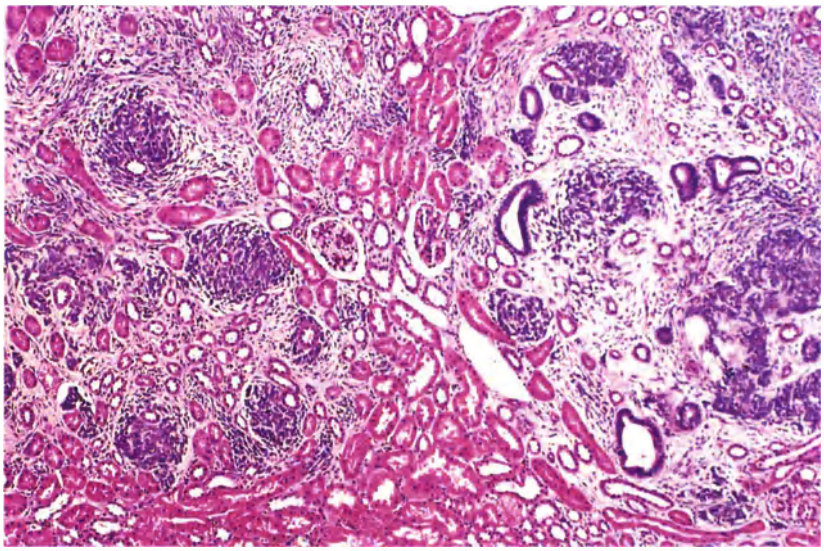

Fig. 64. *Intralobar nephrogenic rest.* Hyperplastic. Brightly staining normal nephrons admixed with the lesional tissue

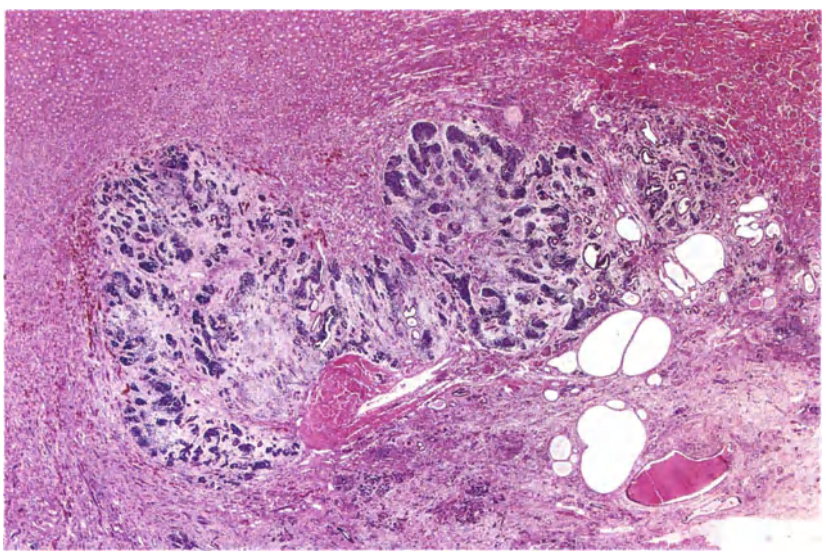

Fig. 65. *Intralobar nephrogenic rest.* Hyperplastic. This example includes a cystic component

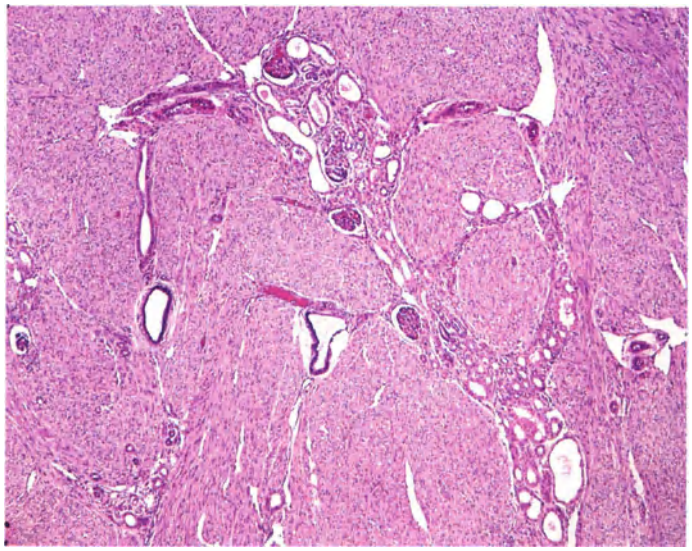

Fig. 66. *Mesoblastic nephroma.* Classic pattern

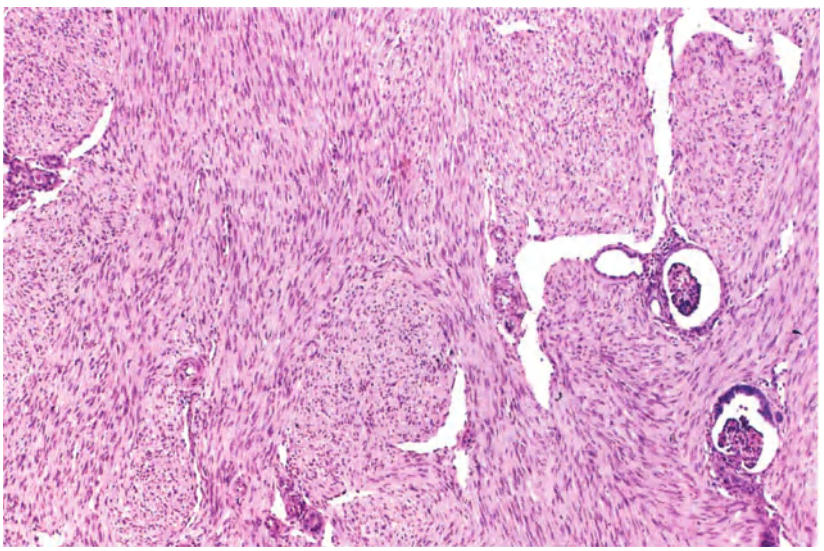

Fig. 67. *Mesoblastic nephroma.* Classic pattern. Embryonal metaplasia of Bowman's capsule (lower right)

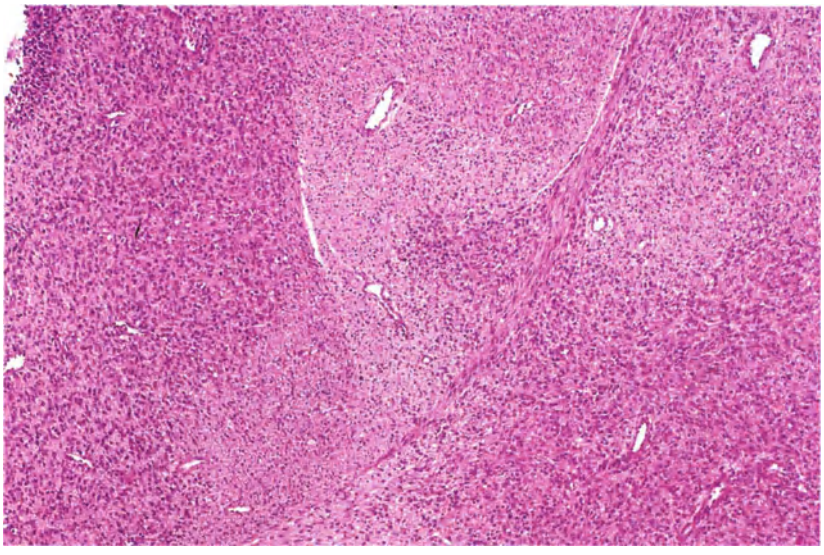

Fig. 68. *Mesoblastic nephroma.* Classic and cellular patterns

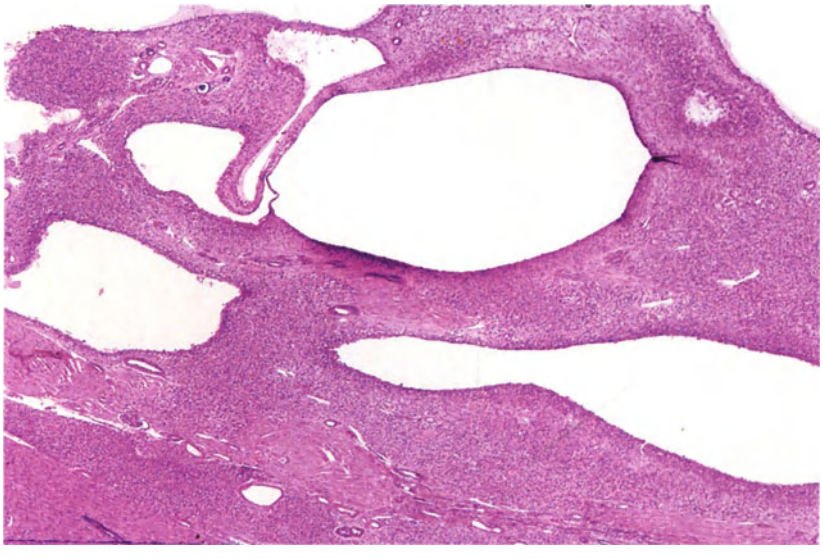

Fig. 69. *Mesoblastic nephroma.* Cellular pattern with extensive cystic change

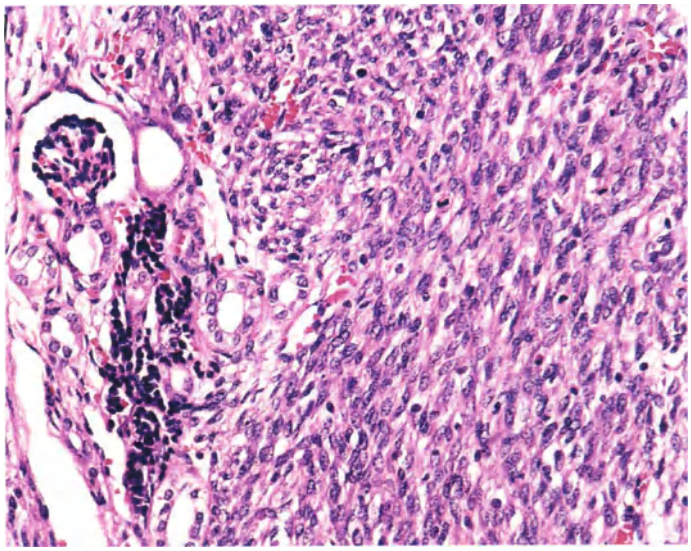

Fig. 70. *Mesoblastic nephroma.* Cellular pattern

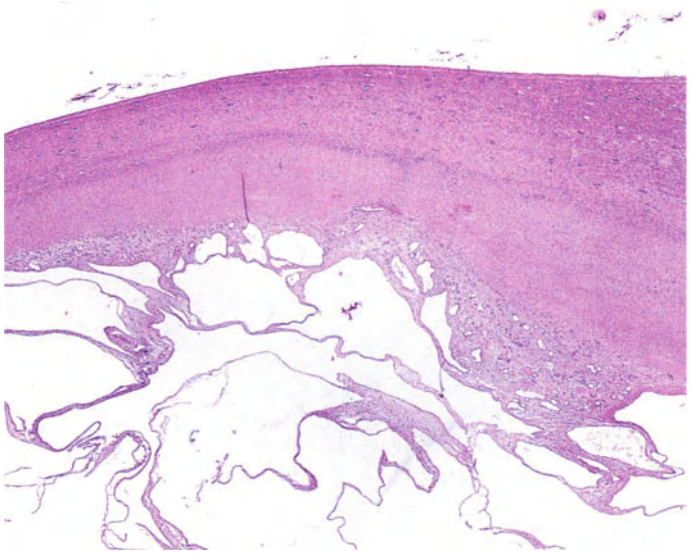

Fig. 71. *Benign cystic nephroma.* Multiple cysts with variable amounts of stroma surrounded by a thick capsule. Renal tissue is compressed

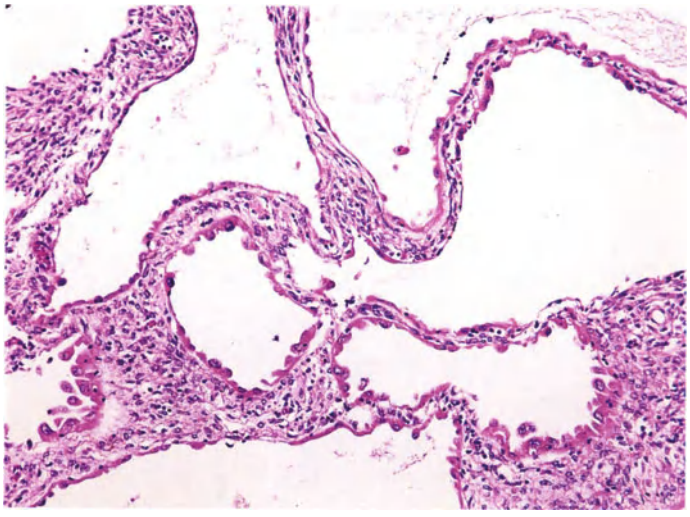

Fig. 72. *Benign cystic nephroma.* Characteristic hob-nail epithelium

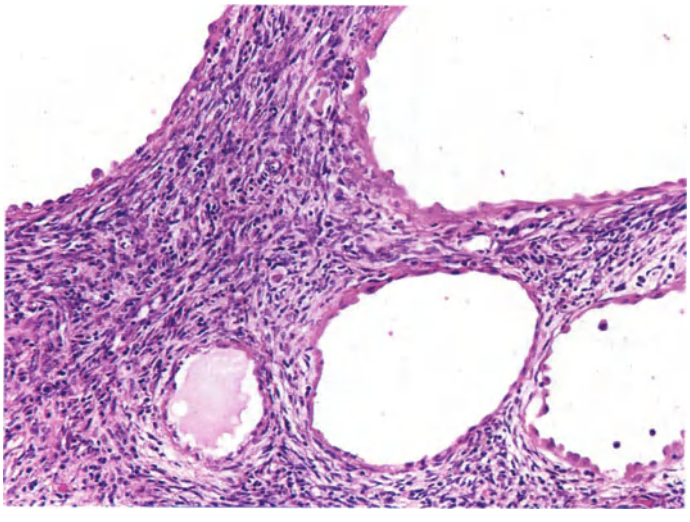

Fig. 73. *Benign cystic nephroma.* Highly cellular fibrous stroma

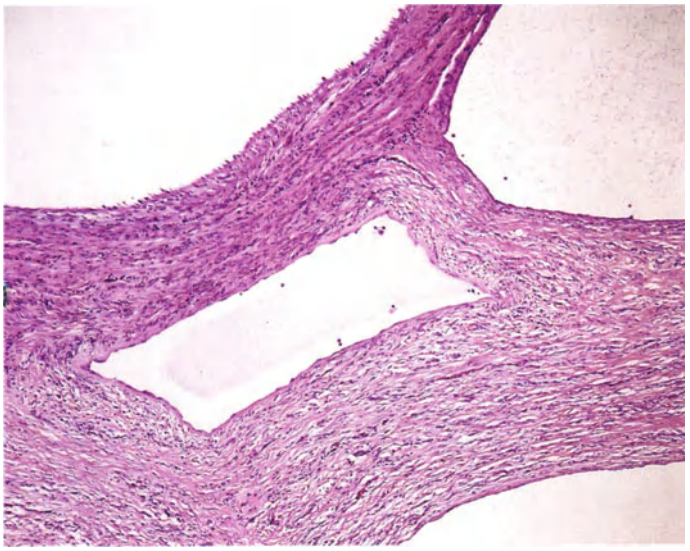

Fig. 74. *Benign cystic nephroma.* The stroma is more collagenous and less cellular than seen in Fig. 73

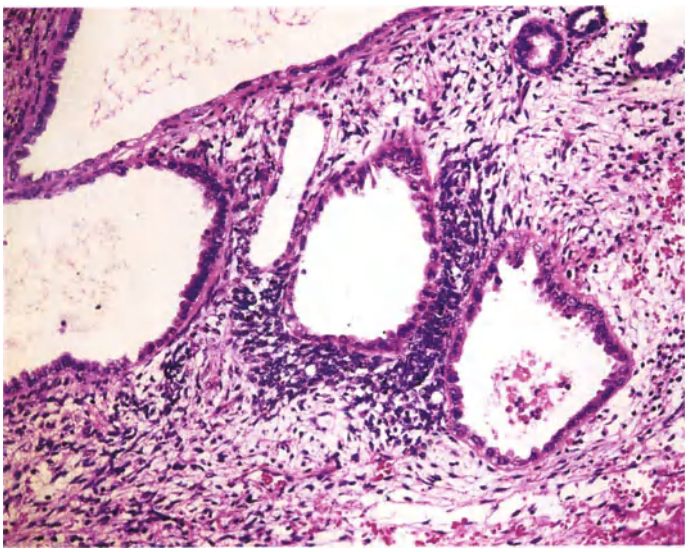

Fig. 75. *Cystic, partially differentiated nephroblastoma.* The tumour resembles the benign cystic nephroma but has foci of blastemal nephroblastoma

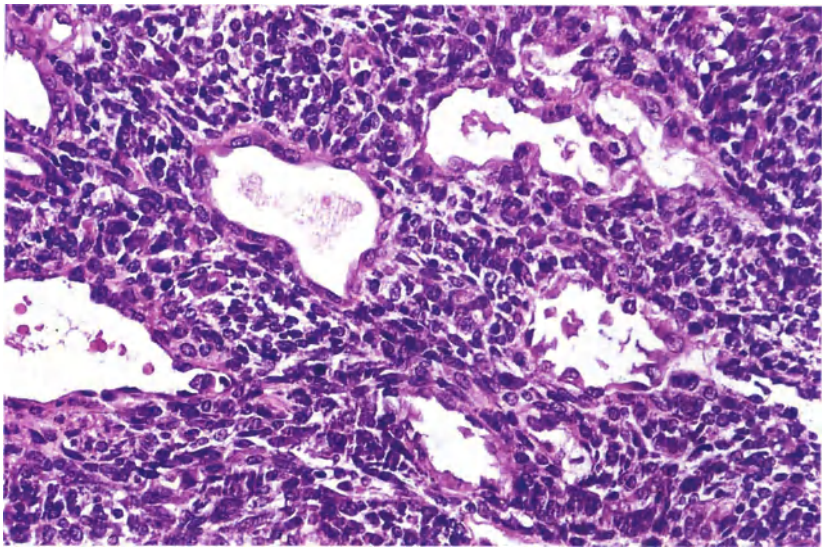

Fig. 76. *Malignant cystic nephroma.* The epithelium is similar to that of the benign cystic nephroma but the stroma is sarcomatous

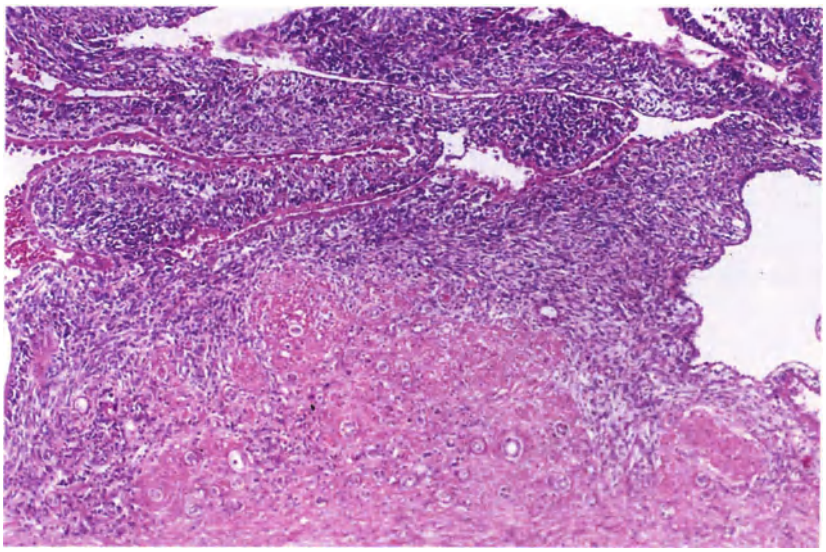

Fig. 77. *Malignant cystic nephroma.* The larger locules are collapsed and the stromal element invades through the tumour capsule (below)

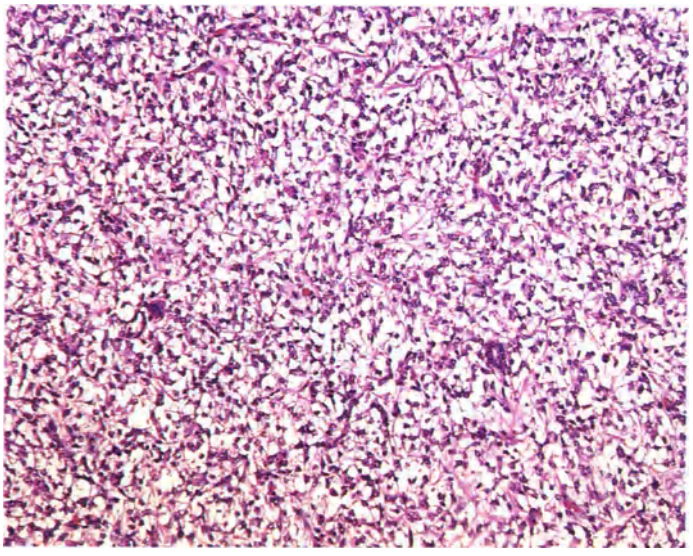

Fig. 78. *Clear cell sarcoma.* This is the classical pattern showing vacuolated tumour cells and delicate vascular channels. Several entrapped tubules are also shown

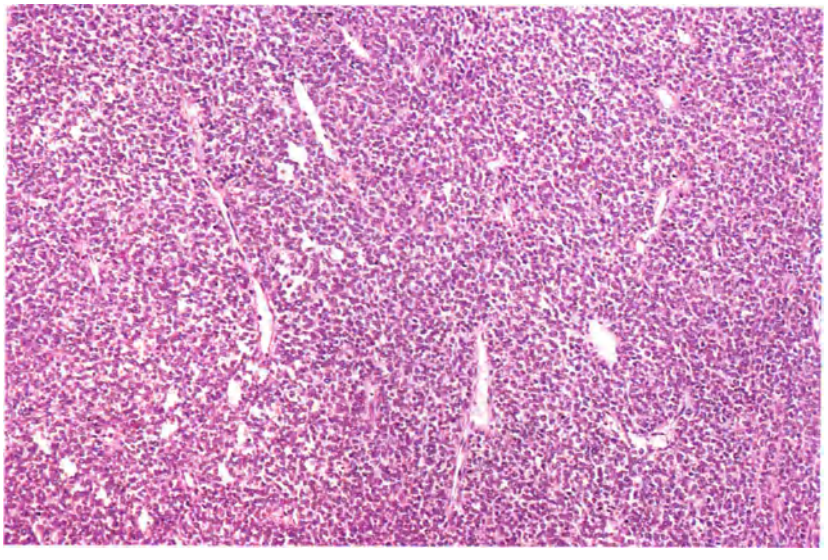

Fig. 79. *Clear cell sarcoma.* In this example, cellular vacuolisation is minimal

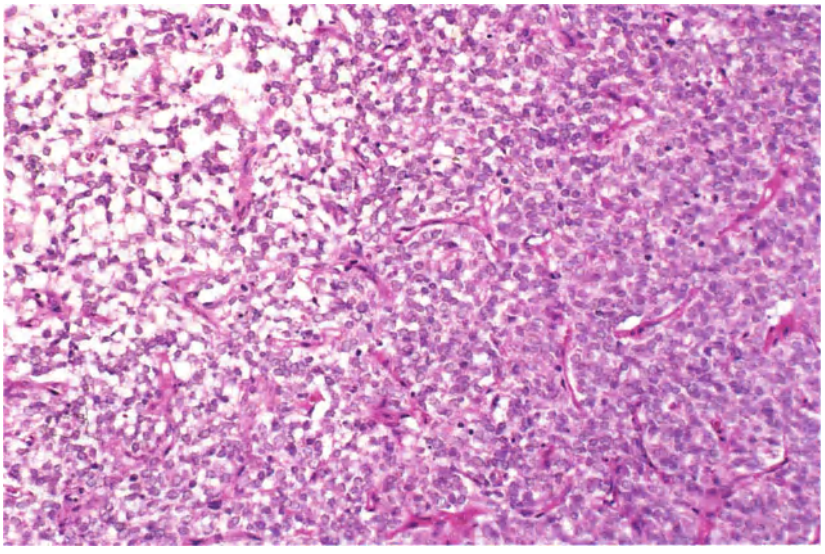

Fig. 80. *Clear cell sarcoma.* Vacuolated and nonvacuolated cells

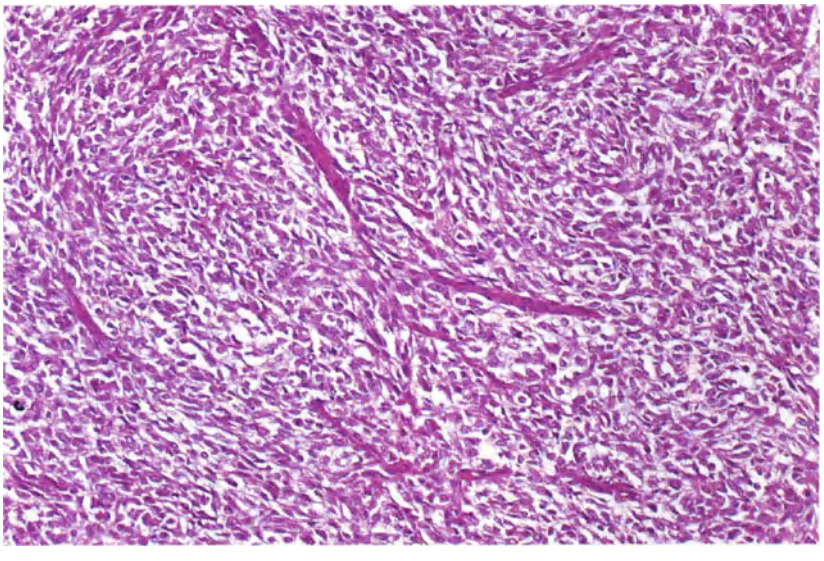

Fig. 81. *Clear cell sarcoma.* Slight spindling of cord cells

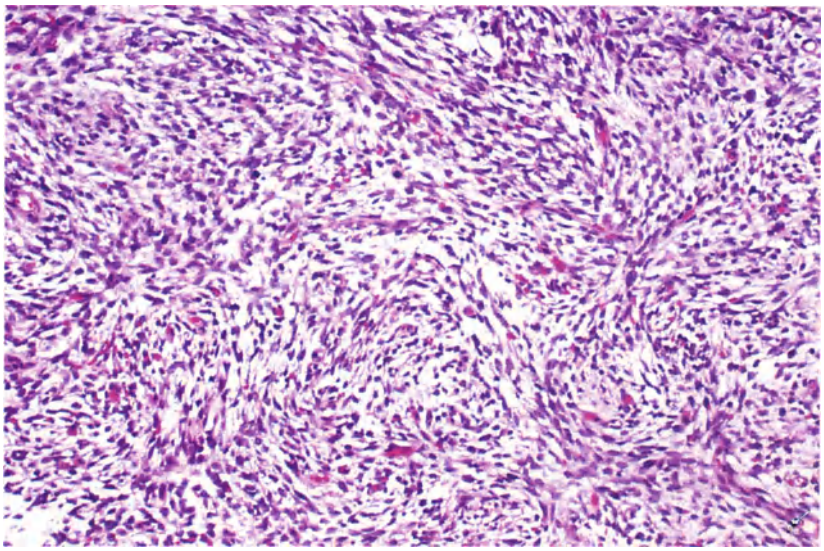

Fig. 82. *Clear cell sarcoma.* Spindled pattern. Spindling of cord cells in this field imparts a storiform appearance

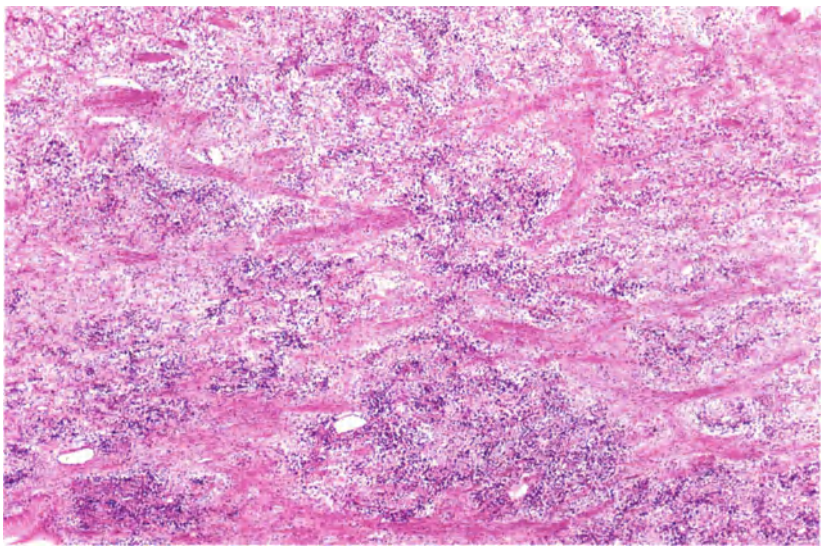

Fig. 83. *Clear cell sarcoma.* Spindled pattern. There is proliferation of spindle cells associated with vessels (septal cells)

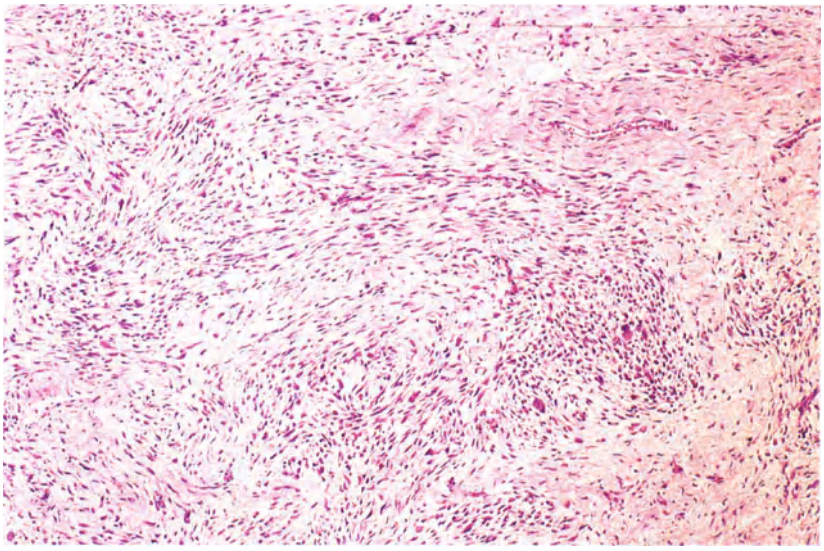

Fig. 84. *Clear cell sarcoma.* There is spindling of both cord cells and septal cells with some pleomorphism

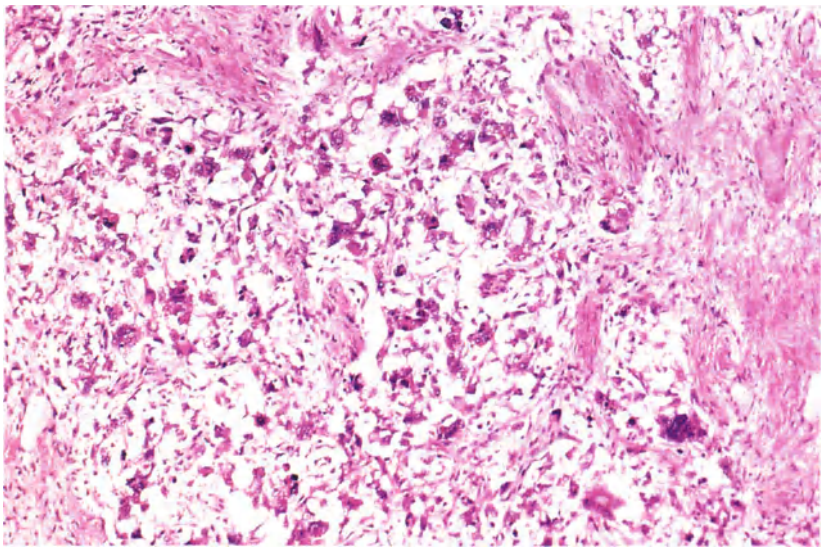

Fig. 85. *Clear cell sarcoma.* Pleomorphic pattern. Rarely, small foci with bizarre cells are present

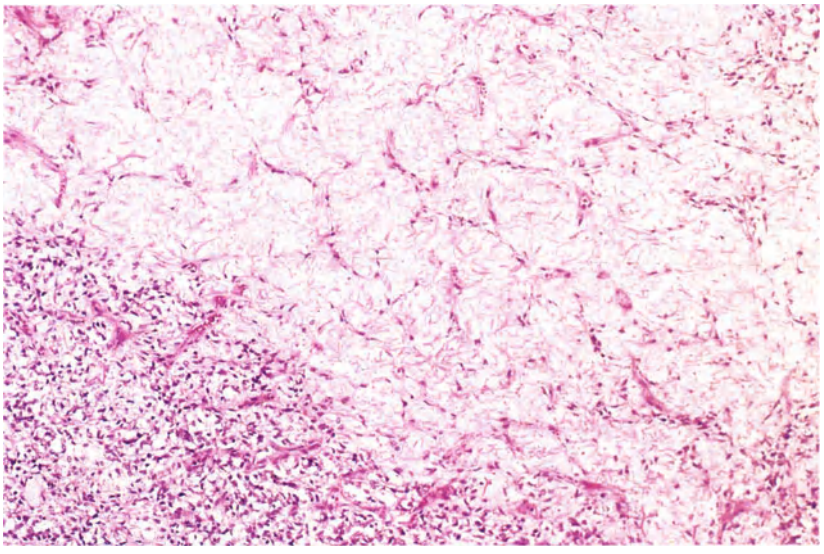

Fig. 86. *Clear cell sarcoma.* Sclerosing pattern. Cord cells have undergone sclerosis, leaving only the vascular element

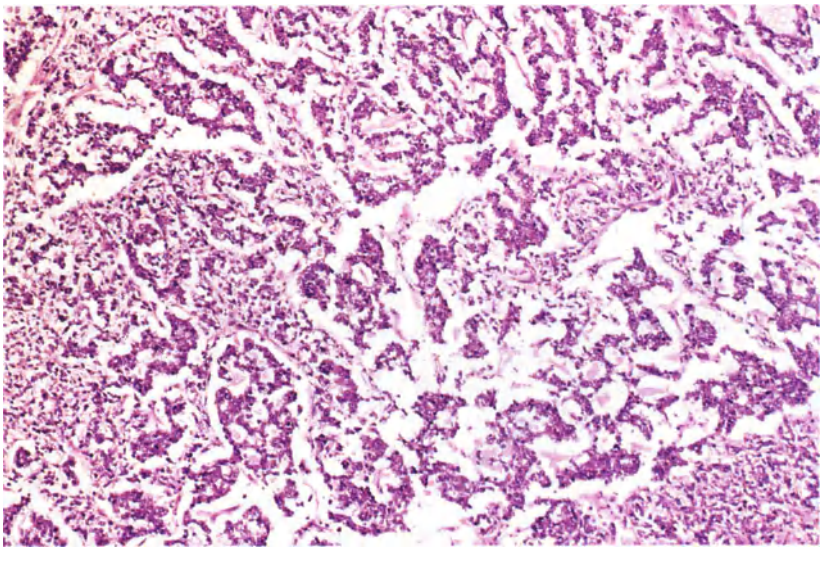

Fig. 87. *Clear cell sarcoma.* Epithelioid pattern. Varied cellular cohesion produces this appearance

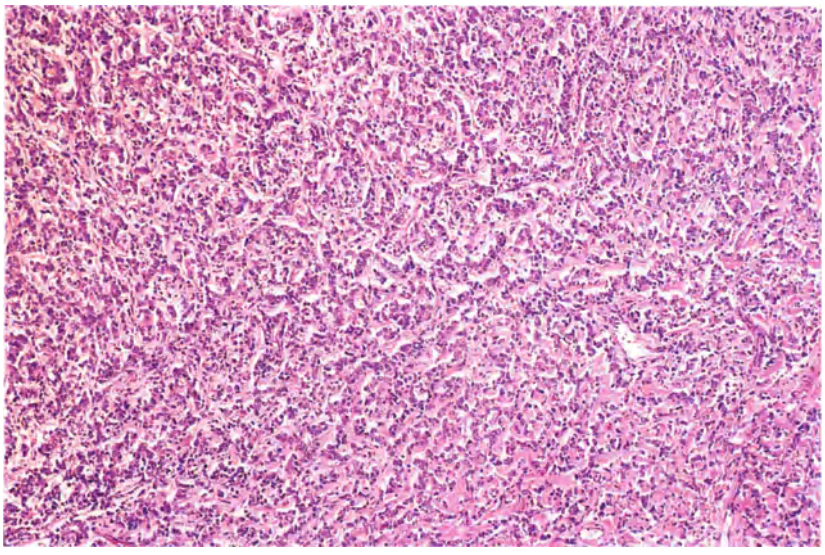

Fig. 88. *Clear cell sarcoma.* Epithelioid or filigree pattern

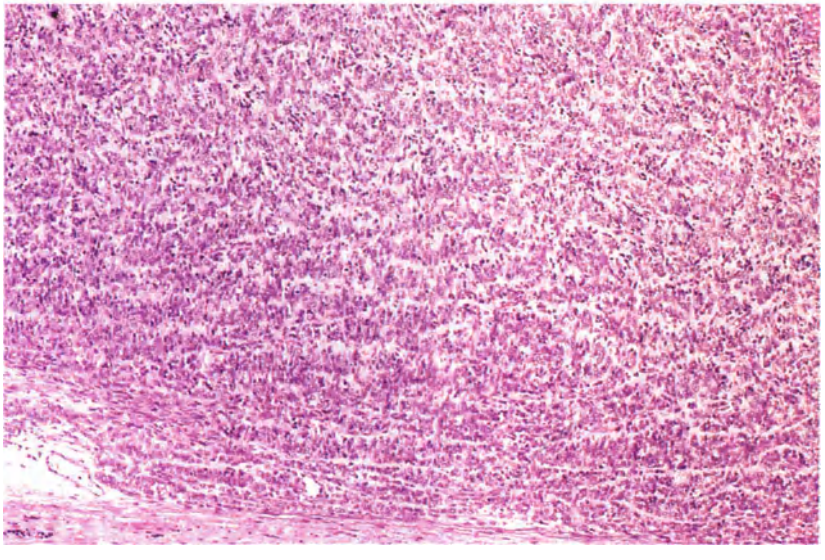

Fig. 89. *Clear cell sarcoma.* Palisading pattern

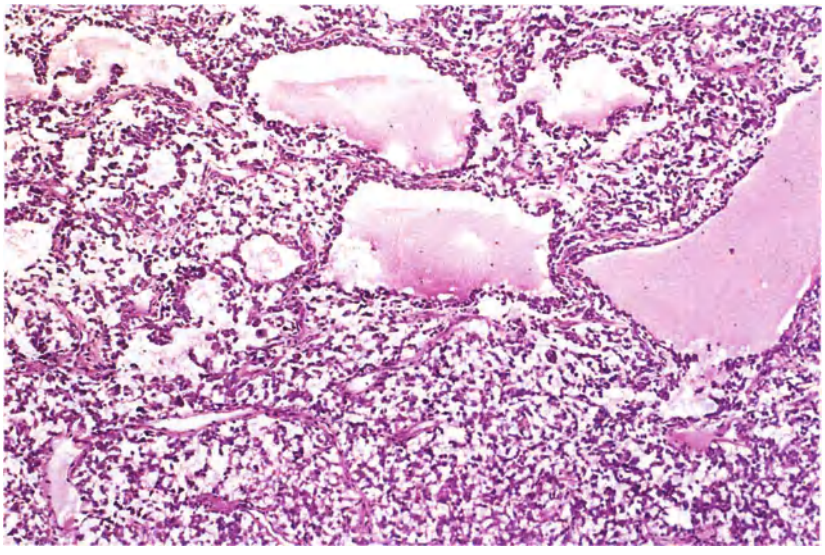

Fig. 90. *Clear cell sarcoma.* Cystic and myxoid pattern

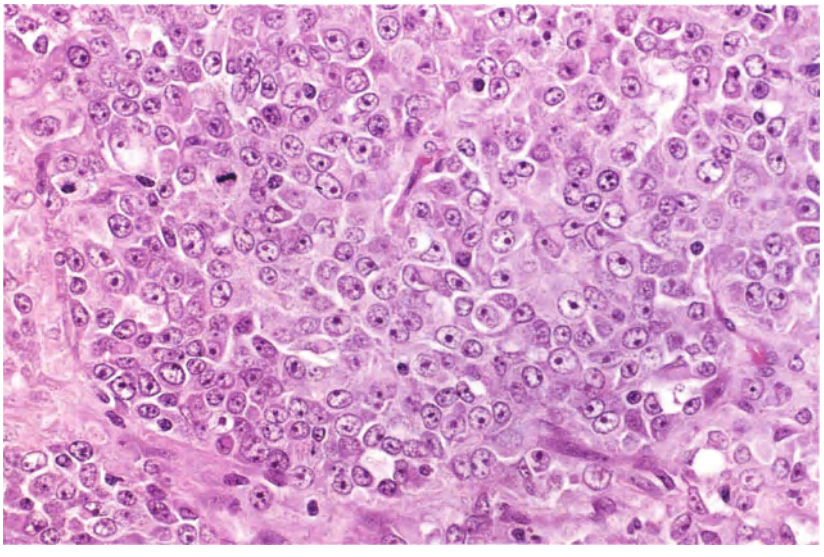

Fig. 91. *Rhabdoid tumour.* This typical field shows loosely cohesive cells and large nucleoli

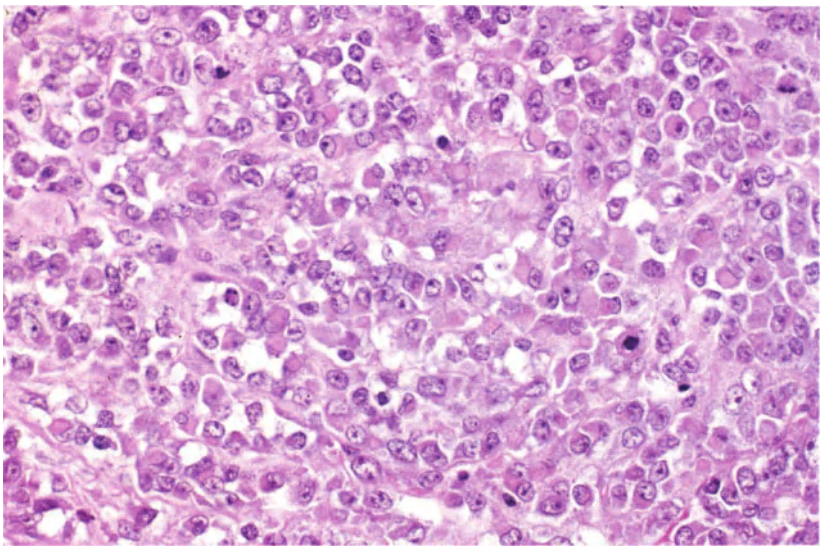

Fig. 92. *Rhabdoid tumour.* Discrete cytoplasmic inclusions are present in many of these cells

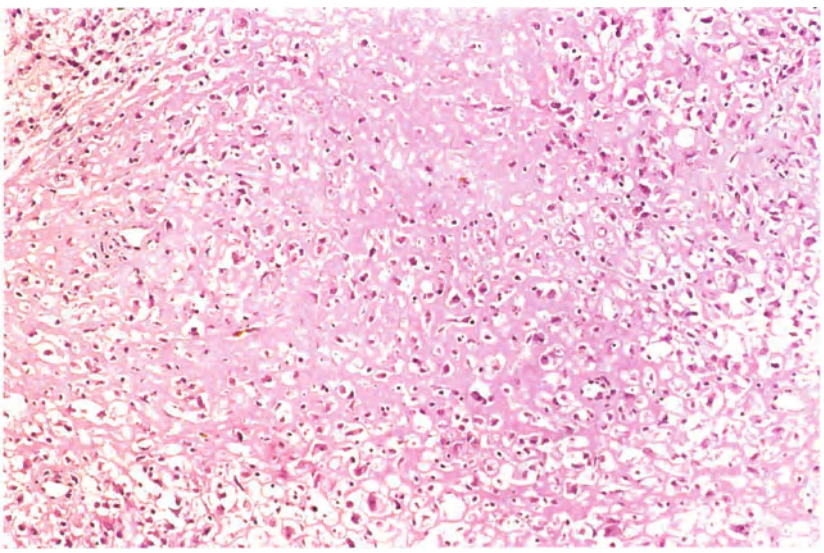

Fig. 93. *Rhabdoid tumour.* Sclerosing pattern. In this field the lesion has an osteosarcomatoid appearance

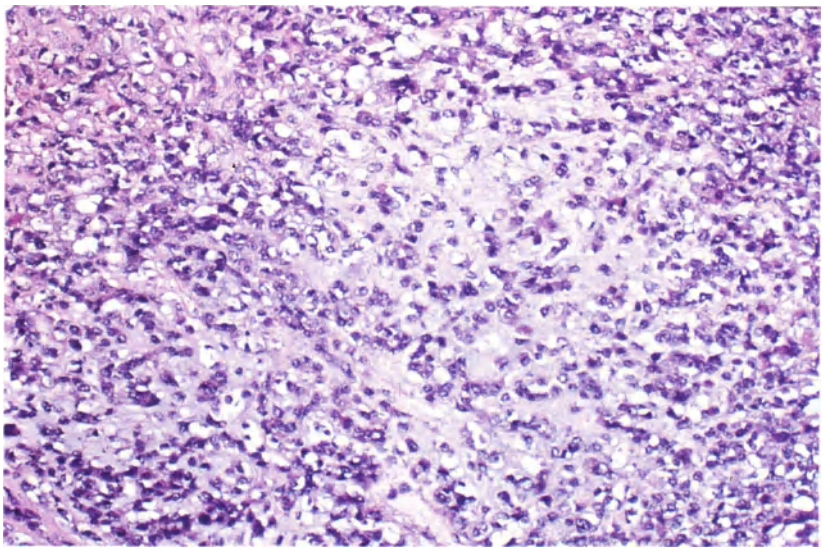

Fig. 94. *Rhabdoid tumour.* Sclerosing pattern. Basophilic ground substance may impart a chondroid appearance

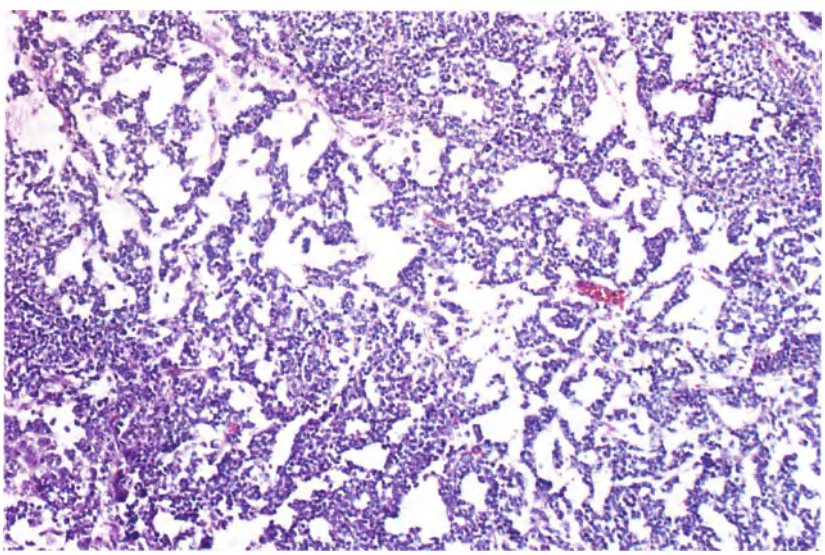

Fig. 95. *Rhabdoid tumour.* Epithelioid pattern

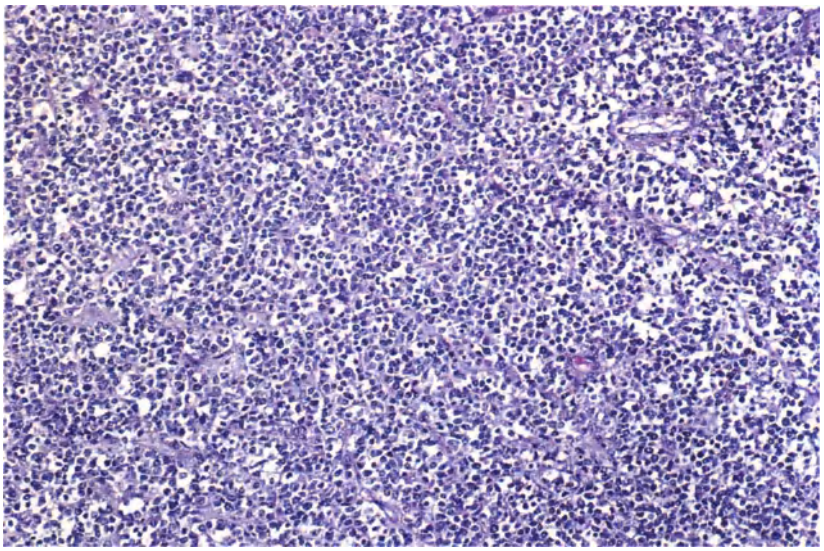

Fig. 96. *Rhabdoid tumour.* Lymphomatoid pattern

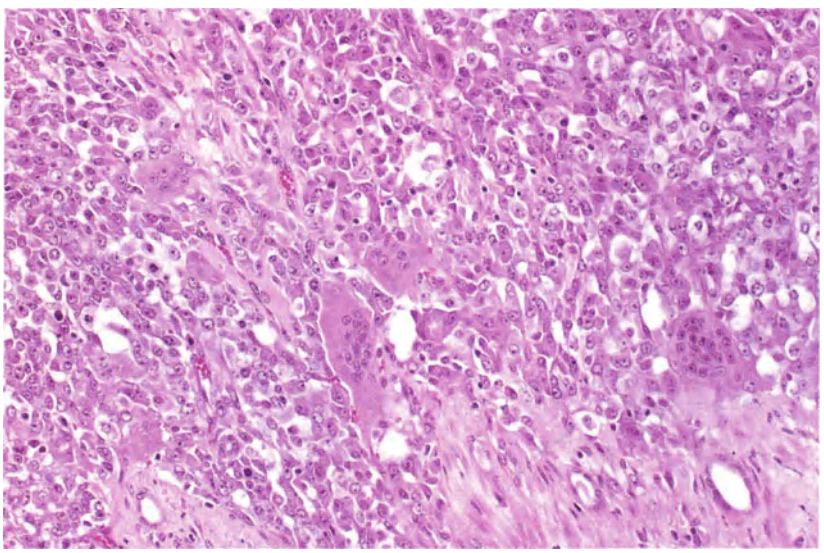

Fig. 97. *Rhabdoid tumour.* Focal histiocytoid pattern

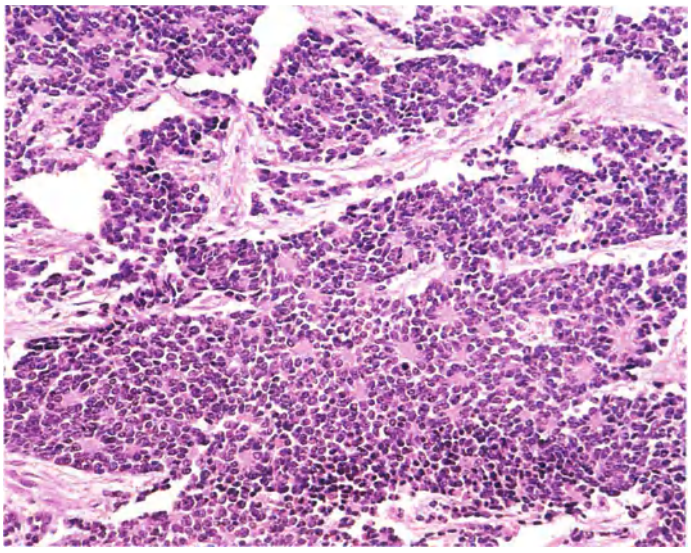

Fig. 98. *Neuroblastoma.* A primary tumour of the kidney

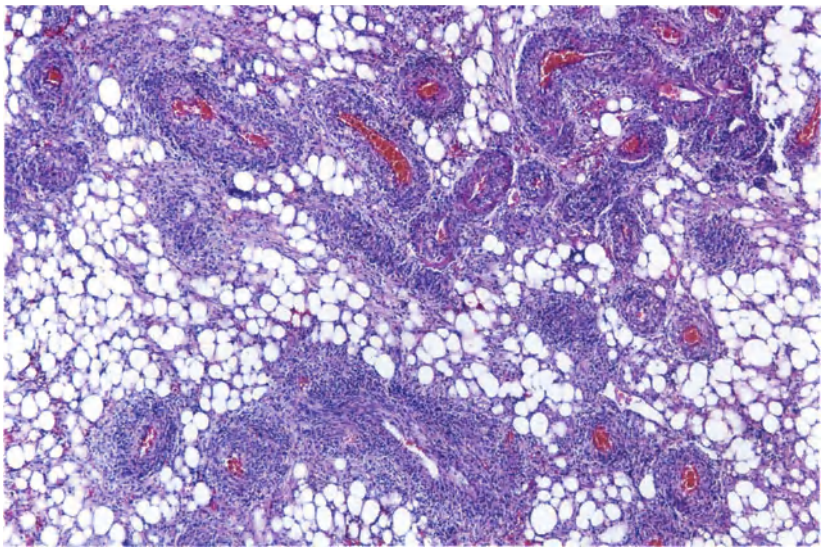

Fig. 99. *Angiomyolipoma*

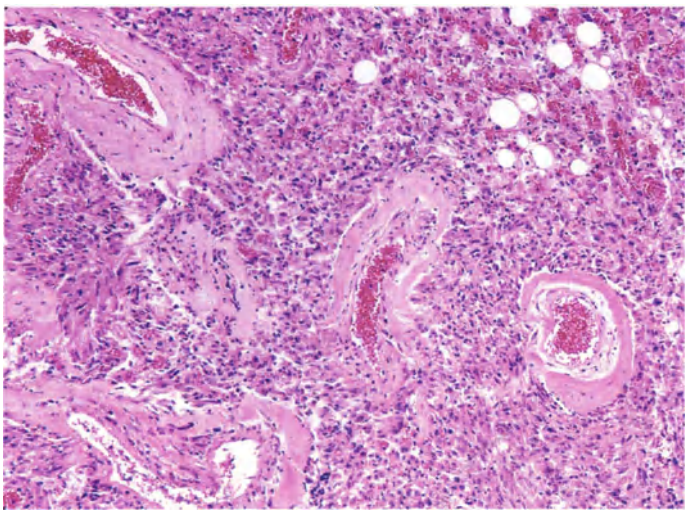

Fig. 100. *Angiomyolipoma.* Vascular sclerosis

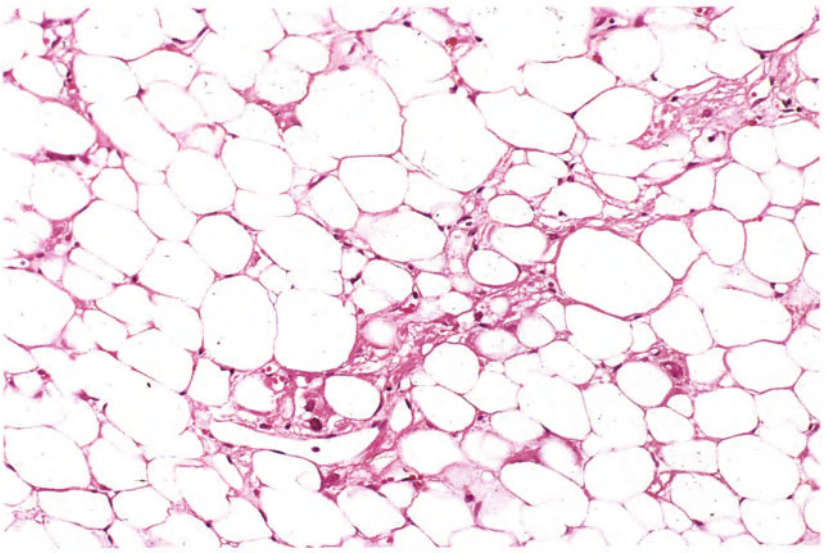

Fig. 101. *Angiomyolipoma.* A myomatous element is centered in this field of a tumour which is largely lipomatous

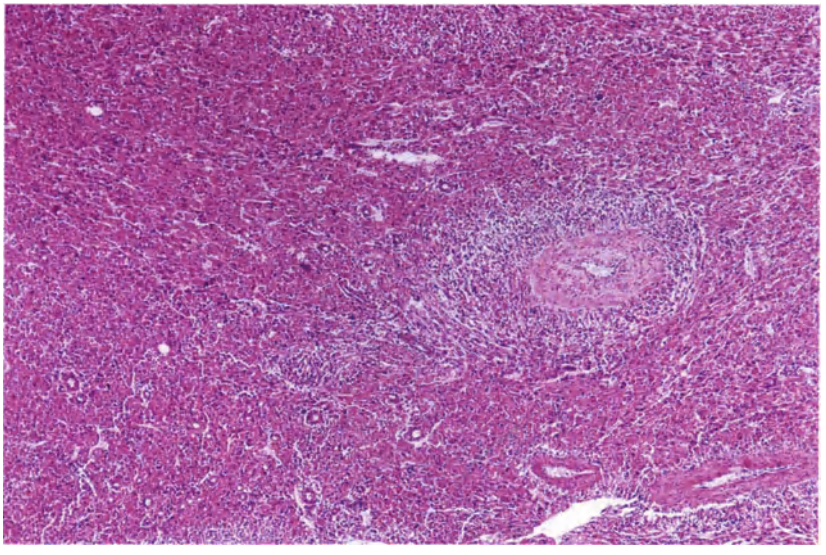

Fig. 102. *Angiomyolipoma.* Largely myomatous. Vascular collar (right)

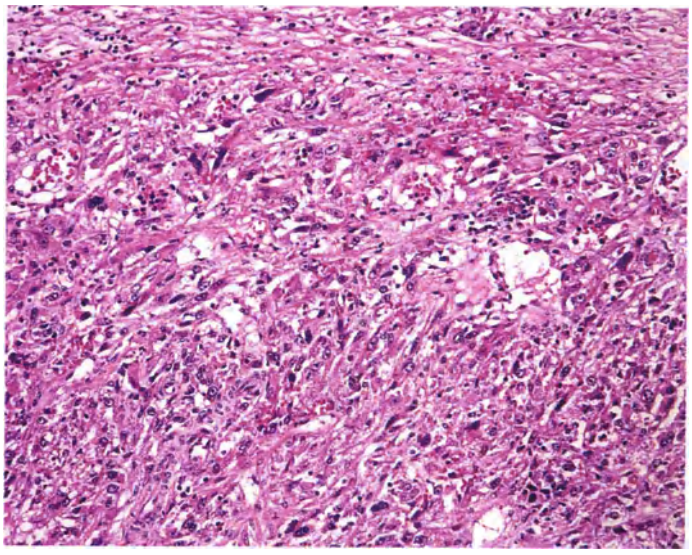

Fig. 103. *Angiomyolipoma.* Muscle cells usually show some degree of pleomorphism, at least focally

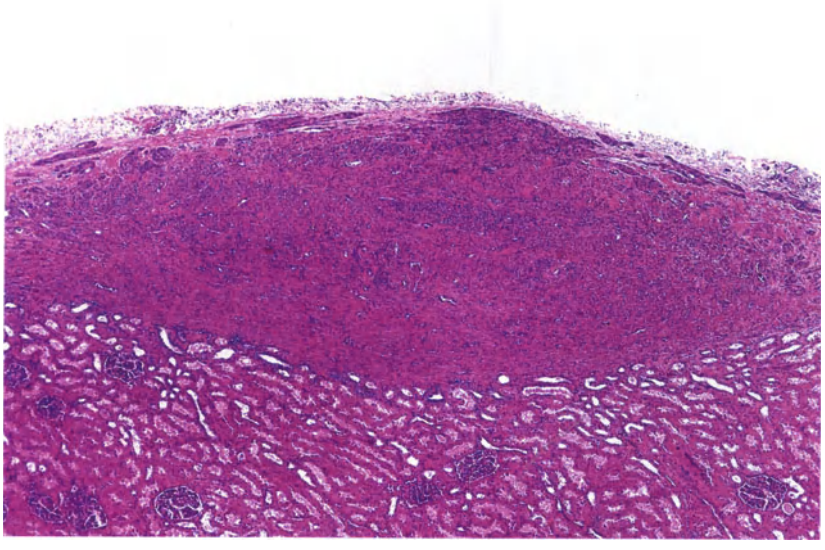

Fig. 104. *Capsular leiomyoma.* This was an incidental finding

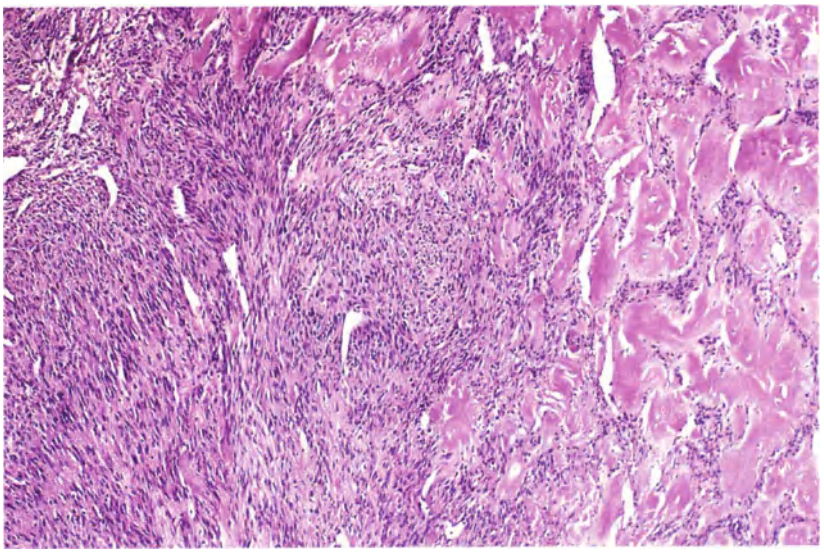

Fig. 105. *Capsular leiomyoma.* Focal sclerosis

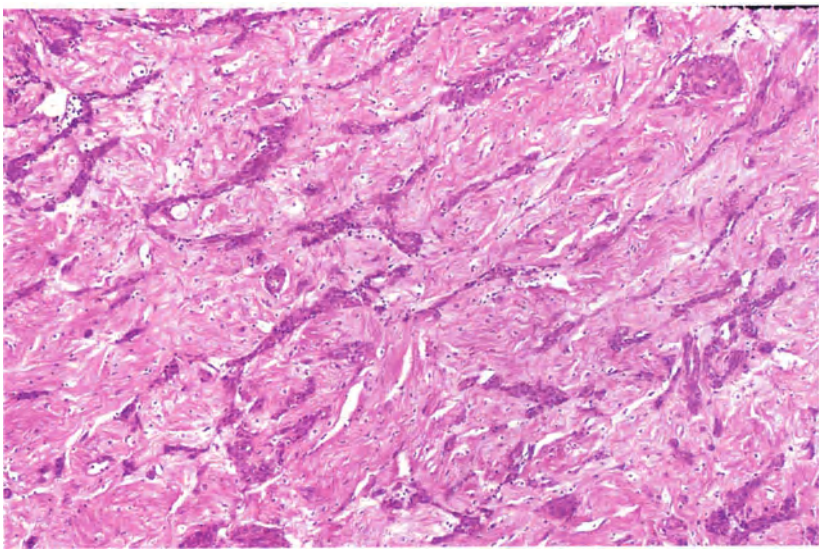

Fig. 106. *Capsular leiomyoma with extensive sclerosis.* The extent of hyalinisation varies markedly in these tumours

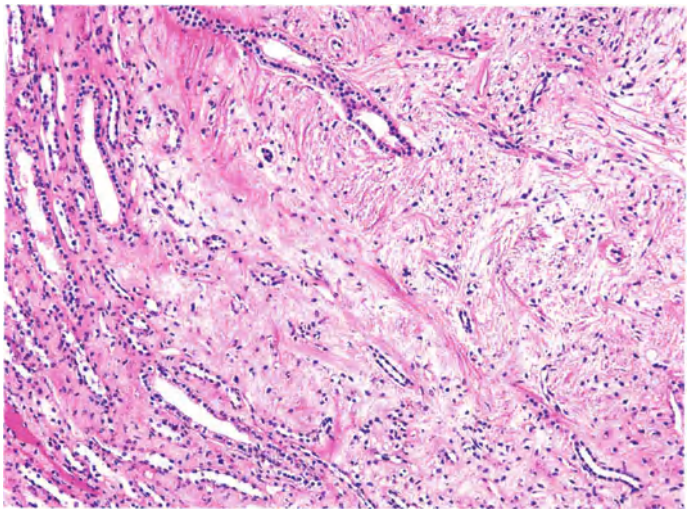

Fig. 107. *Renomedullary interstitial cell tumour*

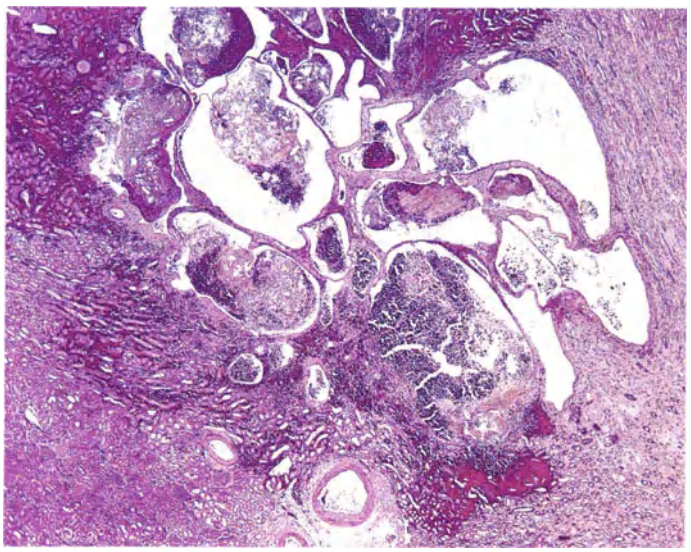

Fig. 108. *Cavernous haemangioma.* This was found in peripelvic soft tissue adjacent to the medulla

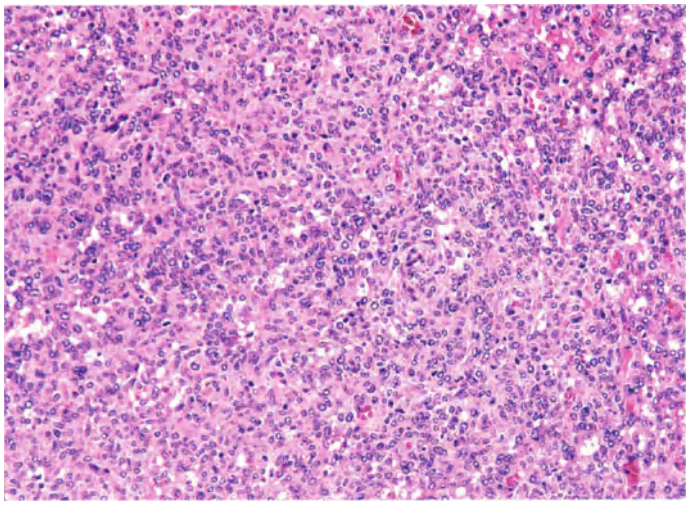

Fig. 109. *Juxtaglomerular cell tumour.* Some tumour cell aggregates have a small, central blood vessel

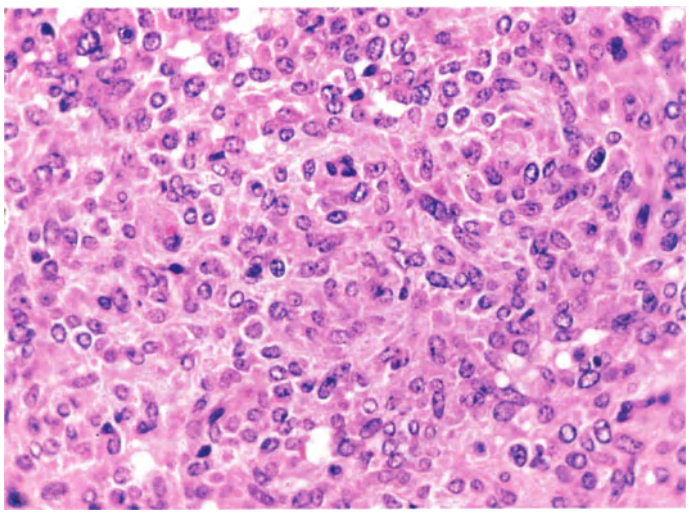

Fig. 110. *Juxtaglomerular cell tumour.* Same tumour as in Fig. 109

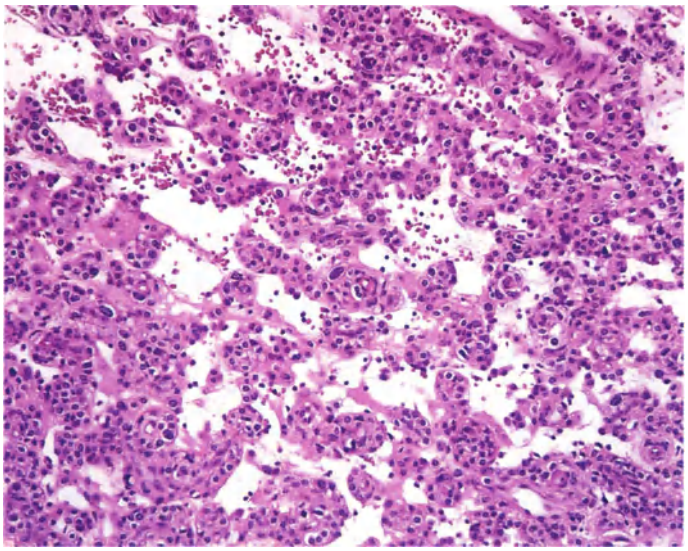

Fig. 111. *Juxtaglomerular cell tumour.* Many of these cell aggregates form the walls of small vessels

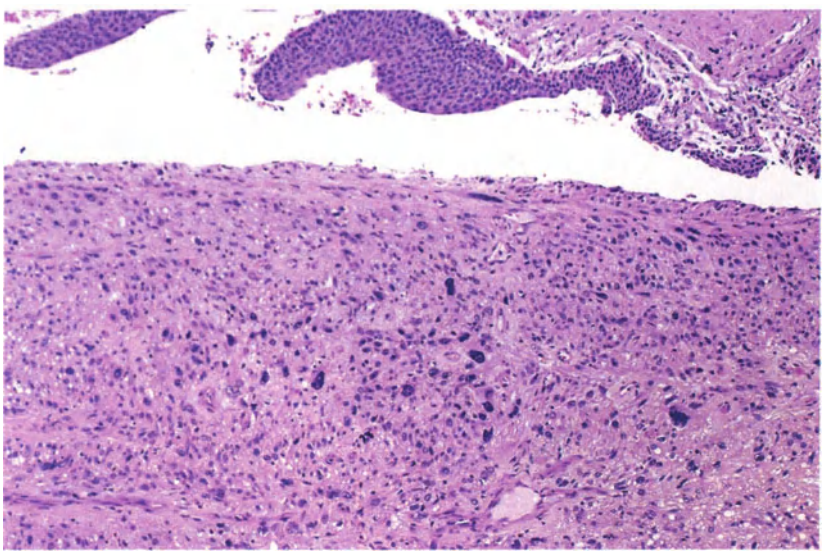

Fig. 112. *Renal leiomyosarcoma.* This high grade tumour protruded into the renal pelvis

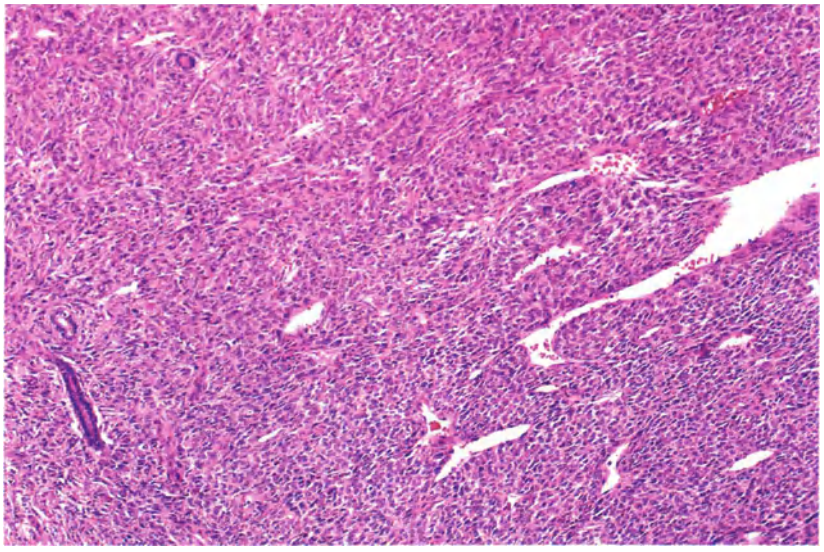

Fig. 113. *Renal haemangiopericytoma*

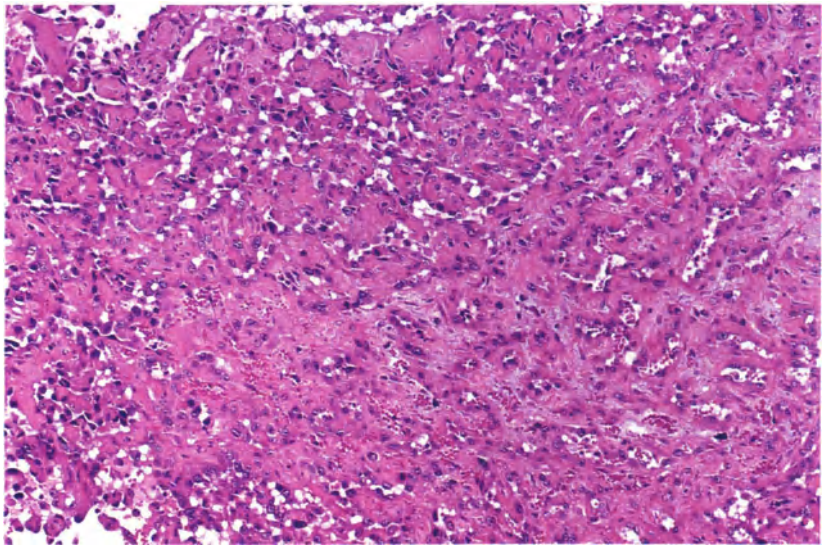

Fig. 114. *Renal angiosarcoma*

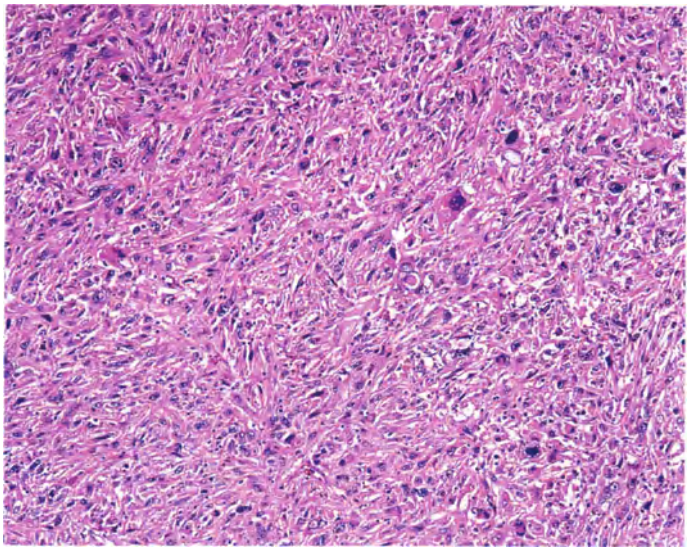

Fig. 115. *Malignant fibrous histiocytoma*

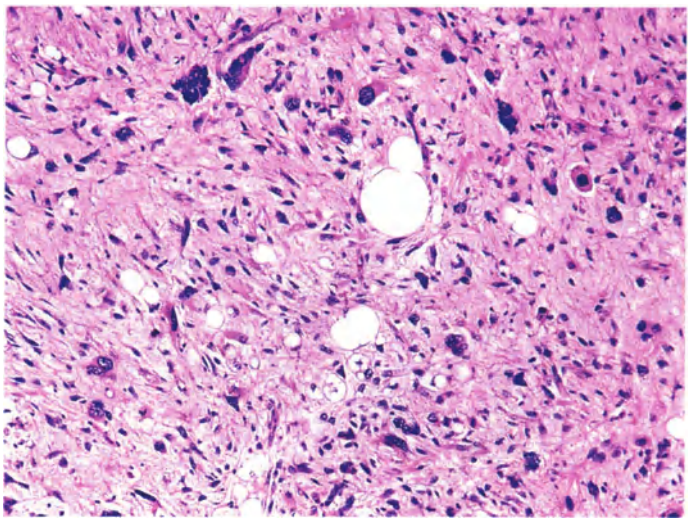

Fig. 116. *Liposarcoma*

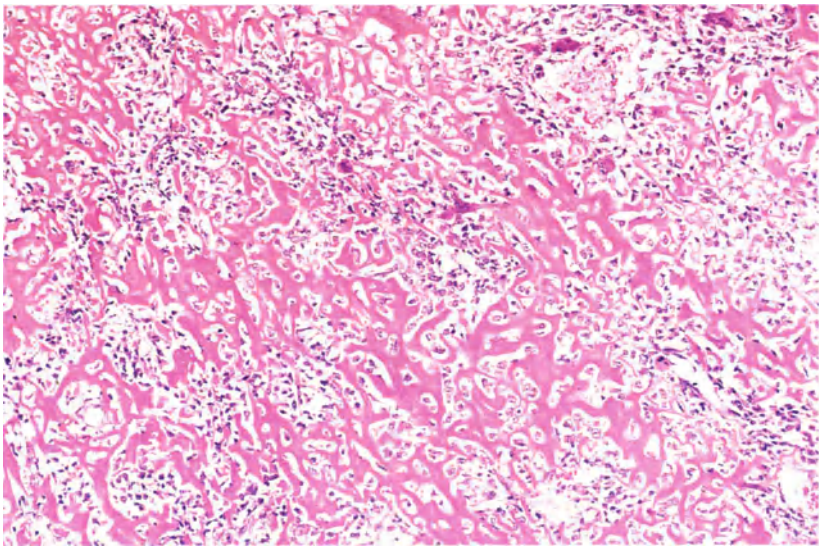

Fig. 117. *Renal osteosarcoma*

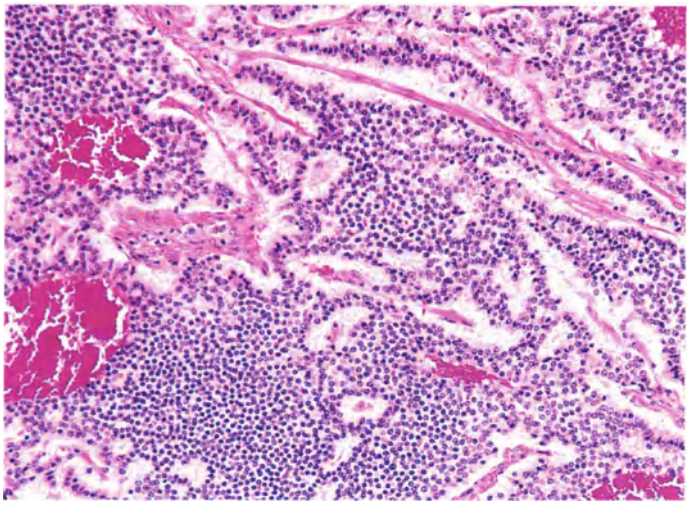

Fig. 118. *Renal carcinoid tumour*

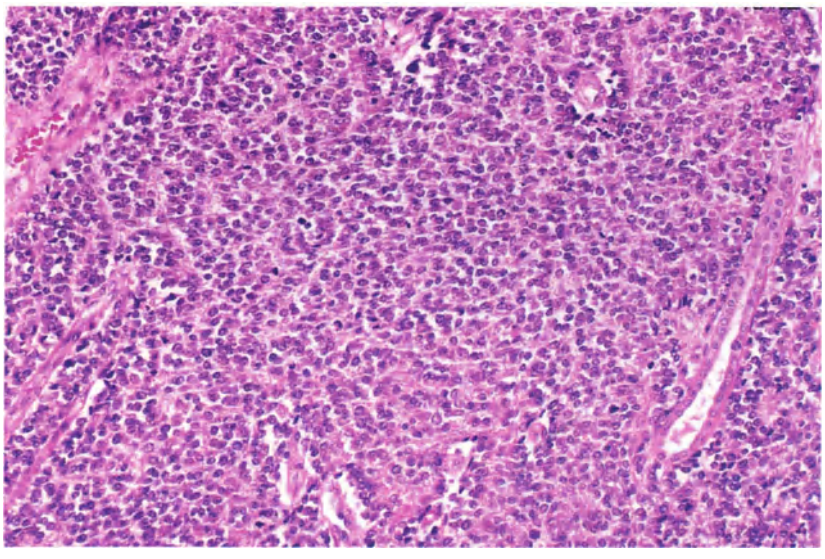

Fig. 119. *Primitive neuroectodermal tumour*

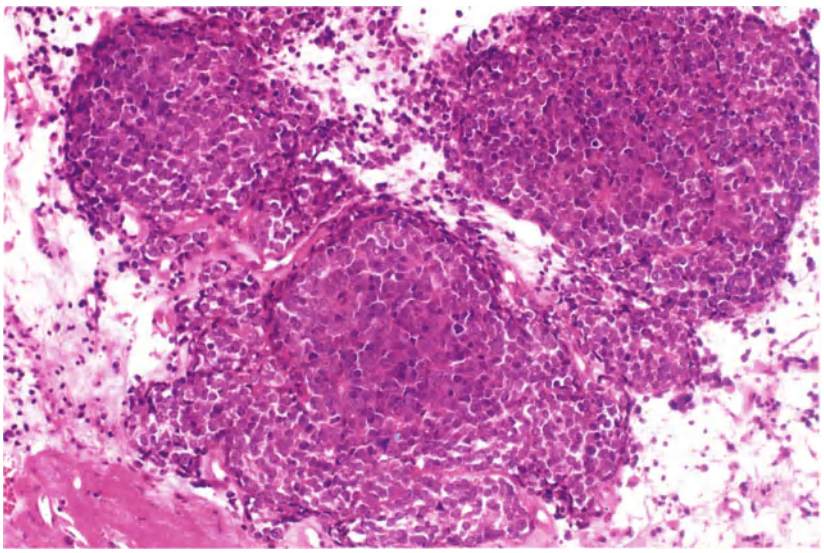

Fig. 120. *Primitive neuroectodermal tumour.* Several poorly formed rosette-like structures are present

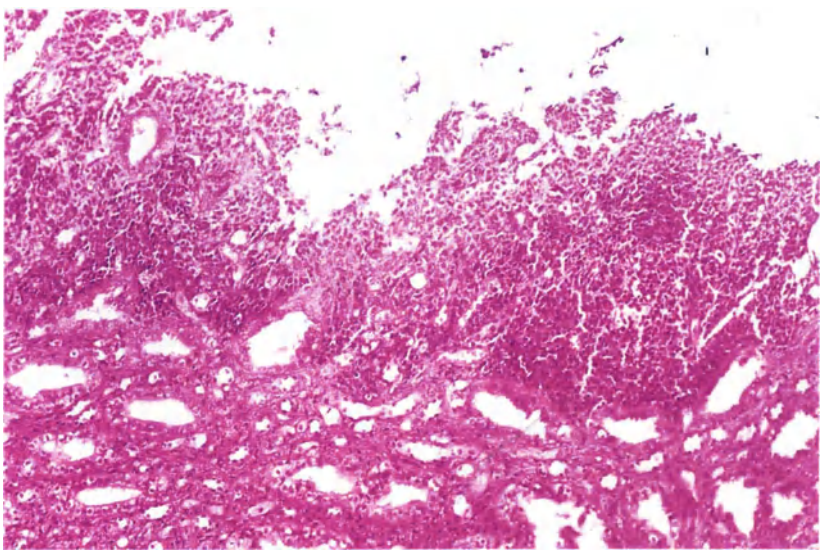

Fig. 121. *Ossifying renal tumour.* This illustrates attachment of the polypoid mass to a renal papilla

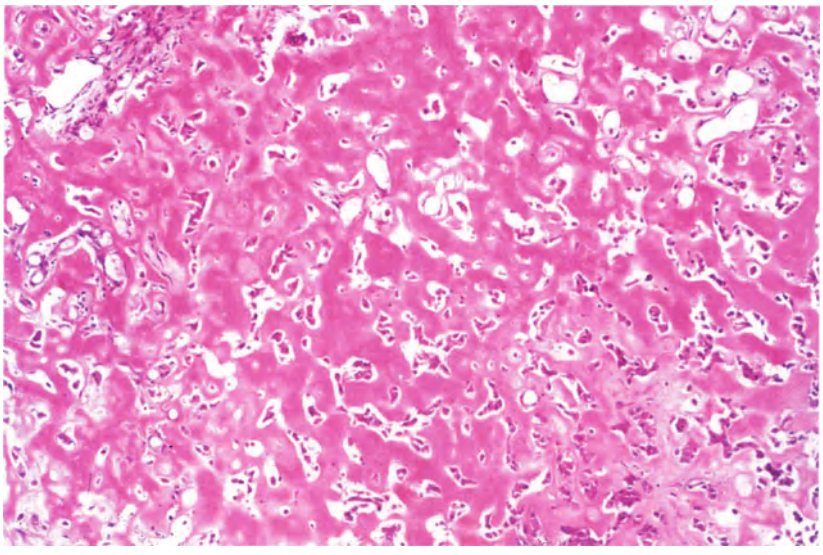

Fig. 122. *Ossifying renal tumour.* Osteoid forms the core of the tumour mass

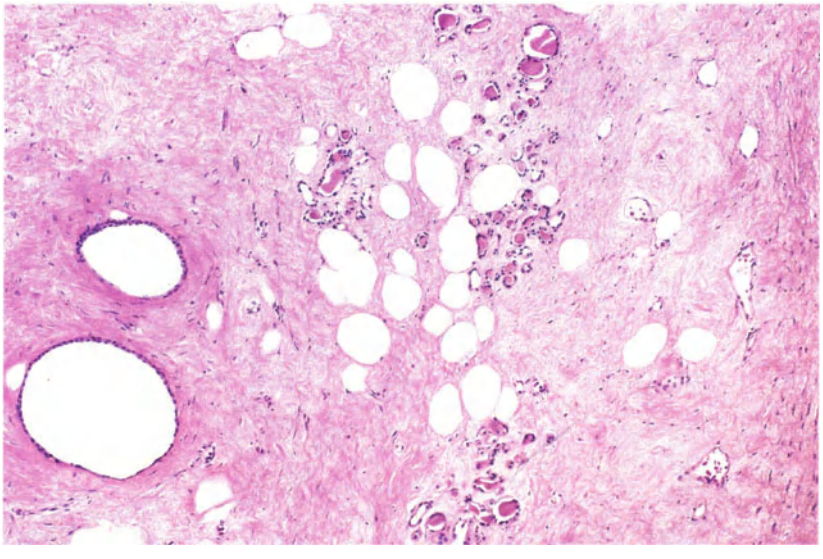

Fig. 123. *Renal cortical hamartoma.* This tumour consists largely of collagen, adipose tissue and adenomatous foci

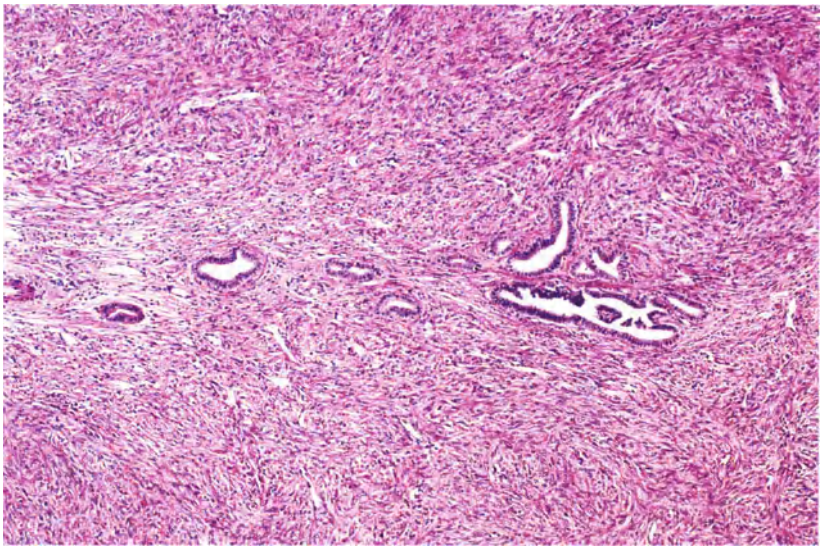

Fig. 124. *Renal cortical hamartoma.* Smooth muscle and adenoma-like areas. Some tumours combine the features of Figs. 123 and 124

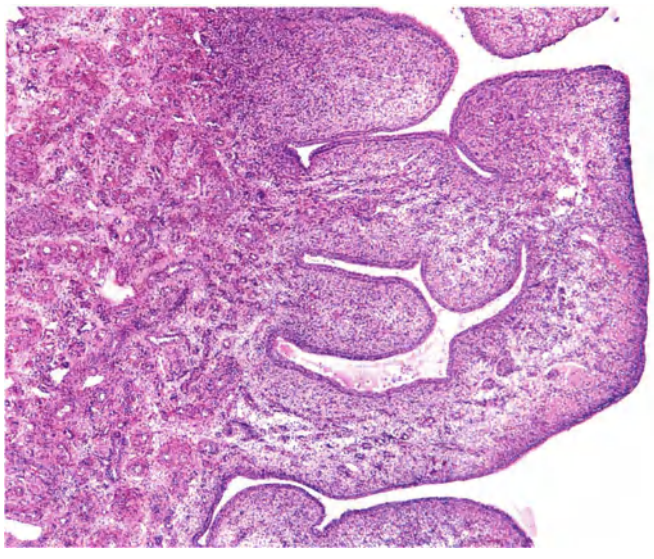

Fig. 125. *Renal pelvic hamartoma.* This shows a very vascular polyp with poorly differentiated stroma

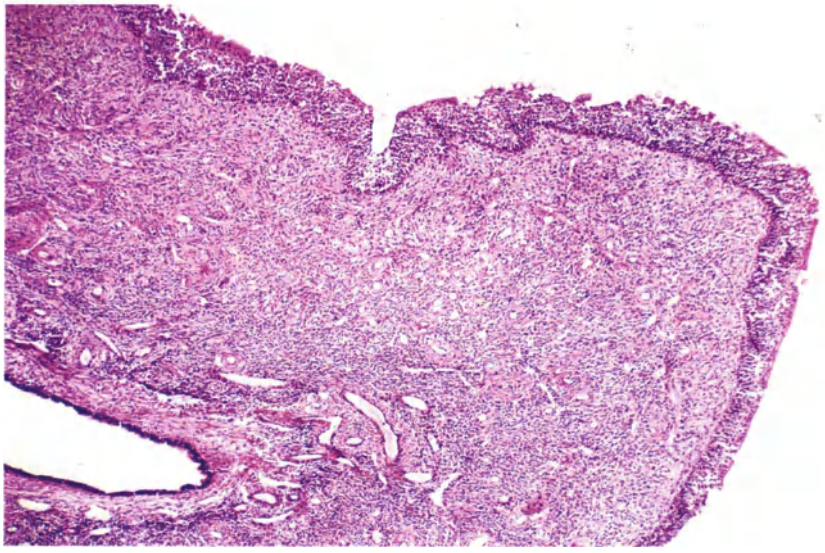

Fig. 126. *Renal pelvic hamartoma.* Microcystic structure (lower left) with poorly differentiated stroma

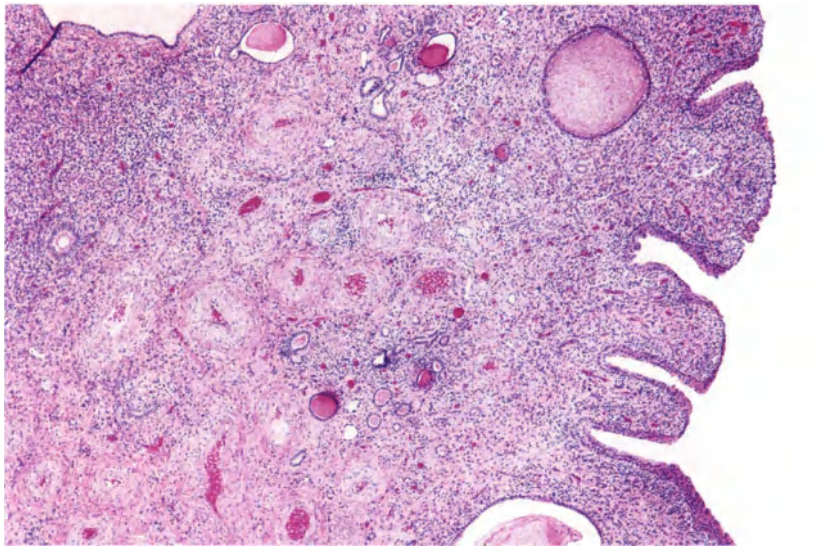

Fig. 127. *Renal pelvic hamartoma.* Aggregate of thick blood vessels

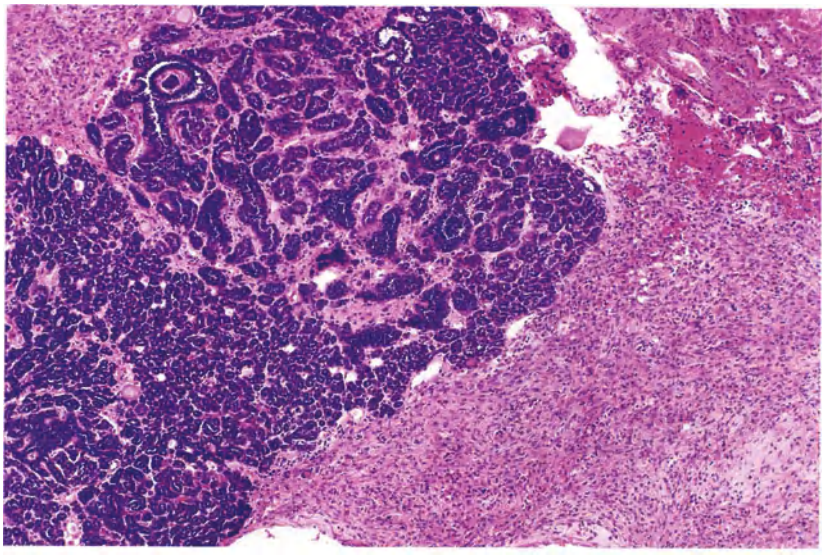

Fig. 128. *Nephrogenic adenofibroma*

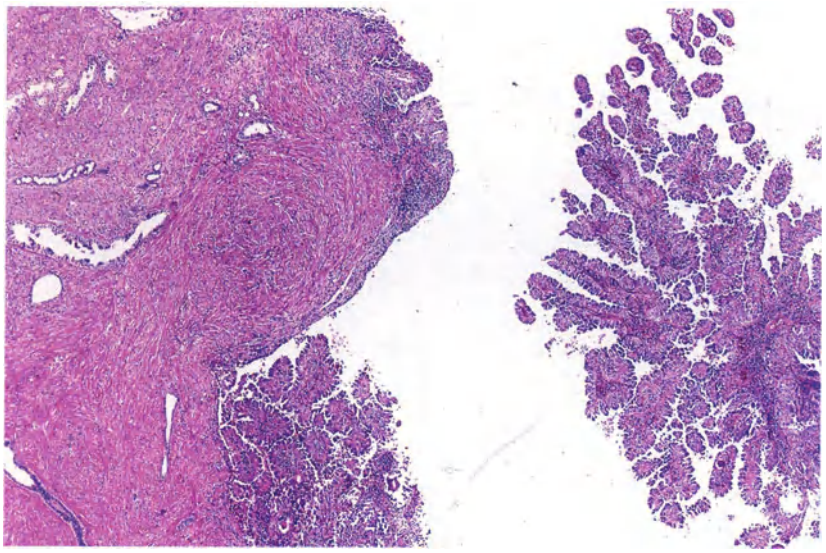

Fig. 129. *Nephrogenic adenofibroma.* This example was similar to Fig. 128 but included a papillary lesion in the pelvis resembling collecting-duct carcinoma

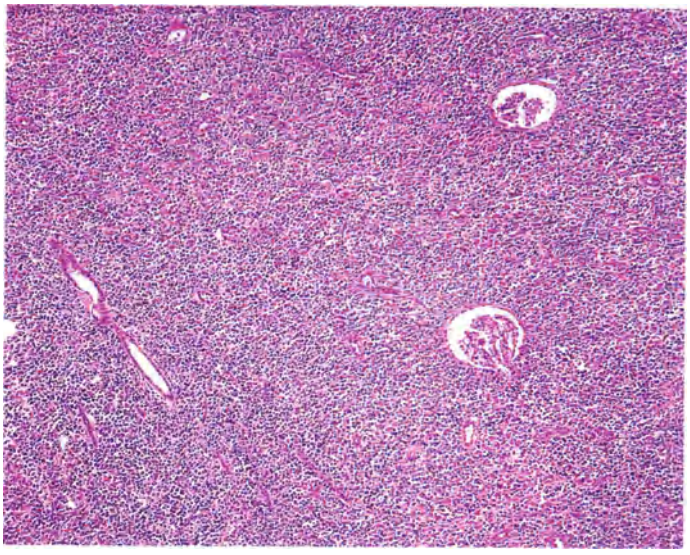

Fig. 130. *Malignant lymphoma*

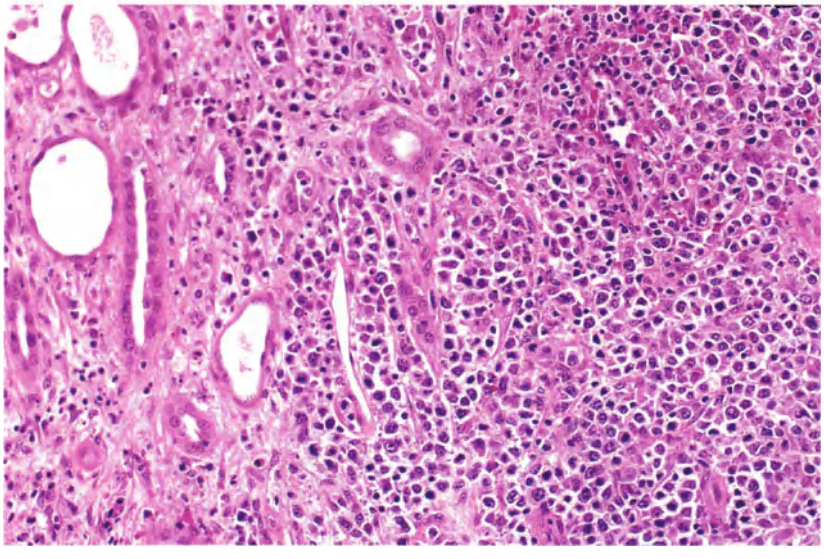

Fig. 131. *Malignant lymphoma.* Diffuse, large B-cell type

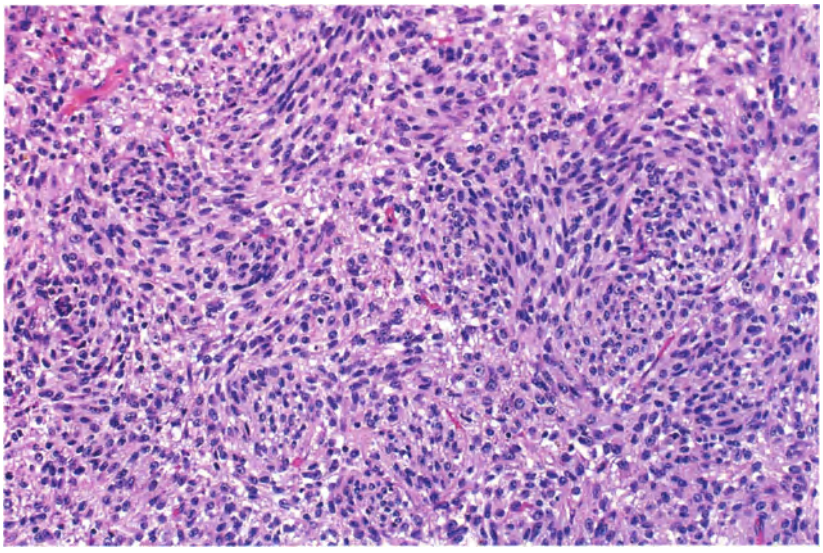

Fig. 132. *Malignant melanoma*

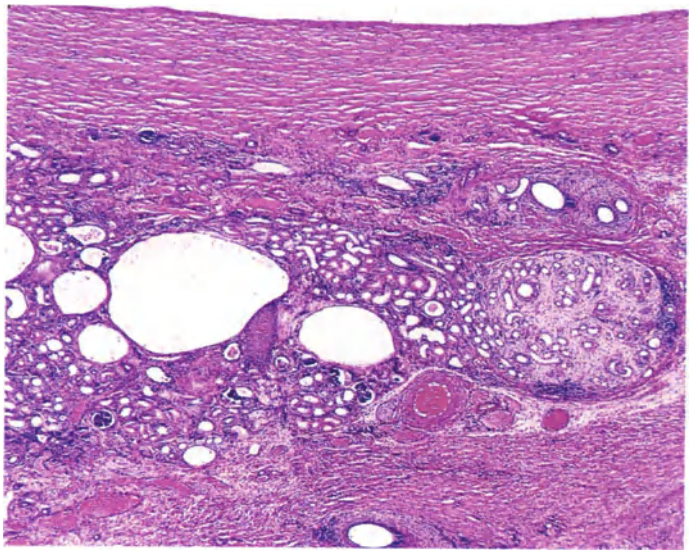

Fig. 133. *Renal dysgenesis.* An abortive renal segment in a malformed kidney. Cartilage just left of centre

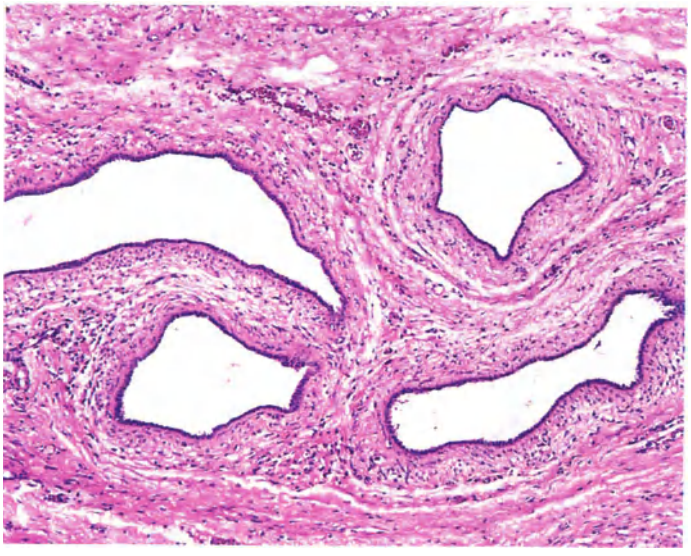

Fig. 134. *Renal dysgenesis.* The tubules and microcysts have connective tissue collars. In this adult patient the tissue is mature

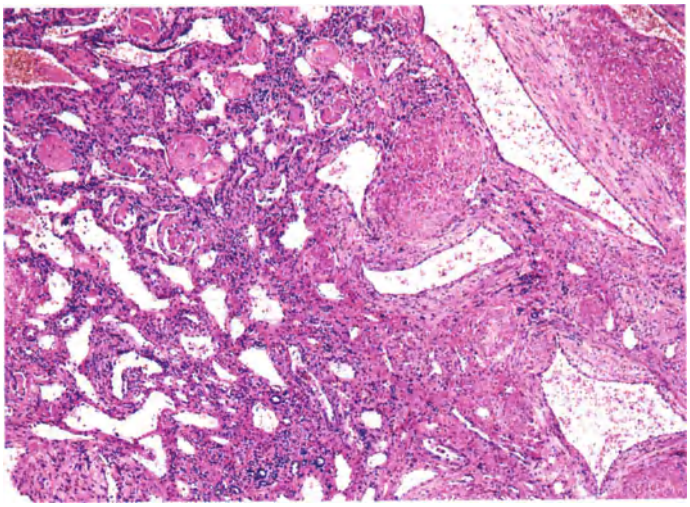

Fig. 135. *Vascular malformation.* Arteriovenous type

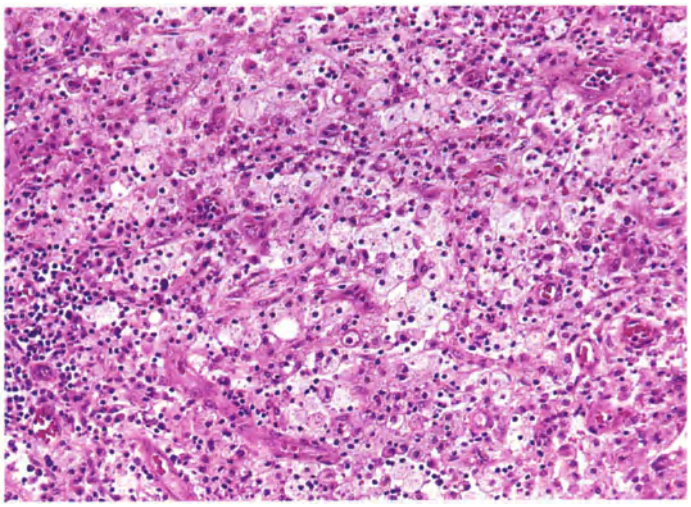

Fig. 136. *Xanthogranulomatous pyelonephritis.* Lipid-laden macrophages are admixed with lymphocytes and plasma cells

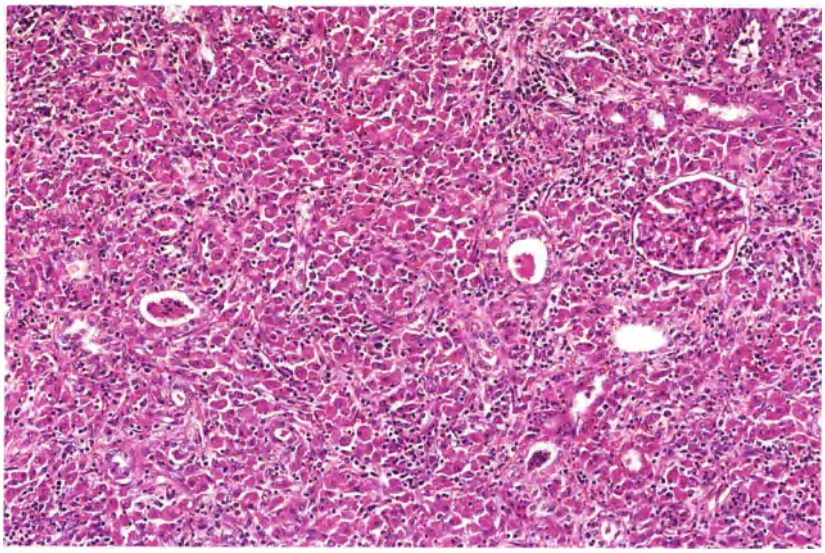

Fig. 137. *Renal cortical malakoplakia.* Eosinophilic histiocytes are admixed with lymphocytes and plasma cells

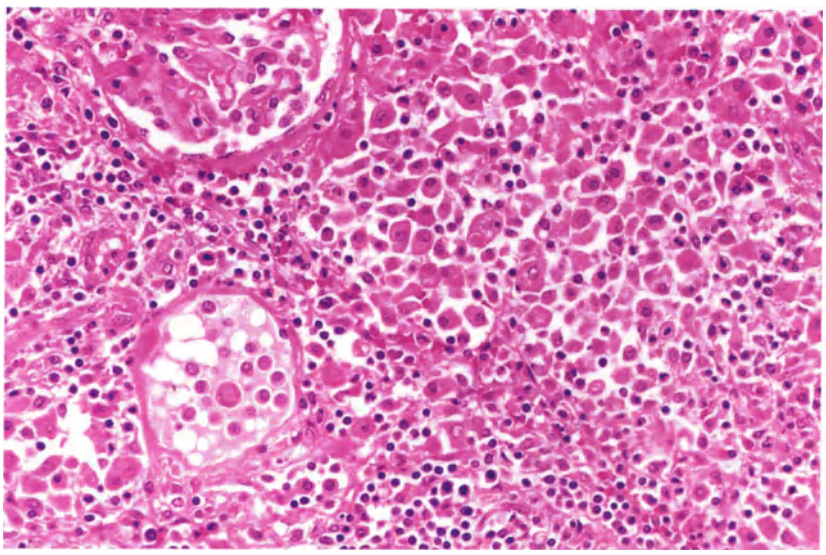

Fig. 138. *Renal cortical malakoplakia*

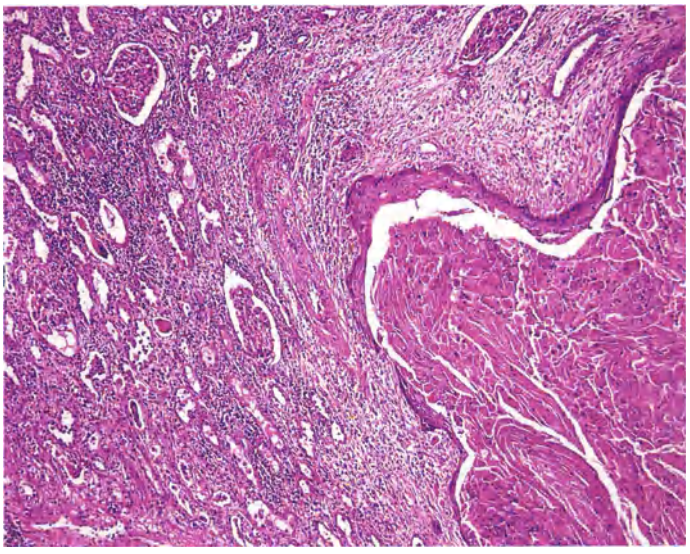

Fig. 139. *Cholesteatoma.* Keratinising squamous metaplasia of calyceal epithelium may produce this appearance

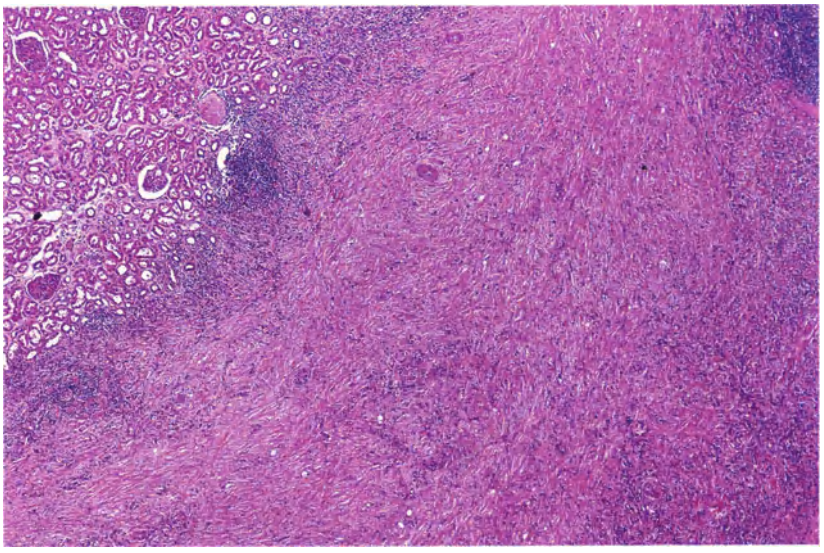

Fig. 140. *Inflammatory pseudotumour.* This tumour-like mass consists of myofibroblastic spindle cells with lymphocytes at the periphery

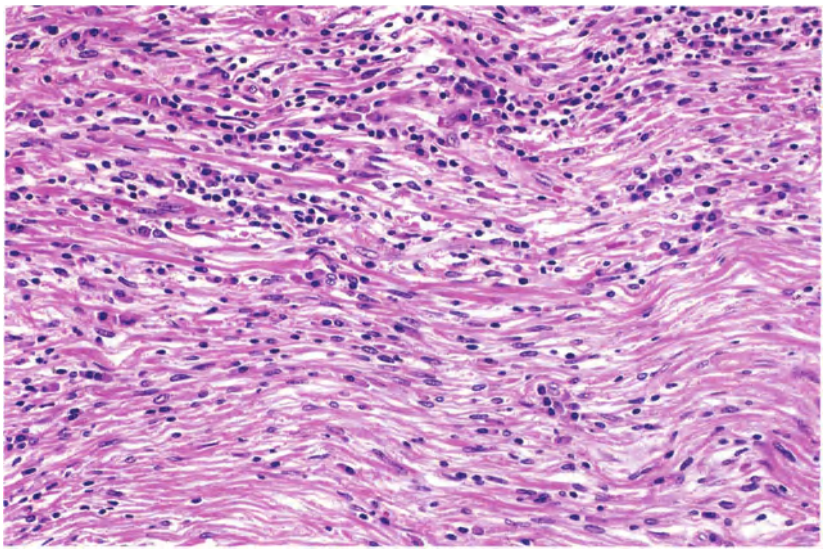

Fig. 141 *Inflammatory pseudotumour.* Higher magnification of Fig. 140

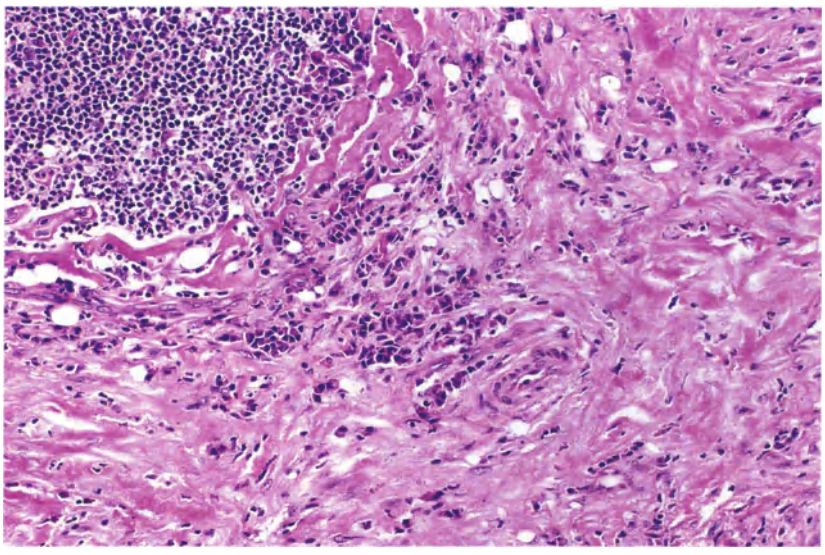

Fig. 142. *Inflammatory pseudotumour.* This lesion consists largely of hyalinised stroma with plasma cells and lymphoid aggregates

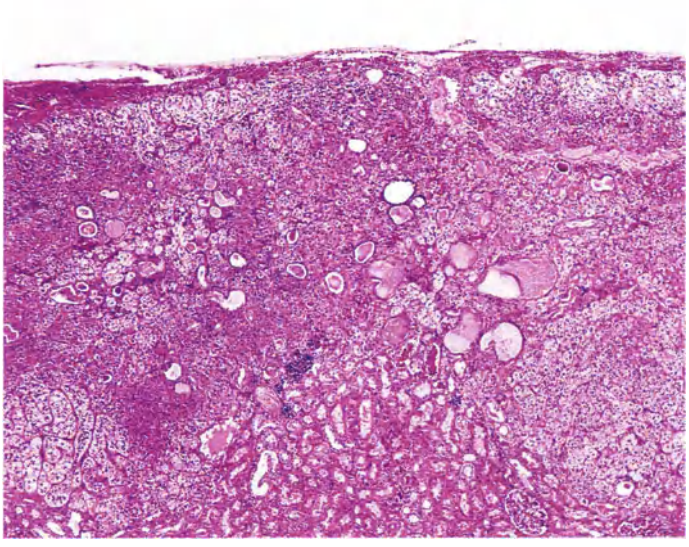

Fig. 143. *Adrenal rest.* Adrenal cortical tissue is located in the superficial renal cortex

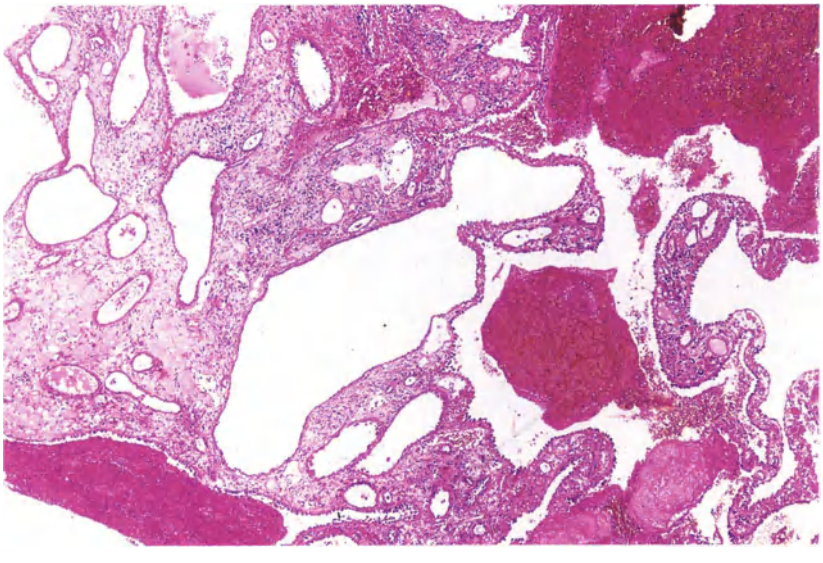

Fig. 144. *Nephrogenic adenoma, renal pelvis*

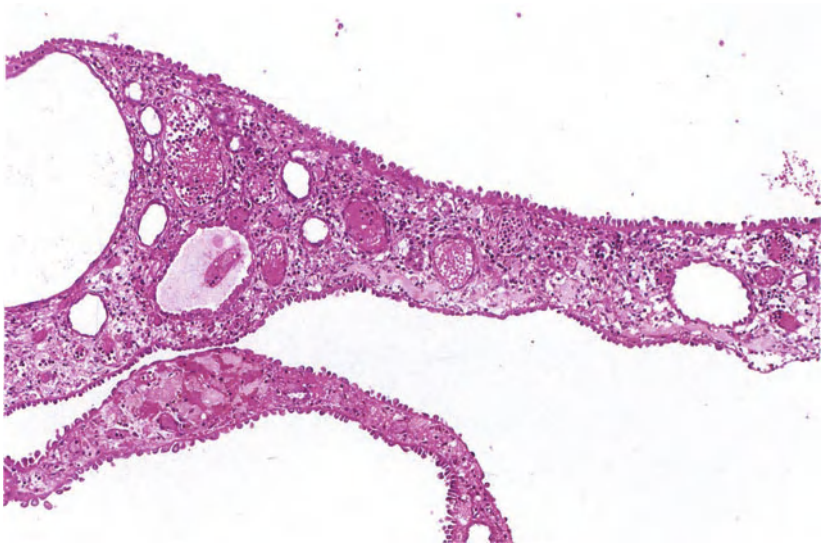

Fig. 145. *Nephrogenic adenoma, renal pelvis.* The stroma shows oedema and inflammation

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