9 Kidney Sarcomas

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9.1 Introduction

Primary sarcomas arising in the kidney, although quite rare, should be considered in the differential diagnosis of renal tumors. These tumors, of which there are various histological types, account for less than 1% of all renal malignancies (Vogelzang et al. 1993). Primary renal sarcomas may arise from one or more of the progenitor cell lines of the connective tissues normally found within the kidney (Table 9.1).

Table 9.1. Cellular origin of primary renal sarcomas

Tissue type	Tumor
Smooth muscle	Leiomyosarcoma
Adipose tissue	Liposarcoma
Skeletal muscle	Rhabdomyosarcoma
Vascular	Angiosarcoma, hemangiopericytoma
Bone	Osteosarcoma
Cartilage	Chondrosarcoma
Myofibroblast	Malignant fibrous histiocytoma
Fibroblast	Fibrosarcoma
Neuroectoderm	Extraosseous Ewing sarcoma/primitive neuroectodermal tumor
Undifferentiated	Malignant mesenchymoma

Leiomyosarcoma is the most common histological subtype accounting for 50–60% of the primary renal sarcomas reported in the literature (Grignon et al. 1990). Less common entities, such as rhabdomyosarcoma, liposarcoma, malignant hemangiopericytoma, angiosarcoma, osteosarcoma, chondrosarcoma, malignant fibrous histiocytoma, fibrosarcoma, extraosseous Ewing sarcoma/primitive neuroectodermal tumor (PNET), and malignant mesenchymoma arising from the kidney, have also been reported but are exceedingly rare. Overall, the age at presentation with primary renal sarcoma ranges from 28 to 70, with the median age of 49 years. Forty percent of renal sarcomas contain central areas of necrosis at the time of diagnosis; average tumor size ranges from 5.5 to 23 cm. Prognosis is generally poor, and 90% of patients present with distant metastasis, pulmonary metastasis being the most common (Levine and King 2000). Radiographically, most primary renal sarcomas are indistinguishable from renal cell carcinoma and histologically may be confused with renal cell carcinomas which contain tissues which have undergone sarcomatoid differentiation. For this reason, a case of sarcomatoid differentiation within a renal cell carcinoma is included in section 9.13, as this tumor is part of the differential diagnosis of primary sarcoma of the kidney. The diagnosis of primary renal sarcoma is made based on a combination of the imaging and histological characteristics of the tumor and often after

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extensive scrutiny to exclude more common tumors such as renal cell carcinoma.

9.2 Leiomyosarcoma

Renal leiomyosarcomas are the most common histological subtype of renal sarcomas, accounting for 58% of all primary renal sarcomas (Levine and King 2000). Arising from smooth muscle, these tumors originate from the renal capsule or from the smooth muscle of the renal pelvis or vasculature, including the renal vein or intraparenchymal vasculature (Moazzam et al. 2002; Ng 1985). These tumors most commonly occur in the sixth decade of life (Chen and Lee 1997). Preponderance in women (7 of 10) has been reported, as well as preference for the right kidney (Deyrup et al. 2004).

Histopathologically, leiomyosarcoma appears as closely packed interlacing spindle-shaped cells with cellular atypia, and therefore can be confused with sarcomatoid differentiation within a renal cell carcinoma which also is composed primarily of spindle-shaped cells. Immunohistochemical stains of the cytoplasm of tumor cells of leiomyosarcoma are positive for vimentin and muscle-specific antigen, whereas those of the sarcomatoid variant of renal cell carcinoma are not (Moudouni et al. 2001). Renal leiomyosarcoma

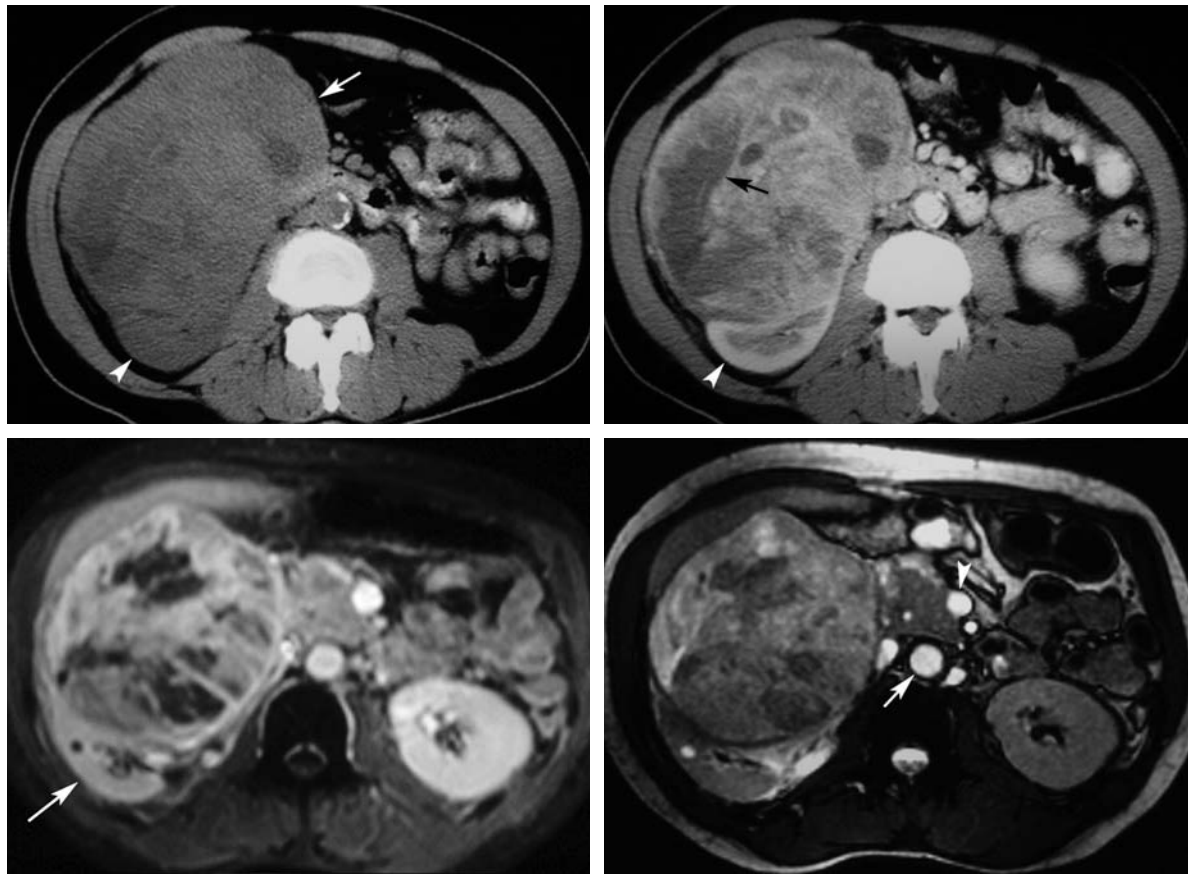

Fig. 9.1a-d. Leiomyosarcoma in a 60-year-old man. **a** Axial unenhanced CT scan demonstrates a large, heterogeneous soft tissue density mass arising from the anterior aspect of the right kidney at the midpolar region (*arrow*). The tumor is slightly hyperdense than the adjacent normal renal parenchyma, seen posteriorly (*arrowhead*). **b** Axial contrast-enhanced CT scan at the same level as **a** shows that the normally enhancing renal parenchyma posteriorly is clearly delineated (*arrowhead*) and the mostly solid mass is obviously arising from and splaying the kidney. Heterogeneous enhancement with hypodense areas is seen. These hypodense areas (*arrow*) are secondary to internal necrosis of the mass as it outgrows its vascular supply. **c** Axial fat-suppressed contrast-enhanced T1-weighted MR image demonstrates a heterogeneous, predominately hyperintense renal mass. The normal-appearing kidney is seen posteriorly (*arrow*). **d** Axial balanced fast-field-echo (FFE) T2-weighted MR image demonstrates a heterogeneous predominately hypointense mass. Extension of the tumor between the aorta (*arrow*) and the superior mesenteric artery (*arrowhead*) is demonstrated.

has been reported in a patient with tuberous sclerosis (Fernandez de Sevilla et al. 1988) and malignant transformation of angiomyolipoma to leiomyosarcoma has also been reported (Lowe et al. 1992).

On unenhanced CT imaging, leiomyosarcomas are often higher in density than adjacent renal parenchyma and similar to paraspinal muscle density (Fig. 9.1). After administration of intravenous contrast, the solid components of these tumors demonstrate variable enhancement (Figs. 9.1, 9.2). Tumor calcification can occur in 10% of cases. Even though presentation with spontaneous rupture of a renal neoplasm is more commonly seen in large angiomyolipomas, leiomyosarcomas can present in this way (Moazzam et al. 2002). In such a case, the imaging findings correspond to those of a retroperitoneal hematoma. Computed tomography shows a large heterogeneous mass in the perinephric space with minimal postcontrast enhancement.

Magnetic resonance imaging characteristics of leiomyosarcoma, like the majority of primary renal sarcomas, include intermediate to hypointensity on T1-weighted MR imaging and intermediate to hyperintensity on T2-weighted MR imaging (Figs. 9.1, 9.2). Internal necrosis, especially when the tumor becomes large, is a common feature of this tumor (Kaushik and Neifeld 2002). On T1-weighted MR images, the area of internal necrosis is hypointense, whereas on T2-weighted images, the area of necrosis is hyperintense (Fig. 9.2).

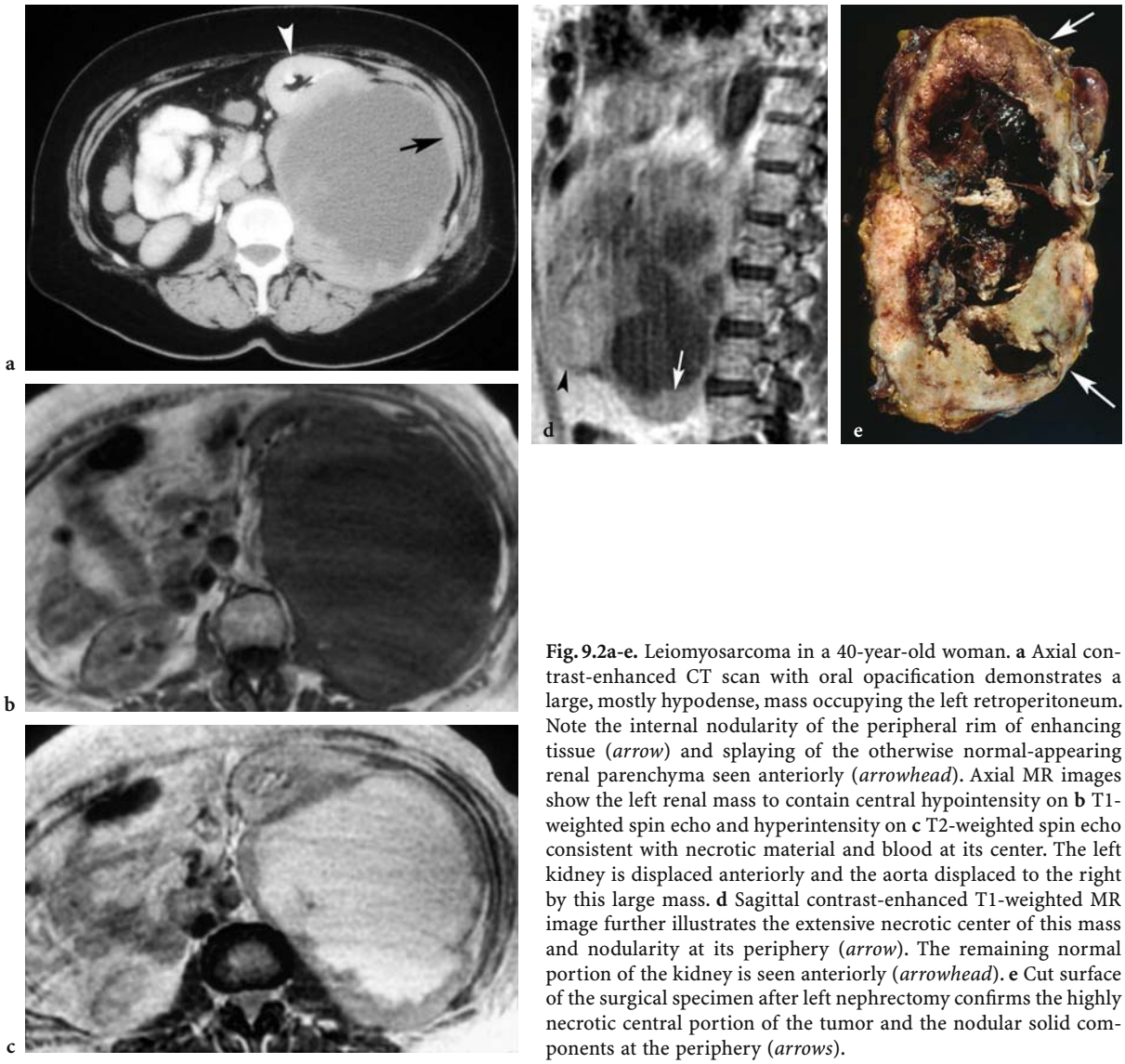

Fig. 9.2a-e. Leiomyosarcoma in a 40-year-old woman. **a** Axial contrast-enhanced CT scan with oral opacification demonstrates a large, mostly hypodense, mass occupying the left retroperitoneum. Note the internal nodularity of the peripheral rim of enhancing tissue (*arrow*) and splaying of the otherwise normal-appearing renal parenchyma seen anteriorly (*arrowhead*). Axial MR images show the left renal mass to contain central hypointensity on **b** T1-weighted spin echo and hyperintensity on **c** T2-weighted spin echo consistent with necrotic material and blood at its center. The left kidney is displaced anteriorly and the aorta displaced to the right by this large mass. **d** Sagittal contrast-enhanced T1-weighted MR image further illustrates the extensive necrotic center of this mass and nodularity at its periphery (*arrow*). The remaining normal portion of the kidney is seen anteriorly (*arrowhead*). **e** Cut surface of the surgical specimen after left nephrectomy confirms the highly necrotic central portion of the tumor and the nodular solid components at the periphery (*arrows*).

At nephrectomy, a tumor composed of nodular solid elements, and commonly areas of internal necrosis, is found (Fig. 9.2). Nephrectomy with adjuvant chemotherapy and postoperative radiation is currently the method of treatment; however, published clinical studies of the effectiveness of treatment and the prognosis are lacking (Moudouni et al. 2001). Intermediate- and high-grade tumors are aggressive neoplasms with poor prognosis.

9.3 Liposarcoma

Liposarcoma is one of the most common retroperitoneal tumors, often occurring in the perinephric space (Israel et al. 2002). Liposarcoma can arise from the retroperitoneal fat, including the perirenal fat within Gerota's fascia, or even the renal sinus fat, but are not usually primary neoplasms of the kidney. Because this tumor arises from the retroperitoneal space, a retroperitoneal liposarcoma will be seen displacing or encasing, but not invading, the renal parenchyma, as is illustrated in the selected CT images of two cases of retroperitoneal liposarcoma in Figs. 9.3 and 9.4. On the other hand, a primary renal liposarcoma will arise from and distort the architecture of the kidney. Rather than merely displacing the kidney, a primary renal liposarcoma will cause a defect in the renal parenchyma as it grows outward from the organ. In some cases, especially when the tumor is large, it can be difficult to determine whether a liposarcomatous mass is arising from or merely encasing the kidney. In a comprehensive review by Mayes et al. (1990), eight cases of liposarcoma with unequivocal involvement of the renal parenchyma were described. The histology of these tumors was reviewed, and the diagnosis of primary renal sarcoma was confirmed. In this case series, of the eight tumors occurring in patients from 33 to 68 years of age, there was no gender predominance.

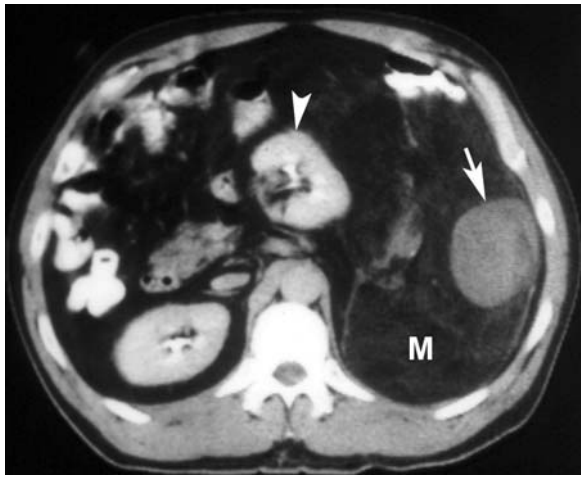

Fig. 9.3. Retroperitoneal liposarcoma in a 63-year-old man. Axial contrast-enhanced CT scan demonstrates a large retroperitoneal fat density mass (*M*) occupying the left renal fossa and retroperitoneal space. Within the mostly fatty tumor is a solid soft tissue component (*arrow*). The left kidney is significantly displaced anteriorly and medially (*arrowhead*), but there does not appear to be any disruption of the renal parenchyma. This tumor is a retroperitoneal liposarcoma, which is much more common than the primary renal liposarcoma.

On CT, both retroperitoneal and primary renal liposarcomas demonstrate a variety of appearances dependent on the differentiation of the fatty component of the tumor, ranging from well-differentiated tumors with fat-density Hounsfield units (HU) values to undifferentiated tumors that are difficult to distinguish radiographically from other sarcomas (Levine and King 2000). The differential diagnosis of these usually prominent fatty neoplasms should also include angiomyolipoma. In a review of 27 fat-containing tumors of the retroperitoneum, Israel et al. (2002) described the major imaging findings which are important to consider when differentiating angiomyolipoma from liposarcoma of the retroperitoneum. They noted that a defect in the renal parenchyma adjacent to the mass is commonly seen in exophytic angiomyolipomas, but not identified in the 12 cases of retroperitoneal liposarcoma reviewed. Liposarcoma arising in the retroperitoneum displaces and distorts the adjacent kidney, but the interface between the two is characteristically smooth. According to the same principle, primary renal sarcomas, because they arise from the kidney itself rather than the retroperitoneal space, disrupt the renal parenchyma, causing a defect similar to that described for angiomyolipomas. Additionally, Israel et al. (2002) noted that angiomyolipomas commonly contain enlarged vessels that can be seen on contrast-enhanced CT. By comparison, well-differentiated liposarcomas are less vascularized and those vessels that are present are not usually enlarged. Magnetic resonance imaging characteristics depend on the fat content of these tumors, which are hyperintense on both T1- and T2-weighted sequences (Fig. 9.5).

At nephrectomy, the lobulated fatty portion of this tumor is evident (Fig 9.5).

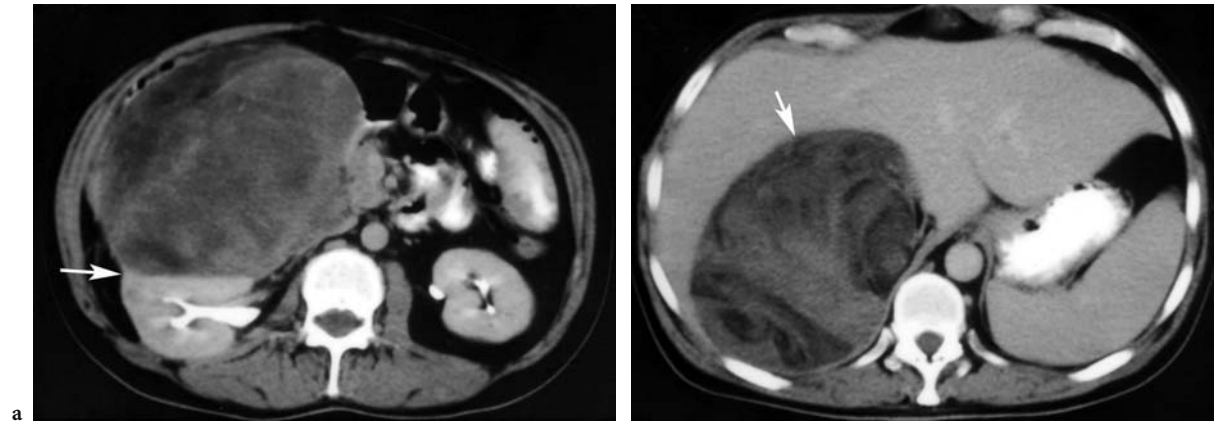

Fig. 9.4a,b. Retroperitoneal liposarcoma in a 53-year-old woman. **a** Axial contrast-enhanced CT scan shows a large, mostly fat density mass, displacing, compressing, and distorting the right kidney. Compared with the previous case of retroperitoneal liposarcoma seen in Fig. 9.3, this tumor is less well differentiated, containing soft tissue density whorls of tissue. Also note that the interface between the mass and the anterior margin of the right kidney is smooth (*arrow*). There is no defect in the renal parenchyma to suggest that the tumor is arising from the kidney, as would be seen in angiomyolipoma or primary renal liposarcoma. **b** This tumor is large, extending superiorly into the hepatorenal fossa (*arrow*) and displacing the right lobe of the liver anteriorly. Retroperitoneal liposarcoma demonstrates a propensity to spread throughout the retroperitoneal spaces.

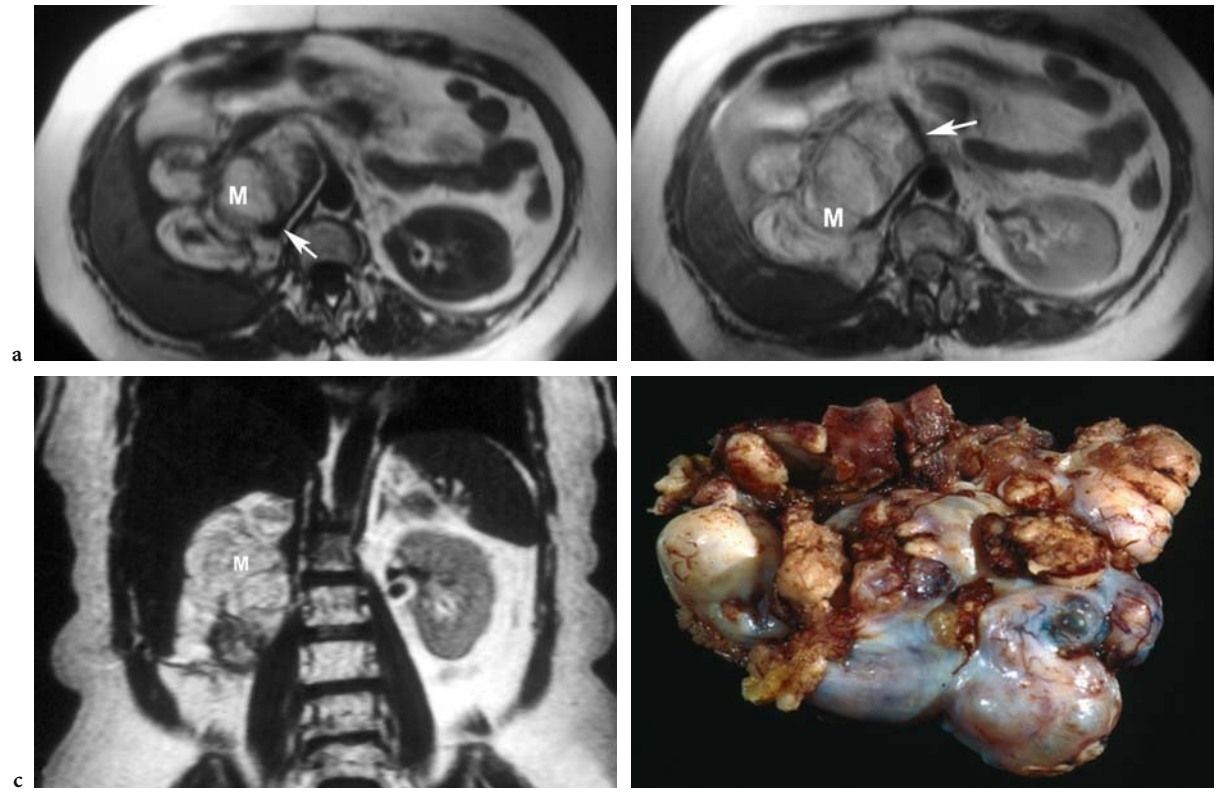

Fig. 9.5a-d. Capsular renal liposarcoma in a 48-year-old man. **a** Axial T1-weighted spin-echo MR image of a fat-containing, lobulated mass (*M*) arising from the right kidney causing significant compression of the inferior vena cava (*arrow*). No normal renal parenchyma remains. **b** Axial proton density-weighted MR image at the same level as **a** shows the inferior vena cava and left renal vein are significantly compressed by the mass (*M*), which extends medially to the level of the superior mesenteric artery (*arrow*). **c** Coronal T1-weighted spin-echo MR image demonstrates the lobulated fatty tumor of the right renal fossa (*M*) which has completely replaced the right kidney, growing superiorly into the hepatorenal fossa where it displaces the right lobe of the liver. **d** Gross surgical specimen shows the lobulated fatty tumor. The fatty components are yellowish, soft, and variegated. Histology is consistent with a liposarcoma arising from the renal capsule and invading the renal parenchyma.

9.4 Angiomyolipoma with Sarcomatoid Transformation

Angiomyolipomas are benign hamartomas of the renal parenchyma found in approximately 45–80% of patients with tuberous sclerosis, often multiple and bilaterally (Lowe et al. 1992). Angiomyolipomas may also present as unilateral, and often large, solitary tumors in a patient with no clinical signs of tuberous sclerosis. Given the prevalence of benign angiomyolipoma, especially in patients with tuberous sclerosis, a tumor arising from the kidney containing CT or MR imaging characteristics consistent with intratumoral fat is most likely a benign angiomyolipoma. The fat content with a negative density value (–10 to –100 HU) of these intraparenchymal renal lesions is readily apparent on thin-slice CT, and the diagnosis of angiomyolipoma is relatively certain except in rare cases of renal cell carcinoma containing intratumoral fat or, very rarely, primary renal liposarcoma.

Reported cases of renal angiomyolipoma containing a large soft tissue component made up of cells characteristic of sarcoma have been reported in the literature (Lowe et al. 1992). Upon histological examination of these tumors, cellular atypia is a common feature. These tumors most commonly contain spindle-shaped smooth muscle cells with variation in nuclear size and occasional mitosis, and may be confused with liposarcoma (Israel et al. 2002); however, aggressive features, such as cellular atypia, can also be seen in benign angiomyolipoma and therefore are not reliable indicators as to the potential malignant nature of these tumors. The propensity to invade adjacent structures, including the renal veins and local recurrence after incomplete surgical resection, is a well-documented feature of benign angiomyolipomas, and also is not helpful in distinguishing a benign angiomyolipoma from those with malignant sarcomatoid transformation. The true malignant nature of sarcomatoid transformation within a renal angiomyolipoma is noted in well-documented cases of distant metastasis to the lung and liver from malignant transformation of angiomyolipomas (Christiano et al. 1999; Cibas et al. 1998; Ferry et al. 1991; Kawaguchi et al. 2002; Martignoni et al. 2000). For this reason, it is important to include sarcomatoid transformation of an angiomyolipoma in the differential diagnosis of primary renal sarcoma. Figure 9.6 illustrates the CT and MR imaging characteristics of an angiomyolipoma with sarcomatoid transformation diagnosed in a 52-year-old woman who subsequently underwent nephrectomy. At the time of surgery, liver metastases consistent with metastatic sarcomatous angiomyolipoma were discovered. The only consistent finding noted in the few cases of previously reported sarcomatoid transformation of angiomyolipoma is the presence of intratumoral fat. Both CT and MR imaging are capable of demonstrating the fat-density component of this tumor.

9.5 Rhabdomyosarcoma

Rhabdomyosarcoma is so named because of the malignant-appearing muscle cells within the tumor, which are identified by the presence of cross striations and immunohistochemical stains positive for desmin, myoglobin, and myogenin (Mainguene et al. 2003). Although rhabdomyosarcoma is primarily a tumor found in the soft tissues, primary rhabdomyosarcoma of the kidney in adults have also been reported but are exceedingly rare (Mainguene et al. 2003; Seabury et al. 1967; Srinivas et al. 1984). Weeks et al. (1991) reported eight cases of primary renal primitive neuroectodermal tumor similar clinicopathologically to malignant rhabdomyosarcoma, although the latter is noted to be more aggressive, with invasion of the renal veins, IVC, and adjacent soft tissues common. In one case series of three patients with primary renal rhabdomyosarcoma, despite aggressive treatment with combinations of radiation therapy (preoperatively or postoperatively), nephrectomy, and chemotherapy, all had distant metastasis at the time of diagnosis and died within 6 months of diagnosis (Srinivas et al. 1984).

Rhabdosarcomatous tumors of the kidney in children are better documented in the literature than those diagnosed in adults. Some authors have noted histologically malignant-appearing muscle cells in up to 50% of nephroblastomas (Wilms tumors) and consider embryonal renal rhabdomyosarcoma to represent an overgrowth of malignant muscle within a nephroblastoma (Beckwith and palmer 1978; Seabury et al. 1967). In one case report, embryonal rhabdomyosarcoma, also referred to in the literature as cystic partially differentiated nephroblastoma, is described in infants with variegated mosaic aneuploidy and central nervous system malformations such as microcephalus, Dandy-Walker malformation, and cataracts (Furukawa et al. 2003).

Rhabdoid tumor of the kidney (RTK) is a rare, highly malignant renal tumor of childhood, recog-

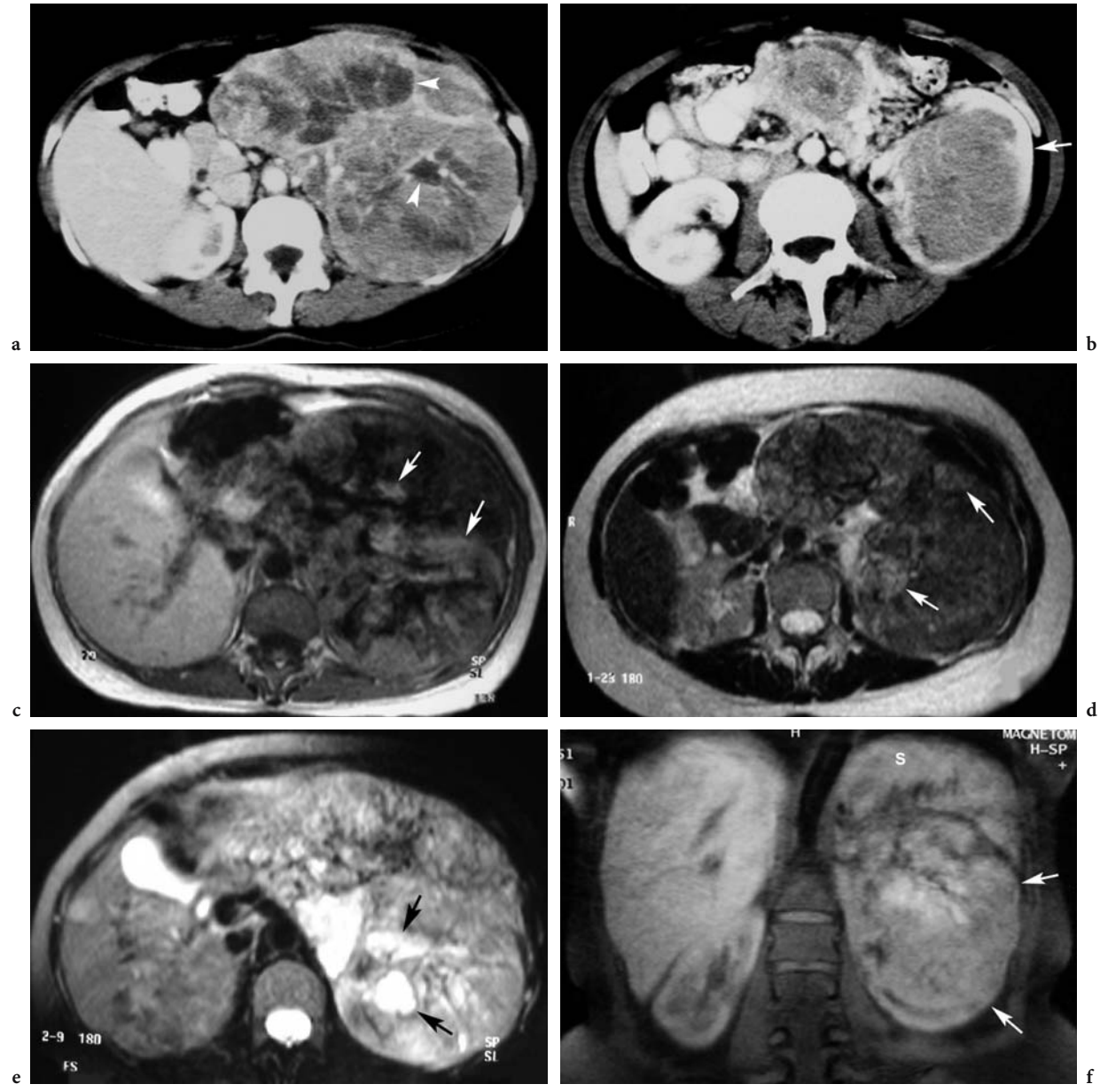

Fig. 9.6a-f. Angiomyolipoma with sarcomatoid transformation in a 52-year-old woman. **a,b** Axial contrast-enhanced CT scans show a large renal mass of the left kidney which extends across the midline of the abdomen. The mass contains tissues of differing degrees of enhancement, including areas of fat-density attenuation (*arrowheads*). There is a rim of normally enhancing renal parenchyma seen at the margins of the tumor in **b** (*arrow*). Axial **c** T1-weighted spin-echo and **d** T2-weighted spin-echo MR images demonstrate the complex nature of the solid left renal mass. Intratumoral fat appears hyperintense on the T1- and T2-weighted images (*arrows*). **e** Axial fat-suppressed FFE T2-weighted spin-echo MR image of the left renal mass demonstrates multiple hyperintense areas consistent with cystic fluid-filled spaces (*arrows*) similar in signal intensity to the fluid-filled gallbladder. Fat suppression is employed in this sequence to better characterize the fatty components of the tumor. **f** Coronal fat-suppressed contrast-enhanced T1-weighted MR image shows the large left renal mass (*arrows*) completely replacing the left kidney and displacing the spleen (*S*) superiorly; compare with the normal-appearing right kidney.

nized as a distinct entity (Agrons et al. 1997). Rhabdoid tumor of the kidney accounts for 2% of childhood renal neoplasms (Lowe et al. 2000). It is unique among other renal tumors of childhood in its well-documented association with a second primary intracranial malignancy (Weeks et al. 1989). As many as 10–15% of patients with RTK also have central nervous system lesions. Primitive neuroectodermal tumor, ependymoma, and cerebellar and brainstem astrocytoma have all been documented (Lowe et al. 2000). The implications of finding a brain lesion in a child with a renal tumor are significant, since most children with RTK die within 1 year of diagnosis (Agrons et al. 1997). At presentation, nearly 80% have metastasis to the brain or lungs, and 18-month survival is less than 20% (Lowe et al. 2000). On the other hand, the diagnosis of Wilms tumor often incurs a favorable prognosis, with cure rates after nephrectomy and, in some cases, adjuvant chemotherapy, near 90% (Lowe et al. 2000). Compared with Wilms tumor, which is most commonly found in children at 36 months of age, and mesoblastic nephroma which is diagnosed at birth or in the neonatal period, the median age of diagnosis of RTK is 11 months (Agrons et al. 1997). The male-to-female ratio is 1.5:1. Although the radiographic appearance of RTK is often indistinguishable from Wilms tumor, the pre-test predictive value for diagnosis of the former is low because of the rarity of RTK relative to Wilms tumor. Rhabdoid tumors can be identified on contrast-enhanced CT by their lobulated appearance. Often the tumor lobules are separated by intervening areas of low density necrosis and/or hemorrhage (Chung et al. 1995; Lowe et al. 2000). A unique feature of RTK is the presence of a subcapsular fluid collection, identified in 44% of the tumors reviewed by Chung et al. (1995).

9.6 Malignant Hemangiopericytoma

Hemangiopericytoma is a rare sarcoma arising from pericytes, the contractile cells that surround blood vessels and function in regulation of blood flow. These tumors occur in multiple areas in the body, the most common being the thigh. In a review of the literature, hemangiopericytoma of the kidney are reported in patients of 16–68 years and no gender predominance is noted (Heppe et al. 1991).

Radiographically, hemangiopericytomas are highly vascular solid tumors which may resemble renal cell carcinoma or be homogeneous in density on CT.

Histopathologically, hemangiopericytomas show dilated branching capillaries referred to as the "staghorn" vascular pattern, lined by normal-appearing endothelial cells and surrounded by nests or whorls of spindle-shaped neoplastic pericyte cells (Chhieng et al. 1999). These findings are not pathognomonic and similar architectural patterns are observed in other soft tissue neoplasms, such as malignant fibrous histiocytoma, chondrosarcoma, and leiomyosarcoma. Metastatic rates has been reported to be from 20 to 50% in hemangiopericytoma arising from all sites (Heppe et al. 1991). Prognosis is generally poor, with an overall mortality rate of 50% (Levine and King 2000).

9.7 Osteosarcoma

Primary renal osteosarcoma is an exceedingly rare tumor. It is believed to arise from undifferentiated mesenchymal cells of the kidney. Extensive ossification in a "sunburst" pattern, usually evident on routine radiographs, may suggest the diagnosis (O'Malley et al. 1991); however, the sarcomatoid variant of renal cell carcinoma and osteogenic sarcoma with metastasis to the kidney may also contain ossification (Levine and King 2000; Micolonghi et al. 1984). Radiographic differentiation of these three malignancies is difficult. The sarcomatoid variant of renal cell carcinoma may be distinguished from primary renal osteosarcoma by the presence of a histologically detectable epithelial component and/or positive cytokeratin immunohistochemical staining in the former (O'Malley et al. 1991).

Renal tumor ossification is seen in osteogenic sarcoma of the bone metastasizing to the kidney, which is by far a more common malignancy than primary renal osteosarcoma. Elevated alkaline phosphatase levels may be helpful in distinguishing primary renal osteosarcoma from metastatic osteogenic sarcoma of the kidney when a calcified renal tumor is found in the absence of a clinical history of a primary tumor elsewhere or abnormal bone lesion on technetium nuclear bone scan (Daneshmand et al. 2003).

9.8 Angiosarcoma

Angiosarcomas can be found anywhere in the body, although most commonly in the skin and subcuta-

neous tissues. There is a known association of vinyl chloride and Thorotrast exposure with an increased incidence of hepatic, cerebral, and pulmonary angiosarcoma (Makk 1974). Few sufficiently well-documented cases of primary renal angiosarcoma exist in the English-language literature (Allred et al. 1981; Prince 1942). Microscopically, the tumor cells form closely packed sinusoidal vascular channels filled with blood and fibrin, lined by polygonal or spindle-shaped tumor cells (Allred et al. 1981). The presence of tumor cells lining the lumen of vascular channels distinguishes this tumor from the spindle cell variety of renal cell carcinoma and from the richly vascular hemangiopericytoma (Allred et al. 1981). Imaging characteristics are nonspecific.

9.9 Chondrosarcoma

First described as a skeletal tumor, reported cases of extraosseous mesenchymal chondrosarcoma arising from the kidney are found in the literature, the first case described by Pitfield et al. in 1981. In all sites, these tumors present in patients, more commonly women, between 20 and 30 years of age (Gomez-Brouchet et al. 2001). Although these tumors are relatively slow growing, they are also unpredictable. Development of metastasis to the liver and lungs 18 years after nephrectomy has been described in a case report of renal chondrosarcoma in a 39-year-old woman by Mehanna et al. (2004). The CT imaging of primary renal chondrosarcoma most often demonstrate a large soft tissue tumor with central calcification. Often, the calcified renal mass can be seen radiographically (Nativ et al. 1985). Histologically, the tumor consists of islands of well-differentiated cartilage surrounded by undifferentiated and spindle-shaped cells (Malhotra et al. 1984).

9.10 Malignant Fibrous Histiocytoma and Fibrosarcoma

Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma occurring in adulthood, accounting for 20–40% of all soft tissue sarcomas (Gibbs et al. 2001). However, the incidence of MFH arising within the kidney is low; fewer than 50 cases have been reported in the literature (Chen et al. 2003). More commonly, MFH arises in the extremities or the retroperitoneum. Histologically, MFH is thought to arise from primitive mesenchymal cells with partial histiocytic differentiation that grow in a storiform or cartwheel-like growth pattern (Scriven et al. 1984). Like other tumors with fibroblastic origin, including leiomyosarcoma, and fibrosarcoma, immunohistochemical stains for vimentin are positive (Chen et al. 2003). The imaging characteristics are nonspecific and often similar to other primary renal sarcomas and renal cell carcinoma. In 50% of cases, necrosis or dystrophic calcification may also be seen. Intratumoral fat is rare. Magnetic resonance imaging is suited best for defining the anatomy of the tumor and its surrounding structures. Despite radical nephrectomy with wide excisional margins, MFH shows a greater than 50% tendency for local recurrence and distant metastases to the lungs and bone are common (Chen et al. 2003).

Another uncommon malignant tumor of the kidney arising from fibrous tissue, usually of the renal capsule, is the renal fibrosarcoma (Valdes et al. 2003). This tumor occurs in adults, and the male-to-female ratio is equal (Kansara and Powell 1980). Sheets of spindle-shaped cells are seen microscopically, which can easily be misinterpreted as the much more common tumor, leiomyosarcoma; however, further evaluation with electron microscopy will reveal a prominent amount of collagen fibers interlaced with primitive-appearing mesenchymal cells suggestive of a fibrous rather than smooth muscle origin of the tumor. Staining with van Gieson and Mallory trichrome stains is helpful to confirm the presence of the collagenous fibers in the tissue and differentiate it from other renal tumors (Kansara and Powell 1980). Fibrosarcoma of the kidney is noted for its rapid rate of growth and aggressive behavior (Ares Valdes et al. 2003). Although it typically remains encapsulated, invasion of the renal veins is noted in 40% of published cases (Kansara and Powell 1980).

9.11 Malignant Mesenchymoma

Malignant mesenchymoma has been described as a tumor composed of two or more unrelated tissue types in addition to any fibrosarcomatous element. Specific tumor types, such as rhabdomyosarcoma with liposarcoma and rhabdomyosarcoma with osteosarcoma or chondrosarcoma, have been reported in the thigh and in the retroperitoneum.

Two previously reported cases of malignant mesenchymoma arising in the kidney demonstrate features of both leiomyosarcoma and osteosarcoma (Quinn et al. 1993).

9.12 Extraosseous Ewing Sarcoma/Primitive Neuroectodermal Tumor

Over 50 cases of renal primitive neuroectodermal tumor (PNET) have been reported in the literature (Pomara et al. 2004). It is often difficult to distinguish these tumors from extra-osseous Ewing sarcoma (EES), and also from malignant rhabdomyosarcoma, as these tumors are thought to arise from a common stem-cell precursor, although they demonstrate variable degrees of differentiation (Rodriguez-Galindo et al. 1997). These tumors are so similar, in fact, that EES and PNET have been shown to arise from a shared and unique chromosomal translocation in more than 90% of cases (Pomara et al. 2004). Primary renal PNET occurs most commonly in adolescents and young adults. The natural history of extra-skeletal PNET in all sites consists of a rapidly growing tumor with a propensity for early metastases to regional lymph nodes, bone, lung, and liver. Prognosis is poor, even for well-defined completely resectable tumors, because local recurrence after resection is common. Less than 55% of patients are disease free after 5 years (Pomara et al. 2004). Histological evaluation of both EES and PNET reveals small blue round cells arranged in perivascular lobules referred to as "Homer-Wright rosettes," interspersed with spindle-shaped cells and tumoral cells within the vascular lumens (Pomara et al. 2004). The appearance is similar to neuroblastoma, and immunohistochemical stains are often necessary for proper diagnosis (Gonlusen et al. 2001). Immunohistochemical stains positive for the monoclonal antibody CD99 are noted in the cytoplasm of the malignant-appearing cells and highly suggestive of the diagnosis of EES/PNET (Jimenez et al. 2002; Pomara et al. 2004).

9.13 Sarcomatoid Differentiation of Renal Cell Carcinoma

Sarcomatoid change has been found to arise in all histological subtypes of renal cell carcinoma which include conventional/clear cell carcinoma, papillary renal carcinoma, chromophobe renal carcinoma, and collecting duct carcinoma, although it is not known whether there is predilection for sarcomatous change in any of these subtypes relative to the others (de Peralta-Venturina et al. 2001). Sarcomatoid change within a renal cell carcinoma is much more common than primary renal sarcoma; therefore, a thorough histological evaluation of multiple tumor sections should be employed to exclude sarcomatoid variant of renal cell carcinoma before a diagnosis of primary renal sarcoma is made.

Pathologically, the sarcomatoid variant of renal cell carcinoma contains the malignant spindle cell appearance similar to primary renal sarcomas. In a study of 101 sarcomatoid renal cell carcinomas by de Peralta-Venturina et al. (2001), the sarcomatoid component of the tumor resembled fibrosarcoma in 54%, malignant fibrous histiocytoma in 44%, rhabdomyosarcoma in 2% and undifferentiated sarcoma in the remaining tumors. The distinction between sarcomatoid variant of renal cell carcinoma and primary renal sarcoma depends on identifying an epithelial component of the tumor in the former. In sarcomatoid variant of renal cell carcinoma, the epithelial component is often easily identified histologically. The use of immunohistochemical stains for cytokeratin is also employed, although sometimes these are also positive in cases of leiomyosarcomas and, rarely, malignant fibrous histiocytoma (Grignon et al. 1990). In a review of more than 2000 cases of radical nephrectomies performed for renal cell carcinoma, Cheville et al. (2004) reported the presence of a sarcomatoid component in 5% of the resected specimens.

Imaging characteristics are variable depending on the degree of differentiation of the tumor but are seldom helpful in distinguishing a renal cell carcinoma with sarcomatoid differentiation from a non-sarcomatous renal cell carcinoma. Diffusely calcified sarcomatoid renal cell carcinoma with osteosarcomatous differentiation has been reported as presenting with CT findings of a large calcified renal mass (Daneshmand et al. 2003). Sarcomatoid change within a renal cell carcinoma indicates a highly aggressive neoplasm which frequently invades adjacent abdominal organs and regional lymph nodes (Fig. 9.7). The presence of a sarcomatous component was reported with a higher mortality and poorer prognosis, with a median survival of less than 1 year in most studies (Cheville et al. 2004).

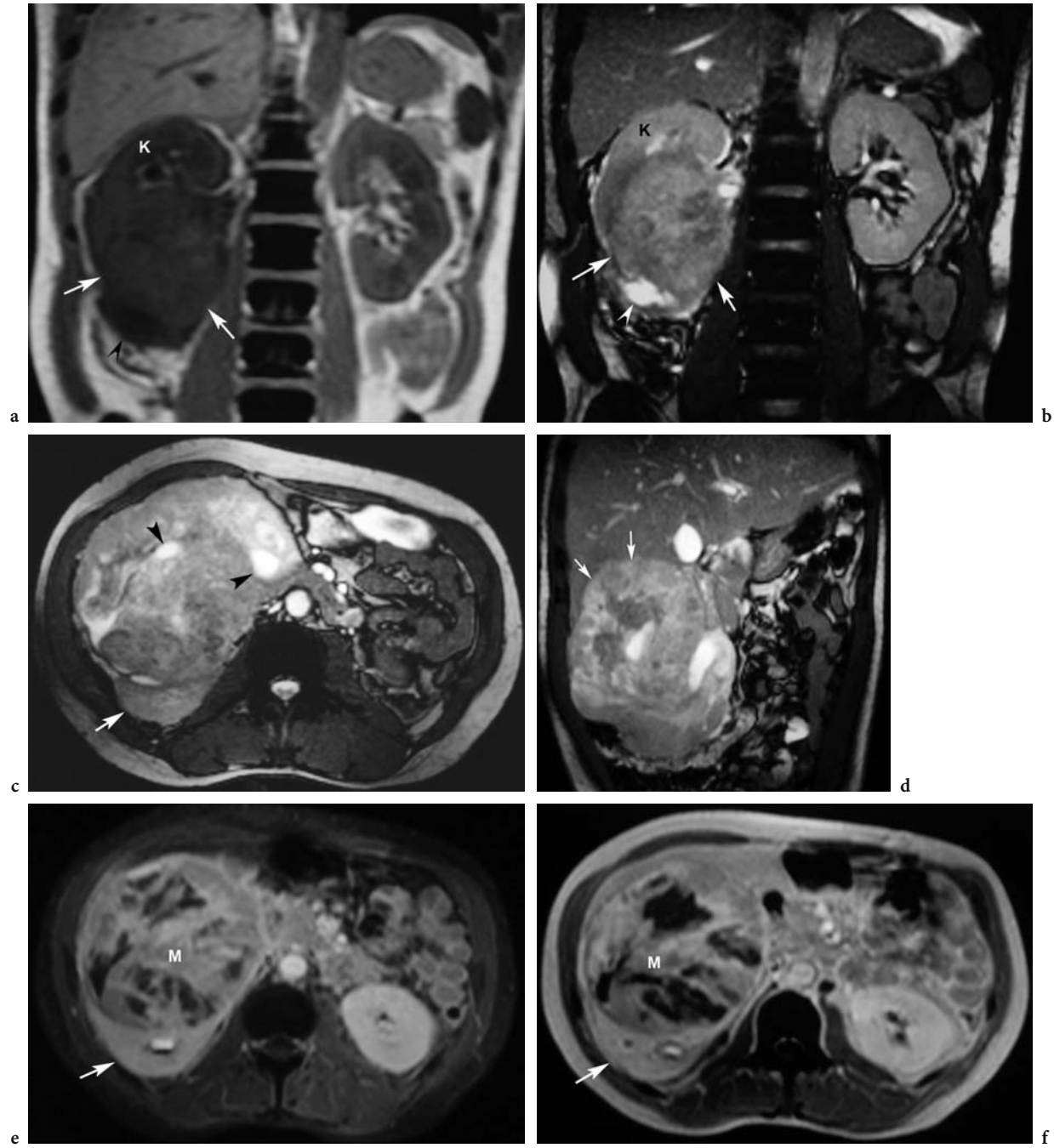

Fig. 9.7a-f. Sarcomatoid differentiation of renal cell carcinoma in a 65-year-old woman. Coronal **a** T1-weighted spin-echo and **b** balanced FFE T2-weighted MR images show the renal mass arising from the inferior pole of the right kidney (*arrows*). The normal renal parenchyma is seen at the superior pole (*K*). This predominately solid tumor contains a small area at its inferior aspect which is hyperintense on the T2-weighted images and hypointense on the T1-weighted images which may represent necrotic tissue or a cystic, fluid-filled space (*arrowhead*). **c** Axial balanced FFE T2-weighted MR image demonstrates the mass extending anteriorly from the right kidney and approaching the midline. A component of normal-appearing renal parenchyma (*arrow*) is seen posteriorly. Again demonstrated are the cystic spaces (*arrowheads*) within the mass. **d** Sagittal balanced FFE T2-weighted MR image shows that the right renal mass is replacing the renal parenchyma and extending into the subhepatic space (*arrows*). Axial **e** fat-suppressed contrast-enhanced T1-weighted and **f** contrast-enhanced T1-weighted MR images demonstrate the mostly solid mass arising from the right kidney. The complex nature of this mass (*M*) is demonstrated by the heterogeneous enhancement which appears as hyperintense areas after contrast administration, as compared with the homogeneous contrast enhancement of the normal contralateral kidney. A rim of normal renal parenchyma and contrast in the renal collecting system is seen posterior to the mass (*arrow*).

9.14 Conclusion

Primary renal sarcomas are rare entities which demonstrate aggressive local behavior and propensity for recurrence after surgical resection. The ominous nature of these lesions necessitates timely diagnosis and treatment. Before the diagnosis of primary renal sarcoma is made, the more common entities, such as sarcomatoid renal cell carcinoma or primary retroperitoneal sarcoma, should be considered. Diagnosis of renal cell sarcoma can be made with accuracy if three criteria, previously described by Grignon et al. (1990), are met:

1. The patient does not have a history of sarcoma elsewhere, to exclude metastatic disease.
2. The gross appearance of the tumor is compatible with origin in the kidney rather than involvement of the kidney by a retroperitoneal tumor.
3. Sarcomatoid variant of renal cell sarcoma is excluded.

Imaging characteristics of these neoplasms are highly variable, depending on the degree of differentiation of the tumor components. Both CT and MR imaging are useful in defining these characteristics, but the final diagnosis most often depends on histological and immunohistochemical evaluation.

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