Primary renal sarcoma is extremely rare and account for less than 1% of malignant renal tumors. We report a 24-year-old female with huge undifferentiated sarcoma arising from renal parenchyma of left kidney. The tumor was confined in the renal cortex without disruption of the renal capsule, which was difficult to be differentiated from renal cell carcinoma due to its unusual location. We present the imaging and histopathological findings of this rare case, and review the literatures.

Key words: Kidney neoplasms, Angiography; Kidney neoplasms, CT; Renal sarcoma

Primary renal sarcoma is a very high grade malignant tumor with various subtypes. The various mesenchymal cells found in the kidney may potentially develop into tumors of different histology. Leiomyosarcoma, fibrosarcoma, and liposarcoma are most frequent malignant ones. Most of the primary renal sarcoma arises from renal capsule or renal sinus. Undifferentiated sarcoma arising from renal parenchyma is extremely rare. We reported a 24-year-old female with a huge undifferentiated renal sarcoma arising from renal cortex with blood supply from renal artery. The pre-operative diagnosis of this unusual tumor was challenging because of its atypical imaging findings on both CT and angiography. The accurate diagnosis strongly depends on histopathologic findings. Despite of its rarity, primary renal sarcoma should be taken into differential diagnostic considerations in huge renal tumors.

CASE REPORT

A 24-year-old woman presented with left flank pain lasting for almost a week. Initially, a routine urine test showed unremarkable findings. The patient then underwent abdominal sonography in a local clinic, showing a large mass in the left kidney. Consequently, she underwent abdominal CT for further evaluation of the left renal mass. CT scan showed a large mass measuring about 8.0 cm in diameter at the upper half of the left kidney. The tumor was an encapsulated, heterogeneous, low-attenuating mass with peripheral enhancement (Fig. 1a, 1b). These findings may suggest malignancy. For preoperative evaluation, angiography was suggested. Angiography of left renal artery revealed a scanty vascular pattern in the main part of the tumor and hypervascularity in the periphery (Fig. 2). These observations also suggested malignancy.

On the basis of the imaging findings, a malignant left renal tumor was highly suspected. The primary conclusion was most likely an atypical renal cell carcinoma or an adult Wilms tumor. Left radical nephrectomy was indicated and performed about 2
weeks later. The operative finding confirmed an encapsulated mass with central necrosis and hemorrhage (size of about 8.0 cm) that occupied the upper half of the left kidney (Fig. 3).

To our surprise, microscopic examination showed that the neoplasm was composed of high-grade spindle cells with frequent mitoses (Fig. 4). The immunohistochemical studies demonstrated negative immunoreactivity in epithelial (CK), neural (S-100 and NSE), vascular (factor 8 and CD34), and muscle (actin and desmin) markers. However, a positive result was noted for vimentin stain, a sarcoma marker (Fig. 5). The pathologic conclusion was a renal sarcoma with poor histologic differentiation. The postoperative course was uneventful.

**DISCUSSION**

Primary sarcomas of the kidney are rare, constituting only 1.1% of all malignant renal tumors [1]. In clinical practice, sarcoma of the kidney is a diagnosis of exclusion, and the following possibilities must be ruled out: renal cell carcinoma, nephroblastoma, renal hamartoma, sarcomatoid carcinoma, and even renal teratoma [2]. In addition, the tumor should arise from nonepithelial mesodermal tissue. Various mesenchymal cells in the kidney can potentially develop into tumors with different histologies. The most

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**Figure 1.** a. Non-contrast enhanced CT shows inhomogeneous low-attenuated mass occupied the upper-half of the left kidney. The relatively hyperdense lesion is probably a hematoma (arrowhead), and the hypodense lesion may be necrosis (arrow). b. After intravenous contrast administration, peripheral enhancement is identified (big arrow), and prior suspected hematoma (arrowhead) and necrosis (arrow) appear more clearly.

**Figure 2.** The renal tumor reveals a hypovascular pattern with minimal neovascularity (arrow) in the periphery on the angiogram.

**Figure 3.** The picture of the surgical specimen shows a large tumor with central necrosis (arrow) and hemorrhage (arrowhead) over the upper-half of the left kidney in the sagittal section.
frequent malignancies include leiomyosarcoma, fibrosarcoma, and liposarcoma.

Other sarcomas, such as rhabdomyosarcoma, fibrosarcoma, angiosarcoma, osteosarcoma, can involve the kidney but are even more rare. In general, the renal sarcomas occur most frequently in persons aged 28-70 years and are high-grade malignancies. Men and women are equally affected, with possibly a slight male predominant. As for clinical symptoms, abdominal mass, pain, hematuria, and occasional weight loss are commonly noted. The prognosis is poor, with 90% of patients developing distant metastases. Pulmonary metastases are the most common site [1-2].

Radiographic features of renal sarcoma have been described in the literature [3-6]. As reported by Shirkhoda and colleagues [3], the CT and angiographic findings may suggest a preoperative diagnosis of renal sarcoma. Such findings include the location of the tumor, the lack of extension of the mass beyond its capsule, a fatty density of the mass in cases of liposarcoma, and a vascular pattern of the tumor. They proposed that CT could be useful in suggesting the diagnosis of renal sarcoma and that angiography could be helpful to confirm the diagnosis. When a well-encapsulated neoplasm originates from the renal capsule or renal sinus and when the tumor is hypovascular or avascular on angiography, the diagnosis of renal sarcoma should be highly suspected. Additionally, in the case of liposarcoma, the negative attenuation value of the tumor is a specific diagnostic characteristic. Furthermore, angiosarcoma of the kidney may appear as a characteristic hypervascular or hypovascular mass in respects of with tumor stains [7-8]. Most primary renal sarcomas possess imaging findings characteristic of malignant tumors but without histologic specificity, and cannot be differentiated from renal cell carcinomas [4].

As for our case, contrast-enhanced CT showed an encapsulated, large, low-attenuated mass with peripheral enhancement that occupied the upper half of the left kidney. The tumor appeared to arise from the renal parenchyma, and a renal cell carcinoma might have been considered. However, angiograms showed that the mass had a hypovascular pattern with peripheral neovascularity, which is similar to the angiographic feature of a primary renal sarcoma, as in Shirkhoda’s description [3]. In our clinical practice, atypical renal cell carcinomas can also have this clinical appearance. From this viewpoint, a renal cell carcinoma or an adult Wilms tumor (nephroblastoma) might be considered first, rather than a primary sarcoma because of its incidence. In this case, the pathologic report indicated a high-grade undifferentiated renal sarcoma with an unusual presentation.

In our case, the final pathologic conclusion was an undifferentiated renal sarcoma that originated from the renal parenchyma. This tumor is rare and further immunohistochemical stains are negative except for vimentin. Methods such as the polymerase chain reaction or electron microscopy may be helpful in this regard [9].

Regarding the imaging findings, CT and angiographic results are nonspecific, but a primary renal sarcoma has characteristic features. Angiography not only helps to differentiate malignant tumors preoperatively but also reveals the possibility of a sarcoma. However, the clinical differentiation of a sarcoma from a carcinomatous malignancy is difficult.

Figure 4. The histopathology of the renal tumor shows high-grade spindle cells with frequent mitoses. (Hematoxylin and eosin stain, 400x)

Figure 5. The cytoplasm of the spindle cell reveals positive stain for vimentin. (vimentin stain, 400x)
REFERENCES

未分化之腎臟肉瘤：病例報告

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原發性腎臟肉瘤極罕見，約佔所有腎臟惡性腫瘤的 1% 不到。我們報告一位 24 歲女性，患有一种种腎組織長出的巨大未分化型的原發性肉瘤。此腫瘤侷限於腎臟皮質內，且未穿出腎包膜外；於此不常見的位置是很難與一般腎臟細胞癌鑑別。在此我們提出這罕見案例的影像與病理組織發現，以及探討相關的文獻資料。

關鍵詞：腎腫瘤，血管攝影；腎腫瘤，電腦斷層；腎臟肉瘤