Case Report

Primary renal leiomyosarcoma: A diagnostic challenge

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Abstract

Primary leiomyosarcoma is an extremely rare entity constituting only 0.5–1% of all invasive renal tumors. It is frequently diagnosed on histological examination because it does not have any specific diagnostic features clinically and radiologically. At times, it is difficult to differentiate leiomyosarcoma from the sarcomatoid renal cell carcinoma even in histopathology as both the tumors have spindle-shaped atypical cells. Moreover, some epithelial markers can be present in pure smooth muscle sarcomas, while some smooth muscle markers are positive in carcinomas. Hence, a diagnosis of primary renal leiomyosarcoma should be made with caution. Since the prognosis for a renal sarcoma is particularly poor, differentiation from sarcomatoid renal cell carcinoma is necessary. The diagnostic challenge of one such tumor is discussed.

Key Words: Leiomyosarcoma, renal, sarcomatoid carcinoma

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INTRODUCTION

Primary sarcomas constitute from 0.8 to 2.7% of all renal tumors in adults.[1] Amongst the renal sarcomas, leiomyosarcoma is the most common histological subtype, accounting for 50–60% of all cases.[2,3] The most common symptoms and signs are like those of renal cell carcinoma, namely pain, palpable mass, and hematuria, all of which are indicators of an extensive local disease. They present as solid or cystic masses. Imaging may not be able to differentiate between leiomyosarcomas and renal cell carcinomas in all cases. Histogenesis remains obscure. Renal sarcomas may arise from the smooth muscle fibers of renal parenchyma, renal capsule, renal pelvis, or renal vessels.[4]

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CASE REPORT

A 62-year-old woman presented with angular stomatitis. She complained of pain abdomen off and on for the past 6 months, which was mild to moderate in severity. She was severely anemic. No other abnormality was found on both general and systemic examination. She denied history of postmenopausal bleeding and any bowel disturbances including fresh or altered blood in the stool and frank hematuria. No other hematological or biochemical abnormality was detected except severely low hemoglobin (6 g/dl). Occult blood in stool and red blood cells in urine were negative.

Ultrasonography revealed left-sided hydronephrosis. All other visceral organs including the right kidney were normal. Intravenous pyelogram showed opacity in the region of left kidney, causing mass effect on the left pelvi-calyceal system. Contrast enhanced computed tomography scan (CT scan) of the kidney, ureters and urinary bladder (KUB) region showed a solid, enhancing soft density mass with focal necrosis, measuring 9 × 7.5 cm, inseparable from the medial wall of the left kidney, which was displaced laterally. The lesion was causing extrinsic pressure effect on the pelvi-calyceal system

and ureter, resulting in an upward/outward displacement and dilatation [Figure 1a]. Urinary bladder was normal. A possibility of retroperitoneal sarcoma was considered on the CT scan. An exploratory laparotomy revealed a mass in left kidney. Left radical nephrectomy was done. It showed an enlarged kidney measuring 17 × 12 × 8 cm. The outer surface was smooth, bosselated and congested with adherent capsule. Cut section revealed a well-circumscribed, lobulated, grayish white, firm mass measuring 10 × 9 × 8 cm in the lower pole, reaching up to the capsule and displacing the pelvis. It had a characteristic whorled appearance [Figure 1b]. Focal areas of necrosis and myxoid degeneration were also noted. Rest of the kidney showed mild dilatation of the calyces, which was otherwise normal.

Microscopically, a well-demarcated, circumscribed and lobulated tumor, composed predominantly of spindle cells, arranged in bundles, interlacing fascicles and whorls [Figure 1c and d] was seen. The tumor cells had moderate, eosinophilic cytoplasm and oval vesicular nuclei showing moderate pleomorphism. Few bizarre and multinucleated forms were interspersed, especially around the necrotic foci. Mitoses were 20–25/10 HPF. Large areas of hemorrhage and necrosis (about 20% of total tumor) were present. Foci of mucinous and hyaline degeneration were also observed. No capsular breach or infiltration into peri-nephric fat was seen. No epithelial area was recognized even after extensive sampling of the tumor. There was no lymphovascular invasion.

Immunohistochemically, the tumor cells showed diffuse and strong positivity with smooth muscle actin (SMA), desmin and vimentin. Epithelial membrane antigen (EMA) was focally positive, while pancytokeratin and high-molecular weight cytokeratin (34β E12) were negative [Figure 2]. High-grade (Grade 3) leiomyosarcoma was diagnosed using French Federation of Cancer Centers System.[5] The patient is doing well 10 months post surgery.

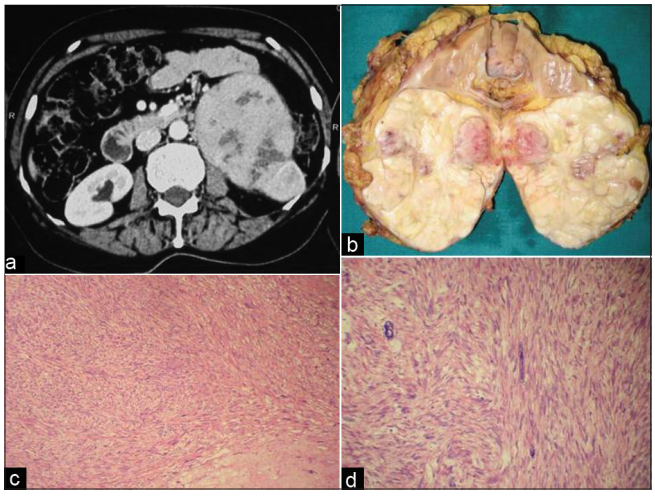

Figure 1: (a) CT scan KUB showing a solid soft tissue density mass inseparable from the medial wall of the left kidney. (b) Gross photograph showing a well-circumscribed, lobulated, grayish white, firm mass with whorled appearance in the lower pole, reaching up to the capsule and displacing the pelvis. (c and d) Photomicrograph of the kidney tumor, composed predominantly of spindle cells with few scattered bizarre cells, arranged in bundles, interlacing fascicles and whorls

DISCUSSION

Leiomyosarcomas are represented primarily as case reports or as components of larger series of renal sarcomas in the literature. They constitute 0.12% of all invasive renal malignancies, as per a study conducted by Kendal.[6] There are only two relatively larger studies of renal leiomyosarcoma, one comprising 10 cases[7] and the other of 27 cases.[8] Both the studies involved cases collected from three large institutions each, over a period of 21 and 23 years, respectively. Miller *et al.*[8] studied only H and E sections without using immunohistochemistry. Hence, effectively, there is only one case series by Deyrup *et al.*, where there was a complete analysis of cases.[7]

Histologically, leiomyosarcoma of kidney has to be differentiated from sarcomatoid renal cell carcinoma, leiomyoma, and angiomyolipoma. Differentiating leiomyosarcoma from leiomyoma is not difficult as mitoses and necrosis are present only in malignant tumor, though cellular pleomorphism can be seen in both. In renal angiomyolipoma, fascicles of smooth muscle cells are admixed with mature fat and thick-walled blood vessels.

Sarcomatoid carcinomas lack the typical alternating fascicles and cytologic features of smooth muscle cells and are composed predominantly of pleomorphic cells, while leiomyosarcoma generally has monomorphic nuclei, although some cases may show nuclear pleomorphism. Diagnosis of sarcomatoid renal cell carcinoma is easier if typical renal cell carcinoma is seen

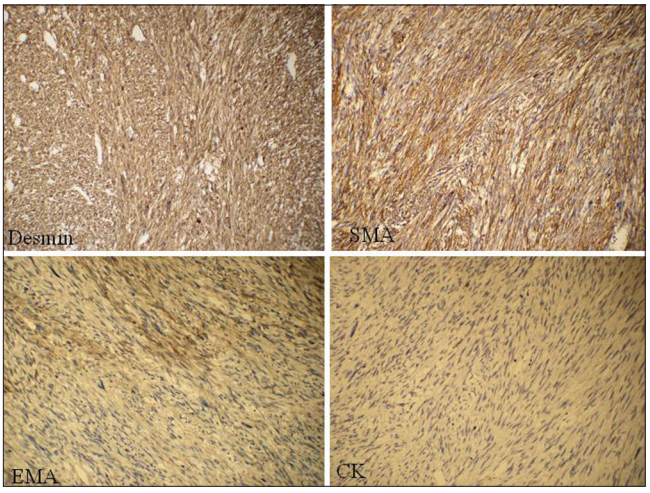

Figure 2: Immunohistochemistry showing tumor cells with diffuse and strong positivity with smooth muscle actin (SMA) and desmin. Epithelial membrane antigen (EMA) is focally positive while cytokeratin (CK) is negative

somewhere within the tumor. Cytokeratin positivity with absence of myoid markers supports a diagnosis of sarcomatoid renal cell carcinoma. Sarcomatoid carcinomas, however, are not uniformly positive for cytokeratins and may express SMA. Some leiomyosarcomas may also express cytokeratin and or EMA.[9,10] In the latter situation, the presence of desmin is diagnostically helpful since this is positive in leiomyosarcoma and not in sarcomatoid carcinoma.[11] Our case had immunohistochemical profiles of smooth muscle differentiation, namely, SMA and desmin.

It is important to grade the tumor as it has great prognostic implications. In the series of Deyrup *et al*.,[7] one patient with a grade 1 tumor was alive with no evidence of the disease, while all three patients with grade 3 tumors died of disease. Outcomes in the seven patients with grade 2 disease were distributed between these two extremes in median 36 months of follow-up. Radical nephrectomy is the primary treatment, but in view of the aggressiveness of this malignancy, triple therapy (surgery, chemotherapy, and radiotherapy) has been advocated.

In conclusion, leiomyosarcoma of the kidney is a rare tumor, which has been ascribed particularly poor prognosis compared to other subtypes of renal malignancy. Frequent immunoreactivity for epithelial markers in leiomyosarcoma and occasional diffuse and strong immunopositivity should be recognized as a potentially serious diagnostic pitfall in the differential diagnosis of leiomyosarcoma and other malignant spindle cell neoplasms. A thorough morphological analysis and careful interpretation of immunohistochemical markers are necessary to arrive at the correct diagnosis for proper management.

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