INTRODUCTION
Primary sarcomas constitute from 0.8% to 2.7% of renal tumours in adults. Of these, 47-60% are leiomyosarcomas, 15% liposarcomas, 9% haemangio-pericytomas, 7% fibrosarcomas, 6% malignant fibrous histiocytomas, and 5% rhabdomyosarcomas. There is no difference in the gender distribution with a mean age at diagnosis in the sixth decade. They typically present with the classical triad of symptoms i.e. flank pain, hematuria, and abdominal mass mimicking renal cell carcinoma. Radiological findings are non-specific and diagnosis is usually made postoperatively. The etiology of renal leiomyosarcoma remains obscure.

We herein, report a case of renal leiomyosarcoma with a follow-up of 36 months.

CASE REPORT
A 57 years old female presented in March, 2007 with complain of left flank pain, hematuria and abdominal mass. On clinical examination, patient was haemodynamically stable, pale looking, with a bimanually palpable left flank mass, extending to right hypochondrium. Initial laboratory workup showed low hemoglobin of 6.1 gm/dl (normal range being 11.5-15.5 gm/dl). Urinalysis showed microscopic hematuria and serum creatinine level was 1.8 mg/dL. Computed tomographic scan of abdomen (Figure 1) showed a large heterogeneous mass measuring 23 x 22 x 13 cm, occupying the left perinephric space with minimal post-contrast enhancement. Rest of the abdominal cavity was unremarkable. Skeletal scintigraphic findings were negative for metastatic involvement of the skeleton. A left radical nephrectomy was performed with removal of left adrenal gland and adequate perinephric fat.

Left radical nephrectomy specimen with left adrenal and perinephric fat was received in 10% buffered formalin and processed routinely. On gross inspection, it was markedly distorted, hugely enlarged kidney measuring 25 x 23 x 13 cm and weighing 3.8 kg. Ureter was identified measuring 5.8 x 1 cm that appeared tumour free. On sectioning, most of the renal parenchyma was replaced by the tumour measuring 23 x 22 x 13 cm having a grayish white firm mucoidy cut surface with foci of necrosis. Scattered areas exhibit gelatinous cut surface. Focally residual renal parenchyma was identified with an incidental solitary cyst measuring 4 x 2 cm. The capsule was intact and perinephric fat was grossly free of tumour. “Adrenal gland”, sent in a separate container measured 5.2 x 2.3 x 1 cm that was grossly unremarkable.

Sections from tumour revealed a cellular neoplasm composed of spindle to markedly pleomorphic cells. In spindle cell areas, the cells were arranged in fascicular array and interlacing bundles whereas pleomorphic cells were arranged in diffuse sheets. The pleomorphic areas exhibited a number of bizarre atypical cells with numerous multinucleated tumour giant cells. In these areas, there was marked nuclear pleomorphism and hyperchromasia with prominent nucleoli (Figure 2). Brisk mitotic activity was appreciated with numerous atypical mitotic figures. Foci of necrosis with intervening hyalinized areas were also noted. Capsule was intact and perinephric fat and ureteric resection margin were tumour-free. There was no renal vein invasion and adrenal gland was also tumour-free.

Immunohistochemistry was performed on the formalin-fixed, paraffin embedded tissue using the streptavidin-
biotin complex with diaminobenzidine (DAB) as chromogen. Following antibodies were used after microwave antigen retrieval: Cytokeratin AE1/AE3 (CK AE1/AE3, dako, Germany, prediluted), Low Molecular Weight Cytokeratin Cam 5.2 (CKCAM5.2, Becton-Dickinson, USA, prediluted), vimentin (dako, Germany, 5:1000), smooth muscle actin (ASMA, dako, Germany, 5:1000), Desmin (dako, Germany, 10:1000) and S-100 protein (dako, Germany, 5:1000).

On immunohistochemical workup, tumour cells were diffusely and strongly positive with smooth muscle Actin and Vimentin while Desmin showed focal positivity. Cytokeratin AE1/ AE3, Cytokeratin Cam 5.2 and S-100 protein were negative (Figure 3). Postoperative course was uneventful and patient was discharged on 10th postoperative day. Neither neo-adjuvant chemotherapy nor radiotherapy was given to the patient. The patient was alive and well 36 months after the operation (dated 10-02-2010) and no evidence of local recurrence or distant metastasis was observed during follow-up with computed tomographic scanning.

**DISCUSSION**

Leiomyosarcoma of the kidney has preponderance in women and is more frequent in fourth decade of life but can be found in almost any age group, with a gradually increasing incidence in the later period of life. Histogenesis remains obscure, as renal sarcomas may arise from the smooth muscle fibers of renal parenchyma, renal capsule, renal pelvis or renal vessels. Neither ultrasonography, tomography nor magnetic resonance are able to differentiate between leiomyosarcomas and renal cell carcinomas. They present as solid or cystic masses. The most common symptoms and signs are pain, palpable mass and hematuria, all of which are indicators of an extensive local diseases. Sarcomas of the kidney are very uncommon and are usually found as incidental tumours. Few renal leiomyosarcomas have presented as a spontaneously ruptured renal tumour. A case of renal leiomyosarcoma with renal vein thrombosis is on record. Rare cases with small sized tumour were treated with partial nephrectomy. Renal leiomyosarcomas have a very poor prognosis with most patients dying within 2 years. It has been difficult to evaluate the true overall survival rate, as most reports do not have long follow-ups. Unfortunately, no role of postoperative chemotherapy or radiotherapy has been determined.

**REFERENCES**


