

Case Report

Primary Leiomyosarcoma of Kidney in a Young Male Treated with Partial Nephrectomy - A Case Report

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Abstract

Primary sarcoma of kidney is very rare. Leiomyosarcoma is the most common histological subtype among all renal sarcomas. We report a rare case of primary Leiomyosarcoma of kidney in a young male. A 39 year old male presented to our hospital with a lump in left abdomen since 1 month. CT abdomen showed a heterogeneous mass arising from lower pole of left kidney. A provisional diagnosis of renal cell carcinoma was made and patient underwent partial nephrectomy. Gross examination showed a grayish white, firm, encapsulated mass with whorled appearance on cut section. Microscopic examination showed pleomorphic spindle cells arranged in interlacing pattern with high mitotic activity. The smooth muscle origin of the cells was confirmed by immunohistochemical positivity for smooth muscle actin and desmin; and negative for cytokeratin, HMB 45, CD117, and CD34.

Introduction

Primary sarcomas of kidney are very rare accounting for only 1-2 percent of all malignant tumors of kidney [1]. Leiomyosarcoma, fibrosarcoma liposarcoma are the most common malignant mesenchymal tumors of kidney. Leiomyosarcoma is the most common histological subtype accounting 60-70% of all sarcomas of kidney. Commonly it is seen in the 6th decade with clinical features of abdominal mass, pain and haematuria. Here we present a case of renal Leiomyosarcoma in a young adult who presented with abdominal lump since one month [2].

Case Presentation

A 39 year old male presented with lump in left abdomen since one month. He had no history of pain, haematuria, or any bowel disturbances.

On examination, vitals were within normal limits. Physical examination revealed a firm mass in the left hypochondrium extending up to left iliac, left lumbar, and umbilical region.

Laboratory investigations revealed normocytic hypochromic anemia with a raised ESR (ESR 56 mm at the end of 1 hour). Serum chemistry levels (renal and liver function tests) were normal. Urinalysis showed 2-5 polymorph nuclear cells per high power field without any evidence of microscopic haematuria.

Abdominal CT scan done showed a well defined exophytic, heterogeneously enhancing mass measuring 15x13x10cm arising from lower pole of left kidney reported most likely as renal cell carcinoma (Figure 1).

Intraoperative frozen section was done and was report as malignant spindle cell neoplasm of left kidney. Radical nephrectomy was done.

On gross examination the tumor was well circumscribed, encapsulated mass measuring 16x 14x 12cm. Cut section was firm

gray white with whirled appearance (Figure 2). Areas of necrosis were seen.

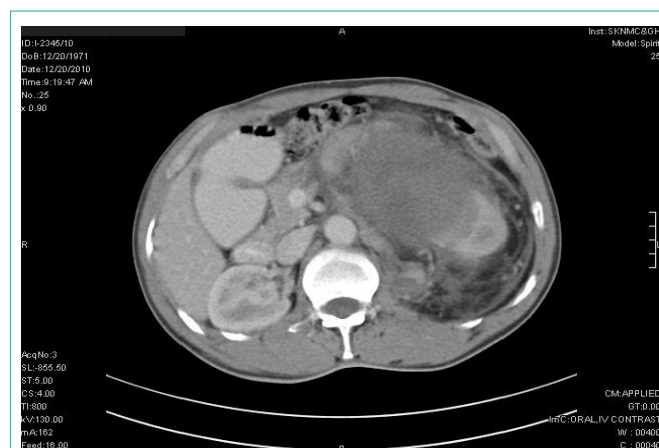


Figure 1: Axial CT scan showing mass arising from left kidney.



Figure 2: Show nephrectomy specimen showing well encapsulated tumor. Cut surface is grayish white with whorled appearance.

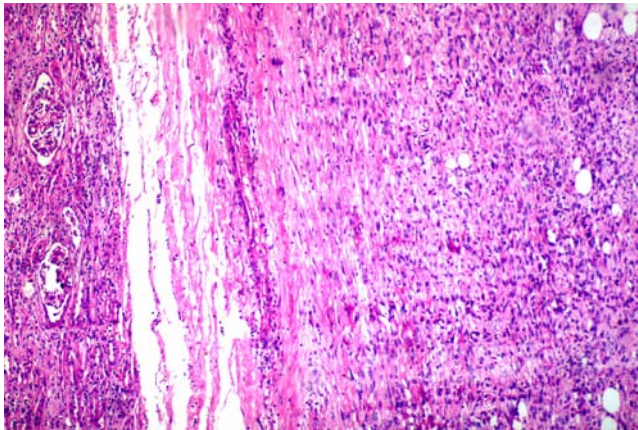


Figure 3: Shows normal kidney and a well encapsulated tumor composed of interlacing fascicles and bundles of spindle shaped cells.

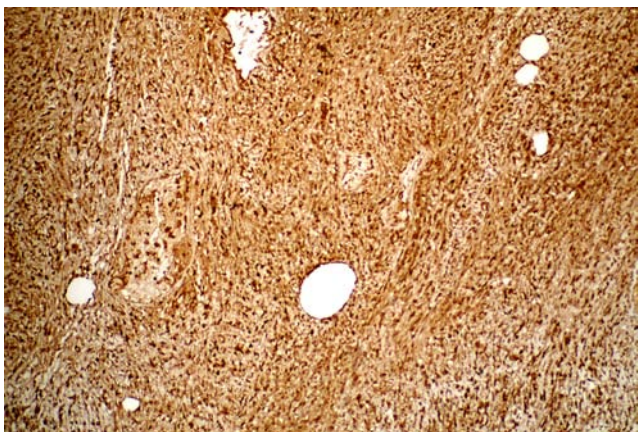


Figure 4: Shows tumor cells are positive for SMA.

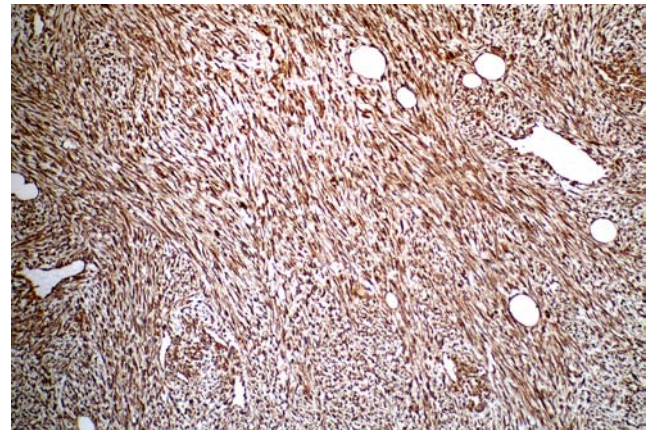


Figure 5: Shows tumor cells are positive for Desmin.

Microscopic examination showed well circumscribed tumor composed of bundles and whorls of smooth muscle cells with elongated hyperchromatic, pleomorphic nuclei (Figure 3). Mitotic activity was 5 -7/10 high power field. Large areas of necrosis were also seen. The cut margin was negative. According to French Federation of Cancer Centers Sarcoma Group the tumor was graded as grade 2. Immunohistochemistry showed tumor cells with diffuse cytoplasmic positivity for smooth muscle actin (SMA, Figure 4) and desmin (Des, Figure 5). They were negative for immunostaining for cytokeratin, HMB 45, CD 117 and CD 34. There was no distant metastasis. As cut margin was negative and metastasis was absent chemotherapy or radiotherapy was not given to the patient. Patient was alive after one year follow up without any evidence of recurrence.

Discussion

Leiomyosarcoma is a malignant tumor of smooth muscle origin. It is more commonly seen in adults and elderly people but rarely has also been reported in children [3]. The common sites include uterus, retroperitoneum, extremities and head and neck region [4]. Leiomyosarcomas of kidney are very rare and represent only 1 to 2% of all malignant tumors of kidney [1]. In a study conducted by Kendal they constitute 0.12% of all tumors of kidney [5]. They appear to arise from renal capsule or smooth muscle tissue of the vessels (i.e. vascular

Leiomyosarcoma) or renal pelvic wall. The mean age at presentation is 50–60 years with a female preponderance. Miller et al. studied 27 cases from three different institutes, the mean age for men was 58.5 and for females it was 59 year [6]. Deyrup et al. studied 9 cases from three institutes, 3 male and 7 females in the age group of 40 to 75 years [7]. Our patient was a 39 year old young male. Leiomyosarcoma usually present with flank pain, haematuria (microscopic or macroscopic) and an abdominal mass. The diagnosis is usually made postoperatively as the radiologic findings are nonspecific. Grossly, the tumors look like leiomyomas with a well-circumscribed margin and whorled cut surface. The malignant counterpart, however, appears fleshy and has areas of necrosis, hemorrhage, and cystic degeneration [7]. Microscopically Leiomyosarcoma shows bundles and whorls of smooth muscle cells with blunt end nuclei [8]. The features of malignancy are necrosis, nuclear pleomorphism and increased mitotic activity [9].

Leiomyosarcoma of kidney should be differentiated from sarcomatoid variant of renal cell carcinoma. But the latter has more pleomorphic appearance, foci of typical renal cell carcinoma and also cytokeratin positivity which will not see in case of Leiomyosarcoma [8]. Monophasic variant of synovial sarcoma can mimic Leiomyosarcoma. It shows plump cells arranged in diffuse sheets with irregular borders. They show entrapped renal tubules with BCL-2 positivity [9].

Leiomyosarcomas metastasize to lung, liver and lymph node. Radical nephrectomy is the treatment of choice, but recently partial nephrectomy has been shown to give good results in these patients [10]. Surgery offers the best chance of cure, and the role of adjuvant chemotherapy and / or radiotherapy remains debatable, due to the paucity of data on the treatment of this rare renal neoplasm.

The patient was treated with radical nephrectomy. No evidence of local or distant metastasis was found on 2 years follow up on computerized scanning.

To conclude renal Leiomyosarcoma is rare tumor should be differentiated with sarcomatoid variant of renal cell carcinoma, monophasic synovial sarcoma and radical nephrectomy may provide cure to the patient.

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