

Primary bilateral leiomyosarcoma of kidneys with metastases at unusual sites- a rare case report

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ABSTRACT

Primary Leiomyosarcoma of kidney is rare entity, constituting 0.5-1 % of all invasive renal tumors and bilateral Leiomyosarcoma makes it extremely rare. One such case diagnosed by Ultrasound Sonography Guided Fine Needle Aspiration Cytology is reported here. Needle core biopsy was performed for Immunohistochemical markers . Renal Leiomyosarcomas are difficult to differentiate from Sarcomatoid Renal cell carcinoma, as both tumors have atypical spindle cells. Also it is difficult to differentiate radiologically and clinically. Immunohistochemical markers were done. Smooth Muscle Actin and Vimentin were positive in this tumor, confirming the mesenchymal nature of the tumor. Desmin showed focal positivity. HMB 45, Cytokeratin, EMA were Negative. Since this being a bilateral primary renal leiomyosarcoma with metastases, supposedly the first case with this presentation to be reported, it turned out to be a diagnostic challenge to differentiate from sarcomatoid renal cell carcinoma. Prognosis of renal sarcoma is poor.

Key words: Leiomyosarcoma, Sarcomatoid Renal cell carcinoma

Introduction

Leiomyosarcomas are malignant aggressive tumors with high potential of metastasis usually originating from the soft tissues and smooth muscle of the uterus. Renal leiomyosarcoma may originate from the smooth muscle fibres of renal parenchyma, renal pelvis, calyces, renal capsule and renal vessels but their most common site of origin is smooth muscle of the renal veins.[1] Primary sarcomas constitute 1-2 % of all renal tumors in adults, out of which 50-60 % of renal sarcomas are leiomyosarcomas. It is very difficult to differentiate them from other renal masses prior to operation.[2]. 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. Imaging may not be able to differentiate between leiomyosarcoma

and Renal cell carcinoma. Renal leiomyosarcomas has a more fatal prognosis and they reach big sizes because of their mesenchymal components and they are devoid of barriers like those in other types of tumor.

Case report

A 32 year female presented with mild abdominal pain and headache. No irregularity in menstrual history was complained. Ultrasound abdomen revealed bilateral renal masses with variegated echotexture (solid and cystic areas). Inferior vena cava showed a huge thrombus. Solid lesions were seen in liver, posterior uterine wall, left ovarian pedicle. Soft tissue lesions (seven) in scalp in no. measuring 1x1 cm to 2x2 cm, tender, firm and also in right popliteal fossa 2x2 cm lesion were present. Whole body CT scan and MRI revealed solid lesions in right lateral parietal lobe, liver, posterior uterine wall, left ovarian pedicle, right popliteal fossa, scalp, 9 cm long hypodense lesion in medullary cavity of right lower end femur. Chest and lungs were free from obvious metastases. Also there were no abdominal lymph nodes. Fine Needle Aspiration Cytology from Right and Left Kidney were done which revealed sheets of spindle shaped cells showing pleomorphism, cigar shaped nuclei and scanty

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mitotic activity(Fig.1) FNAC from other sites like liver , ovary, uterine wall, scalp, right popliteal fossa, revealed the same cytomorphology as that of the kidneys.Other biochemical investigations like renal

function tests, liver function tests were within normal range. Hemoglobin was slightly reduced to 10.0 g/dl. No blood in urine or stool was seen.Urine Cytology showed presence of tumor cells (Fig. 2).

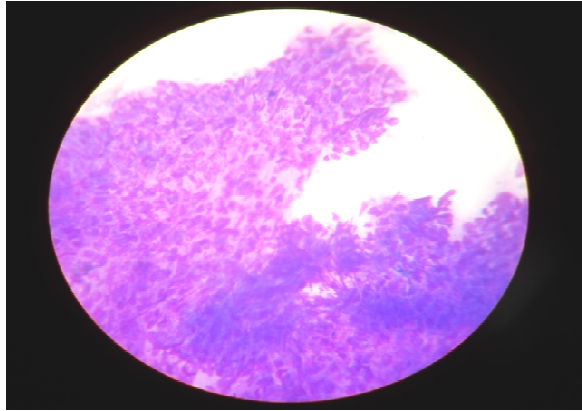


Fig1: Renal Cytology (MGG Stain),40 X

Looking at the widespread pathology, tumor resection was not possible. So , Core needle biopsy was done using Biopsy gun. The histopathology sections revealed normal renal tissue alongwith tumor cells showing pleomorphism, hyperchromatic cigar shaped nucleus, mitotic activity 8-10/HPF [3,4]. Immunohistochemical markers were done to differentiate between Primary Leiomyosarcoma,

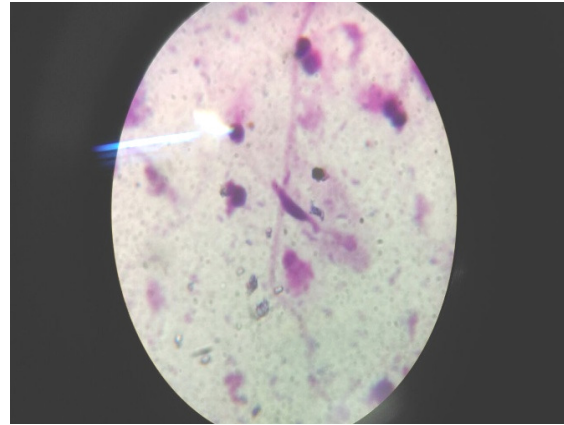


Fig 2:Urine Cytology (MGG Stain), 40X

Sarcomatoid Renal cell carcinoma, Angiomyolipoma and other sarcomas. Smooth Muscle Actin (SMA) (Fig. 4) showed diffuse positivity, Vimentin (Fig 5) also showed positivity confirming the mesenchymal nature of tumor. HMB-45, EMA (Fig. 6) Cytokeratin (Fig 7) were negative , Desmin (Fig 8) was focally positive. No genetic predisposition was detected.

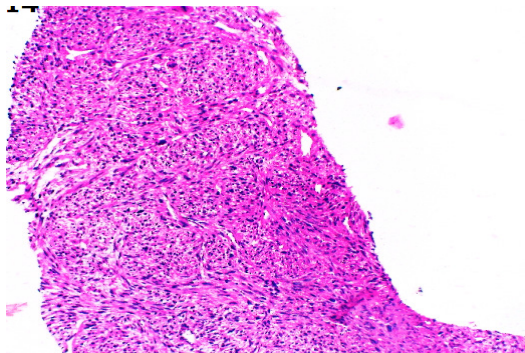


Fig 3:Renal Needle Core Biopsy (H & E),10X

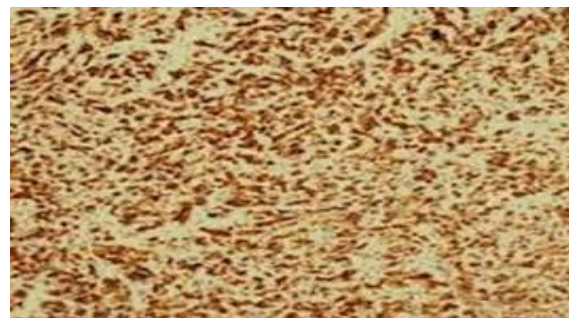


Fig 4:Smooth Muscle Actin (SMA)

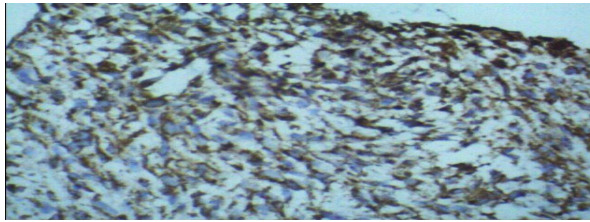


Fig 5: Vimentin

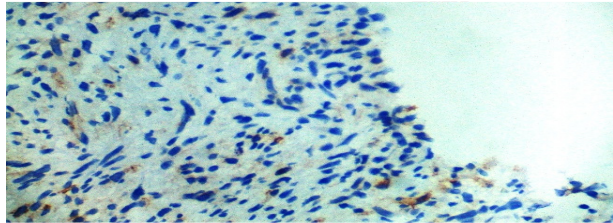


Fig 6:HMB-45, EMA

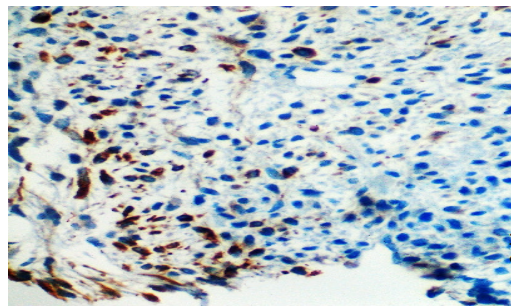


Fig 7:Cytokeratin were negative

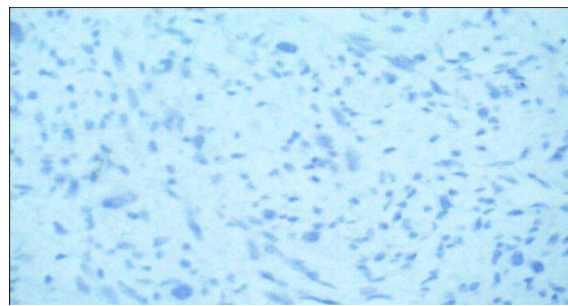


Fig 8: Desmin was focally positive. No genetic predisposition was detected

Discussion

Leiomyosarcomas are represented primarily as case reports or as components of larger series of renal sarcomas in the literature [5,6]. Histologically, Leiomyosarcoma of kidney has to be differentiated from sarcomatoid renal cell carcinoma, leiomyoma, and angiomyolipoma. Cytokeratin positivity with absence of myoid markers supports a diagnosis of Sarcomatoid renal cell carcinoma. Presence of Desmin is diagnostically helpful since this is positive in leiomyosarcoma and not in sarcomatoid carcinoma.[6]

Conclusion

Thus concluding, FNAC is helpful in early diagnosis of such malignancies. It has been confirmed that this is a very rare case of Primary Bilateral Renal Leiomyosarcoma with metastases to unusual sites with poor prognosis. This is supposedly the first case of bilateral primary with no genetic predisposition to be reported. The patient survived for 5 months post diagnosis.

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Conflict of Interest: None

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