African Journal of Urology

CASE REPORTS Open Access

Primary synovial sarcoma of kidney: a rare case report with review of literature

Ravi Lohani[1*], Vishwajeet Singh[1], Atin Singhai[2], Sahil Singla[1] and Manish Kumar Agarwal[1]

Abstract

Background: Synovial sarcoma is a rare type of sarcoma with a predilection for extremities in young adults. Very rarely this tumor can originate from kidney.

Case presentation: A 22-year-old female presented to us with complaints of right flank pain and a large mass in the right side of abdomen for the last 6 months. A provisional diagnosis of renal cell carcinoma was made and right radical nephrectomy was done. However on histopathological examination the tumor was composed of spindle cells arranged in hemangiopericytoma-like vascular pattern. Immunohistochemistry and FISH confirmed the diagnosis of synovial sarcoma of kidney. The patient had no signs of recurrence or metastasis at six months of follow-up.

Conclusion: Renal synovial sarcomas, though rare, should be considered as a differential in young adults who present with extremely large renal mass. Surgical extirpation is the treatment of choice. Ifosfamide- and doxorubicin-based chemotherapy can be given in non resectable and metastatic cases.

Keywords: Primary renal synovial sarcoma, Renal cell carcinoma, Atypical renal mass, Case report

1 Background

Synovial sarcoma is a rare type of sarcoma that accounts for approximately 6–10% of soft tissue sarcomas with a predilection for extremities in young adults [1].Very rarely this tumor can originate from kidney, pleura, lung, mediastinum and ovary [2]. On histology, synovial sarcoma has both epithelial and mesenchymal differentiation. It has three subtypes: monophasic, biphasic and poorly differentiated. This tumor is associated with a characteristic translocation [t(X;18)(p11.2;q11.2)] forming SYT-SSX gene fusion [3]. Owing to its rarity it poses a challenge in its diagnosis and management. We present a case of primary synovial sarcoma of kidney, which was provisionally diagnosed as renal cell carcinoma and managed with radical nephrectomy. However, on histology and immunohistochemistry a diagnosis of renal synovial sarcoma was made.

*Correspondence: ravi_lohani@yahoo.com

[1] Department of Urology, King George's Medical University Lucknow, Lucknow, India
Full list of author information is available at the end of the article

2 Case presentation

A 22-year-old female presented to us with complaints of intermittent right flank pain for last six months. There was no history of hematuria. There was no history of anorexia, significant weight loss, cough, hemoptysis and bone pain. There was no history of similar complaints in any other family members. On physical examination, a large lump involving the right hypochondrium and right lumbar region was palpable.

Ultrasonography whole abdomen revealed a large multi cystic lesion measuring approximately 14.1×8.7 cm in right lumbar region with normal left kidney.

Computed tomography (CT) of whole abdomen with intravenous contrast showed a large solid cystic lesion measuring $13 \times 14 \times 19.1$ cm with thick enhancing septations. No local invasion was seen. The lesion was compressing and displacing the inferior vena cava with no evidence of venous invasion or tumor thrombus (Fig. 1). There were few sub-centimetric mesenteric lymph nodes. The clinical and radiological diagnosis was consistent with renal cell carcinoma.

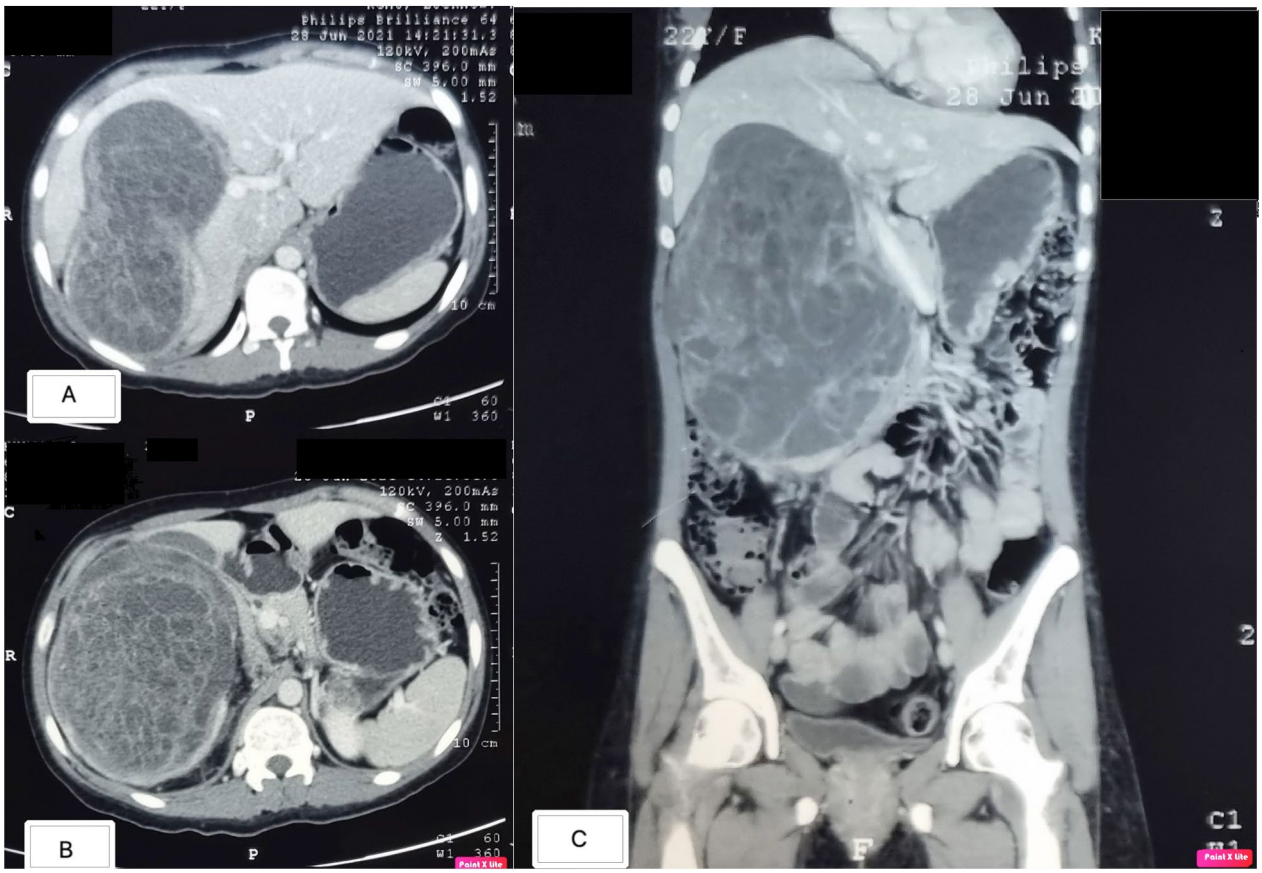

Fig. 1 **A** and **B** Cross section of the abdomen showing a large right renal mass pushing the liver upwards and to the left. **C** Coronal section of the abdomen showing a large right renal mass displacing the liver upwards and to the left. The inferior vena cave is also displaced to the left

Computed tomography of the chest revealed no evidence of metastasis.

2.1 Treatment

Due to the massive size of the tumor, preoperative renal artery angio-embolization was done to facilitate resection of the mass.

Intraoperatively the tumor was found to be well circumscribed without any gross invasion into adjacent structures. Right radical nephrectomy was performed. Grossly the tumor measured 18 × 17 × 7 cm. The cut surface was gray brown with cystic areas (Fig. 2).

Microscopically, the tumor was composed of spindle cells arranged in fascicular pattern. Individual cells were monotonous with vesicular nuclei displaying evenly dispersed chromatin, scant cytoplasm and inconspicuous nucleoli. Tumor cells were arranged around staghorn branching vessels seen in hemangiopericytic pattern. Focal areas of necrosis and mitosis were also seen. Lymphovascular invasion and perineural invasion was absent (Fig. 3).

Immunohistochemistry was positive for Vimentin (diffuse cytoplasmic staining, Bcl-2 and CD99 (showing membranous positivity) (Fig. 4A and B) and negative for CD 34 (Fig. 4C). This was consistent with the diagnosis of synovial sarcoma. Molecular analysis using fluorescent in situ hybridization (FISH) revealed a translocation between SYT gene on chromosome 18 and SSX gene on chromosome X, which further confirmed the diagnosis of primary renal synovial sarcoma.

2.2 Outcome and follow-up

Postoperatively, nasogastric tube was kept for one day. The patient was allowed clear liquids from day 1 when bowel sounds returned and semisolid diet from day 2. Postoperative recovery was uneventful and the patient was discharged after 7 days. We sought opinion from medical oncologist and it was decided to reserve chemotherapy if the patient develops any sign of metastasis or recurrence.

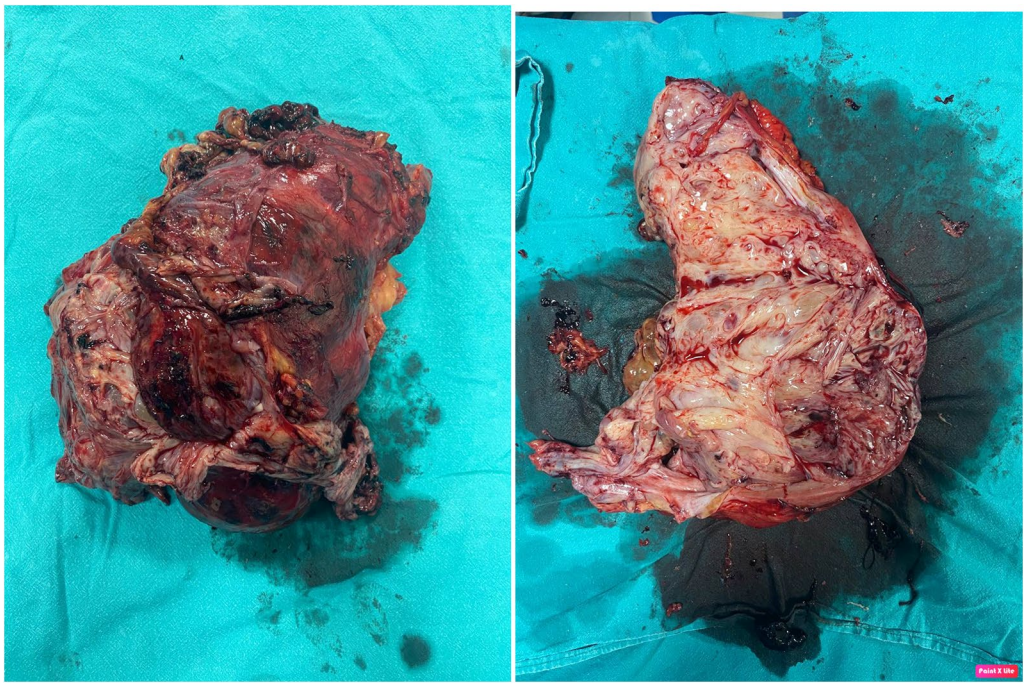

Fig. 2 Gross and cut surface of the renal mass

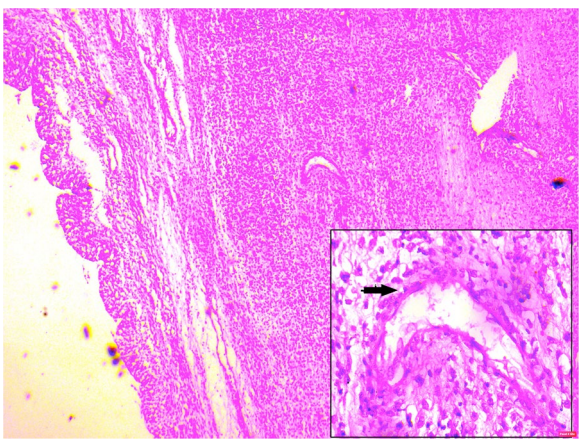

Fig. 3 Histology of the tumor showing monotonous looking stromal cells arranged in a hypercellular fascicular architecture in a hemangiopericytic fashion (H and E × 10). Inset showing arrangement of tumor cells around a blood vessel in hemangiopericytic pattern (arrow) (H and E × 40)

Our patient was doing well and there was no evidence of recurrence or metastasis at 6 months of follow-up MRI whole abdomen (Fig. 4).

3 Discussion

Renal sarcomas account for 1% of malignant renal tumors. Leiomyosarcoma is the most common variant (40–60%) followed by rhabdomyosarcoma, chondrosarcoma, osteosarcoma and liposarcoma [4].

Synovial sarcoma is an exceedingly rare form with very few cases reported in literature [4]. Histologically the tumor is divided into monophasic, biphasic and poorly differentiated variants [5]. The differential diagnosis includes metastatic sarcoma, renal cell carcinoma with sarcomatoid differentiation, adult Wilms' tumor, primary renal primitive neuroendocrine tumors and hemangiopericytoma [6]. Synovial sarcomas usually stain positive for CD99, vimentin, bcl2, EMA and CD 56 while they stain negatively for S100, CK and desmin [7]. It is associated with a very specific chromosomal translocation [t(X; 18) (p11.2; q11.2)], which results in fusion of the SYT gene located on chromosome 18 to SSX gene on chromosome X [3].

Blas et al. in their review of primary renal synovial sarcoma asserted that the mean age of patients was 36.2 years which is half the median age for diagnosis of renal cell carcinoma, with a slight male predominance (1.82:1). The patients with primary renal synovial sarcoma usually present with hematuria (46.3%), either alone or in combination with other symptoms like pain (43%) or palpable mass (37%) which results in common misdiagnosis of renal cell cancer. The most common sites

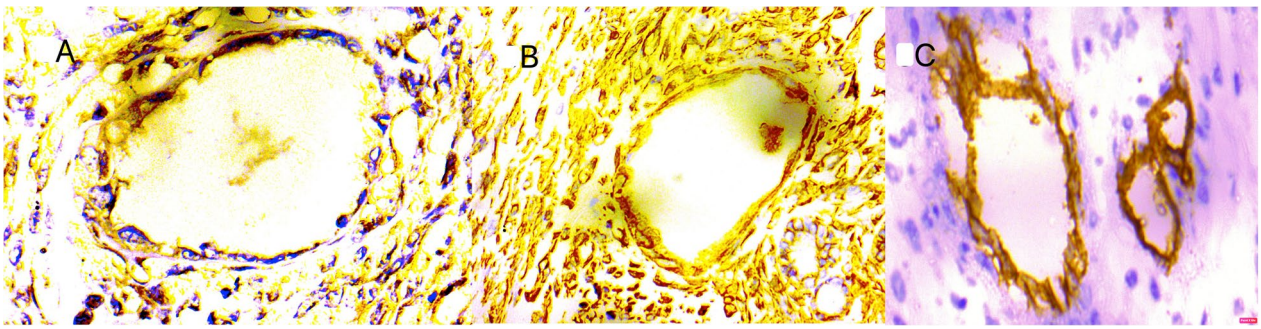

Fig. 4 Immunohistochemistry of tumor cells showing **A** positivity for CD 99 [membranous positivity] **B** diffuse positivity for Vimentin [cytoplasmic positivity] **C** negativity for CD34 [staining showing CD34 negative cells with blood vessel as positive internal control]

of metastasis are lung, liver and bone [8]. There are no definite radiological characteristics which can differentiate renal synovial sarcoma from renal cell cancer preoperatively. Some patients may present very late with caval thrombosis and widespread metastasis (Fig. 5).

Due to the rarity of the tumor, there are no definite guidelines regarding its management. With common consensus surgical extirpation in resectable non metastatic cases is considered the therapy of choice. Few case reports have also described doxorubicin and ifosfamide in the neoadjuvant and adjuvant setting [8]. Chediak et al. reported complete resolution of a metastatic renal synovial sarcoma with surgery and doxorubicin and ifosfamide chemotherapy [2]. Puj et al. described a case of large renal synovial sarcoma in which they used ifosfamide and doxorubicin chemotherapy in the neoadjuvant setting which resulted in down staging of the tumor and subsequent surgical resection possible [9]. We used renal artery embolization in the preoperative setting to facilitate its resection as it was a massive tumor and sub hepatic plane was not clear. There was no evidence of metastasis at the time of diagnosis and at 6 months of follow-up. We decided to keep our patient under close follow-up and to start chemotherapy on the first sign of any metastasis.

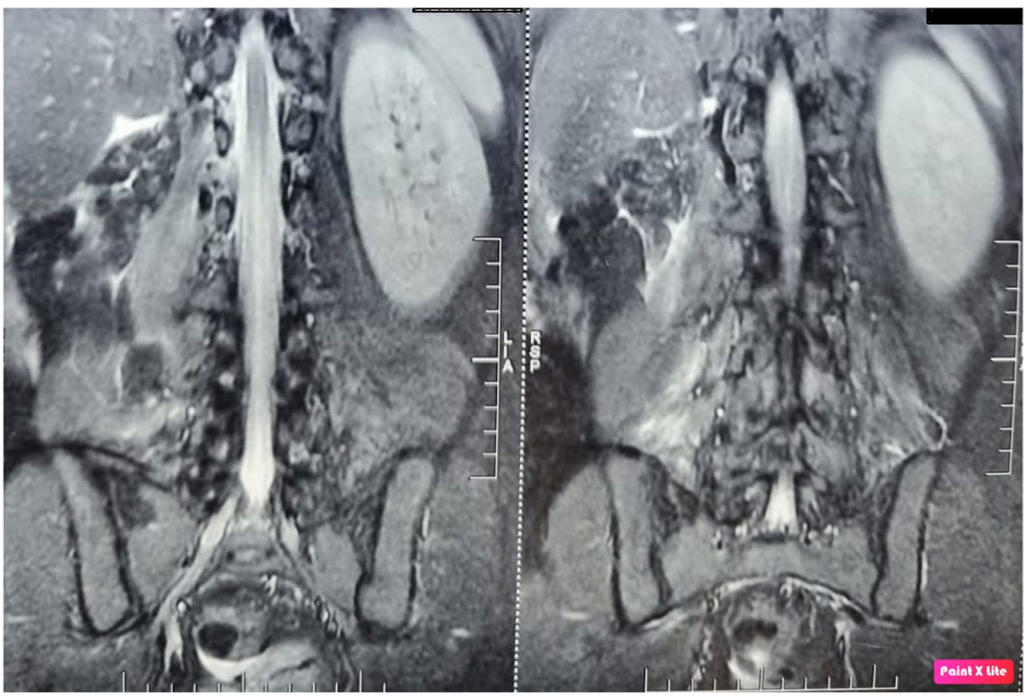

Fig. 5 Follow-up MRI image at 6 months showing no evidence of recurrence

4 Conclusion

Renal synovial sarcomas though rare should be considered as a differential in young adults who present with extremely large renal mass. Preoperative angio-embolization facilitates resection of such large tumors. Surgical extirpation is the treatment of choice. Synovial sarcomas are responsive to ifosfamide- and doxorubicin-based chemotherapy regimens, however treatment with these needs to be individualized.

5 Patient's perspective

I am grateful that the tumor could be resected completely. I have been told that this tumor is rare and have been advised for regular follow-up. I was scared and depressed initially but now I am keeping a positive attitude.

Abbreviations

CD: Cluster differentiation; CK: Cytokeratin; EMA: Epithelial membrane antigen; BCL: B cell Leukemia/lymphoma.

Acknowledgements

I acknowledge the contribution of the operating staff and the anesthetic team as no surgical team can be considered complete without their assistance.

Author contributions

RL was involved in conception and conduct, and acquisition of data. VJS performed analysis and interpretation and reporting. AS reviewed the literature. SS done critical review. All authors have read and approved the manuscript, and they ensure that this is the case.

Funding

The authors declare that this study has received no financial support.

Availability of data and materials

Not applicable.

Declarations

Ethical approval and consent to participate

Ethical approval was taken from Institutional Ethics committee of King George's Medical University, UP, on 10/12/2021.

Consent for publication

Written informed consent for the publication of his clinical history and radiological images has been obtained from the patient herself.

Competing interests

The authors declare no competing interests.

Author details

[1]Department of Urology, King George's Medical University Lucknow, Lucknow, India. [2]Department of Pathology, King George's Medical University Lucknow, Lucknow, India.

Received: 23 December 2021 Accepted: 7 June 2022
Published online: 21 June 2022

References

1. Mishra S, Awasthi N, Hazra SP et al (2015) Primary synovial sarcoma of the kidney. Saudi J Kidney Disc Transpl 26(5):996–999
2. El Chediak A, Mukherji D, Temraz S et al (2018) Primary synovial sarcoma of the kidney: a case report of complete pathological response at a Lebanese tertiary care center. BMC Urol 18:40
3. Krishna Moorthy H, Pillai BS, Varghese J (2014) Primary renal synovial sarcoma: an oncologic surprise. Urol Case Rep 2(5):152–153
4. Huned D, Kam JH, Lee LS et al (2021) Primary renal synovial sarcoma presenting with a retroperitoneal bleed. BMJ Case Rep 14:e237099
5. Bei Z, Chao A, Yanjiao Z et al (2020) Primary renal synovial sarcoma. Medicine 99(42):e22706
6. Dutt UK, Manikandan R, Dorairajan LN et al (2018) Biphasic renal synovial sarcoma with extensive venous tutor thrombosis: a rare presentation. Uro Ann 10:339–341
7. Yang L, Wang K, Hong L et al (2012) The value of immunohistochemistry in diagnosing primary renal synovial sarcoma: a case report and literature review. Int Surg 97:177–181
8. Blas L, Robert J (2021) Primary renal synovial sarcoma and clinical and pathological findings: a systematic review. Curr Urol Rep 22:25
9. Puj KS, Pandya SJ, Warikoo V et al (2018) Primary synovial sarcoma of the kidney: a rare presentation. Urology 116:e7–e8

Publisher's Note