CASE REPORT

Primary Synovial Sarcoma of the Kidney with Inferior Vena Caval Thrombus

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Abstract Synovial sarcomas occurs primarily in the soft tissues, mostly in para-articular regions of the extremities. These tumors have been described in other unusual locations like the pleura, lungs, mediastinum, and kidneys. Primary synovial sarcoma of the kidney is a recently described entity; to date, a total of approximately 35 cases have been reported. Only three cases of primary synovial sarcoma of kidney with IVC thrombus have been reported so far. Here we present the case of a 44 year old gentleman, with primary synovial sarcoma of the left kidney with caval thrombus.

Keywords Synovial sarcoma · Renal · Tumors · Kidney · IVC · Thrombus

Case Summary

A 44 year old gentleman presented with an incidentally detected left renal mass during routine evaluation. On examination, he had a 7*7 cm mass palpable in the left hypochondrium, which was ballotable. CT and MRI scan of abdomen done elsewhere, revealed a 9*9 cm mass lesion involving the upper pole of the left kidney with associated left renal vein thrombosis. Doppler ultrasound at presentation revealed left renal vein thrombosis with no evidence of Inferior

vena cava (IVC) thrombosis. His renal parameters were deranged with blood urea 40 mg/ml and serum creatinine 1.7 mg. Renogram revealed glomerular filtration rate of 17.8 ml/min on the left side and 30.7 ml/min on the right side (Figs. 1, 2, 3 and 4).

PET CT scan (done without IV contrast in view deranged renal functions) showed a tumor in the upper pole of the left kidney with tumor thrombus in the left renal vein with a SUV of 9.2. He underwent left radical nephrectomy. Intraoperatively, the thrombus was extending into the infrahepatic inferior vena cava and left lumbar vein. The IVC was mobilised, both renal veins were exposed, and slings were passed around these structures for control, taking care not to dislodge the thrombus. Incision performed at the ostium of the tumoral renal vein and the thrombus was completely removed under direct vision.

Grossly there was an ill defined white to brown necrotic tumour 10*8*7 cm, involving almost the entire upper pole and with extension into the midpole. Renal vein was dilated with thrombus and the tumor was covered by a thin capsule with perirenal fat and fascia.

Microscopy revealed mult-nodular tumor composed of sheets and intersecting bundles of spindle cells with elongated and pleomorphic vesicular nuclei. Microscopic foci of mucinous area was noted. Mitosis was 2 per high power field. Tumor thrombus was adherant to the lumen of the blood vessel. Lymph nodes around the ureter and para aortic nodes were free of tumour. Overall features were suggestive of malignant spindle cell tumor, high grade.

Immunohistochemistry study showed strong positive reaction for Vimentin, EMA, CD 99 and bcl-2. 40 % cells showed strong nuclear positivity for KI 67. The tumor cells showed negative uptake for CD10 and Keratin. Histology along with Immunohistochemistry correlation favored Synovial sarcoma, High grade.

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Fig. 1 Tumour in the upper pole of Left kidney with tumour thrombus (Arrow)

Our patient was not offered adjuvant chemotherapy in view of his deranged renal functions postoperatively. He developed pulmonary metastasis and expired with a disease free survival of 24 months.

Discussion

Primary synovial cell sarcoma of kidney is a rare entity. It was first reported in 1999 by Faria et al. [1]. Synovial sarcoma accounts for about 8 % of all soft tissue tumors. The peak incidence is in the third decade and

Fig. 2 Immunohistochemistry of tumour cells with strong CD 99 positivity

males are more commonly affected than the females (1.2:1). Synovial sarcomas are morphologically and genetically distinct characterized by the chromosomal translocation $t(X;18)(p11;q11)$ [2]. Typical of soft tissue origin, they have also been described in other sites such as the kidney, lung and pleura.

Based on morphological appearance, synovial sarcoma has been classified into 1) Monophasic spindle cell 2) Biphasic spindle cell and 3) Poorly differentiated variants. The poorly differentiated synovial sarcoma (PDSS) comprises approximately 20 % of cases and shows poor prognosis. Primary renal sarcomas are rare neoplasms that account for 1 % of malignant renal tumors. Leiomyosarcoma is the most frequent type comprising 40–60 %, followed by rhabdomyosarcoma, chondrosarcoma, osteosarcoma, liposarcoma, angiosarcoma and hemangiopericytoma [3]. Pathological features and immunohistochemistry help arrive at a diagnosis.

Immunohistochemistry studies of primary renal SS cases have consistently shown positivity for Bcl2, CD99/Mic2,

Fig. 3 MRI Abdomen revealing tumour thrombus in left renal vein

Fig. 4 MRI revealing thrombus in left renal vein

CD56, Vimentin and focal positivity for EMA. They do not stain for desmin, actin, CD34, and CD31 [4]. Reverse transcriptase-polymerase chain reaction (RT-PCR) analysis of SS shows a unique chromosomal translocation t(X;18) (p11.2;q11.2) that results in the fusion of *SYT* gene on chromosome 18 with *SSX* family gene on chromosome X and helps to confirm the diagnosis [5]. Although five different variants of the *SSX* gene have been identified, only *SSX1* and *SSX2* have been shown to fuse with the *SYT* gene to create the biphasic and monophasic forms of PSS, respectively.

Conclusion

Primary synovial sarcomas of kidney have shown to have aggressive clinical course and poor outcomes [6, 7]. There are no guidelines for the treatment of primary synovial sarcoma of kidney due to the rarity of cases. Surgical management forms the mainstay of treatment. Although there is no proven benefit of adjuvant chemotherapy, primary synovial sarcoma of kidney may be sensitive to high dose Ifosfamide and Adriamycin based regimens [8].

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