



Primary Synovial Sarcoma Kidney: A Rare Case Report

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Abstract

Primary synovial sarcoma (PSS) of the kidney is a rare entity. Since morphologic differentiation from other tumors like metastatic sarcoma, sarcomatoid renal cell carcinoma, retroperitoneal liposarcoma may be difficult, other diagnostic modalities like immunohistochemistry, cytogenetic & advanced molecular analyses need to be employed.

Despite its rarity & non specific presentation, pathologist and clinicians should consider synovial sarcoma in the differential diagnosis of renal masses for the proper management of patients, especially when histopathological examination shows malignant spindle cell neoplasm of kidney.

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Introduction

Common site for Synovial sarcoma (SS) is proximal lower limb of young adults. Uncommonly it can occur in other locations like pleura, lung, mediastinum, and kidney. In kidney it can also occur as a primary or can be metastasis from other site. Primary synovial sarcoma (PSS) of the kidney is a rare entity; to date, fewer than 60 cases have been reported. The first reported case of primary synovial sarcoma of the kidney was described in 1999. This tumor poses a diagnostic dilemma because it is quite difficult to differentiate it from metastatic sarcoma, sarcomatoid renal cell carcinoma, retroperitoneal liposarcoma, and hemangiopericytoma which may have similar histological features. They are specifically associated with a unique chromosomal translocation (t(X;18)(p11.2;q11.2) that results in the fusion of SYT gene on chromosome 18 with an SSX family gene on chromosome X (SSX1, SSX2, or SSX4). Despite its rarity & non specific presentation, pathologist should consider synovial sarcoma in the differential diagnosis of renal masses, especially when HPE shows malignant spindle cell neoplasm of kidney. Since morphologic differentiation from other tumors may be difficult, other diagnostic modalities like IHC, cytogenetic & advanced molecular analyses need to be employed.

Case Report

A 35-year-old male presented with mild to moderate continuous pain in right lumbar area. There was a lump in the right lumbar region since 3 months. There was no significant previous medical history. On examination, right kidney was palpable bimanually and ballotable. On computed tomography (CT) scan, large vascular mass seen in right kidney, measuring 14.1×12.5×10.5 cm invading, right renal vein, compressing IVC, head of pancreas and right lobe of liver, suggestive as renal cell carcinoma. Patient subsequently underwent trucut biopsy from right lumbar/hypochondriac area, which showed infiltration of renal parenchyma by sheets of malignant cells having marked pleomorphism comprising of oval cells, round cells and spindle shaped cells having ill defined cytoplasm. Impression was 1) Sarcomatoid Carcinoma and 2) Spindle Cell Sarcoma. Immunohistochemistry (IHC) was advised and done outside. On IHC tumor cells expressed EMA, MIC-2, bcl-2, Calponin & CD 56 & were immunonegative for Cytokeratin. Features were in favour of synovial sarcoma. Then nephrectomy was done.

Gross findings: Right radical nephrectomy specimen measured 685 grams & 13×11×10cm. Capsule was ruptured. Cut section showed a tumor mass measures 10×10×8 cm having grey firm cut surface. Normal kidney tissue compressed at the periphery measured 3×1×1cm. Ureter was 2cm in length. (Fig 1, 2)

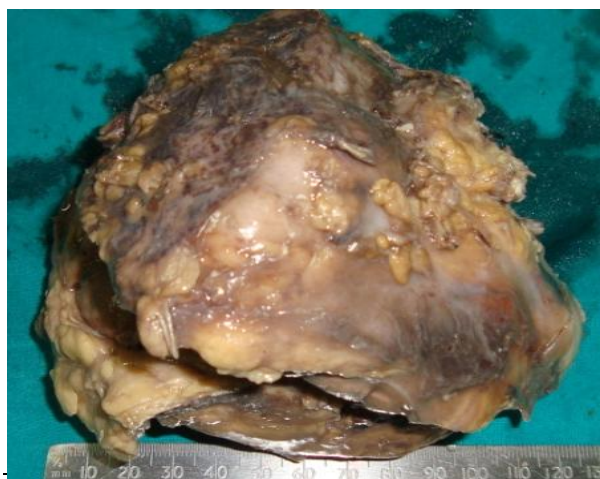


Figure 1: Gross specimen showing renal mass measuring 13×12×10cm. with capsular breach.



Figure 2: Gross specimen of kidney measures tumor mass 10×10×8 cm compressing the normal kidney tissue at periphery.

Microscopic findings: Malignant mesenchymal neoplasm involving right kidney parenchyma, Gerota's fascia, perinephric fat. The histological appearance and immunoprofile was suggestive of primary synovial sarcoma of kidney. (Fig 3, 4) Cytogenetic study done outside and showed chromosomal translocation (t(X;18)), which confirmed the diagnosis of primary synovial sarcoma kidney.

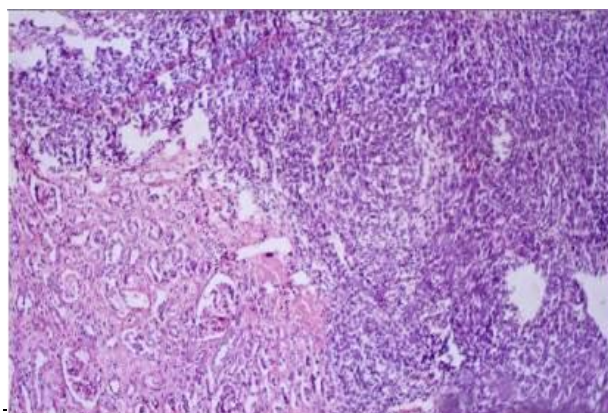


Figure 3: H&E section under 10x shows cellular lesion composed of fascicles of plump spindle cells with hyperchromatic elongated nuclei and indistinct cytoplasm. Normal kidney tissue also seen at the periphery.

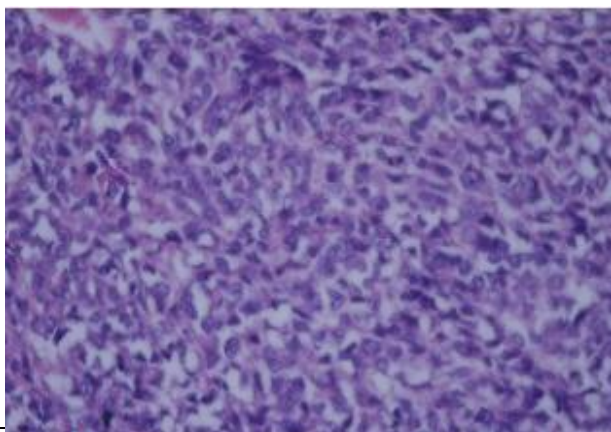


FIGURE 4: H&E section under 40x showing pleomorphic ovoid to spindle cells.

Discussion

Different types of soft tissue sarcoma which can arise in kidney are leiomyosarcoma, rhabdomyosarcoma, liposarcoma, fibrosarcoma, angiosarcoma, hemangiopericytoma and malignant fibrous histiocytoma.^{1,2} leiomyosarcoma is the most frequent type followed by others.^{3,4}

Primary renal synovial sarcoma is a rare entity and the diagnosis is difficult due to rarity of the tumor and its similar presentations as compared to other renal tumors.^{5,6}

Histologically, it is subclassified into biphasic (BSS), monophasic spindle cell (MSSS) and poorly differentiated variants. Amongst these, poorly differentiated synovial sarcoma (PDSS) comprises approximately 20% of cases and shows poorest prognosis.^{7,8,9}

In adults, the most important differential diagnoses include sarcomatoid renal cell carcinoma, adult primitive neuroectodermal tumor (PNET), adult Wilms' tumor, and fibrosarcoma. IHC is useful in differentiating malignancies of spindle cell morphology. Positivity for CK, EMA, BCL-2, and CD99 suggest the possibility of synovial sarcoma. In the study of Wang et al. IHC analysis of 4 cases of primary renal sarcomas revealed positivity for vimentin (4/4), Bcl-2 (4/4), CD99 (4/4), CD56 (3/4), and focally for EMA (3/4) and cytokeratin (3/4).¹⁰

Although IHC markers are highly suggestive but not specific enough to make a firm diagnosis. A characteristic and consistent translocation, t (X; 18) (p11; q11) that results in the fusion of SYT gene on chromosome 18 with SSX family gene on chromosome X and helps to confirm the diagnosis can be identified by conventional cytogenetics (using chromosomal banding studies) or molecular techniques.¹¹

Biphasic synovial cell sarcoma is usually associated with SYT-SSX1, while monophasic synovial cell sarcoma may be associated with either transcript.¹² The

former has a higher rate of proliferation and some reports (but not all) have suggested a poorer outcome.¹³

Although primary surgical resection is the treatment of choice for SS, the prognosis is poor with this treatment alone. As the number of cases of SS of the kidney is less due to its extreme rarity, no clear medical guidelines have yet been established.¹⁴

Conclusion

Synovial sarcomas occurring in the kidney, in analogy to other sites, tend to have an aggressive biologic behaviour. In spite of diagnostic challenge for pathologist the histopathological diagnosis of primary renal synovial sarcoma was based on morphology, IHC and cytogenetics. Because of rapid course and poor outcome, the clinician as well as pathologist should be aware of the existence of this rare tumor, so that timely and appropriate therapy can be applied.

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Competing Interests

None declared.

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