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Case Report

Pathology Section

Primary Renal Synovial Sarcoma- A Diagnostic Challenge

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ABSTRACT

Synovial sarcoma arising primarily in kidney is very rare, accounting for <2% of all malignant renal neoplasms. This tumour poses diagnostic dilemma as it mimics histologically with many other commonly presenting kidney tumours. Here, a 32-year-old female is reported who presented with gross haematuria and flank pain for three months. She complained recent increase in frequency of micturition, poor stream and incomplete emptying of bladder. Clinical possibility of urinary tract infection, urolithiasis, and renal neoplasm were considered. Right lower pole renal mass lesion of soft tissue density measuring 7.2×8.1×6.4 cm was found on radiologic examination. Grossly, well-circumscribed fleshy friable tumour with microcystic and haemorrhagic areas were noted. Monotonous small blue round cells arranged in sheets and in peritheliomatous pattern was noted upon microscopic examination. Histopathological differentials included were ewing sarcoma, adult wilm tumour, Non-Hodgkin Lymphoma (NHL) and rhabdomyosarcoma. A remote possibility of synovial sarcoma was also considered. Immunohistochemistry (IHC) revealed positivity with CD99, CD56, Bcl2 and TLE1. Characteristic histomorphology aided by typical immunohistochemical findings unravelled a diagnostic surprise of a rare renal tumour Primary Renal Synovial Sarcoma (PRSS).

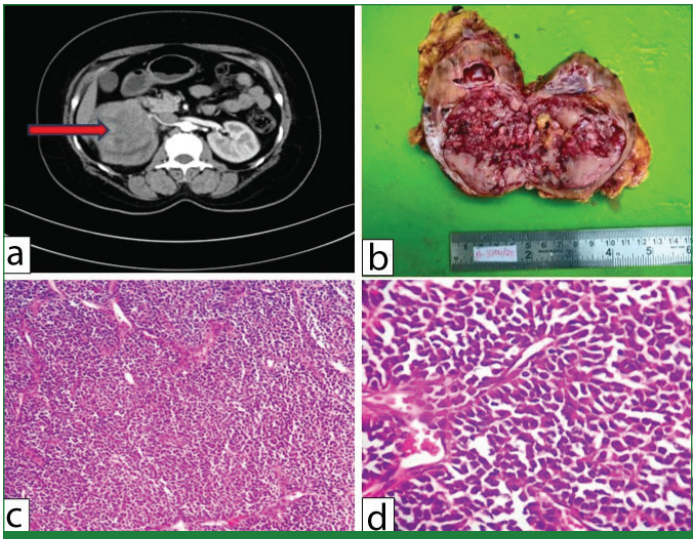
Keywords: Haemangiopericytomatous pattern, Immunohistochemistry, Malignant small round cell tumour

CASE REPORT

A 32-year-old female patient presented with flank pain and gross haematuria for four months with recent increase in frequency of micturition, poor stream and incomplete emptying of bladder. She was non-diabetic, but recently diagnosed to be hypertensive. She had no fever, nausea and was not on any medications. She had undergone total thyroidectomy four years back for multinodular goitre. After obtaining consent from the patient, medical records were reviewed pertaining to clinical data and laboratory investigations.

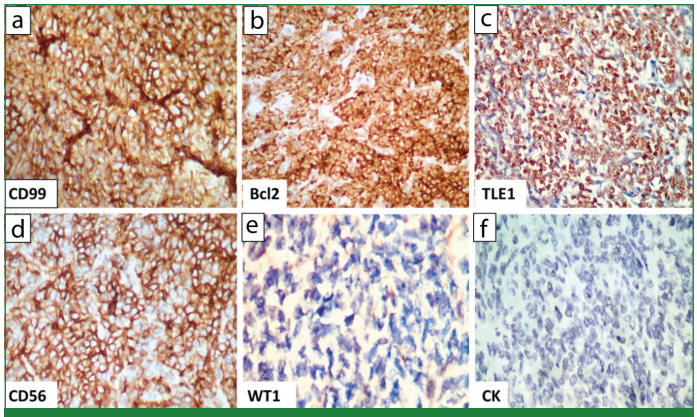
On examination, vitals were stable except for increased blood pressure (160/100 mmHg). No obvious mass was felt on per abdomen examination. Cardiovascular, respiratory and nervous system examination revealed no specific abnormality. Laboratory investigations revealed anaemia (Hb-9.6 gm%) and normal renal function (serum creatinine-0.98 mg/dL, Blood Urea nitrogen (BUN)-14 mg/dL). Prothrombin time was prolonged (PT-29.5 sec) and Erythrocyte Sedimentation Rate (ESR) was high (40 mm/1st hour). Urine examination revealed active sediments like White Blood Cells (WBCs)-3-4 cells/HPF and Red Blood Cells (RBCs)-5-6 cells/HPF. Provisional diagnosis of urinary tract infection, urolithiasis, and remote possibility of renal neoplasm were considered. Further, Computed Tomography (CT) abdomen showed large, well-defined heterogeneously enhancing soft tissue mass in the lower pole of right kidney measuring 7.2×8.1×6.4 cm. Cystic, calcific and necrotic areas were noted, in addition to this, hydronephrosis was present. Based on these findings, final diagnosis of renal cell carcinoma was made. Intraoperatively, exophytic mass involving the lower pole with multiple parasitic vessels on the surface noted. Right radical nephrectomy was performed and specimen was sent for histopathological examination [Table/Fig-1].

Grossly, kidney measured 13×9×6 cm. Lower pole was bosselated. Cut section showed fairly well-circumscribed grey brown, friable, fleshy tumour measuring 7×5×4 cm. The renal sinus and hilum grossly appeared free. The other pole showed dilated calyx. On microscopy, tumour was composed of round to ovoid, small to medium sized cells arranged in sheets, nests, vague lobules and in peritheliomatous pattern [Table/Fig-1]. Cells were largely rounded, monomorphic with hyperchromatic to vesicular nuclei,



[Table/Fig-1]: Radiologic, gross and histopathologic findings. a) CT abdomen-Heterogenously enhancing soft tissue density mass in lower pole of right kidney; b) Well-circumscribed grey white fleshy mass in lower pole of kidney with tiny cystic and haemorrhagic areas; c) Sheet like arrangement of poorly differentiated tumour cells (X40, H&E); d) Peritheliomatous pattern (X400, H&E).

tiny eosinophilic nucleoli and scant cytoplasm. Mitotic activity was evident. Tumour cell nests are separated by delicate arborising fibrovascular septae. Focal necrosis, apoptotic cell debris, cystic areas and calcific specks noted. Extension into perinephric fat and renal sinus was demonstrated microscopically. Uninvolved renal cortex showed obsolete glomeruli and acute tubular injury. Histopathological differentials included were ewing sarcoma, adult wilm tumour, poorly differentiated synovial sarcoma, NHL and rhabdomyosarcoma. IHC clinched the diagnosis of synovial sarcoma with strong positive results for CD56, CD99, Bcl2, and TLE1 [Table/Fig-2]. Absence for epithelial component was demonstrated by negativity for epithelial markers (CK7, PanCK). Other differentials like rhabdomyosarcoma (by desmin-negative), wilm tumour (by WT1, CK7-negative), NHL (by CD45-negative), ewing sarcoma (by positivity for CD56, Bcl2 and TLE1) were ruled out. Genetic analysis was not done due to financial constraints.



[Table/Fig-2]: Immunohistochemistry (IHC) findings. a) CD99- Diffuse strong membranous positivity (X400); b) Bcl2- Diffuse strong cytoplasmic and membranous positivity (X400); c) TLE1- Diffuse strong nuclear positivity (X400); d) CD56- Diffuse strong membranous positivity (X400); e,f) Diffusely negative for WT1 and CK respectively (X400).

A final diagnosis of PRSS {Grade 2-(based on The 'FédérationNationale des Centres de LutteContre le Cancer'2015)} was rendered. Immediate postoperative period was uneventful. Three months later, she presented with right flank mass. Excision biopsy was done and histomorphology revealed similar features as primary tumour. However, no renal tissue was identified. She is yet to receive chemotherapy.

DISCUSSION

Synovial sarcoma is a rare soft tissue tumour accounting for 6-10% of malignant soft tissue neoplasms. It mostly affects children and young individuals, chiefly located near joints and tendons of extremities. Synovial sarcoma arising primarily in kidney is very rare accounting for 1% of synovial sarcomas. First case of renal synovial sarcoma was published in the year 2000 by Argani P et al., [1]. In an extensive review by El Chediak A et al., [2] 114 cases of PRSS have been reported in literature till 2016. PRSS poses diagnostic challenge as it mimics histologically with many other commonly presenting kidney tumours making it difficult to differentiate from them. Here, a rare case of PRSS, which was diagnosed based on characteristic histomorphology aided by IHC is presented.

Synovial sarcoma is one of the rare soft tissue tumours with ambiguous cell of origin with dismal prognosis. Rare sites of occurrence includes head and neck, heart, lung, kidney. Median age for the PRSS is 40.5 years with the age range being 15 to 78 years. Most patients present with lumbar pain and haematuria, majority with local invasion and distant metastasis with rapid disease course [2]. In present case, patient had only haematuria.

Synovial sarcoma of kidney can be solid fleshy growth or can be cystic lesion with mural mass [3]. Definitive diagnosis always rely on histopathology aided by IHC and molecular testing.

Synovial sarcoma is traditionally classified into three subtypes based on histopathology, namely biphasic (20-30%), monophasic (50-60%) and poorly differentiated (15-25%) depending on the spindle cell and epithelial cell differentiation. Poorly differentiated variants are high grade tumours, generally has epitheloid cell morphology with high mitotic rate (10-20/10HPF). Poorly differentiated variant by virtue of its cytomorphology and sheet like arrangement can be confused with Ewings/PNET and adult Wilms tumour. IHC is must to narrow down differentials and to aid in definitive diagnosis. PRSS shows positive results for CD99, vimentin, Bcl2 and CD56. Focal positivity for CK and EMA is also known. Negative results are seen with desmin, WT1, S100, CD34 and CD31 [4].

Peritheliomatous pattern might cause difficulty in differentiating from Haemangiopericytoma, where CD34 will be positive. CD45 negativity rules out diagnostic possibility of lymphoma. Haemangipericytomatous architecture is common with renal synovial sarcoma. TLE1 (transducin-like enhancer of split 1), a transcriptional co-repressor is a useful diagnostic marker of synovial sarcoma [5]. Terry J et al., [6] observed high sensitivity and specificity of TLE1 in synovial sarcoma diagnosis. In one study, 22/28 (79%) of monophasic, 18/23 (78%) of biphasic, and 20/22 (91%) of poorly differentiated synovial sarcomas demonstrated nuclear immunoreactivity for TLE1 [7]. Amongst currently available immunohistochemical markers for synovial sarcoma, TLE1 is more sensitive and specific [8]. Molecular analysis remains the goldstandard in diagnosing synovial sarcoma. t(X; 18) (p11.2; q11.2) SYT-SSX is positive in more than 90% synovial sarcomas. Translocation results in SYT-SSX1 and SYT-SSX2 transcripts, which can be detected by either FISH or RT-PCR [9]. However, when compared to molecular testing, TLE 1 IHC is less expensive with rapid turn around time. With the availability of robust immunohistochemical markers and laboratory settings with limited resources molecular testing becomes almost unfeasible, as in present case.

CONCLUSION(S)

A small round blue cell tumour of kidney invokes several differentials. Keen histomorphologic examination and identification of hemangiopericytomatous architecture should raise a suspicion of PRSS, though rare. Judicious use of IHC based on differential diagnosis will clinch the correct diagnosis.

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