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Case Report

Primary Synovial Sarcoma of Kidney - A Histological Surprise: A Rare Case Report from a Tertiary Care Centre

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Abstract

Renal Synovial Sarcoma (SS) are rare type of tumors arising from the mesenchymal tissue of the kidney. Its presenting features overlap with other renal tumors there by creating a diagnostic dilemma. Very few cases of primary renal SS are described so far in the literature with no defined treatment protocol described. We hereby describe a case report of this rare tumor in a 12 year old girl who presented to us with hematuria and flank pain. Histopathological analysis and the Immunohistochemistry (IHC) confirmed the diagnosis of primary monophasic synovial sarcoma. Follow-up post nephrectomy at six months showed no evidence of metastasis or recurrence.

Keywords: Synovial sarcoma; SYT-SSX; Renal tumours; Renal spindle cell tumour; Pathological response; Survival

Abbreviations

SS: Synovial Sarcoma; CT: Computed Tomography; PCR: Polymerase Chain Reaction; FISH: Fluroscence *In Situ* Hybridization

Introduction

Synovial Sarcomas (SS) are a group of tumors of mesenchymal origin affecting predominantly young adults. SS commonly involves the proximal extremities with male predominance. Other sites of involvement are lungs, heart, head, neck and kidneys but these tumors are exceedingly rare [1]. The term "synovial" is a misnomer as morphology of the tumor and Immunohistochemistry (IHC) does not match with that of a normal synovium of joint [2].

SS can be classified into biphasic or monophasic variety histologically. Monophasic SS of kidney is rare and was initially described by Faria et al. [3]. Monophasic SS consists of only of spindle cells without any epithelial component and biphasic SS consists of spindle cells with epithelial cell components.

Primary SS of kidney is very rare with less than 50 cases being reported till date [4]. It accounts for only 5-10% of adult soft tissue sarcomas and even rare in pediatric population. There is no clinical or imaging findings that are characteristic of the tumor which can clinch the diagnosis [5]. Chromosomal rearrangement studies are required at times for confirmation of diagnosis along with a characteristic histology with IHC. IHC positivity for CD99, bcl2 and TLE11 is characteristic of SS and there is SYT-SSX gene fusion rearrangement, t (X; 18) (p11.2; q11.2) [5]. We hereby report a case of primary SS of the kidney, which was a histological surprise, thought to be a Wilms tumor preoperatively.

Case Presentation

A 12-year-old girl presented with complaints of pain in the left flank which was recurrent since 8 months, insidious in onset and colicky in nature, associated with gross hematuria with passage of clots for a period of 4 days before seeking medical care. Ultrasound imaging showed an upper pole mass indicating a further evaluation of the lesion using a Contrast Enhanced CT scan (CECT). CECT showed presence of heterogeneously enhancing lobulated mass which involved the upper pole of the left kidney along with renal pelvis measuring around 4.7 centimeters (Figure 1a). There was no evidence of metastasis to lung or liver. Based on the above findings, provisional diagnosis of Wilm's tumor was considered and patient underwent left radical nephrectomy (Figure 1b).

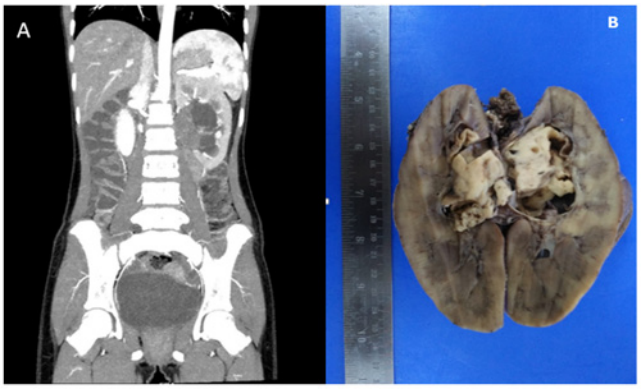

Figure 1: A) Contrast enhanced computed tomography of abdomen and pelvis showing heterogenously enhancing lobulated mass lesion of size 3.9 x 4.0 x 4.7 cm along the walls of left renal pelvis extending into the upper pole calyx and proximal ureter (red arrows) with few interspersed non-enhancing hypodense (likely necrotic) and hyperdense (likely hemorrhage) areas. B) Nephrectomy specimen showing a well circumscribed encapsulated tumor in the hilar region measuring 6 x 5.5 x 5 cm, with grey white fleshy cut surface with cystic change (colour image).

Histopathological analysis of the specimen showed cellular tumour comprising of interlacing bundles of spindle shaped malignant cells suggestive of SS of the kidney (Figure 2a). On IHC, tumour cells were positive for Bcl2, CD 99, TLE 1, negative for S100, synatophysin, SMA, WT-1 (Figure 2b). The tumor showed proliferation of cellular spindle cell without epithelial component.

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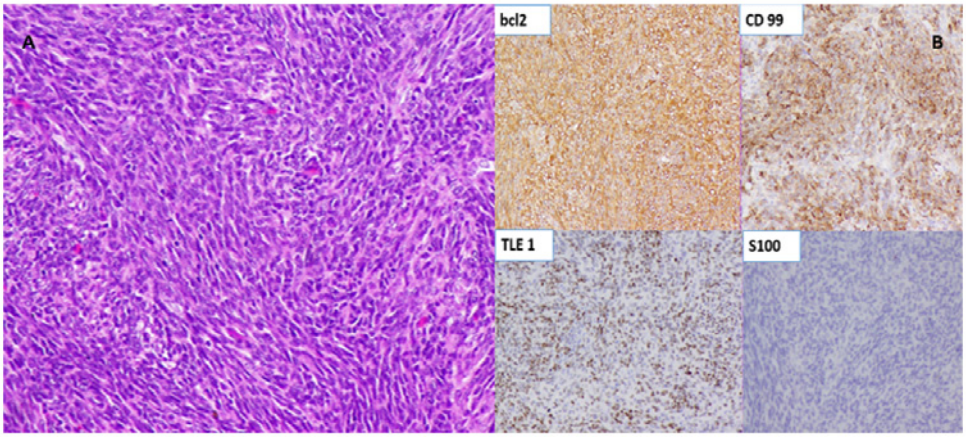

Figure 2: A) Higher power microscopy showing interlacing bundles of spindle shaped malignant cells with elongated to oval mildly pleomorphic nuclei, mitosis of 5-6/10 HPF, 400X, H&E (colour image). B) On Immunohistochemistry, tumor cells were positive for Bcl2, CD 99, TLE 1, negative for S100, Synatophysin, SMA, WT-1 (colour image).

Based on the above findings, primary monophasic SS of kidney was diagnosed.

Patient was further referred to a medical oncologist and based on discussion of pro's and con's with the patient's parents, serial follow up with no adjuvant treatment was chosen. Patient is currently being followed up every three monthly for one year. Our patient has presently completed six months of follow up and CT thorax and abdomen done at six months shows no evidence of metastasis or recurrence.

Discussion

Mesenchymal tumors arising primarily from the kidney are leiomyosarcoma, liposarcoma, rhabdomyosarcoma, fibrosarcoma, and malignant fibrous histiocytoma [6], with leiomyosarcoma being the most common type. SS is a type of mesenchymal tumor of spindle cell origin which displays variable epithelial differentiation. The SS usually occurs in the age groups of 20 to 72 years, median age being 35 years and predominantly involves males [4]. To our knowledge, the case described above was the youngest patient described, with only 5 patients so far described to have SS below the age of 20 years [2].

Primary renal SS is rare, comprising 1-3% of all malignant renal neoplasms [2]. The first case report was published by Argani et al. in 2000 [3]. SS is linked with chromosomal translocation t(X; 18) (p11; q11), leading to the fusion of the SYT gene on chromosome 18 to exon 5 of either SSX1 or SSX2 genes on chromosome X. Some of the recent reports also have identified the involvement of SSX4 gene in the translocation leading to SS [7].

There are no classical clinical symptoms or imaging findings that can differentiate renal SS from other malignant renal tumors. Based on symptoms and imaging, a wide range of differential diagnosis including primary Ewing sarcoma of kidney, sarcomatoid renal cell sarcoma, undifferentiated carcinoma and adult Wilms tumour can be considered. But the presence of both mesenchymal and epithelial markers is suggestive of synovial sarcoma at any site [3].

Difficulty in diagnosing cases can occur even after IHC findings thereby requiring confirmation of rearrangement involving the SYT gene by either RT-PCR or FISH testing [1]. Among SS, monophasic type is frequently observed than biphasic type in the kidney. Renal SS are typically positive for Bcl-2, CD99, CD56, vimentin and focally for epithelial membrane antigen. In a study conducted by Terry et al., TLE1 was found to discriminate SS from other sarcomas excellently [2]. Also TLE1 expression is a strongly predictor of SYT gene rearrangement [1]. TLE1 is one of the most sensitive and specific marker currently available for synovial sarcoma, as it is less frequently detected in other mesenchymal tumors. Hence it is considered an appropriate IHC marker for SS [2].

Flank pain was the most frequent symptom reported at the time of presentation, seen in 55. 5% of cases and 37 patients (44%) had hematuria upon presentation [8]. In a study conducted by Kohle et al., similar data was reported where at the time of presentation, 98% of cases were symptomatic with pain (67%) and hematuria (38%). But the sample size was small [7].

Prognosis of SS in the presence of SYT-SSX fusion type is questionable. But observation suggests presence of SYT-SSX fusion type and metastasis, were important prognostic indicators at the time of diagnosis. SYT- SSX2 fusion type rearrangement was associated with lower prevalence of metastasis and hence had a better overall survival [6].

30 to 50% of the patients were found to have metastasis to lungs or liver at the time of surgical resection of their primary tumor, lungs being the most common site, irrespective of type of fusion [2]. Though SS positive for SYT-SSX 2 fusion has lower prevalence of metastasis, half of these patients had metastasis to the liver. But overall SS with SYT-SSX 1 behaves more aggressively.

On contrary, a retrospective study conducted on Japanese patients with SS, positive for SYT-SSX fusion transcript concluded that unlike tumor size and histological grading, SYT-SSX fusion type was not found to affect prognosis significantly [2].

Primary renal SS behaves aggressively and carries poor prognosis. No standard treatment guidelines have been established owing to the rarity of the tumor. The treatment is upfront radical nephrectomy. Benefit of adjuvant chemotherapy as a part of curative intent is

still controversial. Hence, recent studies limit the use of adjuvant chemotherapy only for patients who are likely to be benefited, which includes younger patients or larger tumors [9]. In a meta-analysis of all randomized clinical trials conducted by The Sarcoma Meta-analysis Collaboration (SMAC) group in 1997, indicated that doxorubicin- based chemotherapy significantly improved time to local and distant recurrence, as well as overall recurrence-free survival in comparison to the control group who were just observed, though it was not statistically significant [10]. For inoperable lesions, recurrent disease after surgery or in the presence of medical contra-indications for surgery, chemotherapy may be given as a part of palliation [4].

Declaration

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Patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient and her parents have given their consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Manuscript has been read and approved by all the authors, that the requirements for authorship as stated earlier in this document have been met, and that each author believes that the manuscript represents honest work, if that information is not provided in another form.

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