ISSN No. 2456-4400
Int J Med Lab Res 2022, 7(2):34-40

IJMLR International Journal of Medical Laboratory Research

CASE STUDY

A RARE CASE REPORT OF PRIMARY SYNOVIAL SARCOMA OF KIDNEY

Kiran Agarwal[1], Neha Suman[2]

[1]Director Professor, Department of Pathology, Lady Hardinge Medical College, New Delhi-110001, India
[2]Senior Resident, Department of Pathology, Lady Hardinge Medical College, New Delhi-110001, India

Received: 18 June, 2022/Revision: 06 July, 2022 /Accepted: 27 July, 2022

ABSTRACT: Synovial sarcoma represents 5-10% of all soft tissue sarcomas. It occurs primarily in the periarticular regions of extremities. Rarely this tumour has been reported in kidney. After extensive search of literature, less than 100 cases have been reported till date. It is difficult to diagnose this tumour because of its rarity and also because of its morphological resemblance to tumours like renal cell carcinoma with sarcomatoid differentiation, adult Wilms' tumour, spindle cell carcinoma and metastatic sarcoma. Thorough histopathological examinations along with immunohistochemical markers are helpful in establishing the diagnosis. Here we present a case of a 27-year-old male who presented with right renal mass and was later diagnosed as primary synovial sarcoma of kidney. We also present a 10-year comprehensive review of the literature.

KEYWORDS: Kidney, rare, synovial sarcoma.

INTRODUCTION:

Synovial sarcomas (SSs) are rare soft tissue tumours, accounting for 5 – 10 % of sarcomas in adults. SSs occur most commonly in the proximity of large joints. It has also been reported in the thoracic and abdominal walls, head and neck region, retroperitoneum, kidney, bone, lung and prostate. [1] Primary SSs of kidney are even rarer with less than 100 cases described in the literature so far.[2] It was first described by Faria et al in 1999. [3] These tumours have characteristic histology and immunohistochemistry, however, due to its rarity, it becomes difficult to diagnose them. Here we describe a rare case of primary synovial sarcoma of kidney in a 27-year-old male and a ten-year comprehensive review of the literature.

CASE REPORT

A 27-year-old male presented with complaint of pain in right flank for last 21 days. The pain was gradually increasing with no associated hematuria or dysuria. His medical history was unremarkable. On general examination, the patient was conscious and oriented. There were no signs of pallor, icterus, oedema or lymphadenopathy. On physical examination, a lump was felt in the right lumbar and right hypochondriac region. Routine blood examination and urine analysis were normal. The chest X-ray was unremarkable. Contrast-enhanced computed tomography of the abdomen and pelvis revealed a heterogeneously enhancing mass of size 14x12.5x3 cm, occupying whole of right kidney. There was no other lesion / evidence of any metastasis.

Corresponding Author:
Dr Neha Suman
Department of Pathology, Lady Hardinge Medical College, Shaheed Bhagat Singh Marg, New Delhi-110001, New Delhi

A diagnosis of renal cell carcinoma was made preoperatively. Intraoperatively, a tumour mass occupying whole of right kidney was seen along with adhesions between abdominal wall and the tumour mass. Right radical nephrectomy was done and the specimen was sent for further histopathological examination. On gross examination, it was a distorted specimen of right kidney, measuring 16.5 x 15 x 3.5 cm. A solid-cystic tumour mass was seen measuring 13.5x12.5x3.5 cm involving the whole of right kidney with focal areas of capsular breach [**Figure 1**].

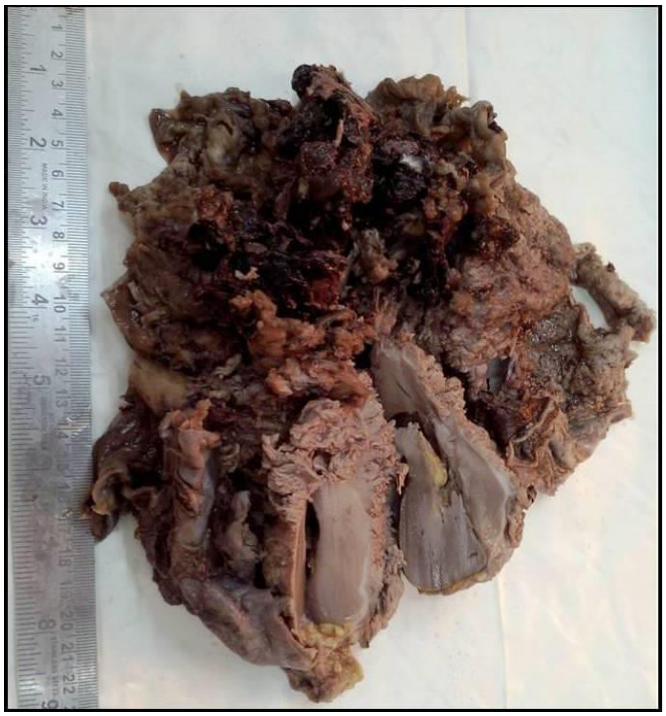

Figure 1: Right kidney with solid- cystic tumor mass

Microscopically, the tumour was composed of plump spindle cells arranged in intersecting

fascicles. The cells had scant cytoplasm and hyperchromatic elongated nuclei [**Figures 2a and 2b**].

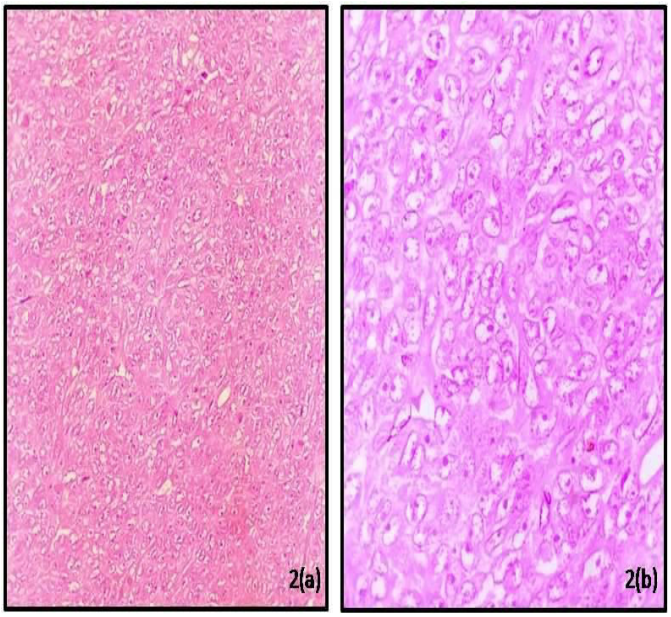

Figure 2(a): Spindle cells arranged in intersecting fascicles (Hematoxylin and eosin stain; X100). Figure **2(b): Spindle cells with scant cytoplasm and hyperchromatic elongated nuclei (Hematoxylin and eosin stain; X400).**

Mitotic figures were 1 per 10 high power fields. Capsule and perinephric fat were involved by the tumour. On immunohistochemistry, tumour cells expressed CK, Vimentin, CD99 and Bcl-2 [**Figures 3 (a), (b), (c) and (d)**].

Tumour staining was negative for SMA, Desmin, WT1 and CD10. This histological appearance and immunohistochemistry confirmed primary monophasic synovial sarcoma of kidney as the diagnosis.

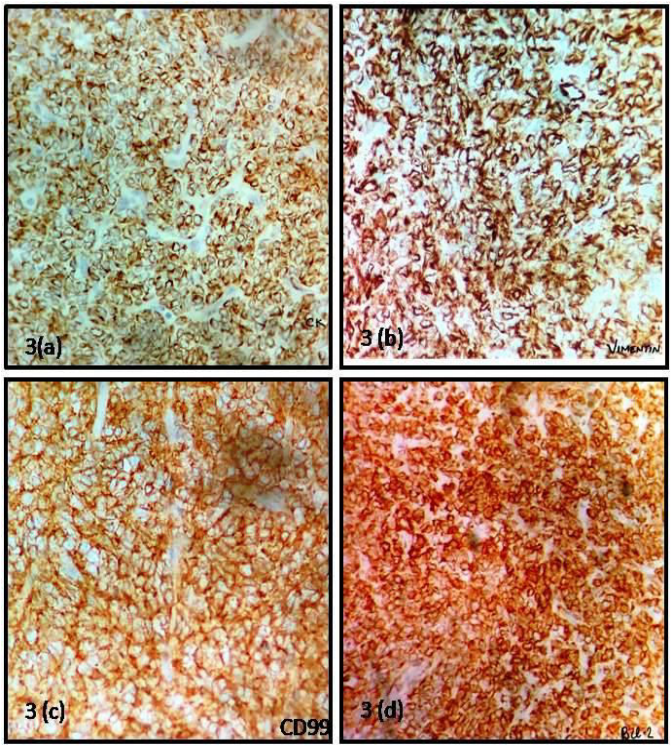

Figure 3: Tumour cells show positivity for (a) CK, (b) Vimentin, (c) CD99, (d) Bcl-2.

DISCUSSION:

Synovial sarcoma is a rare and aggressive soft tissue tumor, occurring mainly in young adults. [1] There are no specific clinical features that point towards primary synovial sarcoma of kidney. Also, the imaging studies are not sufficient for the definitive diagnosis of this entity. The rare incidence of this tumour along with its presentation similar to other renal tumor accounts for the difficulty in its diagnosis. [2] The definitive diagnosis always requires pathological evidence.

Synovial sarcoma usually arises from pleuripotent mesenchymal cells and can be histologically classified into biphasic and monophasic types. In biphasic forms, both epithelial and spindle cells are present in varying proportions. The monophasic form comprises only of epithelial cell or spindle cells, particularly spindle cells. Increased cellular atypia and mitosis are associated with poor prognosis. Poorly differentiated SSs show cells with scant cytoplasm and hyperchromatic high-grade nuclei and lacks the bland spindle cells as seen in the monophasic type. [1] Monophasic type is difficult to diagnose from other spindle cell tumours such as leiomyosarcoma, Wilms' tumor and sarcomatoid renal cell carcinoma. Poorly differentiated SSs show considerable morphological overlap with other round cell sarcomas, especially Ewing sarcoma/ Primitive Neuro Ectodermal Tumour (PNET), adult blastemal Wilms tumour and alveolar rhabdomyosarcoma. [4] Immunohistochemistry is helpful to rule out these differential diagnoses. Synovial sarcoma is usually positive for CK, Bcl2, vimentin and CD99. There is negative expression of Desmin, WT1 and CD10. [1,2,4] Similar expression of IHC was seen in the present case. Wilms' tumor shows expression of WT1. There is positive expression of desmin in leiomyosarcoma. Sarcomatoid renal cell carcinoma is positive for CD10, hence ruled out.

However, molecular or cytogenetic analysis by reverse transcriptase polymerase chain reaction (RT-PCR) to detect SYT-SSX gene fusion has also been used to make a definitive diagnosis of SSs. Most of them are associated with characteristic translocation t(x;18)(p11.2;q11.2) which results in fusion of SYT-SSX gene. [5,6] But very few cases have been confirmed by the molecular technique. Molecular analysis was not performed in our case.

Primary synovial sarcoma of kidney is rare with very few cases reported till date. We present a comprehensive review of the cases reported in the last 10 year and our case in **Table 1**.

Table1: Comprehensive review of the reported cases of primary renal synovial sarcoma

AUTHORS	YEAR	NO. OF PATIENTS	AGE/ GENDER	SYMPTOMS	MAXIMUM TUMOR DIMENSION	METASTASIS	DIAGNOSIS	FOLLOW-UP
Ozkan EE et al[7]	2011	1	68y/ Female	Yes	25cm	None	SS	12 months
Nishida et al[8]	2011	1	63y/ Female	Yes	5cm	None	Monophasic SS	12 months
Grahamp-urohit VU et al[9]	2011	1	21y/ Female	Yes	12cm	None	PDSS	6months
Bakhshi et al[10]	2012	1	33y/ Female	Yes	20 cm	None	Monophasic SS	24 months
Bhatkule et al[11]	2013	1	19y/ Female	Yes	9cm	None	SS	Not mentioned
Majumder A et al[12]	2014	1	46/ Female	Yes	18cm	None	SS	2months
Moorthy HK et al[13]	2014	1	46/ Male	Yes	12.8cm	None	SS	Not mentioned
Modi G et al[14]	2014	1	41/ Male	Yes	5cm	None	SS	Not mentioned
Schoolm-eester J K et al[15]	2014	16	17y to 78y/ Male-9 Female-7	Yes	2.2 to19cm (mean 8.6cm)	Yes in 3 cases (lung)	Monophasic SS	12 to 77months
Abbas M et al[16]	2014	1	33y/ Male	Yes	12cm	None	Monophasic SS	Not mentioned
Wang Z et al[17]	2015	1	54y/ Female	Yes	13cm	None	Monophasic SS	12 months
Vedana M et al[18]	2015	1	76y/ Female	Yes	12cm	None	Monophasic SS	20 months
Mishra S et al[19]	2015	1	60y/ Male	Yes	13cm	None	Monophasic SS	6 months
Kohle O et al[20]	2015	2	52y/Male 50y/Male	Yes Yes	10cm 10cm	Yes (lung & liver) Yes (lung)	PDSS PDSS	18 months 20months
Chandrasekeran D et al[21]	2016	1	44y/ Male	None	10cm	Yes (Lung)	SS	24 months
Pathrose G et al[22]	2017	1	25y/ Female	None	13 cm	None	SS	36 months
Uma Kant Dutt et al[23]	2018	1	21y /Male	Yes	6.5cm	None	Biphasic SS	Not mentioned
Tranesh G et al[24]	2018	1	56y/ Male	Yes	8.5cm	None	SS	Not mentioned
Huang Y et al[25]	2018	1	44y/ Male	None	8cm	None	PD monophasic SS	3months
Yellala et al[26]	2018	1	38y/ Male	Yes	26cm	Yes (lung)	PDSS	16 months
Cao Z et al[27]	2018	2	74y/ Female 49y/ Female	None Yes	18cm Not mentioned	None Yes (lung)	SS SS	3 months 23 months
Chediak A El et al[28]	2018	1	26y/ Male	Yes	6cm	Yes (lung)	Monophasic SS	12 months
Pichler Ret al[29]	2018	1	20y/ Male	Yes	17cm	None	Monophasic SS	Not mentioned
Stamm A et al[30]	2019	1	43y/ Female	Yes	17cm	None	SS	3months
Cai H J et al [31]	2019	1	54y/ Male	Yes	6.8cm	None	SS	24 months
Kiran et al	2020 Present case	1	27y/ Male	Yes	16.5cm	None	Monophasic SS	12 months

SS=Synovial sarcoma. PD= Poorly differentiated

REVIEW OF LITERATURE:

After extensive search of literature, we found only 43 cases including the present case of primary synovial sarcoma of kidney reported in the last ten years. The age of presentation varied from 21 to 78 years with male to female ratio of 1.4: 1. However, only 9 cases (including our case) had been reported in the age range of 21 to 30 years, thus highlighting the rarity of this entity.

Most of these cases had symptoms such as abdominal pain and/or hematuria or dysuria as in the present case, only in 4 cases it was an incidental finding. These symptoms are non-specific for pointing towards the diagnosis as other renal carcinomas also present with these symptoms. Also, radiological investigations such as CT reveal only the malignant nature of the lesion. Definitive diagnosis can be reached only by proper histopathological examination and immunohistochemistry.

The size of the tumour varied from 2.2 cm to 20 cm. The maximum tumour dimension noted in the present case was 16.5 cm. Tumour diameter < 5cm is considered as a good prognostic factor in renal sarcomas. [32]

25 cases had been reported as monophasic SS, 2 were monophasic SS with poorly differentiated type and one was biphasic SS. Rest of the cases were reported as primary renal synovial sarcoma with no further differentiation. The present case was monophasic synovial sarcoma. The importance of subtyping these sarcomas lies with the fact that poorly differentiated types have aggressive behaviour and poor prognosis. [32] However, biphasic versus monophasic morphology has no prognostic relevance.

Metastasis to lung was observed in 8 cases and one had metastasis to liver. However, no metastasis was seen in our case. The presence of metastasis is associated with poor outcome in patients. It had been observed that the mean survival rate is lower in patient with metastatic disease. [32] The most common site of metastasis is lung where late metastases can occur. 5-year survival rate of 50-85% has been reported.[33]

Follow-up was done in 35 cases with a disease-free period ranging from 2 months to 36 months. One year follow up in our patient has shown him to be disease free.

Primary renal SS are treated with complete surgical resection in conjunction with adjuvant therapy as preoperative irradiation for large or initially unresectable primary tumours and chemotherapy for disseminated disease. Favourable results have been observed with high-dose Ifosfamide-based chemotherapy as neo-adjuvant and adjuvant therapy in patients with localized or advanced soft tissue synovial sarcomas. [34]

CONCLUSION:

Primary renal synovial sarcomas are very rare and aggressive tumour that can be mistaken for other types of renal carcinomas. Making the correct diagnosis is challenging, however, morphological and immunohistochemical features are helpful in making the correct diagnosis. Since the disease may have rapid course with poor prognosis, the possibility of this rare entity should be considered for the early initiation of therapy.

REFERENCES:

[1]. Weiss SW, Goldblum JR. Malignant soft tissue tumors of uncertain type. In: Weiss SW, Goldblum JR, eds. Enzinger and Weiss's soft tissue tumors. 4th edition. St. Louis: MO: Mosby; 2001:1483-1565.
[2]. Moch H, Humphrey PA, Ulbright TM, Victor E. WHO Classification of Tumours of the Urinary System and Male Genital Organs. 4th edition. IARC: Lyon; 2016: 61.
[3]. Faria P, Argani P, Epstein J. Primary synovial sarcoma of the kidney: A molecular subset of so

called embryonal renal sarcoma. Mod Pathol Am 1999; 12: 94.

[4]. Wang J, Weiss LM, Hu B, Chu P, Zuppan C, Felix D, et al. Usefulness of immunohistochemistry in delineating renal spindle cell tumours. A retrospective study of 31 cases. Histopathol 2004; 44: 462–1.

[5]. Coindre JM, Pelmus M, Hostein I, Lussan C, Bui BN, Guillou L. Should molecular testing be required for diagnosing synovial sarcoma? A prospective study of 204 cases. Cancer 2003; 98: 2700-7.

[6]. Bella AJ, Winquist EW, Perlman EJ. Primary synovial sarcoma of the kidney diagnosed by molecular dectection of syt-ssx fusion transcripts. J Urol 2002; 168:1092-3.

[7]. Ozkan EE, Mertsoylu H, Ozardali HI. A case of renal synovial sarcoma treated with adjuvant ifosfamide and doxorubicin. Intern Med 2011; 50:1575–80.

[8]. Takeshi N, Teruo I, Hiroshi U. Monophasic primary renal synovial sarcoma accompanied with a haemorrhagic cyst. Urol J 2011; 8: 244-7.

[9]. Grampurohit VU, Myageri A, Rao RV. Primary renal synovial sarcoma. Urol Ann 2011; 3:110-3.

[10]. Bakhshi GD, Khan AS, Shaikh AS, Khan MA, Khan MA, Jamadar NM. Primary renal synovial sarcoma. Clin Pract 2012; 30:44.

[11]. Bhatkule MA, Dhawle MS, Kokandakar HR, Bindu RS. Primary renal synovial sarcoma: Report of a rare case. Clin Cancer Investig J 2013; 2: 353-5.

[12]. Majumder A, Dey S, Khandakar B, Medda S, Chandra Paul P. Primary Renal Synovial Sarcoma: A Rare Tumor with an Atypical Presntation. Arch Iran Med 2014; 17: 726-8.

[13]. Moorthy HK, Pillai BS, Varghese J. Primary Renal Synovial Sarcoma: An Oncologic Surprise. Urol Case Rep 2014; 2:152-3.

[14]. Modi G, Madabhavi I, Panchal H, Anand A, Patel A, Parikh S, Revannasiddaiah S. Primary synovial sarcoma of kidney: a rare differential diagnosis of renomegaly. Case Rep Pathol 2014; 2014: 657497.

[15]. Schoolmeester JK, Cheville JC, Folpe AL. Synovial sarcoma of the kidney: a clinicopathologic, immunohistochemical, and molecular genetic study of 16 cases. Am J Surg Pathol 2014; 38:60-5.

[16]. Abbas M, Dämmrich ME, Braubach P, Meinardus A, Kramer MW, Merseburger AS, et al. Synovial sarcoma of the kidney in a young patient with a review of the literature. Rare Tumors 2014; 6:5393.

[17]. Wang Z, Zhong Z, Zhu L, Xiong W, Pan C, Wang X, et al."Primary synovial sarcoma of the kidney: A case report". Oncol Letters. 2015; 6: 3542-4.

[18]. Vedana M, Fuenfschilling M, Tzankov A, Zellweger T. Primary Synovial Cell Sarcoma of the Kidney: Case Report and Review of the Literature. Case Rep Oncol 2015; 8:128-2.

[19]. Mishra S, Awasthi N, Hazra SP, Bera MK. Primary synovial sarcoma of the kidney. Saudi J Kidney Dis Transpl 2015; 26: 996-9.

[20]. Köhle O, Abt D, Rothermundt C, Öhlschlegel C, Brugnolaro C, Schmid HP. Soft tissue sarcomas of the kidney. Rare Tumors 2015; 7:5635.

[21]. Chandrasekaran D, Narayanaswamy K, Sundersingh S, Senniappan K, Raja A. Primary Synovial Sarcoma of the Kidney with Inferior Vena Caval Thrombus. Indian J Surg Oncol 2016; 7:345-8.

[22]. Pathrose G, John NT, Hariharan P. Renal Synovial Sarcoma in a Young Pregnant Lady: A Case Report and Clinico-Pathological Profile. J Clin Diagn Res 2017; 11:13-4.

[23]. Dutt UK, Manikandan R, Dorairajan LN, Srinivas BH. Biphasic renal synovial sarcoma with extensive venous tumor thrombosis: A rare presentation. Urol Ann 2018; 10:339-41.

[24]. Tranesh G, Cortese C, Thiel D, Zhai QJ. Primary synovial sarcoma of the kidney-A case report and literature review. Int J Clin Exp Pathol 2018; 11: 2241-5.

[25]. Huang Y, Liu D, Chen W. Primary renal synovial sarcoma: A case report and literature

review. J Can Res Ther 2018; 14 Suppl S1: 267-9.

[26]. Yellala A. Primary renal synovial sarcoma – a diagnostic dilemma. J Community Support Oncol 2018; 16: 202-5.

[27]. Cao Z, Lu HZ, Feng XL. Primary renal synovial sarcoma: two cases and review of the literature. Int J Clin Exp Pathol 2018; 11:4346-1.

[28]. El Chediak A, Mukherji D, Temraz S, Nassif S, Sinno S, Mahfouz R, et al. Primary synovial sarcoma of the kidney: a case report of complete pathological response at a Lebanese tertiary care center. BMC Urol 2018; 18:40.

[29]. Pichler R, Heidegger I, Schachtner G, Stuehmeier J, Agaimy A, Maier H, et al. Renal rupture - Not what it seems. Urol Case Rep. 2017; 16:132-4.

[30]. Stamm A, Conti N, Olgac S, Flores JP, Wagner MJ, Kozlowski P. Intraperitoneal Rupture of a Synovial Sarcoma of the Kidney. Rev Urol. 2019; 21(2-3):136-40.

[31]. Cai HJ, Cao N, Wang W, Kong FL, Sun XX, Huang B. Primary renal synovial sarcoma: A case report. World J Clin Cases. 2019; 7(19):3098-3103.

[32]. Öztürk H. Prognostic features of renal sarcomas (Review). Oncol letters: 2015; 1034-8.

[33]. Gabilondo F, Rodriquez F, Mohar A, Nuovo GJ, Domínguez-Malagón H. Primary synovial sarcoma of the kidney: corroboration with in situ polymerase chain reaction. Ann Diagn Pathol. 2008; 12:134–7.

[34]. Park SJ, Kim HK, Kim CK, Park SK, Go ES, Kim ME, et al. A case of renal synovial sarcoma: complete remission was induced by chemotherapy with doxorubicin and ifosfamide. Korean J Intern Med. 2004; 19:62–5.

Cite of article: Agarwal K, Suman N. A rare case report of primary synovial sarcoma of kidney. Int. J. Med. Lab. Res. 2022; 7,2: 34-40. ***http://doi.org/10.35503/IJMLR.2022.7206***

CONFLICT OF INTEREST: Authors declared no conflict of interest
SOURCE OF FINANCIAL SUPPORT: Nil