Discoveries Journals
publishing the future

DISCOVERIES 2026, Jul-Sep, 14(3): e236
DOI: 10.15190/d.2026.15

Original Article - Case Report

Primary synovial sarcoma of the kidney: Unravelling a rare and aggressive tumour

Aashna Gupta[1], Puneet Kaur Somal[2,], Nilay Nishith[2], Aishwarya Sharma[2], Rahul Raj[3], Ravikiran Pawar[2], Deepander Rathore[4], Sankalp Sancheti[2]*

[1] Department of Pathology; Punjab Institute of Liver and Biliary Sciences, Mohali, India
[2] Department of Onco-Pathology; Homi Bhabha Cancer Hospital and Research Centre; Tata Memorial Centre, Homi Bhabha National Institute (HBNI), New Chandigarh, India
[3] Department of Pathology, All India Institute of Medical Sciences, Mangalagiri
[4] Department of Radiodiagnosis; Homi Bhabha Cancer Hospital and Research Centre; Tata Memorial Centre, Homi Bhabha National Institute (HBNI), New Chandigarh, India

* *Corresponding author*: *Dr. Puneet Kaur Somal,* Associate Professor, Department of Onco-Pathology; Homi Bhabha Cancer Hospital and Research Centre; Tata Memorial Centre, Homi Bhabha National Institute (HBNI), New Chandigarh, India; Email: puneet.somal@gmail.com

Submitted: Aug 08, 2026; *Revised:* Sept. 27, 2026; *Accepted:* Sept. 28, 2026; *Published*: Sept. 30, 2026.
Citation: Gupta A, Somal PK, Nishith N, Sharma A, Raj R, Pawar R, Rathore D, Sancheti S. Primary Synovial sarcoma of the kidney: Unravelling a rare and aggressive tumor. *Discoveries* 2026, Jul-Sep, 14(3): e236. DOI: 10.15190/d.2026.15

ABSTRACT

Primary renal synovial sarcoma (PRSS) is an exceptionally rare and aggressive malignancy, comprising only 1–3% of all renal neoplasms. Its clinical and radiological features often mimic renal cell carcinoma, posing significant diagnostic challenges. This case report highlights a case of primary renal synovial sarcoma and reviews the relevant published literature.

A male patient in his late 40s presented with right flank pain and hematuria. Contrast-enhanced CT scan revealed a heterogeneous right upper pole renal mass measuring 7.9 x 7.2 x 6.9 cm with necrosis and vascular involvement, raising suspicion for renal cell carcinoma. He underwent radical nephrectomy with thrombectomy. Histopathological examination showed a high-grade spindle cell tumor, infiltrating the perinephric fat, renal sinus, and adrenal gland. Immunohistochemistry was positive for TLE1, SS18::SSX fusion–specific antibody, CD99, BCL2 and Vimentin, confirming PRSS. Postoperative imaging revealed lung metastases, prompting initiation of ifosfamide and doxorubicin-based chemotherapy which was later modified to gemcitabine and docetaxel due to ifosfamide induced renal tubular acidosis.

PRSS is an aggressive and diagnostically challenging entity that requires a multimodal approach, including histopathology, immunohistochemistry, and molecular testing, where available. Due to the lack of standardized treatment, further studies are essential to establish optimal therapeutic protocols and improve patient outcomes.

Abbreviations

American Joint Committee on Cancer (AJCC); B-cell lymphoma 2 (BCL2), cluster of differentiation molecule 99 (CD99/MIC2); computed tomography (CT); cytokeratin (CK7); epithelial membrane antigen (EMA); immunohistochemistry (IHC); fluorescence in situ hybridization (FISH); Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC); paired-box factor 8 (PAX8); primary renal synovial sarcoma (PRSS); Primitive Neuroectodermal Tumour (PNET); radiotherapy (RT); Renal Cell Carcinoma (RCC); smooth muscle actin (SMA); synovial sarcomas (SS); soft tissue sarcoma (STS); transducin-like enhancer of split 1 (TLE1); TNM; Wilms tumor protein (WT-1).

Keywords
Primary renal synovial sarcoma, PRSS, SS18::SSX fusion–specific antibody, TLE1

INTRODUCTION

Synovial sarcomas (SS) are a type of soft tissue sarcoma (STS) that primarily affect younger individuals, with the extremities being the most frequently involved sites. Diagnosing these rare tumors can pose significant challenges. In addition to standard histological examination and immunohistochemistry (IHC), chromosome rearrangement studies may be required to establish a definitive diagnosis. The incidence of primary renal synovial sarcoma (PRSS) is exceedingly rare, representing only 1-3% of all malignant renal neoplasms in adults[1]. The first case was reported in 1999 and subsequently published by Argani et al. in 2000[2]. Clinical manifestations of primary renal synovial sarcoma typically include the presence of an enlarged abdominal mass, non-specific pain, hematuria, and potential local invasion, along with liver and lung metastases. In this case report, we present the case of a middle-aged male patient diagnosed and treated at a tertiary cancer care hospital.

CASE PRESENTATION

A male patient in his late 40s presented with right flank pain and hematuria. A CT scan of abdomen revealed a heterogeneous mass lesion in the upper pole region of the right kidney, measuring 7.9 x 7.2 x 6.9 cm. This mass was partially exophytic, exhibiting areas of necrosis, hemorrhage and was seen extending superiorly and medially into the perirenal space. T2 weighted MRI demonstrated its heterogenous internal components (**Figure 1A**). The lesion completely involved the upper pole and extended superiorly, showing a loss of fat planes with the right adrenal gland. Additionally, it encroached upon the right renal artery and renal vein. No suspicious retroperitoneal lymph nodes were identified. The left kidney appeared normal in size, with evidence of multiple cortical cysts measuring 16 x 13 mm in the lower pole, without any evident mural nodules. The patient was subsequently scheduled for open right radical nephrectomy and adrenalectomy.

The department of oncopathology received the radical nephrectomy specimen, which measured 24.5 x 10 x 5.5 cm. Upon sectioning, an exophytic tumor measuring 7.5 x 7.2 x 7.0 cm was identified, exhibiting grey-tan firm areas (**Figure 1B**). Additionally, areas of hemorrhage and necrosis were present. The tumor involved the upper pole of the kidney and the interpolar (mid-pole) region. Both the renal capsule and perinephric fat were infiltrated by the tumor. The tumor infiltrated into the renal sinus and the pelvicalyceal system. The adrenal gland was also involved by tumor. A tumor thrombus was noted within the renal vein, while the ureteric and vascular margins were free of tumor involvement.

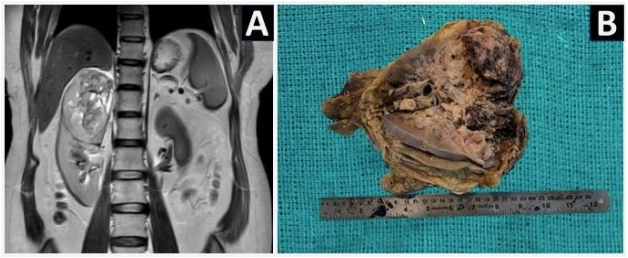


Figure 1. Right renal upper pole mass (A) Coronal T2 weighted image exhibiting cystic components, haemorrhage (T2 dark areas), and fluid levels, as well as the triple sign (the T2-weighted MRI appearance of coexisting high, intermediate and low signal, reflecting solid tissue, haemorrhage/cystic change and fibrous/collagenous or calcified areas); **(B)** Right radical nephrectomy specimen, Gross image showing exophytic tumor with grey-tan, haemorrhagic and necrotic areas

MICROSCOPIC FINDINGS

Microscopic examination revealed a high-grade spindle cell tumor organized in intersecting fascicles within a background of hemangio-pericytomatous vasculature (a distinct branching blood vessel pattern with thin-walled vessels). The spindle cells exhibited moderate to marked nuclear atypia (**Figure 2A-B**), with a brisk mitotic rate of approximately 20-22 per 10 high-power fields and areas of necrosis. The tumor involved the renal capsule, perinephric fat, and renal sinus. The adrenal gland was also infiltrated by the tumor. In addition, a tumor thrombus in the renal vein was also identified. Immunohistochemical analysis showed that the tumor cells were positive for TLE1 (diffuse) (**Figure 2E**), SS18::SSX fusion–specific antibody (diffuse nuclear) (**Figure 2D**), CD99, BCL2, and Vimentin (**Figure 2C**). Focal positivity for AE1/AE3 was noted. The tumor cells were negative for CD34, S100, MUC4, EMA, p63, CD10, SMA, Desmin, PAX8, and CAIX. EMA, PAX8, and AE1/AE3 highlighted the entrapped renal tubules. Six

renal hilar lymph nodes were dissected and were negative for metastasis. Based on the findings, a diagnosis of synovial sarcoma was given. By the FNCLCC system, the tumor corresponded to grade 3 (tumor differentiation score 3, mitotic count score 3, with tumor necrosis present). Kidney sarcoma is staged using the AJCC (American Joint Committee on Cancer) TNM staging system for soft tissue sarcoma, and involvement of another organ (adrenal gland in the present case) would be staged as T3[3].

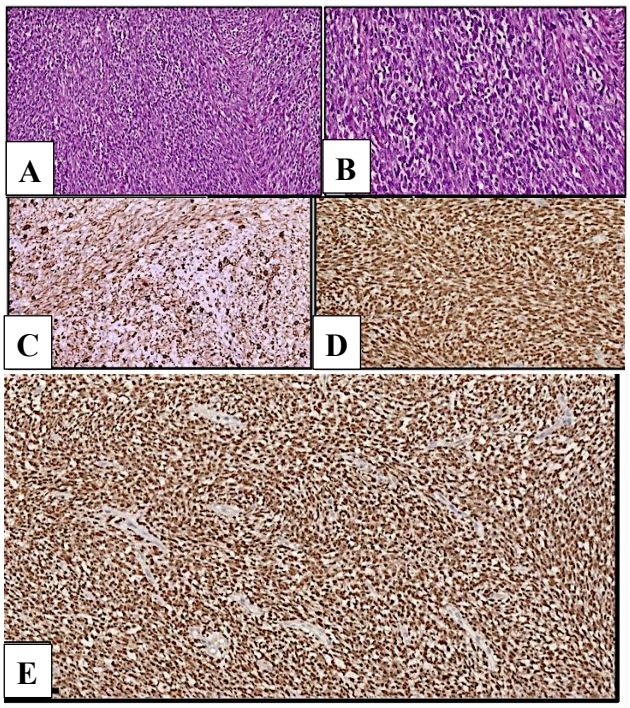


Figure 2. Right radical nephrectomy (A) Cellular spindle cell tumor arranged in intersecting fascicles H&E, X200; **(B)** Monotonous tumor cells with hyperchromatic nuclei and inconspicuous nucleoli H&E, X400; **(C)** High grade spindle cell component highlighted by vimentin, X400); **(D)** Diffuse positivity of tumor cells for SS18::SSX fusion–specific antibody, X400; **(E)** Diffuse positivity of tumor cells by TLE-1, X200

The patient was scheduled for adjuvant chemotherapy (ifosfamide and doxorubicin) followed by radiotherapy (RT). A pre-chemotherapy CT scan revealed multiple metastatic nodules in both lungs. The patient was started on chemotherapy; however, he developed ifosfamide induced proximal renal tubular acidosis after two cycles of chemotherapy. The treatment was then changed to a combination of gemcitabine and docetaxel. The patient remained on follow-up for approximately eight months post-surgery, after which he was lost to follow-up.

DISCUSSION

PRSS is a highly uncommon soft tissue malignancy that typically affects younger patients of both sexes, with a slight male predominance[4]. Soft tissue sarcomas originating from the kidney include leiomyosarcoma, liposarcoma, rhabdomyosarcoma, fibrosarcoma, and malignant fibrous histiocytoma. The age of presentation ranges from 20 to 72 years, with a median age of 35 years[5].

Macroscopically, PRSS typically appears as a yellow-brown, jelly-like, well-encapsulated mass containing cystic, necrotic, and hemorrhagic areas. Tumor sizes range from 1 to 35 cm, with the majority between 5 and 20 cm, frequently replacing the entire kidney and invading the surrounding renal capsule. While lymph node involvement is uncommon, vascular invasion can occur[6].Extension into the inferior vena cava has been reported in three cases. Some poorly differentiated synovial sarcomas exhibit a hemangiopericytoma-like morphology and have frequently been misdiagnosed as hemangiopericytoma at initial evaluation[7].

On histology, synovial sarcoma can present as one of three distinct histologic variants: monophasic, biphasic, and poorly differentiated. Monophasic synovial sarcoma is the most common variant, characterized by spindle cells exhibiting significant cytologic atypia arranged in fascicles. Biphasic tumors contain fascicular spindle cells alongside a substantial epithelial component. Poorly differentiated tumors exhibit high cellularity and rounded tumor cells[8].

There are no specific clinical or imaging features for renal synovial sarcoma; symptoms and CT scan findings often overlap with other malignant renal tumors. The diagnosis of PRSS can be challenging and is typically based on the morphological and immunohistochemistry profile. These tumors exhibit a specific chromosomal translocation t(X;18)(p11.2; q11.2), resulting in the fusion of synovial sarcoma genes SYT-SSX1 and SYT-SSX2, and, more rarely, SYT-SSX4[9]. In current nomenclature, the involved genes are designated SS18 and SSX, and the fusion is written SS18::SSX, denoting a gene fusion of the SS18 gene on chromosome 18 with the SSX gene on the X chromosome[8,9]. Genetic analysis via Fluorescence in situ hybridization (FISH) can confirm the diagnosis by demonstrating the SYT-SSX gene translocation, and is considered the gold standard. Several studies have noted the relatively

poor sensitivity (83-94%) of these molecular tests, along with associated technical and financial challenges[10]. Immunohistochemistry for transducin-like enhancer of split 1 (TLE1) has proven effective in differentiating synovial sarcoma from other soft tissue malignancies. While TLE1 demonstrates robust and diffuse nuclear staining in synovial sarcoma, its specificity is limited, as it can also be positive in up to one-third of non-synovial sarcoma cases[11]. Recently, Baranov et al. assessed SS18-SSX immunohistochemistry in 100 genetically verified primary synovial sarcoma tumors and 300 histological mimics and concluded that SS18-SSX immunohistochemistry exhibited high sensitivity (95%) and specificity (100%) for diagnosing synovial sarcoma[12]. In the present case, however, molecular confirmation of the SS18::SSX rearrangement through FISH or RT-PCR could not be done due to cost constraints.

The commonly positive IHC markers in PRSS include SS18::SSX fusion–specific antibody, epithelial membrane antigen (EMA), cytokeratin (CK7), B-cell lymphoma 2 (BCL2), cluster of differentiation molecule 99 (CD99/MIC2), vimentin, and TLE1. Conversely, CD34, Desmin, Wilms tumor protein (WT-1), smooth muscle actin (SMA), MyoD1, S100, paired-box factor 8 (PAX8), and synaptophysin are typically negative in renal synovial sarcoma and help in differentiation from other differential diagnoses[13].

The common differential diagnoses include sarcomatoid renal cell carcinoma, leiomyosarcoma, solitary fibrous tumors, retroperitoneal sarcomas, fibrosarcoma, malignant peripheral nerve sheath tumors, primitive neuroectodermal tumors, hemangiopericytoma, mixed epithelial and mesenchymal tumors, and Wilms tumor[13].

Distinguishing PRSS from sarcomatoid renal carcinoma can be especially challenging as monophasic synovial sarcoma is more frequently observed in the kidney than biphasic forms. In the largest to date series of 16 cases of genetically confirmed renal synovial sarcoma published by Schoolmeester et al, the authors reported all cases to be of monophasic type[14]. In most cases of sarcomatoid RCC, an underlying histologic subtype of RCC is identifiable with adequate sampling and aids in distinction from PRSS. Solitary fibrous tumors of kidney also share some morphological features with PRSS, however, diffuse positivity of CD 34 and STAT6 IHC along with recurrent NAB2-STAT6 gene fusions help in differentiation from PRSS[14].

PNET is a common mimic of poorly differentiated synovial sarcoma, however, they exhibit more uniform, round cells than do poorly differentiated synovial sarcoma (PDSS). Lack of expression of TLE1 and SS18 fusion antibody, and rearrangements involving the EWSR1 gene helps to distinguish it from PDSS[13,14]. **Table 1** highlights the characteristic IHC and molecular alterations noted in the common differential diagnoses.

Table 1. Differential diagnosis of Primary Renal Synovial Sarcoma with key distinguishing immunohistochemical and molecular features

Entity	Characteristic positive immunohistochemistry	Characteristic molecular features
Primary renal synovial sarcoma	TLE1, SS18::SSX fusion-specific antibody, BCL2, CD99	t(X;18)(p11;q11) translocation
Sarcomatoid renal cell carcinoma	PAX8, EMA, Pancytokeratin	Chromosomal gains and losses, -13q and -4q
Solitary fibrous tumor	CD34, STAT6	NAB2::STAT6 fusion
Leiomyosarcoma	SMA, Desmin, h-caldesmon	Complex karyotype
Malignant peripheral nerve sheath tumor	S100, SOX10, Loss of H3K27me3	Inactivating mutations in NF1, CDKN2A/CDKN2B, TP53, and PRC2
PNET/Ewing family tumors[15]	CD99, NKX2.2, FLI1, ERG	EWSR1 rearrangement

PRSS follows a highly aggressive course, and the prognosis is generally poor. Due to the rarity of these tumors, definitive treatment guidelines are lacking, and management strategies are often based on isolated case reports in the literature. Frontline surgical intervention is usually recommended. Chemotherapy may serve as palliative treatment when upfront surgery is not feasible due to inoperable lesions or when patients are unfit for surgery, or if the disease recurs post-surgery. Limited case reports suggest adjuvant chemotherapy options for soft tissue sarcomas, including doxorubicin, ifosfamide, and etoposide[16].

- **Consider primary renal synovial sarcoma (PRSS) in the evaluation of high-grade spindle cell renal masses, particularly in younger adults**
- **TLE1 marker alone lacks specificity. However, the SS18-SSX fusion-specific antibody, when used in conjunction with morphological assessment, enables a reliable diagnosis and can be used as a surrogate marker when molecular testing is not available**
- **Currently, no standardised chemotherapy regimen for PRSS exists; treatment primarily involves anthracycline- and ifosfamide-based therapies adapted from practices in soft tissue sarcomas. Unfortunately, the prognosis for metastatic disease remains poor**
- **Ifosfamide induced Renal tubular acidosis, can be a significant treatment-limiting toxicity in these patients with prior nephrectomy and diminished renal reserve and should be used with caution**

In practice, renal synovial sarcomas are often managed with ifosfamide-based chemotherapy followed by radical nephrectomy. Kampe et al. reported tumor-free survival in 13 patients with localized soft tissue synovial sarcoma after a 37-month follow-up[17].

Park et al. documented complete remission in a patient with primary renal synovial sarcoma who developed lung metastases four months post-radical nephrectomy after undergoing chemotherapy with doxorubicin and ifosfamide[18].

Recently, Iacovelli et al. reported a meta-analysis of 64 previously published renal synovial sarcomas. The authors reported a median overall survival of 48 months and median disease-free survival of 33.0 months. While 8% patients had metastatic disease at presentation, 36% of patients developed metastases after nephrectomy with lung being the most common site. Patients with metastatic disease had a very poor prognosis[19]. In another case series of 16 cases of PRSS, the authors reported mean age of 46 years and adverse patient outcome in more than 50% of cases[14]. Three patients presented with lung metastases at presentation[13]. In comparison, the present patient presented with lung metastases shortly after surgery, placing him at the more aggressive end of the reported spectrum of published cases.

CONCLUSION

PRSS is a rare and aggressive neoplasm that may mimic other renal cell carcinomas. Accurate diagnosis relies on morphological evidence of spindle cells and the identification of the SS18::SSX translocation, supported by IHC and, where available molecular studies. Effective management necessitates a high index of suspicion combined with the judicious use of immunohistochemistry to exclude common histologic mimics. Published series suggest anthracycline-based regimens may improve outcomes, though the present patient's own course was complicated by toxicity and loss to follow-up, illustrating the unpredictable nature of PRSS.

Conflict of Interest

The authors have no conflicts of interest to disclose.

Acknowledgements

The data that support the findings of this study are available from the corresponding author upon reasonable request. Written informed consent for publication could not be obtained, as the patient was lost to follow-up. All identifying information has been removed from this report, in accordance with ICMJE recommendations. **Source of Funding**: This study was not supported by any funding.

Declaration of Generative AI and AI-assisted technologies

No content, data, interpretation, or conclusion was generated by AI. The text was verified and reviewed by the authors thereby maintaining the integrity and accuracy of the manuscript.

Author Contribution

AG and PS were involved in histopathology reporting, conceptualization, and manuscript preparation. NN, AS, RR, RP and SS were involved

in editing and reviewing the manuscript, whereas DR helped in the radiological correlation.

Author ORCID iDs
Aashna Gupta 0009-0009-9987-427X; Puneet Kaur Somal 0000-0002-5363-3130; Nilay Nishith 0000-0003-2192-7393; Aishwarya Sharma 0000-0003-3978-0027; Rahul Raj 0000-0003-2741-9496; Ravikiran Pawar 0009-0004-0793-062X; Deepander Rathore 0000-0001-7211-4056; Sankalp Sancheti 0000-0001-5810-8733.

Publisher's note
All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article, or claim that may be made by its manufacturer, is not guaranteed or endorsed by the publisher.

References

1. Kataria T, Janardhan N, Abhishek A, Sharan GK, and Mitra S. Pulmonary metastasis from renal synovial sarcoma treated by stereotactic body radiotherapy: a case report and review of the literature. J Cancer Res Ther. 2010;6(1):75–79.
2. Argani P, Faria PA, Epstein JI, Reuter VE, Ali SZ, Man YG, et al. Primary renal synovial sarcoma: molecular and morphologic delineation of an entity previously included among embryonal sarcomas of the kidney. Am J Surg Pathol. 2000;24(8):1087–1096.
3. Amin MB, Edge SB, Greene FL, et al, eds. AJCC Cancer Staging Manual. 8th ed. New York, NY:Springer; 2017.
4. 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(4):2241-2245.
5. Modi G, Madabhavi I, Panchal H, Anand A, Patel A, Parikh S et al. Primary synovial sarcoma of kidney: a rare differential diagnosis of renomegaly. Case Rep Pathol. 2014;2014:657497.doi:10.1155/2014/657497.
6. 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(1):128-132. doi:10.1159/000379740.
7. Grampurohit VU, Myageri A, Rao RV. Primary renal synovial sarcoma. Urol Ann. 2011;3(2):110-113. doi:10.4103/0974-7796.82182.
8. Blay JY, von Mehren M, Jones RL, Martin-Broto J, Stacchiotti S, Bauer S, et al. Synovial sarcoma: characteristics, challenges, and evolving therapeutic strategies. ESMO Open. 2023;8(5):101618. doi:10.1016/j.esmoop.2023.101618.
9. Alzahrani I, Albqami N, Alkhayal A, AlOudah N, Alyami A. Primary Renal Synovial Sarcoma Presenting as Haemorrhagic Shock: A Rare Presentation. J Kidney Cancer VHL. 2021;8(3):1-4. Published 2021 Sep 3. doi:10.15586/jkcvhl.v8i3.187.
10. Sun B, Sun Y, Wang J, Zhao X, Zhang S, Liu Y, et al. The diagnostic value of SYT-SSX detected by reverse transcriptase-polymerase chain reaction (RT-PCR) and fluorescence in situ hybridization (FISH) for synovial sarcoma: a review and prospective study of 255 cases. Cancer Sci. 2008;99(7):1355-1361.
11. Thway K, Fisher C. Synovial sarcoma: defining features and diagnostic evolution. Ann DiagnPathol. 2014;18(6):369–80. https://doi.org/10.1016/j.anndiagpath.2014.09.002.
12. Baranov E, McBride MJ, Bellizzi AM, Ligon AH, Fletcher CDM, Kadoch C, et al. A novel SS18-SSX fusion-specific antibody for the diagnosis of synovial sarcoma. Am J Surg Pathol. 2020;44(7):922–33.
13. Mastoraki A, Schizas D, Karavolia DM, et al. Primary Synovial Sarcoma of the Kidney: Diagnostic Approach and Therapeutic Modalities for a Rare Nosological Entity. J Pers Med. 2022;12(9):1450. doi:10.3390/jpm12091450.
14. 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(1):60-65.
15. Khoury JD. Ewing sarcoma family of tumors: a model for the new era of integrated laboratory diagnostics. Expert Rev Mol Diagn. 2008;8(1):97-105.
16. Ozkan EE, Mertsoylu H, and Ozardali HI. A case of renal synovial sarcoma treated with adjuvant ifosfamide and doxorubicin. Intern Med. 2011;50(15):1575–1580, 2011.
17. Kampe CE, Rosen G, Eilber F, Eckardt J, Lowenbraun S, Foster J, et al. Synovial sarcoma. A study of intensive chemotherapy in 14 patients with localized disease. Cancer. 1993;72(7):2161-2169.

18. 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(1):62-65.
19. Iacovelli R, Altavilla A, Ciardi A, et al. Clinical and pathological features of primary renal synovial sarcoma: analysis of 64 cases from 11 years of medical literature. BJU Int. 2012;110(10):1449-1454.

This article is an Open Access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited and it is not used for commercial purposes; 2026, Gupta et al., Applied Systems and Discoveries Journals.