CANADIAN JOURNAL OF UROLOGY

Tech Science Press

Doi:10.32604/cju.2026.076969
CASE REPORT

Renal Ewing sarcoma with an unusual presentation: a case report

Felix Lübbersmeyer, Paula Lindfeld, Margit Fisch, Roland Dahlem, Markus Von Deimling*

Department of Urology, University Medical Center Hamburg-Eppendorf, Hamburg, Germany

LÜBBERSMEYER F, LINDFELD P, FISCH M, DAHLEM R, VON DEIMLING M. Renal Ewing sarcoma with an unusual presentation: a case report. *Can J Urol* 2026;33(4):1019–1025.

Backgrounds: *Ewing Sarcoma (ES) is an aggressive pediatric bone tumor requiring multimodal treatment. Primary renal ES is extremely rare and often presents with nonspecific symptoms, potentially delaying diagnosis. Imaging aids detection, while histopathological and molecular analyses confirm the diagnosis.*
Case description: *We report on a 31-year-old female who was initially treated for pyelonephritis. As computerized tomography suggested a malignant kidney tumor with an extensive inferior vena cava thrombus, nephrectomy and caval ligation were performed. Histopathology and further staging confirmed primary renal ES, which was subsequently treated with adjuvant polychemotherapy.*
Conclusions: *The present case highlights the variable clinical spectrum of malignant kidney tumors. ES should be considered in the differential diagnosis of atypical renal masses.*

Key Words: Ewing sarcoma, kidney tumor, renal mass, urology, case report

Introduction

Ewing sarcoma (ES) is a high-grade tumor and typically affects adolescents in the second decade of life.[1] As ES predominantly develops in bones, it can also occur primarily extraosseous.[1] The incidence of ES of the bones is approximately 0.1/100.000/year.[2] Primary manifestations at extraskeletal sites, including the kidney, have also been reported but remain exceedingly rare.[3] To date, only a limited number of cases of primary ES of the kidney have been described in the literature.[4]

Patients with primary ES of the kidney are considerably younger than those with more common renal malignancies, such as renal cell carcinoma (RCC), and tumors are characterized by an extremely aggressive biological behavior.[3] Their symptoms often mimic more common urological diagnoses, leading to a delay in adequate treatment. In addition, one-third present with metastatic disease at the time of initial diagnosis, which is associated with poor survival rates.[3] Therefore, clinicians should be aware of this rare differential diagnosis of atypical renal masses, especially in young patients. Herein, we report a case with an uncommon presentation of a renal tumor that turned out to be a primary ES of the kidney.

It was determined that this study did not require consultation with the Ethics Committee of the Hamburg Medical Association (application number 2026-300706-WF). The handwritten informed consent was obtained from the patients. Besides, this study was prepared according to the CARE case report guideline,[5] and a CARE checklist was provided. Please see Supplementary Material S1 for more details.

Case Report

In July 2023, a 31-year-old woman presented with fever and left-sided flank pain at an emergency room in a non-academic hospital. The flank pain had started a month ago and was resistant to oral analgesics. Prior to her first presentation, the patient was treated with ciprofloxacin, as a urinary tract

Received date 30 November 2025
Accepted for publication 28 February 2026
Published online 21 August 2026

*Corresponding Author: Markus Von Deimling.
Email: markus.von-deimling@uke.de

infection was suspected, and reported that she had been diagnosed with an acute deep vein thrombosis in her right leg. Therapeutic anticoagulation was already initiated. There were no pre-existing medical conditions or further medications. Family history was negative for malignancies. The physical examination revealed an overall reduced general condition and percussion tenderness of the left kidney. Abnormal laboratory findings were a hypochromic, microcytic anemia (hemoglobin 7.7 g/dL) and an elevated C-reactive protein (CRP 120 mg/L), while the leukocyte count and glomerular filtration rate were within normal limits.

On initial abdominal ultrasound, the left kidney showed a hyperechogenic structure accompanied by general renal hyperperfusion with suspicion of a kidney abscess. According to a computerized tomography (CT) scan of the abdomen (Figure 1), the patient was initially diagnosed with nephrolithiasis with abscessing pyelonephritis, and consequently transferred to a department of urology in another non-academic hospital. Primary management included ureteral stent placement and intravenous antibiotic treatment with piperacillin/tazobactam. Reassessment of the CT scan on the following day led to reinterpretation of the inhomogeneous mass as a tumor of the left kidney (largest extent 8.1 cm), highly suspicious of a renal malignancy. A triangular hyperintensity next to the tumor was identified as calcification (Figure 1). Moreover, a tumor thrombus extended into the hepatic segment of the inferior vena cava (IVC) (Mayo III tumor thrombus) and caused thrombosis of the left external iliac vein and beyond the right superficial femoral vein. The IVC showed multiple collateral veins, and infiltration of the IVC wall was suspected. Chest CT showed no lymphatic or distant metastases. In summary, staging indicated a locally advanced malignant kidney tumor.

Upon diagnosis, the patient was referred to our urological department for further diagnostic work-up and surgical treatment. Because of extensive thrombosis, full therapeutic anticoagulation with low-molecular-weight heparin was continued. The patient had a preoperative hemoglobin level of 9.4 g/dL. Transthoracic echocardiography revealed only mild left ventricular hypertrophy. Furthermore, a cardiothoracic surgery consultation was obtained, but the use of a heart-lung machine was not deemed necessary. Following interdisciplinary case discussion, radical tumor nephrectomy, attempted removal of the tumor thrombus, and diagnostic lymphadenectomy via a Chevron incision were performed by an experienced surgeon (RD). Intraoperatively, extensive tumor infiltration of the IVC wall, together with

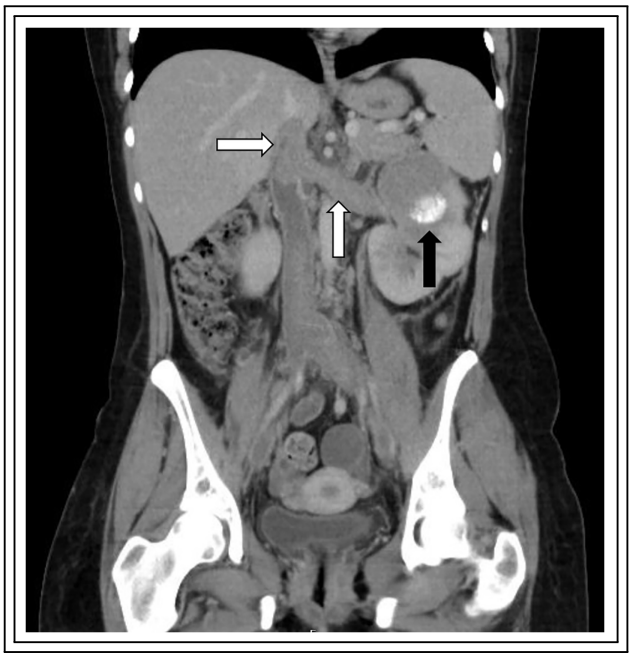

FIGURE 1. CT scan of the abdomen, portal venous phase. Coronal view displaying a mass of the left kidney with a calcification (black arrow) and a tumor thrombus (white arrows) with extension into the inferior vena cava

the unresectable thrombus extending beyond the left external iliac vein and the associated high risk of postoperative thromboembolic complications, led to the decision to ligate the IVC. Due to an intraoperative blood loss of 3.5 L, the patient received five red blood cell concentrates and twelve units of fresh frozen plasma. She spent the next two days in the intermediate care unit and was treated with meropenem for a postoperative infection. No further grade II or higher complications according to the Clavien-Dindo classification occurred. Hemoglobin levels reached a nadir of 7.5 g/dL on postoperative day 3 and subsequently recovered spontaneously without the need for further transfusion. The patient was discharged on the 8th day after surgery with a hemoglobin level of 8.8 g/dL. Four weeks after surgery, hemoglobin levels had increased to 11.5 g/dL.

Pathological examination revealed a sharply demarcated, multinodular, whitish tumor with soft consistency, showing fine encapsulation and partial necrosis (Figure 2). Microscopic examination showed a malignant small round blue cell tumor with marked hemangioinvasion (Figure 3). Immunohistochemistry showed positive staining for cluster of differentiation 99 (CD99) (Figure 4), vimentin, NK2 homeobox 2 (NKX2.2), CD117,

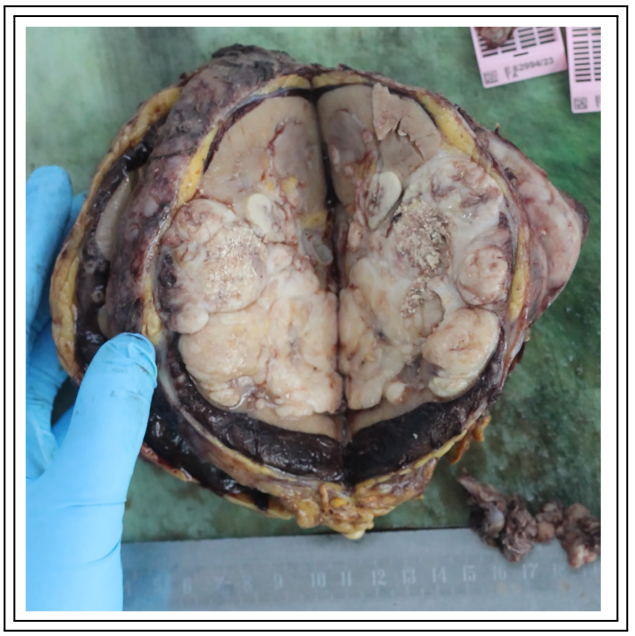

FIGURE 2. Macroscopic appearance of the surgical specimen. Incised, partially necrotic tumor with sharply defined, multinodular, whitish appearance

synaptophysin, and retinoblastoma protein (Rb). Markers for carcinoma (AE1/3, cytokeratin 7 [CK7]), melanoma (S100 protein, melanoma antigen recognized by T cells 1 [MelanA]), lymphoma (CD3, CD20), and neuroendocrine tumors (chromogranin, insulinoma-associated protein 1 [INSM1]) were negative. Marker of proliferation Ki67 index was higher than 80%. *EWS RNA-binding protein 1 (EWSR1)* translocation was detected by fluorescence *in situ* hybridization (FISH). After exclusion of a secondary manifestation by positron emission tomography/computed tomography (PET/CT) and bone marrow biopsy, the final diagnosis of a primary ES of the kidney was established.

Following the Euro Ewing 2012 protocol, adjuvant polychemotherapy was initiated four weeks after surgery.[2] The Euro Ewing 2012 protocol encompasses nine cycles of induction chemotherapy (alternating vincristine, doxorubicin, cyclophosphamide, and ifosfamide, etoposide) and five cycles of consolidation chemotherapy (alternating ifosfamide, etoposide, and vincristine, cyclophosphamide). The administration of the first cycle of ifosfamide and etoposide, and the second cycle of vincristine, doxorubicin, and cyclophosphamide, was postponed by two weeks due to nausea/fatigue, as well as neurotoxicity and thrombocytopenia, respectively. All other cycles of chemotherapy were administered

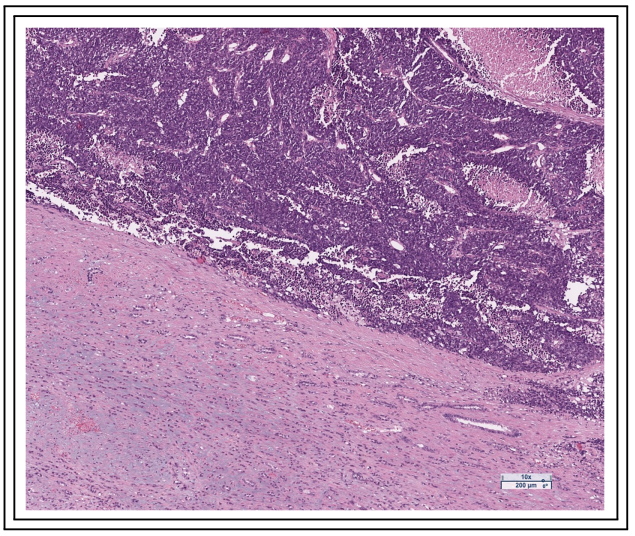

FIGURE 3. Cross section of the Ewing sarcoma (hematoxylin and eosin stain)

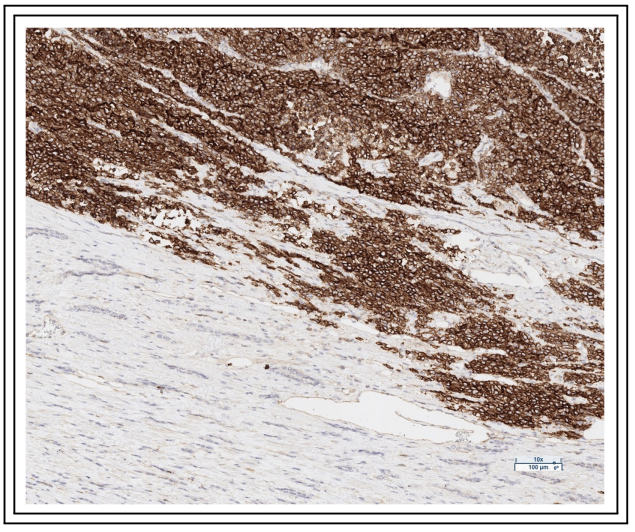

FIGURE 4. Cross section of tumor margin of the Ewing sarcoma. Tumor cells show strong CD99 immunostaining

with a 14-day interval. After the ninth cycle, the patient refused further treatment. Oncological follow-up was initiated. CT scan of the chest and abdomen performed 14 months postoperatively demonstrated no signs of recurrence (Figure 5). The previously described preoperative thrombosis had resolved completely, as imaging at five weeks postoperatively had already shown regression of the tumor thrombus of the IVC with extension into both external iliac veins. The patient is recurrence-free 26 months after surgery and 22 months after the end

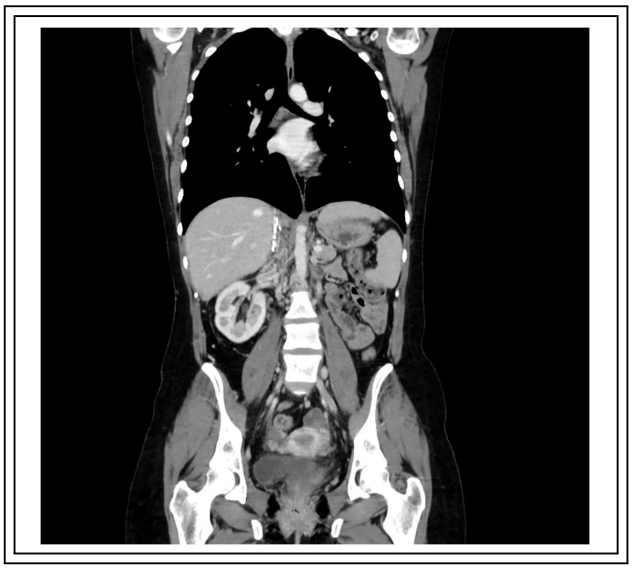

FIGURE 5. CT scan of the abdomen 14 months postoperatively

of adjuvant therapy. The case is summarized as a timeline (Figure 6).

Discussion

We report the case of a 31-year-old female patient with a renal ES and a tumor thrombus of the IVC. An unusual presentation with fever and flank pain delayed the correct diagnosis, which was established by imaging and histopathological examination. Interdisciplinary treatment, including radical nephrectomy and adjuvant chemotherapy, resulted in a long-term remission. However, the following aspects warrant further discussion.

Establishing the final diagnosis of renal ES is challenging. Kidney tumors are usually detected by non-invasive conventional imaging; however, this may not be sufficient for detecting ES. As such, PET/CT or whole-body magnetic resonance imaging are the preferred modalities for staging.[2] Moreover, the final diagnosis is often established after an extensive pathological work-up. Tumor cells are small and round, contain fine chromatin, and exhibit strong membranous expression of CD99 in immunohistochemistry. Nesting or trabecular growth with collagenous septa, perivascular or pseudopapillary formations, and multicystic change may appear as rare architectural features in extraskeletal ES.[1] As performed in this case, molecular diagnostics are mandatory (e.g., FISH, reverse transcription-polymerase chain reaction) to detect specific gene fusions of ES (usually involving *EWSR1*).[1] Our patient harbored an *EWSR1* translocation, which is present in approximately 85% of ES cases (recurrent t(11, 22; q24; q12) translocation).[1] The development of ES-specific liquid biopsy-based biomarkers has high potential for guiding diagnosis, risk stratification, and therapy monitoring.[6]

Primary renal ES is extremely rare. While the age of the patient in our case and tumor size are similar to cases reported in the literature (median age at diagnosis of 28, median size of 13 cm),[3] the initial presentation with fever, left-sided flank pain, and a recently diagnosed acute deep vein thrombosis combined with elevated CRP was very unusual and led to initial diagnosis of nephrolithiasis with abscessing pyelonephritis. According to Risi et al., who summarized clinical data on 116 primary ES of the kidney from a period of almost 40 years, the most common symptoms include pain, hematuria, and renal mass.[3] Of note, no patient was asymptomatic, and 55% of patients were male.[3] Although one-third of patients have metastatic disease at the time of initial presentation, with lungs (60%) and liver (37%) being the most common metastatic sites, staging of the patient in our case showed no signs of distant metastases.[3] Moreover, while Risi et al. reported a median overall survival (OS) across all stages of 26.5 months, our patient is without any signs of disease recurrence 26 months after surgery.[3]

There is a discrepancy between guideline recommendations for preoperative biopsy and clinical practice in patients with renal ES. According to guidelines, a diagnostic biopsy is required before starting neoadjuvant treatment.[2] Yet Rowe et al. reported that only 28 of 85 patients (33%) with renal ES received a diagnostic biopsy, while the remaining patients were diagnosed after surgical treatment.[7] Reasons for avoiding biopsies in suspected renal malignancies reported in the literature include risk of bleeding, injuring adjacent structures, tumor cell seeding, and diagnostic delay due to equivocal histology.[7,8] In addition, sarcomas and localized RCCs may present with similar radiologic features. Because localized RCCs are often managed without preoperative biopsy, clinicians may proceed directly to surgery under the presumptive diagnosis of RCC. Nevertheless, omitting preoperative biopsy and neoadjuvant treatment for primary ES of the kidney is contrary to guideline recommendations and may worsen the prognosis.[2] Furthermore, evidence suggests that the risk associated with renal tumor biopsy may be overestimated. A systematic review and meta-analysis by Marconi et al. on percutaneous renal tumor biopsy reported only one case of tumor cell seeding and three cases

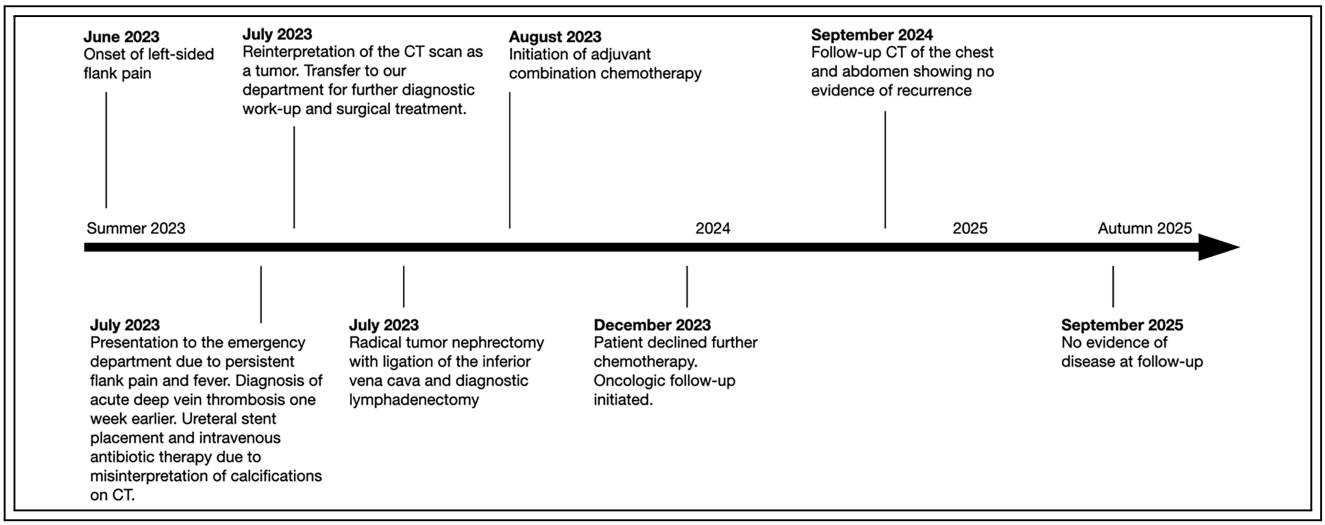

FIGURE 6. Information from this case report organized as a timeline

with Clavien-Dindo grade ≥ 2 complications among 1.919 cases across 17 studies.[8] Hence, renal biopsy represents an important diagnostic tool, particularly in patients younger than 40 years, in whom non-hereditary renal cell carcinoma is less common.[7] This diagnostic dilemma is reflected in our case. We performed a nephrectomy without a preoperative biopsy, as the radiographic features prompted a malignant tumor, with RCC being the most likely differential diagnosis, and to avoid hemodynamic complications from the tumor thrombus.

Guidelines recommend a multimodal treatment for extraskeletal ES, consisting of induction chemotherapy, local therapy (surgical resection and/or radiotherapy), followed by consolidation chemotherapy.[2] The chemotherapy regimen with up to nine cycles of vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide showed superiority over vincristine, ifosfamide, doxorubicin, and etoposide in terms of effectiveness, toxicity, and duration.[9] However, there are no prospective studies on the treatment of renal ES. Therefore, prospective registration of cases is encouraged to optimize treatment.[2]

Given the rarity of renal ES, investigating prognostic factors is very valuable. Risi and colleagues noticed that the median OS in patients with metastatic disease (24 months) was significantly shorter than in patients without metastases (median survival not reached).[3] Using univariable Cox regression analysis, the authors reported a fourfold increase in the relative risk of death in patients with M1 disease compared to patients with M0 disease (HR 4.2; 95% CI 1,8-9,9; $p = 0.001$).[3] Interestingly, patient age (greater or lower than 15 years) and primary tumor diameter (above or below the median diameter) were not associated with OS.[3] Moreover, a retrospective study of 30 patients with primary Ewing sarcoma of the kidney treated at the MD Anderson Cancer Center between 1990 and 2013 found that the use of perioperative chemotherapy was significantly associated with improved event-free survival ($p < 0.0001$) and OS ($p = 0.036$).[10] In addition, guidelines for osseous ES list tumor volume, elevated lactate dehydrogenase levels, axial localization, older age, and incomplete resection as further adverse prognostic factors.[2]

A key strength of this case report is the demonstration of interdisciplinary collaboration in diagnosis and treatment, as well as the provision of long-term follow-up data for this rare entity, with the patient achieving sustained remission. Nevertheless, certain limitations in diagnostic accuracy and therapeutic management should be acknowledged. The patient was initially treated for a suspected urinary tract infection due to nephrolithiasis complicated by pyelonephritis with abscess formation, leading to ureteral stent placement and intravenous antibiotic therapy. Reassessment of the CT imaging on the following day led to the suspicion of an underlying renal malignancy. This diagnostic evolution may have contributed to a delay in establishing the correct diagnosis, with potential implications for prognosis. However, the present case highlights the pitfalls encountered in the clinical management of rare diseases.

In summary, primary Ewing sarcoma of the kidney is a rare disease, and the present case highlights the complex path to final diagnosis due to

its presentation mimicking more common urological conditions. At first glance, deep vein thrombosis was not connected with fever and left-sided flank pain caused by a suspected pyelonephritis. However, further diagnostic work-up revealed that the thrombosis resulted from a primary Ewing sarcoma of the kidney with vena cava involvement. The tumor was treated with radical surgery and adjuvant polychemotherapy.

Conclusion

The present case illustrates the variable clinical spectrum of malignant kidney tumors. As such, renal Ewing sarcoma is a rare disease that requires complex diagnostics and interdisciplinary treatment, including surgical resection and perioperative polychemotherapy. Despite aggressive treatment, the median overall survival is poor. Further case series and prospective registries are needed to gain a greater understanding of this entity.

Acknowledgement

Language editing assistance was provided using ChatGPT (OpenAI).

Funding Statement

The authors received no specific funding for this study.

Author Contributions

The authors confirm the following contribution to the paper as follows: study conception and design: Felix Lübbersmeyer, Markus von Deimling; data collection: Paula Lindfeld, Felix Lübbersmeyer, Markus von Deimling; analysis and interpretation of the results: Roland Dahlem, Margit Fisch, Paula Lindfeld, Felix Lübbersmeyer, Markus von Deimling; draft manuscript preparation: Felix Lübbersmeyer, Markus von Deimling. All authors reviewed and approved the final version of the manuscript.

Availability of Data and Materials

Not applicable.

Ethics Approval

The hospital where the patient initially received treatment is located in the rural surroundings of Hamburg, Germany. The name of the institution has been deliberately omitted. Disclosure of the institution's identity would not contribute to the scientific interpretation or educational value of this case and is not necessary for understanding the clinical course. This approach reflects our commitment to professional fairness and the avoidance of potentially unwarranted attribution or reputational implications. In view of the fact that the patient data that are the subject of this study can no longer be attributed to an identifiable individual, the study does not constitute a "research project involving human beings" as defined in Section 9(2) of the Hamburg Chamber Act for the Medical Professions and therefore does not fall within the scope of research projects requiring consultation pursuant to Section 15(1) of the Professional Code of Conduct for Hamburg Physicians. Accordingly, it was determined that this study did not require consultation with the Ethics Committee of the Hamburg Medical Association (application number 2026-300706-WF). Written informed consent was obtained from the patient. This case report was prepared in accordance with the CARE case report guidelines, and a completed CARE checklist is provided. Please see Supplementary Material S1 for further details.

Conflicts of Interest

The authors declare no conflicts of interest.

Supplementary Materials

The supplementary material is available online at https://www.techscience.com/doi/10.32604/cju.2026.076969/s1.

References

1. WHO—classification of tumours editorial board: Ewing sarcoma. In: *WHO classification of tumours: soft tissue and bone tumours*. Geneva, Switzerland: OMS; 2020. p. 323–325. [cited 2026 Feb 27]. https://publications.iarc.who.int/Book-And-Report-Series/Who-Classification-Of-Tumours/Soft-Tissue-And-Bone-Tumours-2020
2. Strauss SJ, Frezza AM, Abecassis N et al. Bone sarcomas: ESMO-EURACAN–GENTURIS-ERN PaedCan clinical

practice guideline for diagnosis, treatment and follow-up. *Annal Oncol* 2021;32(12):1520–1536. doi:10.1016/j.annonc.2021.08.1995.
3. Risi E, Iacovelli R, Altavilla A et al. Clinical and Pathological features of primary neuroectodermal tumor/ewing sarcoma of the kidney. *Urology* 2013;82(2):382–386. doi:10.1016/j.urology.2013.04.015.
4. Findlay BL, Shinder BM, Fatima A et al. Primary renal ewing sarcoma: a case report and review of the literature. *J Ren Cancer* 2019;2(1):15–19. doi:10.36959/896/603.
5. Riley DS, Barber MS, Kienle GS et al. CARE guidelines for case reports: explanation and elaboration document. *J Clin Epidemiol* 2017;89:218–235. doi:10.1016/j.jclinepi.2017.04.026.
6. Janssen FW, Lak NSM, Janda CY et al. A comprehensive overview of liquid biopsy applications in pediatric solid tumors. *npj Precis Oncol* 2024;8(1):172. doi:10.1038/s41698-024-00677-9.
7. Rowe RG, Thomas DG, Schuetze SM, Hafez KS, Lawlor ER, Chugh R. Ewing sarcoma of the kidney: case series and literature review of an often overlooked entity in the diagnosis of primary renal tumors. *Urology* 2013;81(2):347–353. doi:10.1016/j.urology.2012.10.016.
8. Marconi L, Dabestani S, Lam TB et al. Systematic review and meta-analysis of diagnostic accuracy of percutaneous renal tumour biopsy. *Eur Urol* 2016;69(4):660–673.
9. Brennan B, Kirton L, Marec-Bérard P et al. Comparison of two chemotherapy regimens in patients with newly diagnosed Ewing sarcoma (EE2012): an open-label, randomised, phase 3 trial. *Lancet* 2022;400(10362):1513–1521. doi:10.1016/s0140-6736(22)01790-1.
10. Tarek N, Said R, Andersen CR et al. Primary ewing sarcoma/primitive neuroectodermal tumor of the kidney: the MD anderson cancer center experience. *Cancers* 2020;12(10):2927. doi:10.3390/cancers12102927.

