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Case Report

# Renal Ewing sarcoma with inferior vena cava (IVC) tumor thrombosis; A case report ☆

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ABSTRACT

Ewing sarcoma (ES) is a rare group of undifferentiated tumors that originate from neuroectoderm. Although the overall prognosis is poor, early diagnosis and treatment by a multidisciplinary team with multimodal therapy can improve outcomes. Therefore, we present a 22-year-old female patient with primary renal ES with tumor thrombosis up to the vena cava who had radical nephrectomy and IVC tumor thrombectomy followed by adjuvant chemotherapy because a preoperative percutaneous biopsy was confirmed the diagnosis.



## Introduction

Ewing sarcoma (ES), also known as primitive neuroectodermal tumors (PNET), is a rare group of undifferentiated tumors that originate from neuroectoderm and affect young adults with rapid clinical progress and a poor prognosis, secondary to delayed diagnosis and early-stage metastasis [1]. However, kidney involvement is rarely seen and has a worse prognosis. Renal ES presents highly malignant, grows rapidly, and metastases early to the lung, bone and lymph node [2]. The fundamental challenge is the precise diagnosis allowing the patient's appropriate management, as this kind of kidney neoplasm mandates more aggressive treatment compared with other primary kidney tumors [3]. It is required to prospectively monitor these patients in order to get additional information about the biology of tumors and the function of different therapeutic options when specific tumor characteristics are considered, such as the association between tumor thrombosis and pulmonary metastasis [4]. Therefore, we present a rare case of renal ES with with inferior vena cava (IVC) tumor thrombosis.

## Case presentation

A 22-year-old female patient was admitted with chief complaints of swelling in the left upper quadrant and palpable abdominal mass. After several visits to different doctors, according to the normality of the patient's laboratory tests, she was diagnosed with muscle mass. However, after 4 months, the patient was referred to our clinic due to worsening swelling and slight weight loss.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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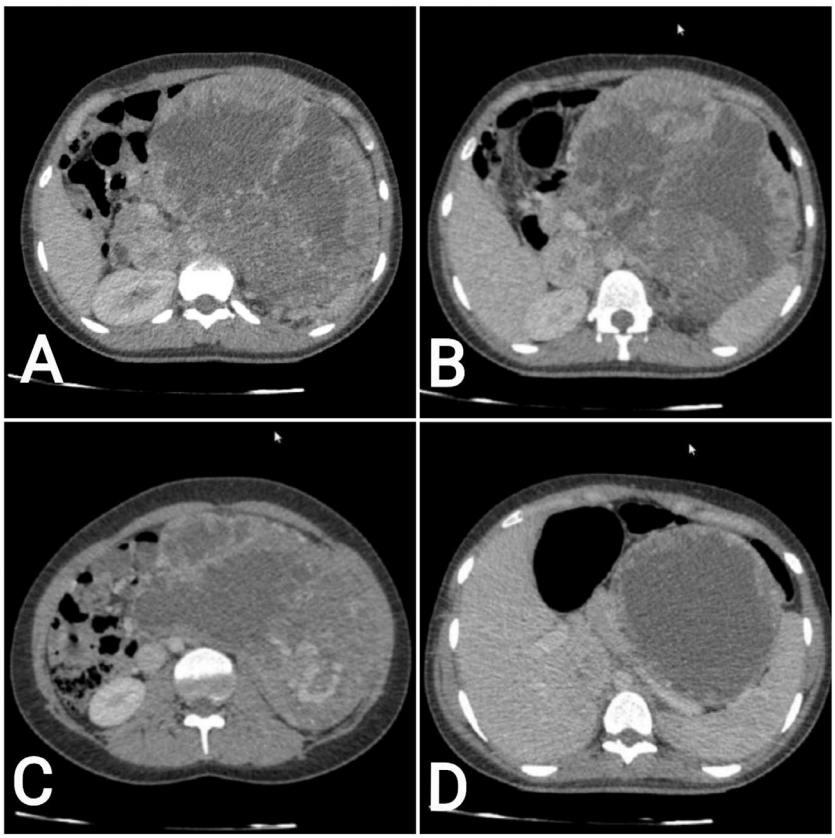


**Fig. 1 – (A) Contrast-enhanced abdominopelvic CT scan reveals a large heterogeneous enhancing mass lesion containing necrotic areas occupying the left renal fossa; (B, C) Heterogeneous enhancement and dilatation of the IVC, which indicates tumor thrombosis due to IVC invasion; (D) Extension of the tumor into the epigastric region is detectable, causing a significant pressure effect on the stomach. CT, Computed tomography; IVC, Inferior vena cava.**

No urinary symptoms such as hematuria and dysuria were found. Her past medical history and family history of malignancy were unremarkable.

Results of routine blood and urine examination as well as tumor markers including AFP, BHCG, CEA, CA199, and CA125 were within the normal range. However, a slight increase in lipase and bilirubin was observed. Ultrasound was performed for the patient, and a large heterogeneous solid cystic mass was observed that extended from the epigastrium to the lower part of the abdomen. Therefore, the patient underwent enhanced computed tomography (CT) for further evaluation. Enhanced CT demonstrated a very large heterogeneous mass (280 mm × 180 mm × 200 mm) from the left kidney with areas of necrosis and high vascularity and aneurysmal changes in the internal vascular structures (Fig. 1).

The remarkable thing is the expansion of the tumoral tissue through the left renal vein to the IVC (Fig. 2). Thrombus was seen in the IVC with a length of approximately 150 mm, which extended to the vicinity of the suprahepatic IVC. Adhesion of the mass was observed to the pancreas, but no evidence of pancreatic invasion was observed. Also, the adhesion of the mass on the front of the splenic vein was evident, but no invasion was observed. According to findings of CT, these suggested kidney tumoral lesion with extension to the IVC, and the diagnosis of malignant perivascular epithelioid cell tumor (PEComa) was proposed.

Further clinical investigations, including a chest CT, were performed for evaluation of extension of tumoral thrombosis in IVC which shows sparing of suprahepatic IVC. Ultimately, an ultrasound-guided biopsy was performed on the patient in order to confirm the diagnosis. The biopsy of the kidney mass revealed Ewing sarcoma/primitive neuroectodermal tumor.

Finally, due to the diagnosis of renal Ewing sarcoma, the patient underwent some procedures, including tumor resection surgery, left nephrectomy, distal pancreatectomy, left colon resection and colorectal anastomosis, and IVC repair. After these surgeries, the patient underwent chemotherapy.

## Discussion

In current study, we present a 22-year-old female patient with chief complaints of swelling in the left upper quadrant. Results of routine blood and urine examination as well as tumor markers were normal. Enhanced CT showed the expansion of the tumoral tissue through the left renal vein to the IVC. The biopsy of the kidney mass revealed Ewing sarcoma/primitive neuroectodermal tumor.

ES rarely presents as a primary kidney tumor. Ewing's sarcoma of the kidney is seen in the young age group (mean age of 28-34 years) [5]. They are initially asymptomatic and

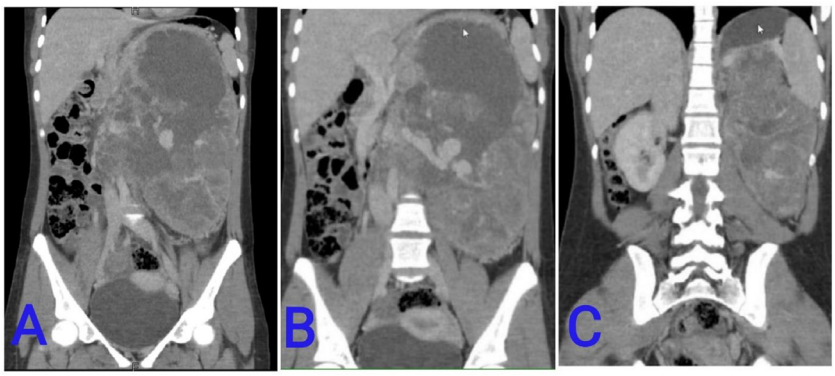


**Fig. 2 – (A, B) Dilated intraregional artery originated from left renal artery branches is visible; (C) The left kidney has been entirely replaced by a hypodense, heterogeneous mass lesion that contains necrotic areas.**

when large enough, usually present with symptoms like flank pain and hematuria. Preoperative diagnosis of the disease is challenging as its clinical symptoms are nonspecific, including pain (54%), hematuria (29%), and renal mass (28%) [6].

Furthermore, radiologic modalities such as CT and MRI are ineffective at distinguishing renal ES from renal cell carcinomas (RCC). Imaging evaluations frequently reveal unclear indications as well as central necrosis and hemorrhage, which makes a preoperative diagnosis of renal ES more challenging [7]. The most frequently, the diagnosis was based on postoperative pathology findings. The renal ES is histologically constituted of uniformly small, round cells. The major differential diagnosis for this histomorphology in the kidney consists of Wilms tumor, malignant lymphoma, synovial sarcoma, solid variant of alveolar rhabdomyosarcoma, clear cell sarcoma of the kidney, small-cell neuroendocrine carcinoma, desmoplastic small round blue cell tumor, and small cell carcinoma. Poorly differentiated small round cell tumors have similar morphologic features but differing prognoses, making diagnosis notoriously difficult but crucial [8]. Improvements in immunohistochemistry and molecular-genetic approaches are beneficial, as various positive and negative immunohistochemical markers provide a better means of differentiation. CD99 and FLI-1 were frequently detected in ES/PNET, including those that originated in the kidney. CD45, WT-1, and Desmin have all been found to be negative. These immunohistochemistry markers created a helpful, albeit imprecise, diagnosis panel for renal ES because several of the markers are common to small, round blue tumors [5]. Additional molecular testing is advised in undefinable instances. A fixed chromosomal translocation t (11:22) between the genes EWS (22q12) and FLI-1 characterizes ES/PNET (11q24) [9].

Chew et al. [10] reported a 33-year-old man with left flank pain developed insidiously over several months. Abdominal computed tomography showed a large multilobulated heterogeneous enhancement lesion in left kidney with an extension of tumor thrombus in the left renal vein. The positive immunohistochemically staining for CD99 confirm the diagnosis of renal ES. For treatment approach, patient was undergoing aggressive treatment with radical nephrectomy, and adjuvant combination chemotherapy.

The results of the survival analysis indicate that the disease is aggressive. Murugan et al. [6] found that, despite surgical treatment combined with systemic chemotherapy, 57% of patients with localized or locally progressed disease developed metastasis in a mean of 14 months. Furthermore, some investigations indicate that renal ES is more aggressive than its nonrenal counterparts. It has been demonstrated that more than 65% of patients with renal ES have metastatic disease.

In contrast, metastasis is observed in 25% of patients with nonrenal ES [5,6]. Moreover, it appears plausible to believe that a considerable majority of patients with localized disease have subclinical metastasis, given that up to 90% of individuals with nonmetastatic bone ES relapsed after local treatment [11].

Due to the extreme rarity of renal Ewing's sarcoma, there is no conventional treatment for this disease; instead, clinical protocols for osseous Ewing's sarcoma are primarily utilized. At the moment, radical nephrectomy followed by chemotherapy is regarded favorably. In the presence of venous tumor thrombus, which was frequently documented in renal ES and was linked to the occurrence of pulmonary metastases, surgical management may include cavotomy. Chemotherapy has been proven to improve renal ES survival [12]. Most chemotherapy regimens include numerous drugs, such as ifosfamide, doxorubicin, vincristine, cyclophosphamide, etoposide, or actinomycin D. In recent years, neoadjuvant chemotherapy after biopsy has been tolerated; however, its survival benefits must be confirmed in the future. As a salvage therapy for specific lesions, radiation has shown modest success. Nonetheless, there are differing opinions on whether radiotherapy should be used as the primary treatment technique [13]. Unfortunately, most renal ES cases are poorly managed, with barely one-third of patients with primary renal ES undergoing biopsy prior to surgery and less than 70% of biopsied patients receiving neoadjuvant therapy before nephrectomy. Most of this poor management is due to the inadequacy of noninvasive preoperative examinations to distinguish ES from prevalent malignant diseases (e.g., RCC) [12,13]. As a result, doing a percutaneous biopsy in young patients with large renal tumors is at the very least reasonable.

## Conclusion

Primary renal ES is a very rare but aggressive renal tumor. We present a 22-year-old female patient with primary renal ES with tumor thrombosis up to the vena cava who had radical nephrectomy and IVC tumor thrombectomy followed by adjuvant chemotherapy because a preoperative percutaneous biopsy was not performed due to a lack of suspicion of such a rare condition. Immunohistochemistry is essential to the diagnosis given its close histologic resemblance to other renal round cell tumors. Although the overall prognosis is poor, early diagnosis and treatment by a multidisciplinary team with multimodal therapy can improve outcomes.

## Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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