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CASE REPORT

A Rare Case of Extraosseous Ewing's Sarcoma of the Kidney Accompanied by a Gastric Adenocarcinoma

Soner COBAN [1], Atilla SATIR [1], Caglar BOYACI [1], Muhammet GUZELSOY [2], Ali Riza TURKOGLU [1], Murat OZTURK [1], Akif KOC [1], Anil ERKAN [1], Metin KILIC [1], Oguzhan AKPINAR [1], Nazmi MUTLU [3]

[1] Department of Urology, University of Health Sciences Turkey Bursa Yuksek Ihtisas Training and Research Hospital, Bursa, TURKEY
[2] Department of Urology, Bursa Cİty Hospital, Bursa, TURKEY
[3] Department of Pathology, Bursa Cİty Hospital, Bursa, TURKEY

ABSTRACT

Primitive neuroectodermal tumors (PNET) are most common in the central nervous system. Peripheral placements are extremely rare. PNET is from the Ewing sarcoma family. Ewing's sarcoma is evaluated in two groups: bone and non-bone origin. Renal PNET is extremely rare. Peripheral PNETs of renal origin are usually seen in young adults. They progress with an aggressive clinical picture and have a poor prognosis. In the follow-up imaging of a 62-year-old female patient after gastrectomy due to stomach cancer (adenocarcinoma), a renal mass was detected incidentally in the left kidney. The patient had no identified complaints. In the middle segment of the left kidney, computed tomography (CT) examination revealed a 42.5x44.6 mm nodular mass lesion, which was noticed as hypodense with contrast material. The patient underwent laparoscopic left radical nephrectomy. On pathological examination, a diagnosis of PNET/Ewing sarcoma was made. Immunohistochemically, membranous/cytoplasmic and nuclear staining with FLI1 and CD99 were observed in the tumor sections. However, staining with LCA was not seen.The patient was scheduled for chemotherapy by oncology. At the end of the fourth course of chemotherapy, no recurrence was found in the radiological controls of the patient.

Corresponding Author: Merve UZBAS mervekesgin265@homail.com

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Introduction

Ewing's sarcoma is a neuroectodermal tumor that develops from long and flat bones. Extraosseous Ewing sarcoma is a very rare condition (1). Renal Ewing sarcoma is a malignant, fast-growing tumor that rapidly metastasizes to the lungs, bones and lymph nodes (2). We present a rare case of renal Ewing sarcoma in a 62-year-old female patient who had a gastrectomy due to stomach cancer and underwent radical nephrectomy with suspicion of Renal Cell Carcinoma (RCC) on imaging.

CASE

Three years ago, the patient underwent gastrectomy for gastric adenocarcinoma. In the follow-up imaging of a 62-year-old female patient after gastrectomy due to stomach cancer, a renal mass was detected incidentally in the left kidney. The patient had no identified complaints including bone pain. In the middle segment of the left kidney, computed tomography (CT) examination revealed a42.5x44.6 mm nodular mass lesion, which was noticed as hypodense with contrast material (Image1). On MRI imaging, a lesion in the left kidney with a size of 5x4 cm in the middle pole, iso-light hyperintense on T1 examination, heterogeneous hypo-hyperintense on T2 examination, showing moderate staining with contrast material with a millimeter hypointense wall on the periphery, was noted. The right kidney was normal. The patient underwent a right laparoscopic radical nephrectomy with a preliminary diagnosis of RCC. No postoperative complications were observed.

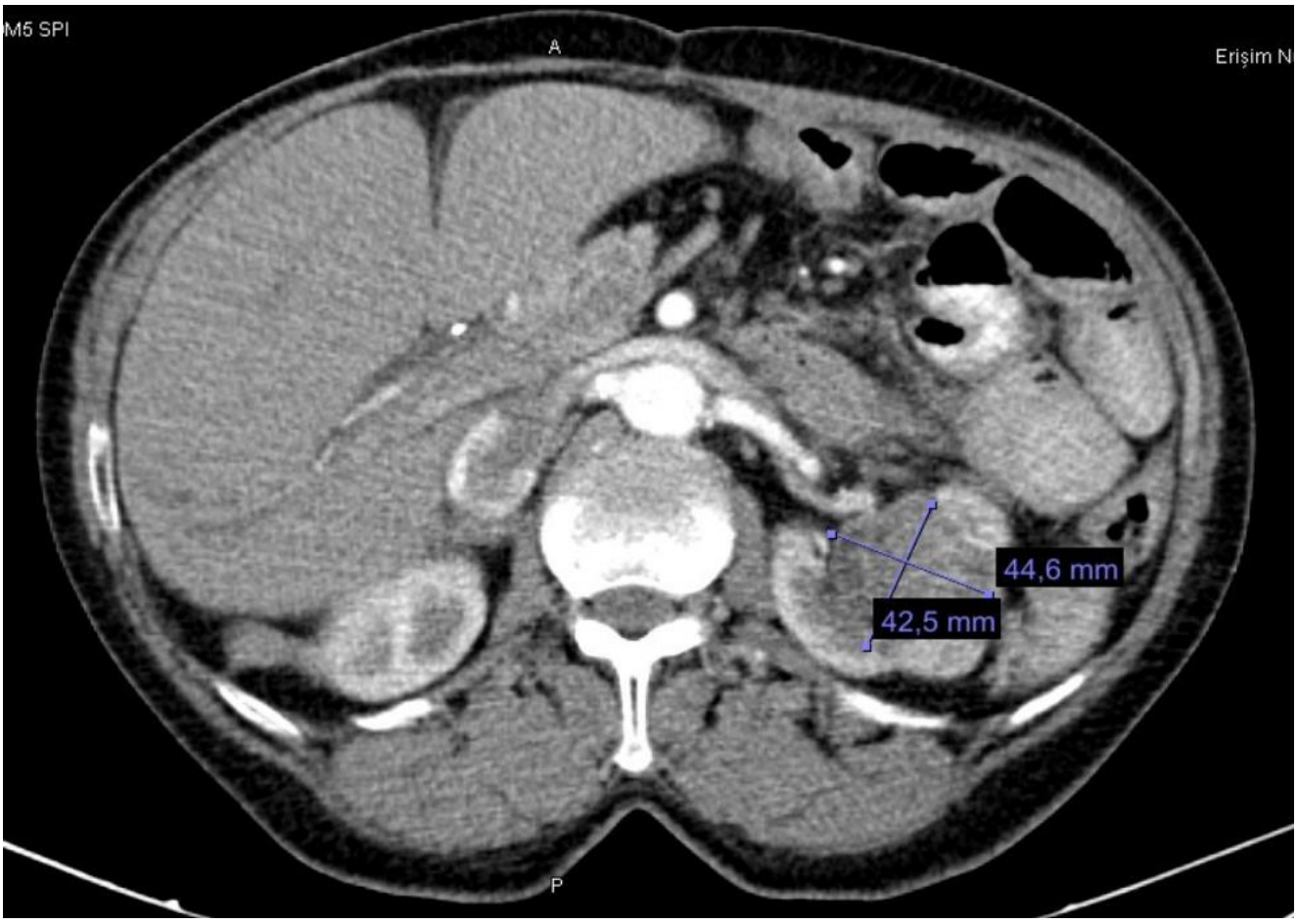

Image1. CT image of the case.

On pathological examination, a diagnosis of PNET/Ewing sarcoma was made. Hematoxylin eosin staining of the samples is seen in Image 2. Immunohistochemically, membranous/cytoplasmic and nuclear staining with FLI1 and CD99 were observed in the tumor sections (Image 3 and Image 4). However, staining with LCA was not seen (Image 5).

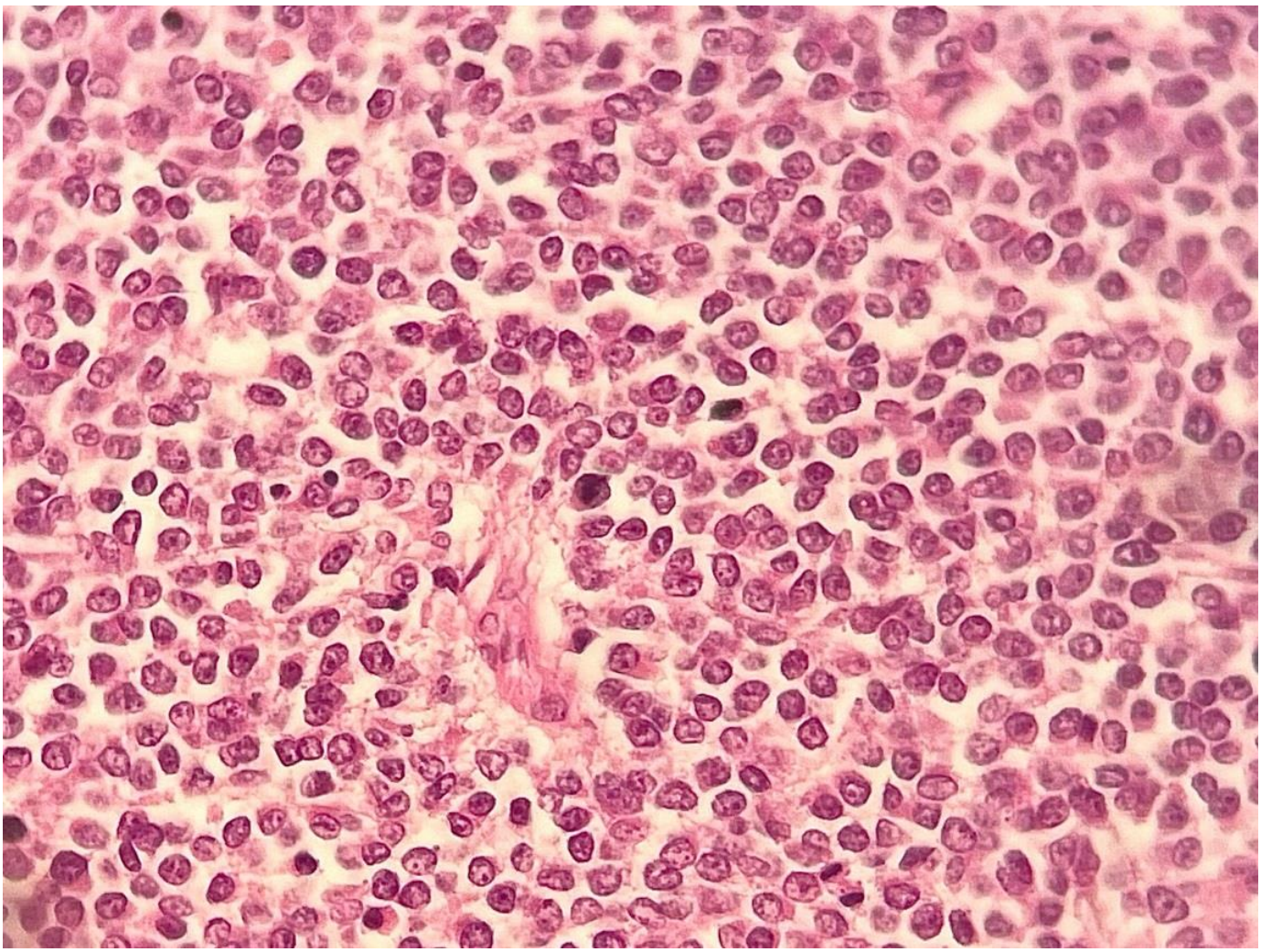

Image2. Hematoxylin Eosin staining.

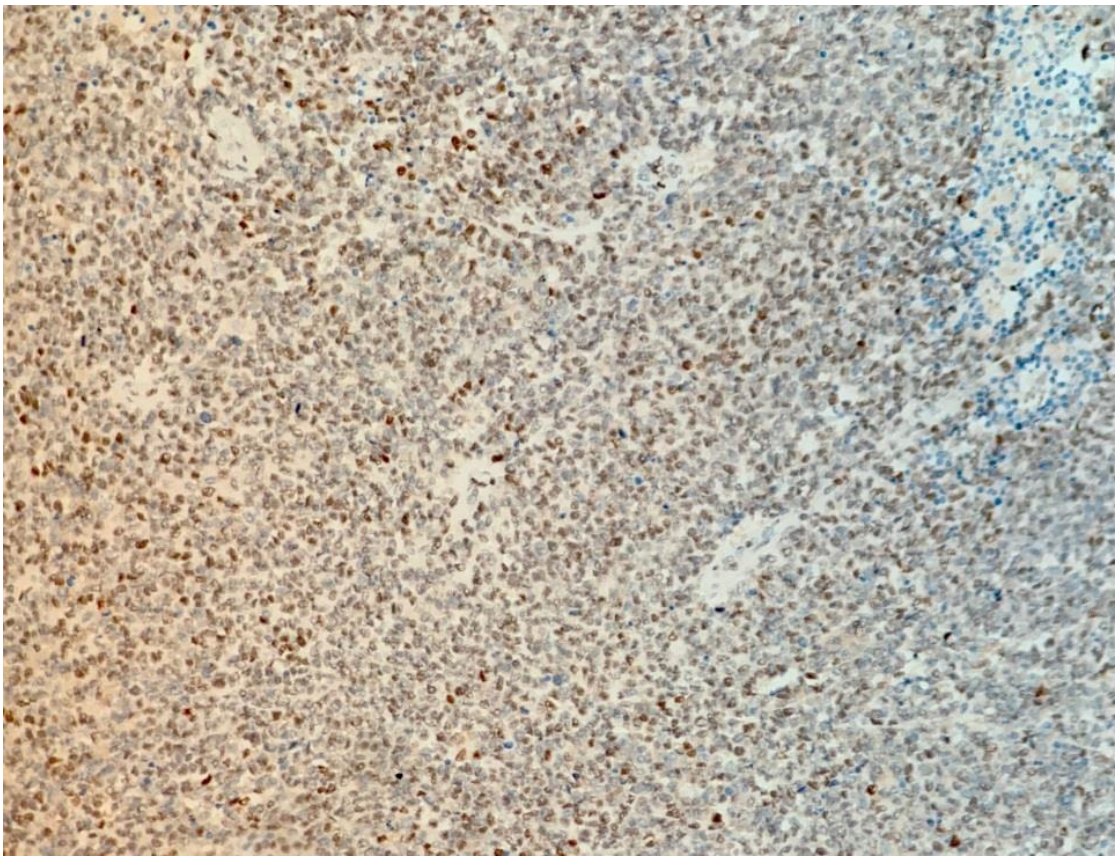

Image3. FLI1 stained cells.

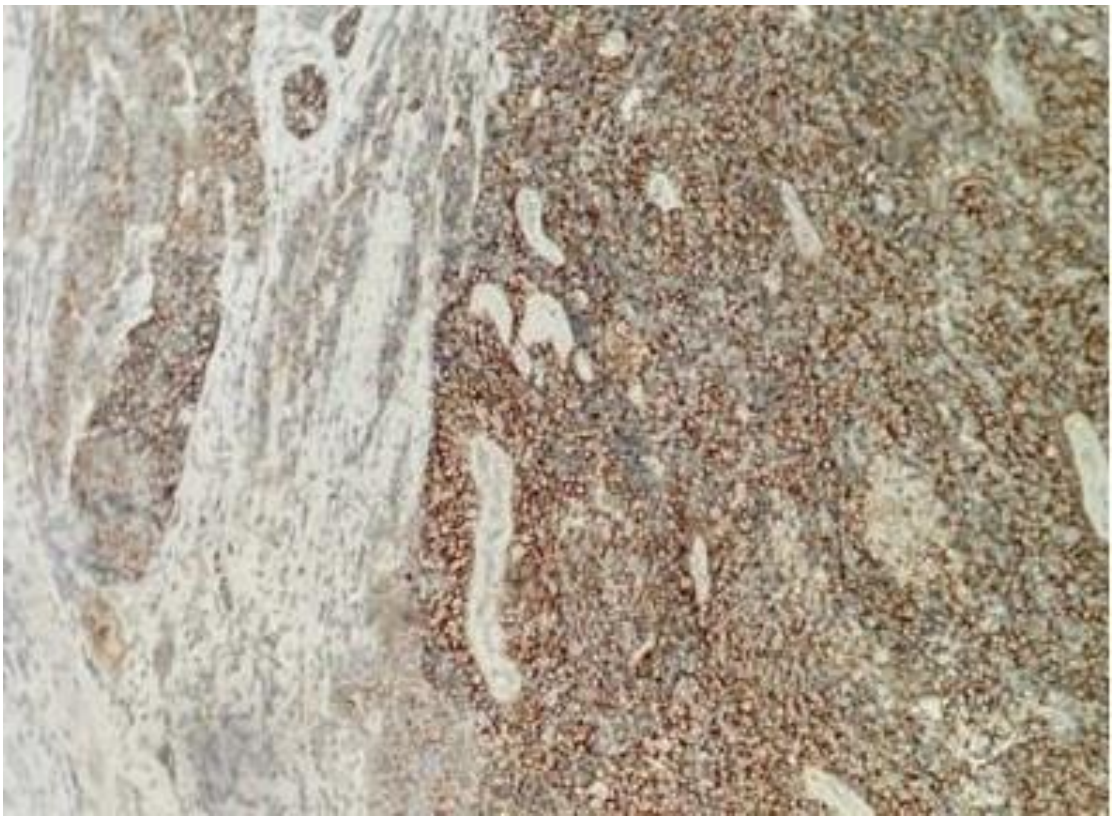

Image4. CD99 stained cells.

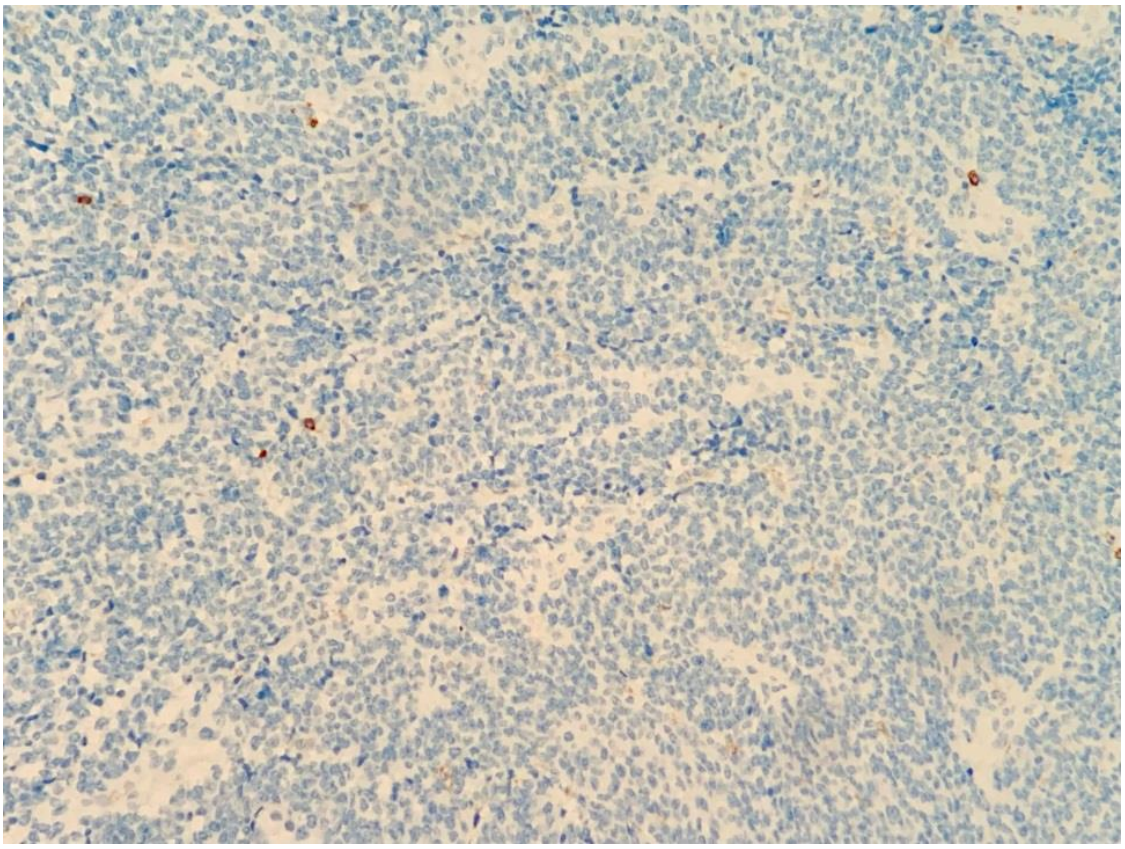

Image5. LCA staining.

No bone lesion or metastasis was detected in postoperative fluorine-18-fluorodeoxyglucose positron emission tomography.

DISCUSSION

Renal Ewing sarcoma is clinically indistinguishable from RCC. About 80% of kidney masses are renal cell carcinoma. RCCs originate in the renal cortex. Although the cellular origin of renal Ewing's sarcoma is not fully known, it is believed that it originates from neural crest cells (3). Renal Ewing's sarcoma usually has an aggressive course, and most patients are metastatic at the time of admission. Surgical excision is the most important treatment modality. The prognosis of this disease is poor, despite aggressive treatments with surgery, chemotherapy and radiotherapy. 5-year survival rates are between 45 and 55% (2,5).

Renal Ewing's sarcoma is mostly observed in the younger age group (28-34 years). Our patient was 63 years old. Initially, it is usually asymptomatic, as in our patient. In the later period, symptoms such as flank pain or hematuria can be seen (4).

It is almost impossible to distinguish renal Ewing sarcomas from RCC by imaging. RCC imaging features are indistinguishable from other renal tumors (6). The main difficulty in diagnosing Ewing's sarcoma is its morphological similarity with other small round blue-cell tumors. In children, Wilm's tumor, neuroblastoma and primary renal rhabdomyosarcoma should be ruled out. The main differential diagnoses in the adult age group are non-Hodgkin lymphoma, monophasic synovial sarcoma and desmoplastic small round cell tumors (7).

There is no clear consensus for the treatment of Renal Ewing's Sarcoma. However, the treatment protocol currently used involves a combination of surgical resection, adjuvant chemotherapy and/or radiotherapy. The standard chemotherapy regimen includes a three-drug combination of vincristine, doxorubicin and d-actinomycin, as well as the addition of etoposide and ifosfamide. In order to treat local relapses or residual tumor, radiotherapy has been considered. However, this treatment protocol seems to provide less benefit in these patients due to the fact that the disease usually presents as advanced. Renal Ewing sarcoma has an aggressive course characterized by early metastasis (8,9). However, our patient has received 4 cycles of chemotherapy and has been followed without recurrence for 1 year.

CONCLUSION

Primary renal Ewing's sarcoma is a very rare but highly aggressive kidney tumor. There is no specific imaging technique that can differentiate it from RCC and, therefore, the possibility of Ewing's sarcoma should always be kept in mind as a differential diagnosis in aggressive kidney tumors seen in young adults. The definitive diagnosis is made by histopathological diagnosis. Early diagnosis is very important so that chemotherapy can be started as soon as possible.

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