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Oncology

A rare case of Ewing's sarcoma of the kidney

Mohammed Hasen A. Alghamdi [a,*], Saud Abdullah Alawad [a], Mohammed Ghazi Alharbi [a], Abdulrahman K. Alabdulsalam [b], Fayez Almodhen [c], Ahmed Alasker [a,c,d]

[a] College of Medicine, King Saud bin Abdulaziz University for Health Sciences (KSAU-HS), Riyadh, 11426, Saudi Arabia
[b] Department of Pathology and Laboratory Medicine, King Abdulaziz Medical City, Riyadh, 11426, Saudi Arabia
[c] Division of Urology, King Abdulaziz Medical City, Riyadh, 11426, Saudi Arabia
[d] King Abdullah International Medical Research Center (KAIMRC), Riyadh, 11426, Saudi Arabia

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ABSTRACT

We report the case of a 15-year-old girl presenting with distended abdomen, left flank pain, and a history of weight loss. Computed tomography showed a large tumor involving the left kidney that was initially diagnosed as renal cell carcinoma. She underwent exploratory open laparotomy and left radical nephrectomy followed by chemotherapy and showed good response. Histology of the resected tumor revealed features of Ewing's sarcoma of the kidney which was confirmed by molecular studies. This disease is rare, particularly in the pediatric population, and this report will help better understand the potential disease course and response to treatment.

Introduction

Ewing's sarcoma of the kidney (ESK) is a remarkably rare malignant tumor of the primitive neuroectodermal tumor family. It is a genetic disease resulting from a translocation mutation fusing the *EWS* and *FLI1* genes of chromosome 22 and chromosome 11, respectively. The treatment of choice is total nephrectomy and adrenalectomy followed by combination chemotherapy with or without adjuvant radiotherapy.[1] However, the overall prognosis and survival of ESK is still poor. Nonetheless, younger patients tend to respond better.[1] We report the case of a young girl with ESK.

Case presentation

A 15-year-old girl referred to the urology clinic presented with left flank pain and abdominal distention. She was healthy until 10 months ago when she noticed mild abdominal distention followed by gradually increasing pain in the left flank. She also lost 16 kg in the last 6 months without fever, nausea, vomiting, hematuria, changes in bowel motions, or lower urinary tract symptoms. Physical examination revealed that she was stable, but had abdominal distention and left flank tenderness.

Computed tomography (CT) scans of the abdomen and pelvis showed a mass of 22 × 20 × 25 cm on the left kidney significantly impinging on adjacent structures, which was highly suggestive of a neoplastic pathology, without metastasis or lymphadenopathy (Fig. 1). The images were not specific. Chest X-ray; ultrasound-guided biopsy; and tests for renal function, liver profile, serum electrolytes, and complete blood count were unremarkable except for normocytic hypochromic anemia with 9.4 g/dl hemoglobin. Therefore, our differential diagnoses were renal cell carcinoma, Wilms' tumor, or neuroblastoma.

We performed an exploratory open laparotomy and left radical nephrectomy. A multidisciplinary team including urologists, medical oncologists, and radiation oncologists started the patient on combination chemotherapy comprising vincristine, doxorubicin, and cyclophosphamide (VDC) alternating with ifosfamide/etoposide (IE). Chemotherapy started 13 days postoperatively and included 17 cycles (nine VDC and eight IE) for 18 months which she tolerated well. As per the radiotherapy protocol, postoperative radiation was not required because patient showed negative resection margin.With no surgical complications and an uneventful postoperative period, the patient was discharged 16 days postoperatively. Histopathology showed small round cell tumor with extensive necrosis, hemorrhage, fibrosis, and scattered white spongy areas (Fig. 2). The tumor approximately measured 25 cm in greatest dimension. It was attached to and invaded the renal pelvis but not the renal parenchyma. The vascular and ureter margins of the kidney were unremarkable. The adrenal gland was tumor-free and unremarkable. Two lymph nodes without metastasis were identified. Immunohistochemical staining was positive for diffuse and membranous CD99 (12E7) and showed retained INI-1 expression. Fluorescence in situ hybridization studies were positive for *EWSR1* (22q12) rearrangement.

* Corresponding author. P.O. Box 105910, Riyadh, 11656, Saudi Arabia.
E-mail address: m.alghamdi@me.com (M.H.A. Alghamdi).

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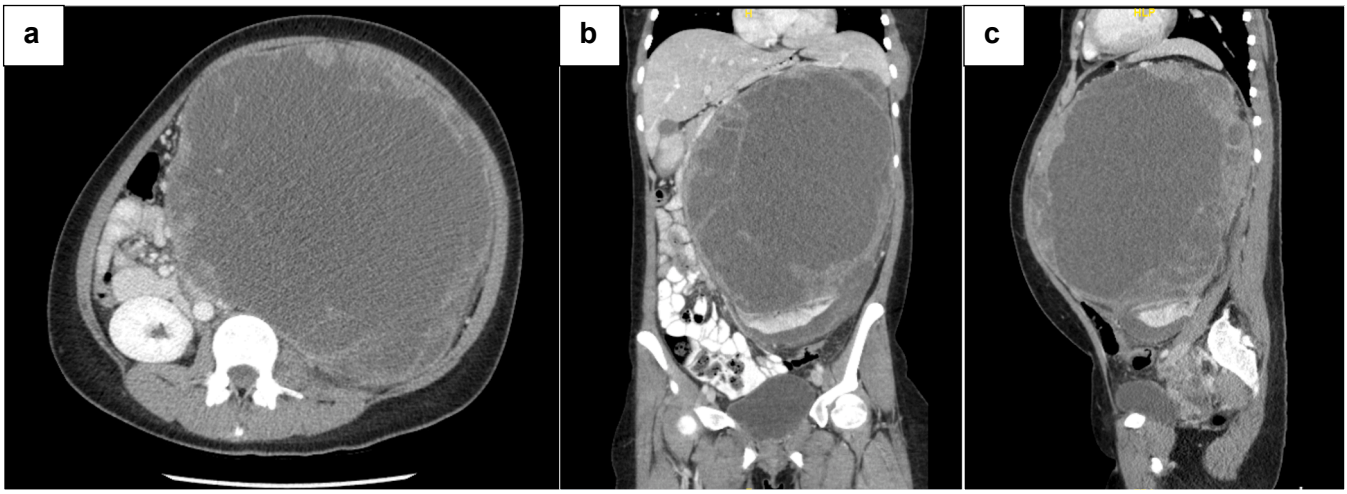

Fig. 1. Abdominal and pelvic computed tomography (CT) scans with contrast showing a large heterogeneous mass on the left kidney significantly impinging on adjacent structures. Renal vessels are patent with no evidence of invasion; (a) transverse view, (b) coronal view, (c) sagittal view.

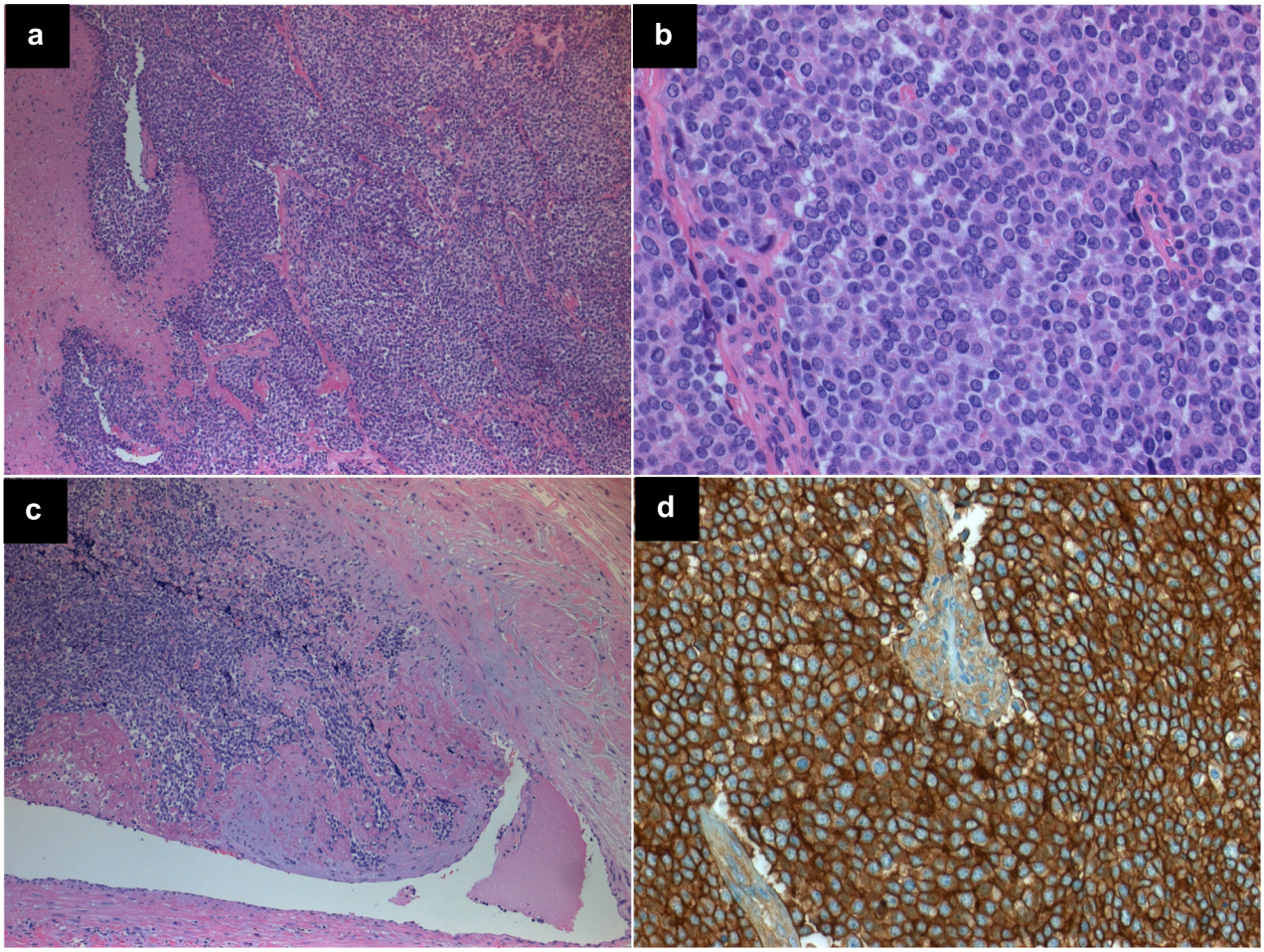

Fig. 2. (a) The tumor shows a solid and diffuse growth pattern, with areas of necrosis (hematoxylin and eosin (H&E) stain, 100 × magnification), (b) this section shows the typical small round blue cell morphology (H&E stain, 400 × magnification), (c) this shows an intravenous tumor thrombus (H&E stain, 200 × magnification), (d) tumor cells show positive diffuse and membranous CD99 (immunohistochemical staining, 400 × magnification).

Subsequent follow-ups using CT at 3 (Fig. 3), 9, 15, and 18 months postoperatively showed a clear surgical bed with no signs of recurrence or metastasis.

Discussion

ESK usually affects young adults aged 30.4 years on average at diagnosis; Approximately 60% of those affected are male.[2] Patients' signs and symptoms mimic that of kidney stones including flank pain

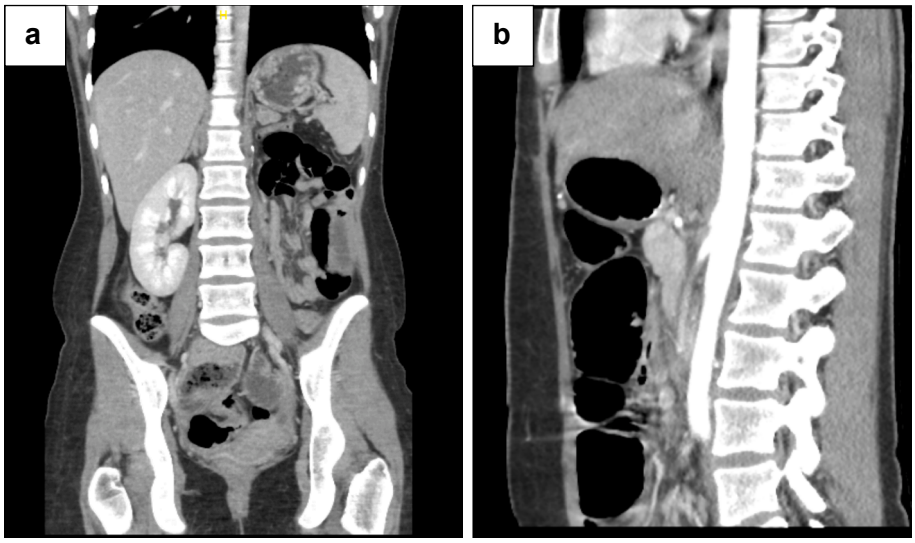

Fig. 3. (a) Coronal and (b) sagittal abdominal CT with contrast 3 months postoperatively. The surgical bed is clear with no signs of recurrence or metastasis.

(84%), palpable neoplasm (60%), and hematuria (38%).[2] Histological and radiological studies do not provide a definite diagnosis because they are ambiguous; the features observed in these reports are seen in other neoplastic pathologies. Diagnosis is more accurate with immunochemistry and cytogenetic tests. One such test is molecular testing for the *EWSR1* gene.[3] Around 80–95% of patients with ESK show the chromosomal translocation t(11;22) (q24;q12) rearrangement whereas 5–20% often display *EWS-ETS* gene mutation.[3] ESK tends to be aggressive and can metastasize to different organs. More than 65% of patients present with distant metastasis.[4] Common sites for ESK include regional lymph nodes, the lungs, and the liver with the lungs being the most common site.[4] The overall patient survival rate is low with most dying within a year after diagnosis.

ESK treatment uses a multimodality approach including radical nephrectomy followed by combination chemotherapy with or without radiotherapy. There is neither a unified protocol for treating ESK nor any evidence about the effectiveness of different treatment modalities. The combination chemotherapy used in ESK treatment includes VDC or vincristine, adriamycin, and cyclophosphamide alternating with IE.[5]

Adjuvant radiotherapy can be offered in case of incomplete resection, positive resection margins, or recurrence. Follow-ups with laboratory and imaging tests are essential to assess recurrence and metastasis.

Conclusion

We reported a rare case of ESK in a 15-year-old girl aiming to clarify the potential disease course and its treatment response. Although a multimodality approach is helpful, survival rate is mostly limited to 1 year. In children, this tumor is rare and mimics common types of tumors such as Wilms' tumor, neuroblastoma, and lymphoma. Precise diagnosis of the tumor is essential because of its poor prognosis. Histological and imaging findings are not specific and have to be supported by molecular studies for accurate diagnosis. Early detection with proper intervention is vital for ESK treatment. More studies are needed to establish treatment protocols that can reduce recurrence rates and improve patient survival.

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Declaration of competing interest

There is no conflict of interest in our case report.

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