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Adrenal mass of unusual etiology: Ewing sarcoma in a young man

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ABSTRACT

Ewing sarcoma and peripheral primitive neuroectodermal tumor belong to the Ewing sarcoma (ES) family of tumors originating from a primitive neural tube. We report a 31-year-old man who was admitted to the urology clinic with complaints of fever, nausea, and dysuria. A right-sided adrenal mass was detected during ultrasonography. The lesion was then evaluated with magnetic resonance imaging, which showed areas of necrosis amid heterogeneous solid areas. Whole body scan with 2-deoxy-2-[fluorine-18]fluoro-D-glucose integrated with computed tomography and bone scan studies showed pulmonary and osseous metastatic foci. The mass and right kidney were removed by an open approach. An immunohistochemical and molecular workup enabled the diagnosis of ES. The patient also underwent radiotherapy and chemotherapy. The patient remained in fairly good health during the 18-month follow-up period, but showed progression of all metastatic foci and died 26 months after treatment. In conclusion, adrenal ES should be included in the differential diagnosis of nonfunctional adrenal lesions despite its rare occurrence.

Introduction

Ewing sarcoma (ES), peripheral primitive neuroectodermal tumor (PNET), extraskeletal ES, and Askin tumor (thoracopulmonary PNET) constitute a group of tumors collectively called Ewing sarcoma family of tumors (ESFT), which originates from a primitive neural tube [1,2]. Ewing sarcoma and peripheral primitive neuroectodermal tumor (ES/PNET) comprises a group of small, round, and blue cell tumors that are related to each other on a molecular and immunopathologic basis. These tumors usually occur in long or flat bones, in the chest wall, in soft tissues, and, to a lesser extent, in paravertebral locations and along the genitourinary tract, including

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the kidney, the urinary bladder, and the vagina [2–4]. Involvement of the adrenal gland in ES/PNET is an extremely rare occurrence. To our knowledge, only about 19 cases of primary adrenal involvement have been reported in the literature, with most cases in the adolescent population and in young adults [5]. Despite its rarity, ES/PNET should be included in the differential diagnosis of an adrenal mass among the more common entities such as adenoma, pheochromocytoma, neuroblastoma, and adrenal cancer. Additionally, ES/PNET must be distinguished from renal tumors [5]. Adrenal ES/PNET is usually managed with surgical intervention followed by chemoradiotherapy, called multimodal treatment; however, no established guidelines exist so far regarding the optimal treatment [6].

In the present study, we report an adrenal ES/PNET case of a young man with distant metastases who was treated with multimodal treatment with some progression in the metastases during an 18-month follow-up and died 26 months after treatment. Literature in English pertinent to the cases of primary adrenal ES/PNET presented so far has also been reviewed at the end.

Case report

In this case report, we present the case of a 31-year-old man who was referred to the urology clinic because of complaints of mild dysuria and fever reaching 104°F for the last 4 days. The patient also complained of nausea and vomiting since 3-4 months and a nonspecific pain in the right hip. The family history was not remarkable, with no history of tuberculosis or malignancy. Blood tests showed leukocytosis (white blood cell, 21,000/mm^3) with neutrophil predominance (16,900/mm^3). All other hematologic parameters were within normal limits. The physical examination was also normal. The ultrasonographic examination revealed a heterogeneous solid right adrenal mass measuring $20 \times 12 \times 14$ cm. Testing for the functionality of the adrenal mass showed no excess metabolites both in blood and urine, including vanillylmandelic acid, 24-hour cortisol, catecholamines, aldosterone, and dehydroepiandrosterone sulfate. Magnetic resonance imaging was then performed to further delineate the mass, which showed areas of increased intensity on T2-weighted imaging, consistent with necrosis and heterogeneous contrast enhancement of solid areas on T1-weighted imaging. Diffusion-weighted imaging with a b value of 1000 showed heterogeneous areas of enhancement with a corresponding decrease in apparent diffusion coefficient values, which was consistent with the restricted diffusion, and areas that were hyperintense on both diffusion-weighted imaging and apparent diffusion coefficient, suggesting necrosis (Fig. 1). Whole-body scan with 2-deoxy-2-[fluorine-18]fluoro-D-glucose integrated with computed tomography (F18-FDG-PET/CT) showed areas of increased metabolic activity in the lingular segment of the left lung, and a bone scan with single-photon emission computed tomography (CT) showed suspicious foci on the S1 vertebra and on the left femoral head, suggestive of lytic-sclerotic metastases.

The patient's fever and nausea were likely attributed to the malignancy.

An open right nephrectomy, along with adrenalectomy, was performed. The patient was discharged on postoperative day 3 without immediate complications.

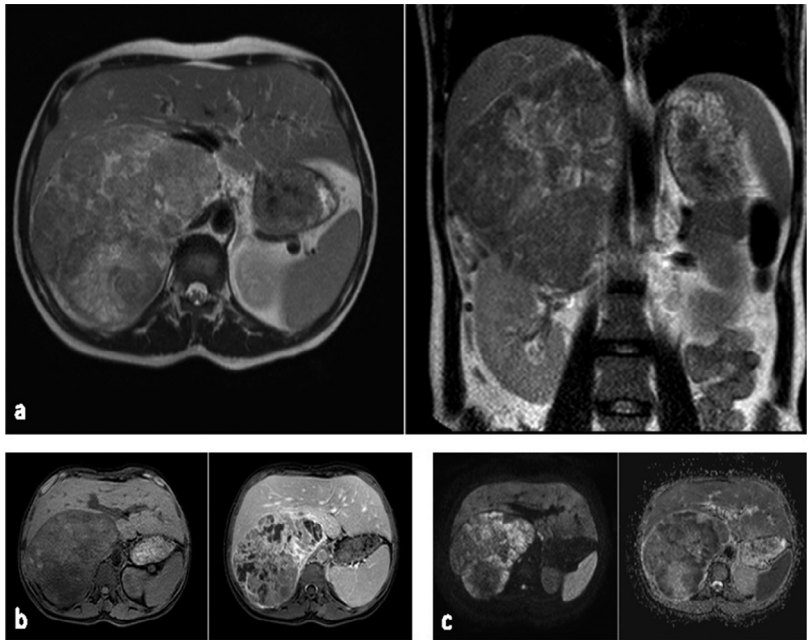

Fig. 1 – (A) Axial and coronal T2-weighted images show a large right adrenal mass partially displacing the liver. The mass has heterogeneous areas of low and high signal intensities, suggestive of solid and necrotic areas. (B) Precontrast and postcontrast fat-saturated T1-weighted images show heterogeneous areas of contrast enhancement. (C) Diffusion-weighted imaging shows that the solid areas restrict diffusion, consistent with high cellularity. The corresponding apparent diffusion coefficient image shows these areas as hypointense, confirming the restriction of diffusion.