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Ewing Sarcoma/Peripheral Primitive Neuroectodermal Tumor in the Adrenal Gland of a Child

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Ewing sarcoma/peripheral primitive neuroectodermal tumor (ES/PNET) typically occurs in long or flat bones, soft tissues, or less often, solid organs. Ewing sarcoma/peripheral primitive neuroectodermal tumor arising from the adrenal gland is extremely rare, especially in children, and only limited cases are reported previously. Herein, we review a case of a 22-month-old girl who presented to our department with abdominal pain, bulging of the left flank, and a nonfunctioning adrenal lesion which was found to be an adrenal ES/PNET. The patient was successfully treated with surgery and adjuvant chemotherapy. Since delayed diagnosis may result in metastatic lesions, this case underscores the importance of considering ES/PNET in the differential diagnosis of large adrenal masses.

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Case Report

INTRODUCTION

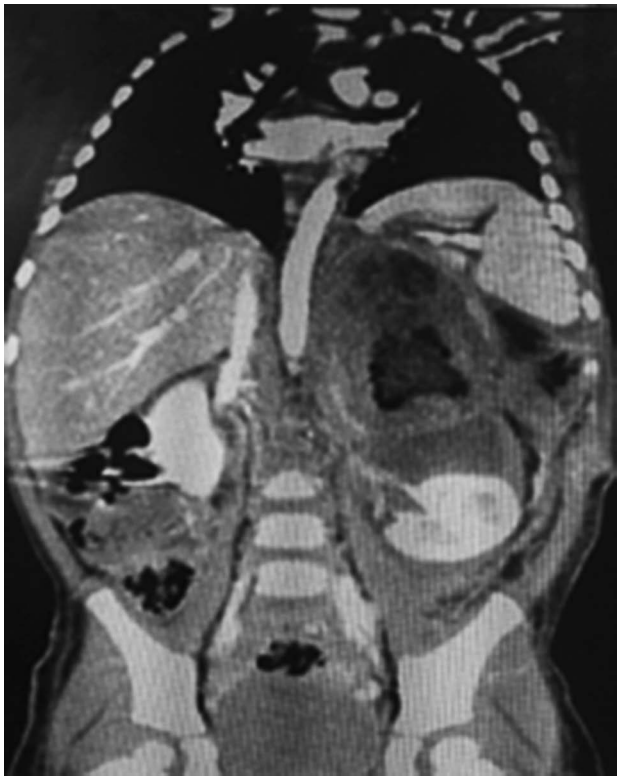
Ewing sarcoma/primitive neuroectodermal tumors (ES/PNET) compromise a family of histopathologically related small round blue-cell tumors, derived from the neural crest.[1] The common sites of this tumor include long or flat bones, soft tissues, or less often, solid organs.[1] To our knowledge, only one previous study has reported ES/PNET arising from the adrenal gland in toddlers.[2] Herein, we report a 22-month-old girl with primary ES/PNET in the left adrenal gland.

CASE REPORT

The patient was a 22-month-old girl referred to our department complaining of abdominal pain and bulging of the left flank. Physical examination revealed a tender mass in the left flank. A computed tomography scan further defined a 117 × 116-mm heterogeneous mass with central hypoechogenicity, suggestive of necrosis, originating from the left adrenal gland with mass effect and displacement of the spleen and the left kidney (Figure). No signs or symptoms of catecholamine excess or hormonal disturbance were present, and all hormonal studies, including cortisol and urine vanillylmandelic acid, were within normal limits. No sites of abnormal uptake were detected in the metaiodobenzylguanidine scan. In the bone survey, bone density was normal and no remarkable changes were observed.

With the impression of retroperitoneal neuroblastoma, she was taken electively to the operating room. A 10 × 10-cm mass adhesive to the spleen, left kidney, stomach, transverse colon, and left lobe of the liver was observed in the operation room. Approximately, 90% of the tumor with the left lobe of the liver, a part of the diaphragm and a part of the upper pole of left kidney were resected. The remaining tumor was resected in a second operation.

The Intraoperative pathologic evaluation revealed the mass as monotonous patterns of sheets of small cells and inconspicuous cellular boundaries arranged in loose fibroconnective tissue. Small round cell tumors were reported to be compatible with ES/PNET. Staining was positive for cluster of Differentiation-99, neuron-specific enolase, and vimentin, and negative for chromogranin and Wilms



Computed tomography scan defining a 117 × 116-mm heterogeneous mass with central hypoechogenicity, suggestive of necrosis, originating from the left adrenal gland with mass effect and displacement of the spleen and the left kidney.

tumor 1 gene product, which further confirmed the ES/PNET diagnosis.

The patient underwent 12-course chemotherapy including vincristine, adriamycin, and cyclophosphamide, alternating every 3 weeks with ifosfamide and etoposide. Radiotherapy was not indicated in this patient because after 6 courses of chemotherapy, a biopsy revealed complete necrosis of the remaining tumor. She was free of recurrence at 17 months postoperative and 8 months off therapy.

DISCUSSION

To our knowledge, only 6 cases of children or adolescents with a certain diagnosis of ES/PNET are reported in the literature.[2-6] Our report adds to the one recently published case report of primary adrenal ES/PNET presenting in toddlers age.[2] Since adrenomedullary tumors, such as pheochromocytoma and neuroblastomas, are more prevalent in children, they are commonly considered as a potential diagnosis in children with large adrenal masses. However, they are usually accompanied by increased catecholamines and/or abnormal uptake on metaiodobenzylguanidine scan.[7] Adenoma or adrenocortical carcinomas are also other potential diagnoses of an adrenal masses. While adrenocortical carcinomas are most often functional, adenomas are non-functional.[8] Similar to our case, all the previous reports of ES/PNET arising from the adrenal gland in children or adolescents reported no catecholamines excess associated with the tumor.[2-6] Compatible with findings of computed tomography scan in our case, a recent study showed that an adrenal ES/PNET usually presents as a large, well defined and heterogeneous mass with necrosis or cystic degeneration.[2] Hence, ES/PNET should be considered in approaching to non-functional large adrenal masses especially when the imaging findings of the tumor is consistent with aforementioned features.

Since ES/PNET is a highly aggressive neoplasm, multimodal treatment including surgery and chemotherapy are essential for a better outcome.[9] Similar to our case, previous studies have used both chemotherapy and surgery to manage patients with adrenal ES/PNET[2-6]; however, only 2 cases survived after treatment.[3,4] Both of these cases had no metastasis and were treated with chemotherapy including ifosfamide and etoposide.[10] Although patients who had adrenal ES/PNET with metastasis had a poor prognosis,[11,12] these results further support the opinion that the most important prognostic factor of ES/PNET is the presence of metastatic lesions.[3]

This case presents a rare diagnosis in a pediatric patient with adrenal mass. Despite its rarity, an ES/PNET should be included in the differential diagnosis of adrenal tumors. Delayed diagnosis may result in metastatic lesions which weakens the prognosis of these patients.

CONFLICT OF INTEREST

None declared.

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