Case Report

Differential Diagnosis of a Large Size Tumor in the Retroperitoneum: A Case Report of Retroperitoneal Lipoblastoma

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Abstract

Lipoblastoma can arise almost anywhere within the soft tissues, especially in the extremities. It is rarely seen retroperitoneally. A 3-year-old girl presented with a palpable abdominal mass underwent an exploratory laparotomy with resection of the retroperitoneal mass. Based on histopathologic and cytogenetic features, a final diagnosis of lipoblastoma was rendered. At 18-month follow-up, she had no evidence of recurrence.

Keywords: Abdominal mass, diagnosis, magnetic resonance imaging, retroperitoneal lipoblastoma, teratoma

Introduction

Lipoblastoma is a rare benign soft tissue tumour of embryonal fat occurring almost exclusively in infancy and early childhood.[1,2] Histologically, the tumors appear to be well-circumscribed and multilobular with fibrous septa.[3] Lipoblastomatosis is a more severe, diffuse form of the disease, demonstrating infiltrative growth with a greater likelihood of entrapping surrounding structures and may involve several different sites.[3] Most commonly, lipoblastomas occur in the extremities and trunk. Unlike liposarcoma, in which approximately 25% of all tumors occur in the retroperitoneum, lipoblastomatosis was rarely found in this region.[3]

Case Report

A previously healthy 3-year-old girl presented with a palpable abdominal mass that was noted on her well-child examination. Her parents reported a gradually increasing abdominal distention for 1 year with no other symptoms. On physical examination, the child had a soft, grossly distended, and nontender abdomen, with a large firm palpable mass on the left side [Figure 1]. Hemoglobin, alpha-fetoprotein, and human chorionic gonadotropin beta subunit levels were normal. An abdominal ultrasound showed a heterogeneous soft-tissue mass with septal walls in the left lower quadrant. A computed tomography (CT) scan revealed a well-circumscribed, heterogeneous, and fatty mass with thin internal fibrous septations in the left lower quadrant measuring 13.5 cm × 10.5 cm × 17.5 cm. The mass appeared to be intraperitoneal and to be causing mass effect on the spleen, left kidney, small intestine, and pancreas. A magnetic resonance imaging (MRI) scan confirmed a multilobulated mass suggestive of a lipomatous tumor of retroperitoneal origin, and a laparotomy was planned for both diagnosis and therapy [Figure 2]. The preoperative diagnosis of this case was a retroperitoneal teratoma.

Subsequently, an exploratory laparotomy with resection of the retroperitoneal mass was performed under general anesthesia. A well-encapsulated mass was loosely attached to

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the retroperitoneum. A thorough exploration of the abdomen was carried out without identifying other abnormalities. On the superior pole of the tumor, the left diaphragm was adherent and carefully dissected free from the mass. The tumor was easily dissected free from the retroperitoneal space without injury to adjacent structures. Grossly, the tumor weighed 1015 g and was well circumscribed, with a thin fibrous capsule. The specimen had a yellow lobulated fatty parenchyma separated by thin fibrous septa. Histologically, the lipomatous tumor consisted of mature adipocytes mixed with numerous lipoblasts in a focal background. No evidence of cellular atypia, mitotic activity, necrosis, or hemorrhage was seen within the mass. Based on histopathologic features, a final diagnosis of lipoblastoma was rendered [Figure 3]. The patient tolerated the surgical procedure well without complication and was discharged home on postoperative day six. At 18-month follow-up, she had no evidence of recurrence by abdominal ultrasound.

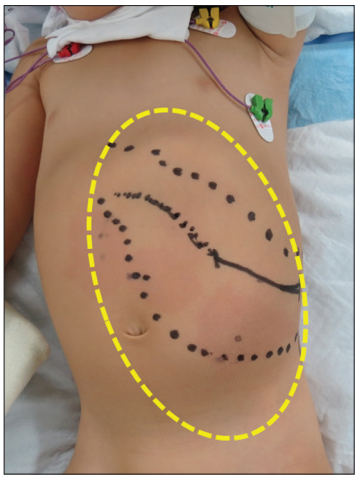

Figure 1: On physical examination, the child had a soft, grossly distended, and nontender abdomen with a large ›rm palpable mass on the left side (a dashed circular shape)

Discussion

Lipoblastomas are rare benign mesenchymal tumors that occur almost exclusively in children, with 88% of the cases reported during the first 3 years of life.[1-3] Nonetheless, cases in older patients have also been reported.[2,4,5] Chung and Enzinger, who have reported the largest series to date, have proposed the subdivision of the tumor according to growth its pattern: "benign lipoblastoma" for localized, well-circumscribed, relatively superficial, and encapsulated forms of the tumor; and "benign lipoblastomatosis" for multicentric, poorly defined, and deeply located diffuse forms.[5] The majority of lipoblastomas have been found on the trunk or extremities, with only 5% of cases reported in the retroperitoneum. Although the retroperitoneum is not a common site for lipoblastomas (5% of the 186 tumors reviewed), when they do occur, they tend to grow very large (range, 13.5–21 cm).[6]

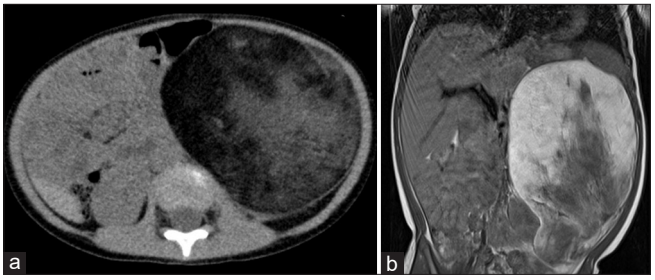

Figure 2: Computed tomography scan revealed a well-circumscribed, heterogeneous, and fatty mass (a). Magnetic resonance imaging con›rmed a multilobulated mass suggestive of a lipomatous tumor of retroperitoneal origin (b)

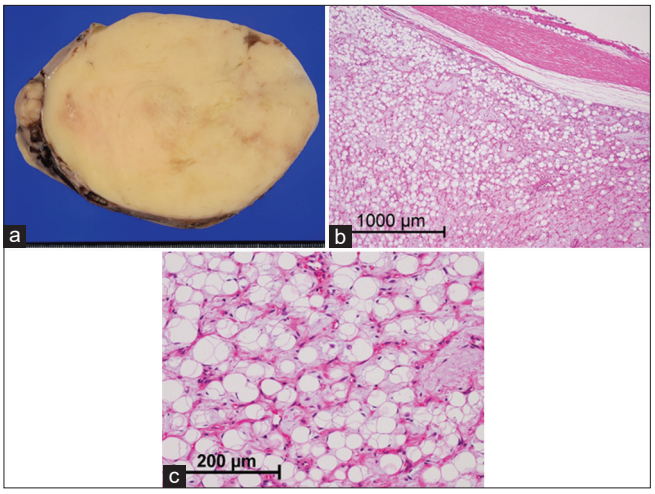

Figure 3: Cut surfaces had a yellowish tan and were commonly mottled with pale-pink, homogeneous, or gelatinous areas (a). The mass was separated into lobules by well-de›ned ›brous septa (b). Higher magni›cation of the tumor cells demonstrated the character of the lipoblasts cytoplasm by hematoxylin and eosin stains (c)

Preoperative diagnosis of the tumor remains a challenge. For example, the initial preoperative diagnosis was accurate in only five cases (16%).[6] The differential diagnosis in young female pediatric patients with large intra-abdominal masses is broad and includes sarcomas, ovarian tumors, lipomas, lymphomas, hepatoblastomas, Wilm's tumors, and neuroblastomas.[7] Workup of the mass may include investigation of tumor biomarkers, CT scans for mass characterization and lymphadenopathy detection, and MRI for specific masses and anatomic delineation.[7] In our case, a CT scan was not able to determine the diagnosis of the tumor in the patient, so the preoperative diagnosis of this case was retroperitoneal teratoma. Currently, MRI remains the most sensitive method for both preoperative planning and diagnosis of soft-tissue masses.[7] Specifically, MRI has the highest sensitivity for the pathology of the tumor, as the increased vascularity in lipoblastomas, compared with lipomas, shows a lower intensity on T1-weighted images.[8] Furthermore, fine-needle aspiration may aid in preoperative diagnosis.[9] However, resection and pathologic examination of the specimen are needed for definitive diagnosis.

Histologically, lipoblastomas appear to be well-circumscribed lobulated lesions that are separated by septa with mesenchymal cells and lipoblasts in the periphery.[5] The recent use of cytogenetics has proven to be helpful for diagnosis, as translations involving the long arm of chromosome 8, in particular 8q11–13, with or without pleomorphic adenoma gene 1 oncogene rearrangements have been found to be associated with lipoblastomas.[9,10] Cytogenetic analysis has been shown to be helpful in distinguishing myxoid lipoblastomas from myxoid liposarcomas.[7] While the tumors may appear clinically and histologically similar, karyotype analysis differentiate the tumors, as myxoid lipoblastomas usually have translocations.[7]

Conclusion

We propose that retroperitoneal lipoblastoma of the pediatric patient could be an important differential diagnosis when there is a large size tumor in the retroperitoneum at each imaging examination.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Confiicts of interest

There are no conflicts of interest.

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