Chest wall lipoblastomatosis in a 2-year-old girl: a case report and literature review

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Lipoblastoma is an uncommon tumor of adipose tissue that usually occurs in infancy and early childhood. They predominantly locate in the extremities, but are less frequently found in the head and neck region, trunk and various other locations. The most common sign is a rapidly growing mass. In spite of their potential for local invasion and rapid growth, they are benign tumors and have no distant metastasis. Although more than 180 cases of lipoblastoma distributed over various parts of the body have been reported, only nine cases were located in the thoracic wall. We present the case of a two-year-old girl with lipoblastomatosis of the chest wall that was identified by histologic examination. In this article, the English literature on this disorder is briefly reviewed.

Key words: lipoblastoma, lipoblastomatosis, chest wall, adipose tumor.

Lipoblastomas are benign soft tissue tumors originating from white embryonic fat[1,2]. They are classified into two forms: benign lipoblastomas are localized, capsular and well-circumscribed tumors, while benign lipoblastomatosis is the occurrence of deeply embedded, noncapsulating, multicentric, and infiltrative tumors[3-5]. These tumors typically occur before three years of age and usually develop in the extremities[6,7]. However, chest wall involvement has been reported rarely. The tumor can behave in a locally aggressive manner and invade the surrounding tissues. In spite of its local invasion and rapid growth properties, it is not considered malignant[1,2]. Treatment of lipoblastoma consists of surgical excision and a close follow-up period of two to five years[3]. In this report, we discuss the rare occurrence of chest wall lipoblastomatosis in children and briefly review the English literature of this rare tumor.

Case Report

A two-year-old girl was admitted to our clinic with a rapidly growing mass on the right side of the chest wall first noticed by her parents a month earlier. Past medical history was unremarkable. Physical examination revealed a nontender, irregular, firm, and fixed mass measuring 3x2 cm over the right hemithorax approximately 1-2 cm lateral to the right areola. The remainder of the physical examination was unremarkable. Serum chemistry, complete blood count, human chorionic gonadotropin, alpha-fetoprotein, and carcinoembryogenic antigen levels were within normal limits. The posteroanterior and right lateral chest radiographs demonstrated a 4x5 cm opacity on the right anterior hemithorax. A computerized axial tomography scan of the chest showed a soft-tissue mass measuring 5x3x6 cm with irregular demarcated borders, including adipose tissue components. The mass occupied the anterior part of the right thoracic cage between ribs 7 to 11, anteriorly compressing the middle pulmonary lobe, inferiorly compressing the anterior right lobe of the liver and extending to the anterolateral chest wall through the intercostal muscles (Fig. 1). An abdominal computerized tomography scan was normal.

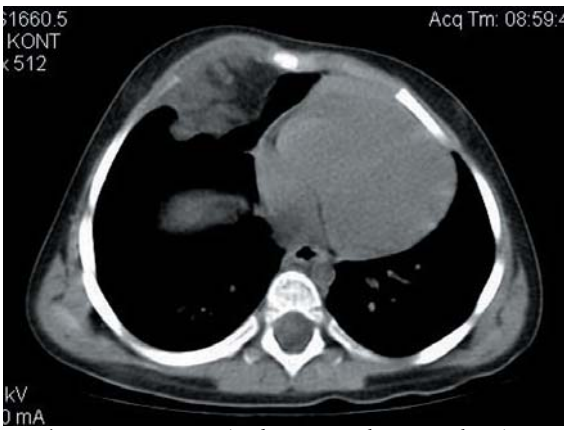

Fig. 1. A computerized tomography scan showing a mass occupying the anterior portion of the right thoracic cavity and compressing the middle pulmonary lobe.

Fine needle aspiration biopsy revealed no specific diagnosis or atypical cells. Planned surgical resection was made to accomplish both diagnostic and treatment goals. At thoracotomy, a multilobulated, irregular and encapsulated firm fatty mass was seen arising from the anterior chest wall and protruding into the right thoracic cavity (Fig. 2). Frozen section examination revealed an adipose tumor with no atypical cells. A complete excision of the mass was achieved successfully without resection of the chest wall. Results of the histological examination of the specimen revealed lobules of mature and immature adipose tissue, separated by well-vascularized fibrous septa. Myxoid stroma was present, and fatty cells showed a spectrum of maturation from primitive stellate cells to spindle cells (Fig. 3). Extension of the lesion to striated muscle tissues was observed in some areas; therefore, a diagnosis of lipoblastomatosis was made. The postoperative course was uneventful and the patient was symptom-free with no evidence of recurrences during the one-year period after the operation.

Discussion

Lipoblastoma, which was originally described in 1973 by Chung and Enzinger[6], is a rare benign mesenchymal tumor of adipose tissue. Although the exact etiology of lipoblastoma is unknown, immunohistochemical and ultrastructural studies demonstrated that lipoblastoma was related with continued proliferation of immature embryonic white fat cells and lobules in the postnatal period[2,8,9]. Clinicopathologically, there are two forms of lipoblastoma: the well-circumscribed, capsular and localized type is called benign lipoblastoma, and the deeply embedded, irregularly confined and noncapsulating type with the tendency to an infiltrative growth is called lipoblastomatosis[1-3,5]. Both forms usually occur in infants and young children and have a male preponderance, with a varying sex ratio[2,7]. However, four male and six female (including the current case) patients affected by chest wall lipoblastomas have been reported (Table I)[1,2,5,7,10-14]. Although they may arise almost anywhere within soft tissue, the trunk and extremities are the most common sites. The head and neck region, mediastinum and retroperitoneum have been reported rarely[15-17]. Chest wall involvement is also extremely

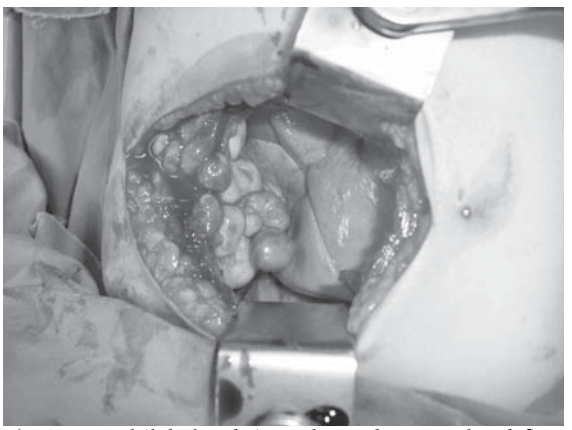

Fig 2. A multilobulated, irregular and encapsulated firm fatty mass, protruding into the right thoracic cavity.

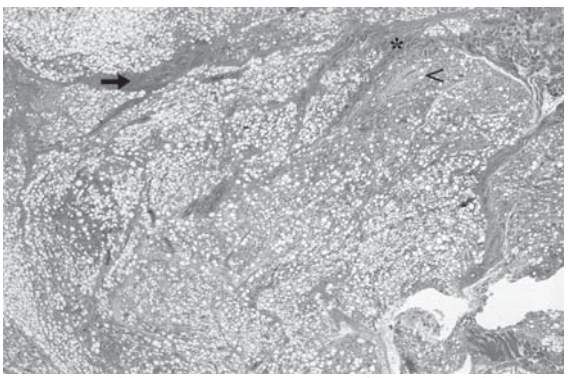

Fig 3. Lipoblastomatosis with its infiltrative and lobular pattern of growth (*). Note the presence of myxoid matrix (arrowhead) and fibrous septum between lobules (arrow) (hematoxylin & eosin stain, x20 magnification).

Table I. Lipoblastomatosis of the Chest Wall in Pediatric Patients

Cases	Authors	Age	Gender	Follow-up	Recurrences
1	Gülhan SS, et al.	7 months	female	24 months	no recurrences
2	Spinelli C, et al.	3 months	male	6 years	no recurrences
3	Cowling MG, et al.	6 months	NA	NA	two recurrences
4	Yuan RH, et al.	22 months	male	17 months	no recurrences
5	Samuel M, et al.	13 months	female	18 months	no recurrences
6	Collins MH, et al.	3 years	male	13 years	no recurrences
7	Jung SM, et al.	9 months	female	14 months	no recurrences
8	Chang PF, et al.	5 days	male	NA	no recurrences
9	Vellios F, et al.	8 months	female	30 months	no recurrences
10*	Yiğiter M, et al	2 years	female	12 months	no recurrences

NA: Not available. *: Our case.

uncommon. We could find only nine cases of thoracic wall lipoblastoma with four prior cases of lipoblastomatosis[1,2,5,7,10-14]. Lipoblastoma/lipoblastomatosis usually exhibits as a painless mass. However, some patients are symptomatic at the time of initial diagnosis because the rapidly growing mass causes compression effects on surrounding structures[3,17]. Our patient exhibited a rapidly growing but painless mass on the chest wall.

These tumors are macroscopically lobulated and soft. Histologically, lobules of mature and immature fat cells are found, including varying degrees of differentiation from primitive stellate cells to spindle cells. Lobules are separated by fibrous septa containing vessels, collagen and fibroblasts[2,3,18]. The diffuse form may differentiate from lipoblastoma with a less pronounced lobular pattern and an infiltrative growth pattern indicated by skeletal muscle fibers[2].

The differential diagnosis of lipoblastoma includes lipoma, hibernoma and myxoid or well-differentiated liposarcomas[2,5,13]. Histologic examination is almost always definitive. Lipomas are less cellular than lipoblastoma and lack lipoblasts, whereas hibernomas consist almost entirely of brown fat cells with mitochondria-rich, eosinophilic, granular cytoplasm[2,5]. However, the most important differential diagnostic consideration is myxoid liposarcoma because of the presence of lipoblasts and the plexiform vascular pattern in a myxoid background. The patient's age and histologic appearance are fundamental to distinguish lipoblastoma from myxoid liposarcoma. Younger patients, the lobulation and the absence of the pleomorphic and atypical cells with abnormal mitoses are more strongly correlated with lipoblastomas[1-5,10]. Histologically, borderline lesions may be differentiated with cytogenetic analysis. Characteristically, lipoblastomas have rearrangements in the long arm of chromosome 8 (8q11–13), affecting PLAG1, whereas a distinctive translocation t(12;16)(q13;p11) is observed in myxoid liposarcoma[2,19,20]. Nevertheless, despite the frequent presence of chromosome 8 anomalies, lipoblastomas may fail to show a karyotypic aberration. For this reason, the indication to perform cytogenetics was not specified. However, if the pathologist is unable to differentiate between lipoblastoma and myxoid liposarcoma, cytogenetic investigations should be performed. A brief summary of the chest wall lipoblastomas including results of cytogenetic analysis is presented in Table I. Because we resected the mass completely and lipoblasts were observed in all developmental stages on the pathologic examination, we had no doubt regarding the diagnosis.

Radiological studies may be helpful in the differential diagnosis of soft tissue tumors, but are usually not reliable in differentiating between the different lipomatous tumors. There are no known imaging findings associated with lipoblastoma; therefore, imaging is best used for surgical planning and postoperative surveillance rather than for diagnosis[3,19].

In spite of their potential for local invasion and rapid growth, lipoblastomas have an excellent prognosis with no report of any distant metastases. The surgical excision is usually curative if the tumor is removed completely[11,21]. Therefore, the treatment of lipoblastoma must be surgical excision with negative margins

except for large invasive lesions requiring mutilating resection[2,3,19]. In the chest wall lesions reviewed, only two reports revealed a chest wall excision including ribs 7 to 9 together with the parietal pleura[1,14]. In our patient, intra- and extrathoracic components of the tumor could be removed completely, avoiding potentially disfiguring procedures. When incompletely resected lesions exist, close clinical follow-up to monitor progression with various imaging methods is advised. All patients should have careful surveillance and a follow-up period of at least five years in light of the documented recurrence rate of these lesions[3,10]. Appropriate length of follow-up is controversial because there is a report of a recurrence seven years after the primary tumor removal[10]. In this respect, we planned a close follow-up with both clinical examinations and regular radiologic tests with ultrasonography over a five-year period. On the other hand, magnetic resonance imaging can be helpful to monitor size/progression in patients with multiple recurrences or incomplete tumor resections.

In conclusion, benign lipoblastomatosis differs from other benign pediatric lipomatous tumors due to the reported recurrence rate of these tumors. However, lipoblastoma has no risk of metastasis. The goal is complete resection of the tumor with negative margins though this is not always possible. We think that this is a benign tumor and that radical cancer surgery should be avoided even in cases where the tumor was not completely removed.

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