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Case Report

Intrathoracic extension of a chest wall Lipoblastoma in an infant: A rare case report[☆]

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ABSTRACT

We report the case of a 9-month-old female infant who presented with a history of enlarging chest wall mass secondary to a lipoblastoma with intrathoracic extension. The baby was successfully treated with a surgical resection. Chest wall lipoblastomas with intrathoracic extension are quite uncommon, with few reports available to date.

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Introduction

Lipoblastoma is a rare, benign mesenchymal-encapsulated tumor originating from embryonic fat cells [1]. It occurs almost always in infants and at a younger age, less than 3 years, and usually involves the extremities and torso [1,2]. Even if it has the capacity to grow rapidly, causing local invasion, it often has an excellent prognosis after complete surgical resection [3].

The chest wall is an uncommon location for the disease. Intrathoracic extension is even rarer. Symptoms depend on its size and growth rate [4–7]. Imaging defines the underlying at-

tenuation of the lesion and its extent. Overall, MRI (magnetic resonance imaging) is a better describer of the internal features. CT (computed tomography) is superior for assessing internal calcifications and rib changes [3].

Case presentation

A 9-month-old female infant presented with right posterior chest wall swelling noticed by her parents 3 months earlier, which progressively increased in size. The baby was in good health prior to her current examination. Her perinatal history

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

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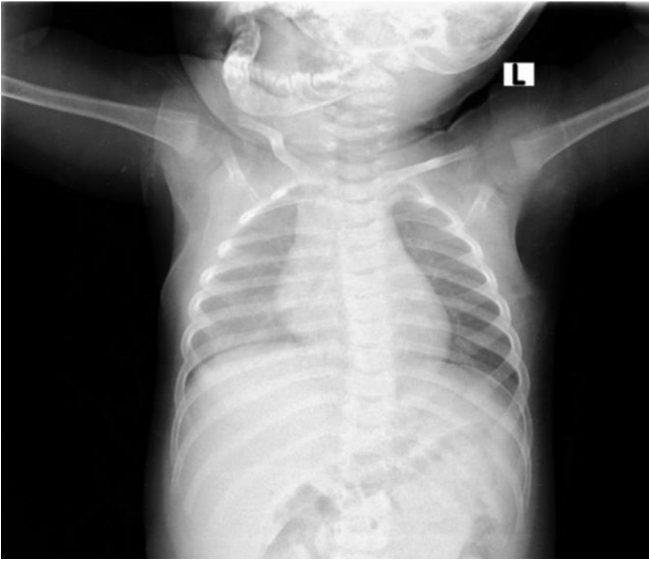


Fig. 1 – Anteroposterior chest radiograph depicts a hazy opacity veiled over the right hemithorax and ipsilateral scapular regions.

was also unremarkable. On physical examination, the child was stable-looking. There was a large, firm, fixed, and non-tender right posterior chest wall mass. Her vital signs were unaffected, and there were no signs of respiratory distress. The complete blood count was within the normal limits for age. The imaging evaluation was stated with a frontal chest radiograph (Fig. 1), which showed a large area of nonspecific right hemithoracic opacity. The lungs were normally expanded. According to sonography, the lesion was homogeneously echogenic (not shown). We performed a non-contrast chest CT (Fig. 2) to better visualize the lesion and the intrathoracic compartment. Contrast was not administered to decrease the radiation dose. The CT revealed a $9.7 \times 6 \times 3.3$ cm (CCxTRxAP) measuring right posterolateral chest wall lesion spanning from T1-T10 levels. It is well encapsulated, predominantly fat in density, with thick strands of soft tissue components. It showed a small intrathoracic extra-lung extension. Otherwise, there were no noted calcifications or adjacent bone destruction. The subcutaneous fat was not disturbed. After initially considering a fatty mesenchymal chest wall lesion, the patient underwent surgery, excising a large lesion and sending it for pathology.

The gross specimen consisted of grey, white, and yellowish relatively well-delineated multinodular globular tissue measuring 9.5 cm by 5.5 cm by 4 cm. The cut surface was yellowish and lobulated, with areas of myxoid change. The low-power microscopic examination of the represented samples showed lobulated sheets of adipocytes separated by a thin fibrous stroma (Fig. 3A). The adipocytes are mature-looking and found at the center, while myxoid areas are found at the periphery of the lobules containing primitive mesenchymal cells (Figs. 3B and C). No mitosis or marked pleomorphism were seen. Foci of growth into skeletal muscle were seen. The histologic findings were consistent with lipoblastomatosis.



Fig. 2 – Non contrast CT images in axial (A, B) and reconstructed coronal (D) and right parasagittal (E) planes show a non-calcified large predominantly fat attenuating mass containing thick internal soft tissue components in the right posterolateral thoracic wall. An anteromedial intrathoracic extension (arrow in B) is seen between the second and third ribs (arrow in B). The adjacent ribs are unaffected. C shows the craniocaudal extension of the lesion clearly.

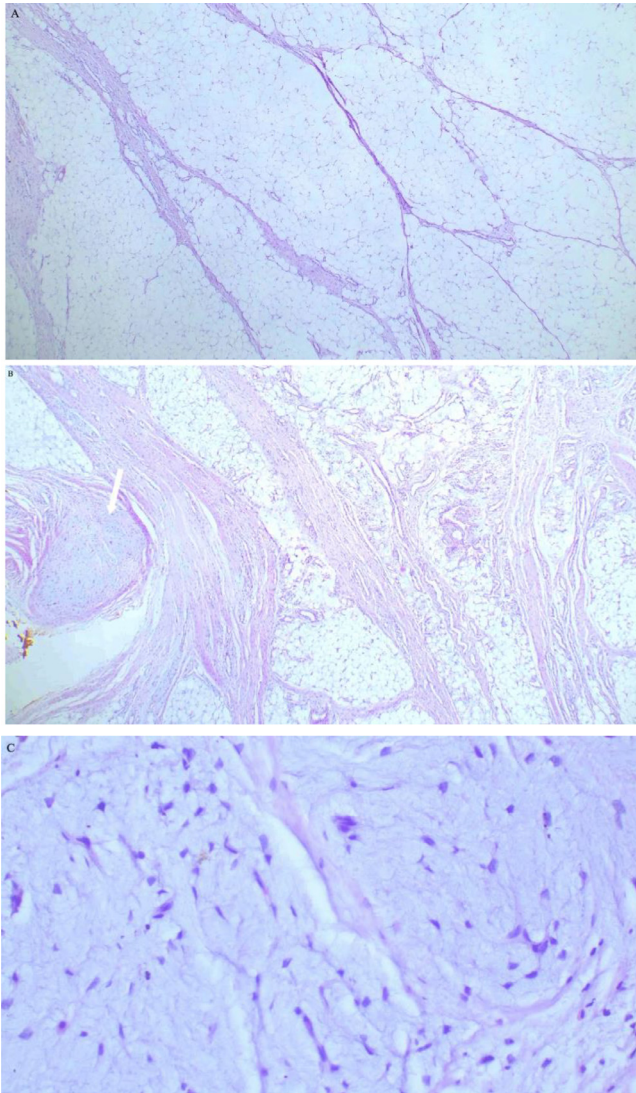


Fig. 3 – Low power magnification-40x (A and B) and high power magnification- 100x (C) photomicrographs demonstrate mature adipocytes in lobulated pattern separated by fibrous septa (A). B shows lobulated adipocytes with peripheral myxoid areas (arrow). In the high power magnification image (C), the myxoid area containing primitive mesenchymal cells is seen.

Discussion

Lipoblastoma is a benign tumor of embryonic white fat cells called lipoblasts or adipoblasts. Unlike a lipoma, it lacks mature adipocytes [1,2]. Lipomatous lesions are uncommon in children and account for only 10% of pediatric soft tissue tumors occurring at an age below 20. Lipoblastoma is even rarer and is responsible for 1% of all childhood tumors [1]. Infants receive 50% of the diagnoses, and 90% occur before the age of 3 [8]. Lipoblastoma predominates in boys by a ratio of almost 4:1 [5]. Two forms are described: The majority, accounting for 66% of the cases, are lipoblastomas, which are more superficial,

well-defined, and non-infiltrative. Lipoblastomatosis, a closely related entity, has a similar histologic make-up but is deeper, has a non-encapsulated border, and tends to be infiltrative. Metastasis to distant organs does not occur with lipoblastoma or lipoblastomatosis [1–3]. The only aggressiveness seen in these lesions is local invasion, which is dependent on their growth rate and the sizeable dimensions they can attain. As a result of the white fat predominance in the trunk and extremities, lipoblastoma occurs more commonly in these locations. Head and neck, mediastinum, abdominopelvic, perineum, and genital regions are other regions where lipoblastoma has been described [3].

Chest wall Lipoblastoma is rare. Yigitar et al. [9] compiled 10 cases of chest wall lipoblastoma, including one case from the author's institution. Moreover, intrathoracic extension, such as in our case, is extremely rare [10]. Symptoms depend on the size and location of tumors. Although lipoblastomatosis have an infiltrative nature, they are still considered benign. Chest wall lesions present with a swelling that has a relatively fast growth rate [7]. With intrathoracic extension, symptoms include dyspnea and recurrent infections [4,6].

Imaging helps in defining the origin, size, and extent of lesions. Cross-sectional imaging using CT and MRI shows the fatty component of these tumors admixed with a variable amount of myxoid stroma, fibrovascular tissue, and soft tissue parts. The adipose component predominates in older children, whereas in infants, the myxoid portion is in excess. CT is better at identifying intra-tumoral calcification or ossification, which is not a feature of lipoblastoma but of a teratoma. The relationship to adjacent ribs is important to review, as rib enlargement caused by benign lipoblastoma can make surgical removal difficult [7]. MRI is better for the internal description of these heterogeneous lesions. The fatty components are isointense with the subcutaneous fat. Myxoid parts, due to their high water content, are low and high in signal intensity on T1W and T2W sequences, respectively. Fibrous strands and soft tissue nodularities can show minimal enhancement. In a similar pattern, sonography shows the fatty part as homogeneously hyperechoic and the myxoid and fibrovascular components as mixed echogenic materials. On chest radiographs, lipoblastomas are commonly soft tissue in density without bone destruction. Contrary to the usual lipoma, they do not contain organized mature fat cells to appear radiolucent [2,11].

Other fat-containing mesenchymal lesions can be considered a differential diagnosis. Lipoma is a well-defined, superficial tumor that displaces rather than infiltrating. But imaging fails to adequately differentiate lipoblastoma or lipoblastomatosis from liposarcoma, especially the myxoid type, which is the most common type of liposarcoma in the pediatric age group. The latter is rare in children, but the final diagnosis rests with a pathologic examination [1,11,12].

Surgery is the best treatment option, with a complete but conservative resection undertaken that diminishes the chances of recurrence. Even though both lipoblastoma and lipoblastomatosis can recur, the risk is higher in the latter [1,3].

In conclusion, the chest wall and intrathoracic regions are very rare locations for lipoblastoma but should be considered in fat density lesions, and intrathoracic extension should be evaluated.

Patient consent

Complete written informed consent was obtained from the patient's parents for the publication of this study and accompanying images.

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