Abstract

Lipoblastoma is a rare benign mesenchymal tumor of embryonal fat that occurs almost exclusively in infants and children below the age of 3. It is a benign tumor with high recurrence rate. We present a case of mediastinal lipoblastoma in a 21-month-old girl patient who presented with respiratory infection and progressive dyspnoea. Complete surgical excision of the mass was achieved via lateral thoracotomy. Post operative course was uneventful and histopathological examination proved the mass to be lipoblastoma.

We emphasize that this rare mediastinal tumor should be included in the differential diagnosis of infants having mediastinal mass.

Keywords: Lipoblastoma, Mediastinum, Surgery.
Introduction

Lipoblastoma and lipoblastomatosis are very rare benign mesenchymal tumors most often found in children less than 3 years old. They may present as a localized well-circumscribed lesion, the so-called lipoblastoma, or as a multicentric, deep and infiltrative type, the lipoblastomatosis. They may arise almost everywhere within the soft tissues, the trunk and the extremities being the most common predilection sites; however, neck, mediastinum are an uncommon location for these tumors [1].

Mediastinal Lipoblastoma can be presented with progressive dyspnoea as in our case, or with neurological symptoms resulting from spinal cord compression [2]. It is important to completely excise the tumor to avoid leaving residual tumor and to prevent recurrences which mostly occur within 2 years.

Case report

A 20-month-old girl presented with respiratory infection and progressive dyspnoea. A chest x-ray showed that she had a right-sided mass involving the upper zone of the thorax, causing displacement of the trachea to the left side (Fig.1A).

The patient was treated for pneumonia that resolved few days later with intravenous antibiotics. Computerized chest tomography scan showed a large fat containing lesion with some septation in mediastinum and right side chest and engulfing some of the main vessels especially branches of the aortic arch (Fig.1B).

Lateral thoracotomy was done. Intra-operatively the mass was found to be engulfing branches of the aortic arch but with clear plane of dissection. Complete surgical excision was achieved without sacrificing any vessel. Post operative course was uneventful.
Histopathological examination revealed adipose tissue composed of mature and immature fat cells in myxoid stroma (Fig.2).

Comment
Lipoblastoma is a rare tumor of infancy and early childhood. It was first described by Jaffe in 1926,[3] and since then many cases were reported; Chung and Enzinger, in 1973, suggested the term benign lipoblastoma for the circumscribed type, and the term benign lipoblastomatosis for the diffuse multicentric type of this neoplasm.[4].
Although lipoblastomatous tumors are benign, they may grow very fast, attain large sizes, and become a painless mass, making it the most common symptom. Tumors located at the mediastinum may present with stridorous respiration, and may also present with neurological symptoms resulting from spinal cord compression like paraparesis, hemiparesis,[5,2].

Although CT scan may show a tumor of fatty origin, preoperative differential diagnosis between other fatty tumors such as lipoma, liposarcoma and myxoliposarcoma is quite difficult[6]. However, lipoblastomatous tumors differ from lipoma or lipomatosis by their cellular immaturity histologically, and they have close resemblance to low-grade liposarcomas. On the contrary, liposarcomas are tumors that occur in adults, and are extremely rare in infants. Histologic differential diagnosis between lipoblastomatous tumors and liposarcomas is based on the prominent lobular architecture of lipoblastomatous tumors in addition to the lack of nuclear atypia unlike the myxoid liposarcomas. In addition, myxoid liposarcomas show a characteristic t (12; 16) translocation, which is not present in lipoblastomatous tumors, and recent studies have defined rearrangements of chromosome 8q11-q13 observed as deletions in lipoblastomatous tumors to distinguish from myxoid liposarcomas [7].
Complete surgical resection of lipoblastomas yields an excellent prognosis. Lipoblastomas are benign tumors, and no malignant degeneration has been documented. Recurrence has been reported in 14% to 25% of cases, usually due to incomplete resection or diffuse disease. Lipoblastomatosis exhibits higher recurrence rates partly due to its propensity to localize within deep soft tissues, which makes complete excision more difficult to achieve [8].

In conclusion, Mediastinal lipoblastoma is an uncommon site of this tumor, but it should be considered in the differential diagnosis any rapidly growing soft fatty mass in children. Surgical excision is the best therapeutic option and gives excellent results. However, high recurrence rate demands follow up.
References


Fig 1 (A) Chest x-ray showing the mass within the right hemi thorax.

Fig 1(B) CT scan showing the mediastinal tumor.
Fig 2. Histological view showing mature and immature adipose tissue.
(Hematoxyline & eosin stain ×600)