An Hourglass Type of Intrathoracic Lipoblastoma Manifested by Edema in Right Upper Limb

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Introduction

Lipoblastoma is a rare benign tumor that arises from the fetal- embiyonal fat tissue. This entity is almost exclusively seen in infants and children and has two morphologic forms: a localized well-circumscribed lesion (hpoblastoma), or a multicentric type (lipoblastomatosis)[1-4]. . Most of them originate from extremities but several other sites have been reported[1,5]. To our knowledge, there are only 4 previously reported intrathoracic lipoblastoma cases in the literature[5-8]. A case of an intrathoracic lipoblastoma of hourglass type is presented and discussed.

Case Report

A 2-year-old boy was admitted to our department with a history of swelling of the right upper extremity beginning 3 months ago. Before admission, lie had been managed conservatively by compression with elastic banthge for 3 months in a different center.
Physical examination revealed that the diameter of the right upper extremity was greater than that of the left. A Chest x-ray and ultrasound (US) showed a mass in the anterior axillaiy region and right hemithorax. Computerised tomography (CT) demonstrated a right upper thomeic solid fatty tumour with extrathoracal extension to axillaiy region (Figure 1).

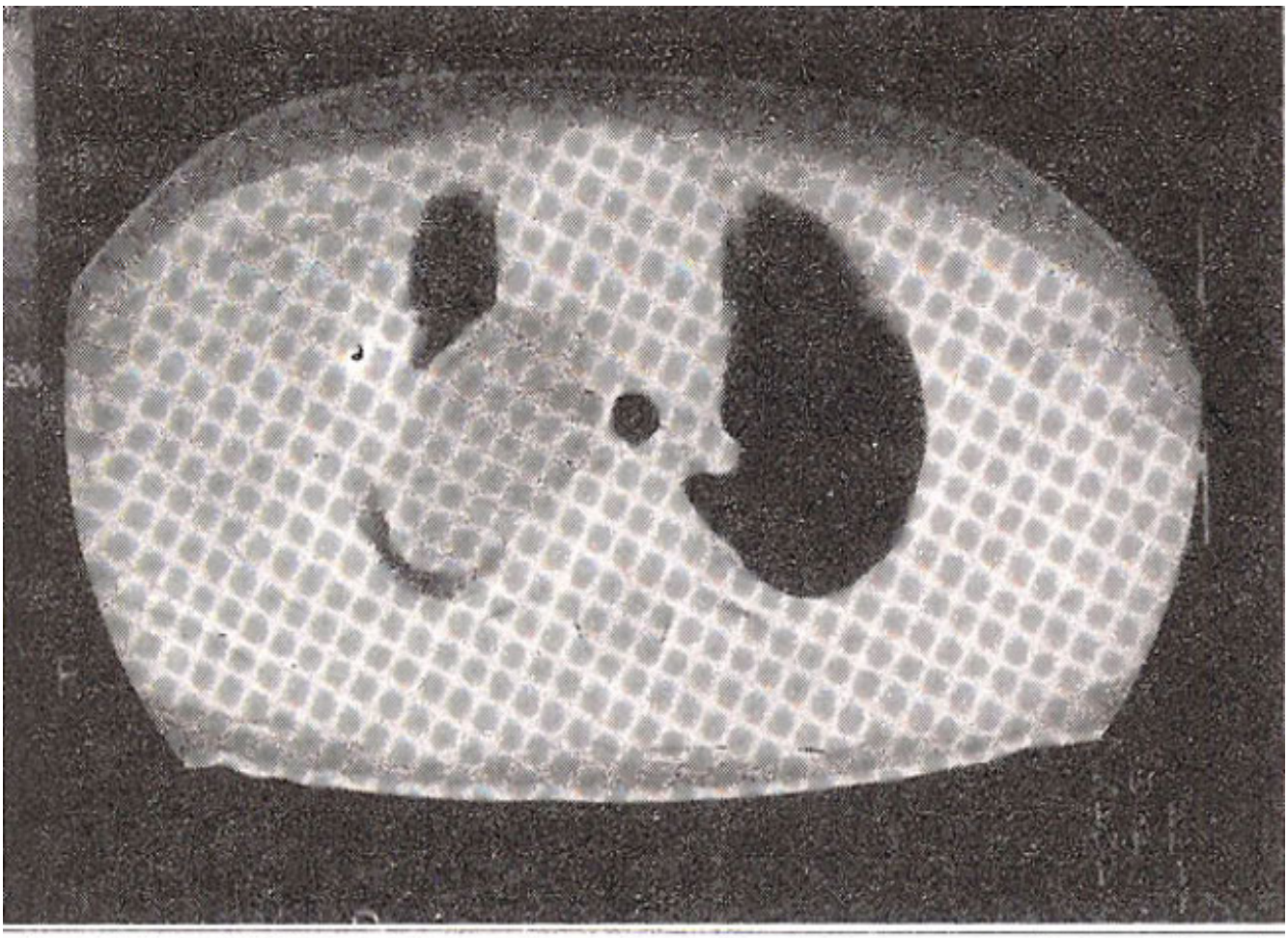

Figure 1. Computed tomography revealing right intrathoracic lipoblastoma with its extrathoracic extension.

Laboratory tests including complete blood count, blood and urine biochemistry, hepatic function tests were nonnal. Tumour markers of vanillylmandelic acid (VMA).
Homovanillylic acid (HVA), neurone- specific enolase (NSE), ferritin showed normal levels in the serum. Bone marrow aspiration biopsy, seintigraphic study of long bones and chromosome analysis were also normal.
Right thoracotomy was performed. An intrathoracic solid, soft, fatty tumor - 5 cms in diameter each, intra- and extrathoracie part and 3 cm in thickness - extending through the 3rd intercostal space was found. The tumour was completely removed and weighed 100 g (Figure 2).

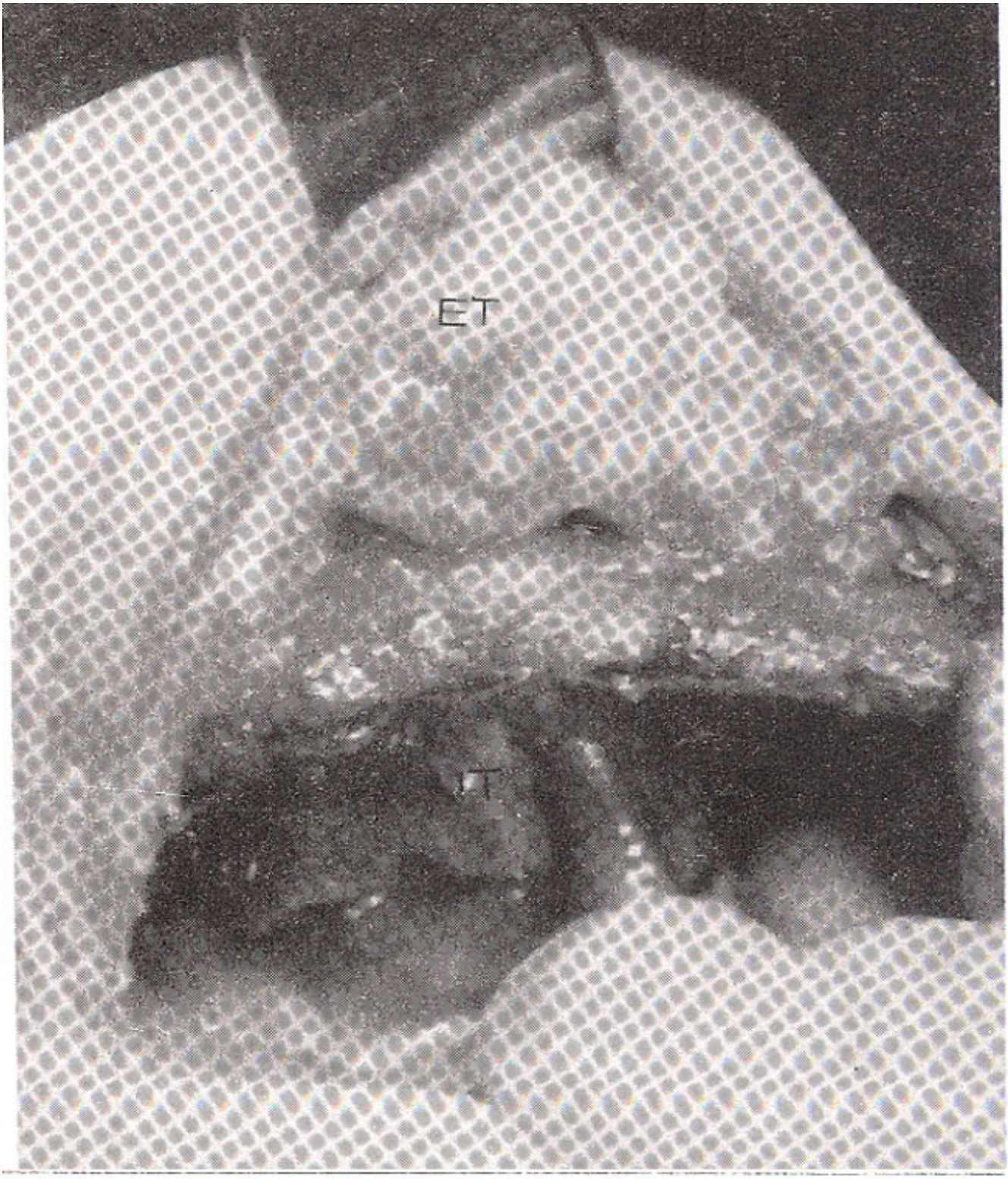

Figure 2. Appearance of intra- and extrathoracic portions of lipoblastoma during operation (IT: Intrathoracic portion, ET: Extrathoracic portion).

Grossly, the tumor was an encapsulated, lobulated mass (l0x6x4 em). On cut section, it was pale (yellow-white), septate and partly myxoid. Microscopically, it was multilobular, septatedby loose fibrous bands and composed of partly differentiated lipoblasts, a prominent vascular pattern and

abundant mucoid matrix. The cells were quite uniform round to spindle-shaped, with fine vacuoles. There were no mitotic figures. The histopathological diagnosis was 'lipoblastoma' (Figure 3a,b).

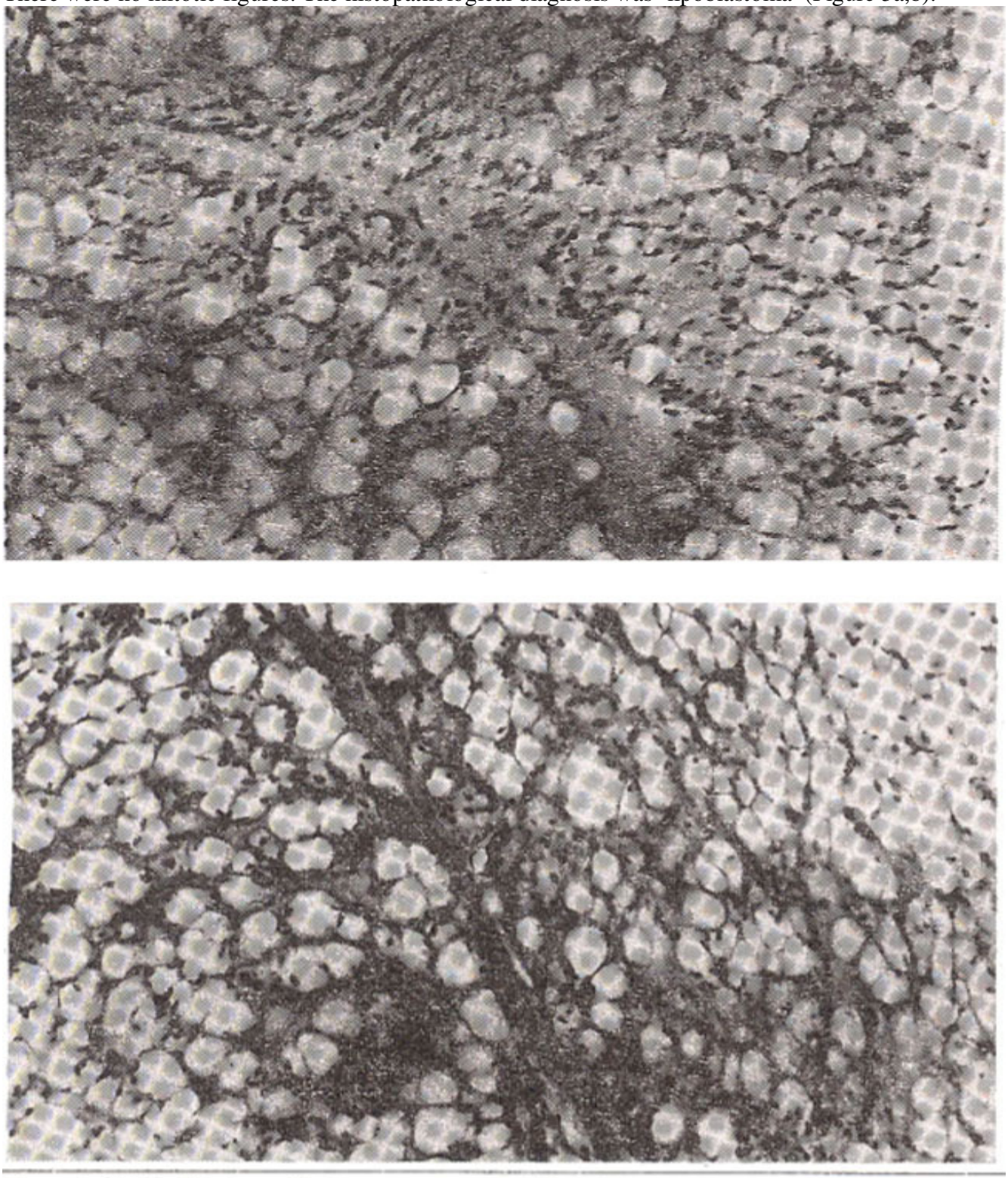

Figure 3a, b. Lipoblastoma-lobular pattern, partly differentiated lipoblasts, prominent vascular pattern, abundant mucoid matrix (HEx275).

Postoperative recovery was uneventful. With a follow-up of 2 years no recurrence was observed.

Discussion

Lipoblastoma and lipoblastomatosis are uncommon types of benign tumors arising from mesenchymal tissues. Of reported 190 cases in the first year of life, 3% were lipoblastomas[9]. Only rare examples in older children and young adults have been reported, but some of these almost certainly represent liposarcoma[10].

In previous reports, lipoblastornas have been noted to have a predilection for sites that possess the most primitive adipose tissue in the newborn such as axilla, neck, chest wall and prevertebral soft tissue[4]. However, in a series of35 cases, 70% of the tumors occurred in the extremities[1]. Lipoblastomas, thoracic in location, are extremely rare[5-7].

Intrathoracic lipomatous tumours were classified into two groups by Keeley[11], 1) Pure intrathoracic type. 2) Hourglass type. Pure inirathoracic type may be mediastinal or extmpleural in location without extension out of the thoracic cavity. Hourglass type has both an intra- and an extrathoracic component with an extension into either the cervicomediastinal area or transmurally through the chest wall[11].

Most patients with intrathoracic lipomatous tumours are clinically asymptomatic. However, a large tumor compressing vital structures in thorax may cause pressure symptoms even death. Transmural lipomatous tumors may impinge on the ribs, separate them and produce pressure necrosis[12]. In our case, the unique symptom was the generalized edema in the right upper extremity because of compromised venous return. Detailed history of our patient revealed that respiratoiy symptoms like chest pain or coughing preceded the swelling of the extremity. At that time, a roentgenogram of the chest disclosed an opacification in the right upper thoracic cavity and the patient was referred to our clinic for further investigations and treatment. The patient could riot be treated at that stage due to non-compliance of the family and he was admitted to ourclinic whenthe symptom of generalized edema in the right upper extremity was added to the clinical picture. Although axillaiy region is a more favoured site compared to thoracic cavity, the history suggested this was the case of an intrathoracic tumor with extrathoracal extension through the axillaiy region.

In radiological evaluation of these tumours, plain radiographs and US disclosed a soft tissue mass. In addition, to chest x-ray and US, CT was done in our case and found to be very useful because it not only demonstrated the limits of the tumor, but also showed that it was fonned mainly of low density tissue suggestive of fat.

It is notable that consistent, apparently diagnostic chromosome re-arrangements have been described in several varieties of soft tissue tumors[13]. Lipoblastorna is one of these tumors in which cytogenetic analysis may be helpful in diagnosis. The most common cytogenetic aberration found in pediatric lipoblastornas is re- arrangement of the chromosome 8 long arm, in more than 25% cases[2,14,15]. Chromosome analysis in our case was found to be normal.

Intrathoracic lipoblastomas should be managed by total surgical removal without the need of a wide “radical” type of operation. Complete resection of tumoral mass was possible in our case.Metastases have not been reported, but local recurrence is a distinct possibility, so careful follow-up is essential. Recurrence is not considered likely after an interval of 12 months[5]. With a follow-up of two years after resection, our patient is asymptomatic and there is no evidence of tumor recurrence.

It is thus suggested that in differential diagnosis of a child with asymmetric appearance in upper extremity due to generalized edema, an intrathoracic tumor with extrathoracal extension through the axillary region should be kept in mind, investigated and treated appropriately.

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