O. Tahbaz MD[1].
M. Bakhshayesh Karam MD[1].
S. Zahirifard MD[1].
K. Kaynama MD[2].
S. Haghighi MD[3].

Mediastinal Lipoblastoma: Report of a Case

Lipoblastomas are rare benign tumors arising from fetal-embryonal fat that almost always occur in children and about two thirds of them occur in the superficial or deep layers of soft tissue on extremities. To our knowledge there are only 6 case reports of mediastinal lipoblastomas in the English medical literature. We report a case of mediastinal lipoblastoma in a 2.5-year-old child. A mediastinal mass was found on the chest x-ray, CT (Computed Tomography) scan and MR (Magnetic Resonance) imaging. We found a fatty density mass with no evidence of septation, strading and soft tissue density without enhancement after contrast injection. On MRI, the mass was high-signal intensity on T_1 and T_2. Complete surgical resection was performed and the pathological examination proved lipoblastoma.

Keywords: mediastinal neoplasms, lipoblastomas, imaging.

Introduction

Lipoblastomas are rare benign mesenchymal tumors of embryonic white fat that occur in infancy and early childhood. They constitute only 3% of the soft tissue tumors in the first year of life. Approximately two thirds of them are found in the extremities. [1, 2] Most lipoblastomas arise in the extremities, although some originate in the trunk, head or neck. [3] We present a case of mediastinal lipoblastoma who was admitted in our hospital. So far, there have been only 6 case reports of mediastinal lipoblastomas in the English medical literature. [4, 5]

Case presentation

A 2.5-year-old boy presented with cough, fever, nausea, vomiting and abdominal pain of two months' duration. He did not have dyspnea, poor feeding, weight loss and falling of the growth curve. Past medical, family and drug histories were unremarkable and no history of malignancy or benign tumors was found in his family either. The child appeared normal on physical examination and no fever was detected.

The laboratory findings on complete blood count (CBC) differentials, and biochemical tests were within the normal limits; however, the erythrocyte sedimentation rate (ESR) was 18 mm/h.

Posteroanterior and lateral chest radiographs were obtained. A large mediastinal mass was evident on the right side of trachea causing tracheal deviation (Figure 1).

He was admitted in our hospital (NRITLD) for further work-up including gastroduodenoscopy, thorax CT scan and MRI.

Gastroduodenoscopy was performed for the gasterointestinal symptoms and revealed severe gasroduodenitis.

Thorax CT scan showed a 5×4 cm well-defined fatty density mass on the right side of superior mediastinum with inferior extension to the right side of the middle and posterior parts of the mediastinum. It extended from base of the neck

1. Assistant professor, department of Radiology
2. Department of Radiology
3. Department of Pathology

National Research Institute of Tuberculosis and Lung Disease (NRITLD)
Maseeh Daneshvary Hospital
Shaheed Beheshti University of Medical Sciences

Corresponding Author:
Mohammad Omid Tahbaz
Address: Maseeh Daneshvary Hospital,Shaheed Bahonar Ave., Darabad, Tehran., Iran
PO Box: 19575/154
Tel: 0098-21 22282111
Fax: 0098-21 22285777
E-mail: omidtahbaz@yahoo.com

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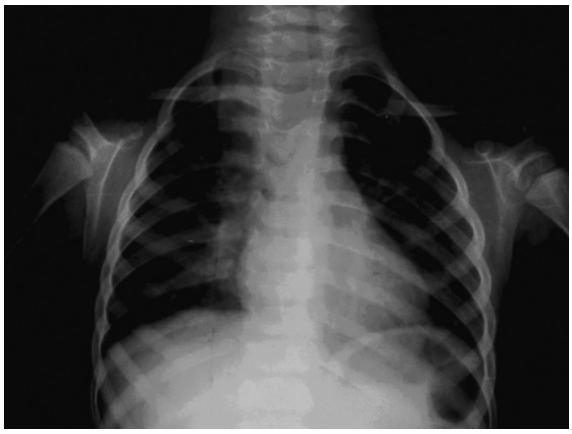

Fig 1. Posteroanterior chest radiography shows the superior mediastinal mass causing tracheal displacement .

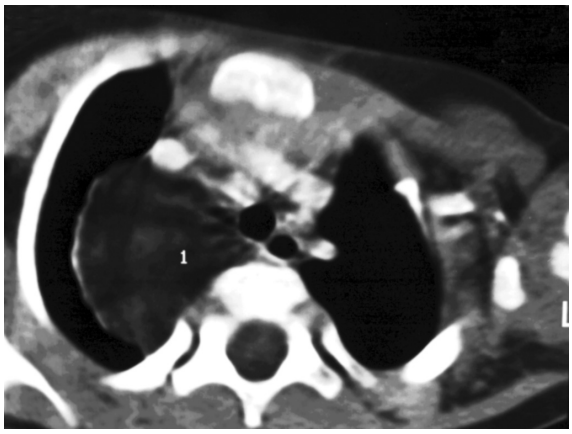

Fig 2. CT scan shows a well-defined fatty density mass.

and the thoracic inlet down to the level of carina. No septation and strading was detected within the lesion (Figure 2).

On MRI, a large mediastinal mass was found at the right side of the superior and posterior mediastinum. It had a well-defined margin, and showed high-signal intensity on T_1 and T_2 in favor of a fatty lesion. Findings were suggestive of lipoma (Figure3).

Right posterolateral thoracotomy was performed. Surgical exploration revealed a large yellow encapsulated mass with soft consistency which was located at the right superior and posterior mediastinum. The mass was excised and sent for histopathological examination.

On pathological examination, microscopic view revealed a neoplastic proliferation of adipocytes at different stages of maturation (mostly mature adipocytes), mixed with vacuolated and nonvacuolated spindle adipocytes and rarely lipoblasts. Foci of myxoid changes and patchy areas of hypervascularity were also seen. No nuclear anaplastic mitotic figures were identified. The pathological diagnosis was lipoblastoma (Figure 4).

No radiological evidence of recurrence was found 1.5 years after the surgical resection.

Discussion

The clinical course, radiological findings and pathological diagnosis regarding the patient's age were compatible with a diagnosis of mediastinal lipoblastomas.

To our knowledge, only six patients with mediastinal lipoblastomas have been reported in the literature. [4, 5, 9-12] Irgau I et al in 1998 and Rao KL et al in 2003 in 2 disticnt case reports noted that there were respectively only 4 and 5 reports of patients with mediastinal lipoblastomas in the literature. [4, 5]

Lipoblastomas are rare, usually encapsulated, benign neoplasms of the embryonal fat. They are composed of both mature and immature fat cells and occur almost exclusively in infants and children. Ninety percent arise before the age of 3 years. The male: female ratio is 3:1. Most lesions present as a subcutaneous soft tissue mass on the extremities or in the head and neck area. Trunkal lesions are less common. [6] Lipoblastomatosis is the unencapsulated diffuse form of the lesion. [3]

Histologically, the differentiation of lipoblastoma from liposarcoma, especially the myxoid variant may be difficult. These benign immature adipocytic neoplasms have a more uniform growth pattern and typically show more striking lobulations. [7] Although, lipoblasts at different stages of maturation are present in lipoblastoma, there is no nuclear atypia or pleomorphism, in contrast to that seen at least focally in liposarcoma. [1]

When arising from the neck or mediastinum in younger patients, these tumors can grow rapidly, leading to recurrent respiratory infections or asyphyxia, occasionally causing death if not removed. [8] As we mentioned, coughing was the sole respiratory symptom in our patient. However, previous case reports mentioned other respiratory signs and symptoms such as wheezing, hoarseness, severe respiratory

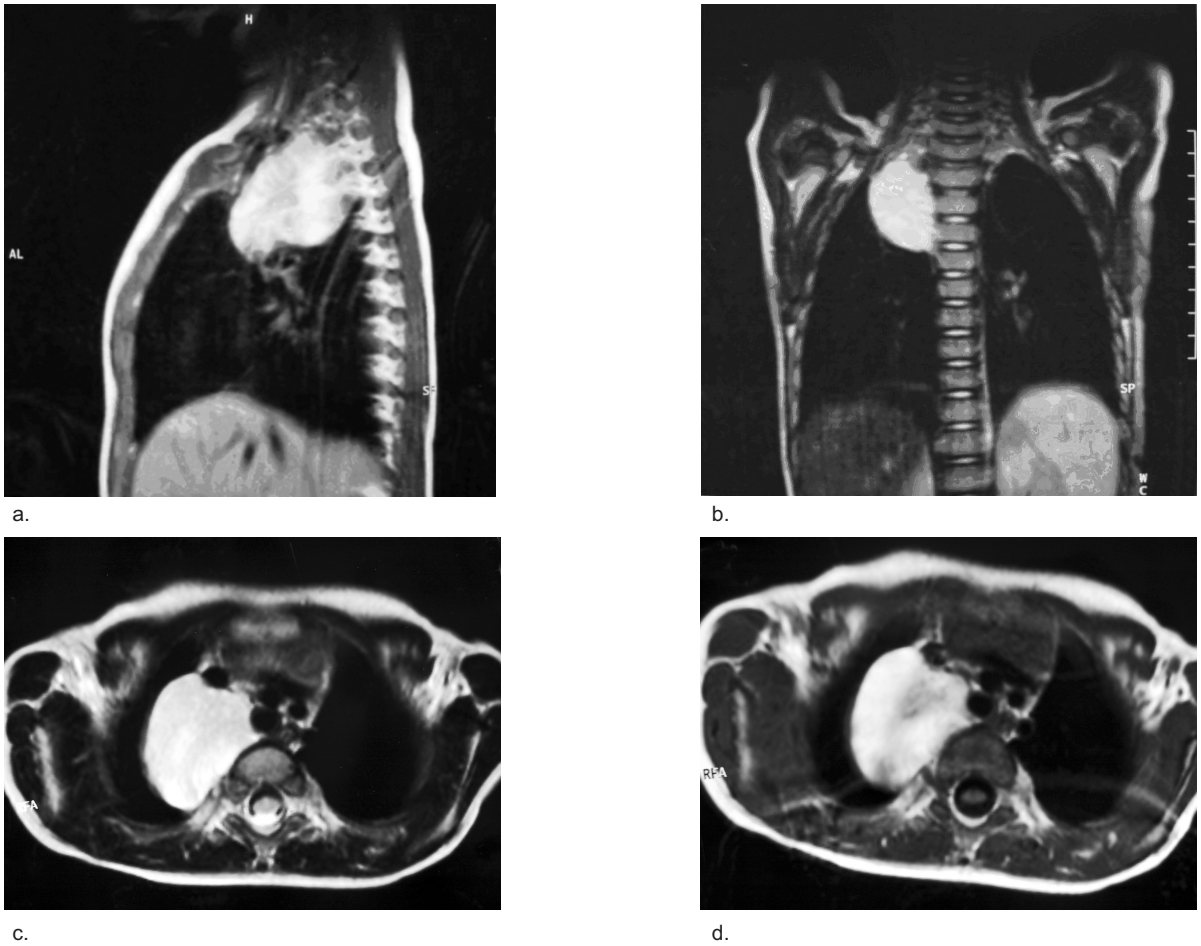

Fig 3. MRI; a, b) Mediastinal mass is shown. c) T2-weighted image. d) T1-weighted image . A high-signal intensity mass on T1 and T2 with a well-defined border is seen.

distress and pleural effusion (once ruptured) .[7, 9]

On conventional chest radiography, lipoblastoma appears as a non-specific soft-tissue density mass.[7]

On CT scan, lipoblastomas contain fat separated by septa of soft-tissue attenuation and do not enhance after administration of contrast media. [13] In 2003, in a case report by Rao KL et al, the tumor had mixed fat and soft tissue attenuation on CT scan which did not enhance significantly. [5] Cowling MG et al, in 1995 reported a case of ruptured lipoblastoma that showed areas of inhomogeneous contrast enhancement on CT.[7] Also CT scan of a patient reported by Whyte Am et al, demonstrated a mass of principally fat attenuation with enhancing hyperdense soft tissue whorls[12] However, on CT scan of our patient, no septation or strading was detected and the mass did not reveal enhancement.

MRI finding of a patient reported by Ko SF et al. in 1998 were low intensity to fatty on T_1-wieghted images and high-signal intensity on T_2-weighted images. They thus concluded that this atypical manifestation could be ascribed to the presence of an excessive amount of immature fat and myxoid tissue, intratumoral infarction and extensive mucoid and cystic degeneration. [2] In a review article by Castellote A et al in 1999, according to the previous case reports, it was noted that unlike lipomas, which have a characteristic appearance on MR images (high-signal intensity on T_1-wieghted images), lipoblastomas can be heterogeneous and have intermediate to high-signal intensity on T_1-wieghted images depending on the amount of immature lipoblasts. [3]

On MR imaging of the young boy we presented, high-signal intensity on T1-weighted and T2-weighted images were shown, although mature adipocytes predominated on histopathological examination, with some lipoblasts that were also detected.

In the case of ruptured lipoblastoma reported by Cowling MG et al, MR images showed linear streaks and whorls of low-signal intensity in the lateral chest

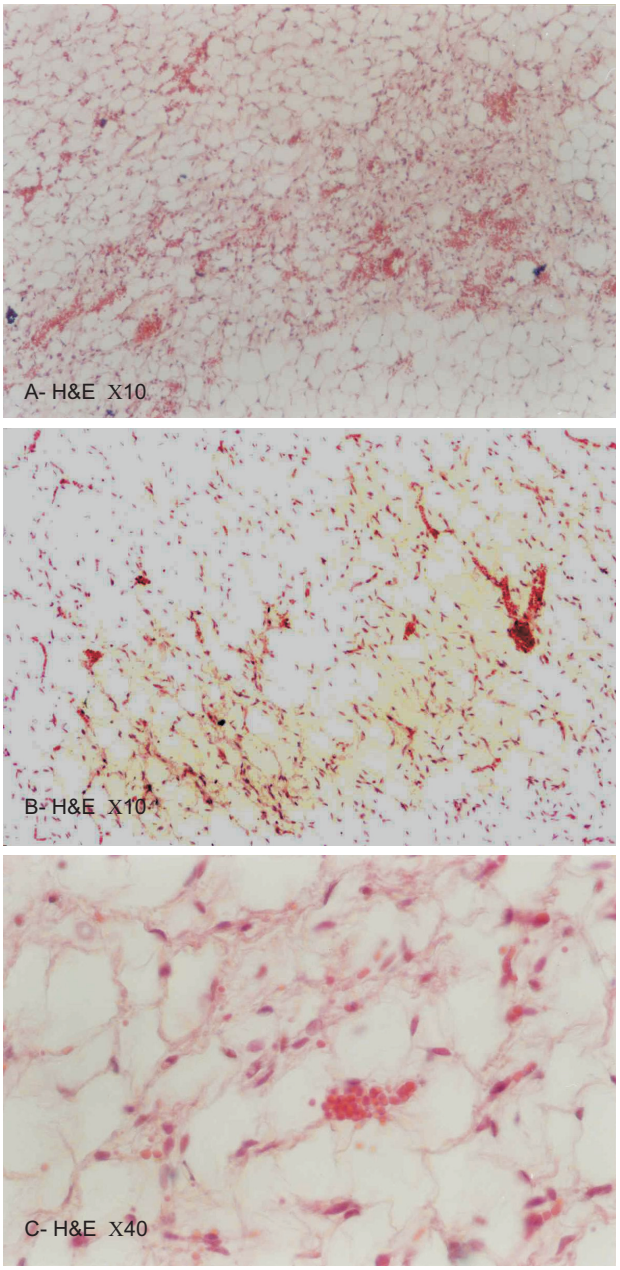

Fig 4 A, B, C) Different stages of adipocyte maturation in the richly vascularized loose myxoid matrix background.

wall, which corresponded with the area of contrast enhancement on CT scan.[7]

In our patient, based on the patterns visible on CT and MR images and the information from the previous articles, we initially assumed that the mass could be a lipoma until the pathological examination of the resected tumor revealed a lipoblastoma.

The main differential diagnosis is liposarcoma which may well resemble the clinical and radiological presentation of lipoma. Liposarcoma in patients under 10 years of age is exceedingly rare and its myxoid type is nearly impossible to be histologically distinguished from lipoblastoma. [1]

As in our patient, local recurrence following surgical excision of lipoblastoma is uncommon. Metastases have not been reported either. [8]

We could not explain the gastroduodenitis detected by endoscopy that was performed to evaluate nausea, vomiting and abdominal pain of our patient.

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