

Intrathoracic lipoblastoma in a 15-month-old infant

Bitá Geramizadeh,^{1,2} Farshid Javadi,² Hamid-Reza Foroutan³

¹Transplant Research Center, ²Department of Pathology and ³Department of Surgery, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

Lipoblastoma is a rare tumor of infancy. It originates from the white fetal fat in soft tissue. The most common location of this rare tumor is extremity and to best of our knowledge less than 10 cases of intrathoracic and mediastinal lipoblastoma has been reported in the English literature. Herein we present our experience with a 15-month-old boy infant who presented with severe dyspnea. Imaging studies showed a mass in the thoracic cavity and mediastinum which was diagnosed as lipoblastoma after pathologic examination of the resected mass. Lipoblastoma has been considered as a tumor of soft tissue, but it should also be considered as a rare cause of intrathoracic masses of young children.

Introduction

Lipoblastoma is a rare benign tumor of embryonic fat. It most commonly occurs in infancy and early childhood.¹ Although it is a benign tumor, but it has the capability to grow rapidly with recurrence rate of 12-24%.² Less than 200 cases have been reported in the literature since its first description in 1926; and it is exceptionally rare in the thoracic cavity.³ Herein we report a 15-month-old infant with intrathoracic and mediastinal mass which turned out to be a lipoblastoma after surgery and pathologic examination.

Case Report

A 15-month-old infant was brought to our pediatric ward with chief complaint of lethargy, poor feeding, dyspnea and tachycardia since 10 days prior to admission. He has had recurrent respiratory infection in the last 4 months.

His past medical history was unremarkable and he was born after normal vaginal delivery (NVD) with uneventful neonatal period. Physical examination showed blood pressure: 120/70 mmHg, pulse rate: 120/min, respirato-

ry rate: 40/min, and T: 36.5°C. Chest examination showed decreased breathing sound in right side of chest. Abdominal examination revealed palpable liver 4 cm below costal margin, otherwise unremarkable. Chest X-ray showed total opacification of right hemithorax (Figure 1).

Chest CT scan revealed a large hypoattenuated lesion in right hemithorax with significant shift of the heart and mediastinum. Total collapse of right lung was noticed with downward displacement of right hemidiaphragm (Figure 2).

Preoperative tru-cut needle biopsy was failed and no adequate tissue was obtained for diagnosis.

There was high degree of suspicion regarding possible tumor, so decision was made to perform surgery and excise the tumor.

With a right posterolateral thoracotomy incision, a large firm and solid mass was identified involving the entire mediastinum and right thoracic cavity. Frozen section of the mass was diagnosed as benign myxoid tumor. The mass was attached to the pericardium and right pleura, but was easily detached and separated from the surrounding tissue. There was no attachment to phrenic nerve. The entire mass was resected without any complication and sent for pathologic study.

The received specimen in the pathology laboratory was a large creamy white oval-shaped and myxoid to fatty tumor, measuring 20×15×6 cm. Cut section of the mass was the same as surface (Figure 3).

Histologic examination showed low cellularity with delicate fibrous bands demarcating lobules composed of mature white fat and myxoid changes (Figure 4). The tumor cells showed spindle to stellate shape with vacuolated cytoplasm. Many chicken-wire type small blood vessels were also present. No cellular atypia, necrosis or mitosis was identified. Immunohistochemical study showed reactive S100, and CD34 in the tumor cells. Nuclear MIB-1 index was very low. (<1%). Figure 5 shows his postoperative chest X-ray. With the diagnosis of lipoblastoma, the patient discharged 7 days after surgery and now after 3 months he is doing well and symptom-free to be followed by physical examination and imaging studies in the next few months.

Discussion

Lipoblastoma is rare tumor of infancy and early childhood.⁴ This term was first described by Jaffe in 1926 as a tumor of immature white fat.⁵ However, the morphologic criteria for the diagnosis of this tumor was first introduced by Enzinger and Chung in 1973.⁶

According to the pathogenesis, a close rela-

Correspondence: Bitá Geramizadeh, Transplant Research Center, Pathology Department, Shiraz University of Medical Sciences, Shiraz, Iran, PO BOX: 71345-1864, Telefax: +98.711.6474331. E-mail: geramib@sums.ac.ir

Key words: Intrathoracic lipoblastoma, infant.

Received for publication: 26 May 2011.

Revision received: 16 July 2011.

Accepted for publication: 2 November 2011.

This work is licensed under a Creative Commons Attribution NonCommercial 3.0 License (CC BY-NC 3.0).

©Copyright B. Geramizadeh et al., 2011
Licensee PAGEPress, Italy
Rare Tumors 2011; 3:e51
doi:10.4081/rt.2011.e51

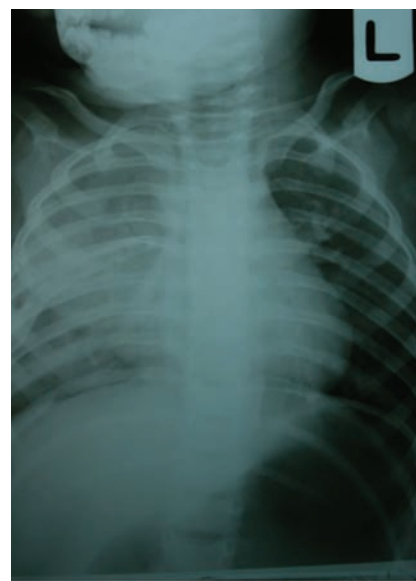


Figure 1. Chest X-ray showed total opacification of right hemithorax.

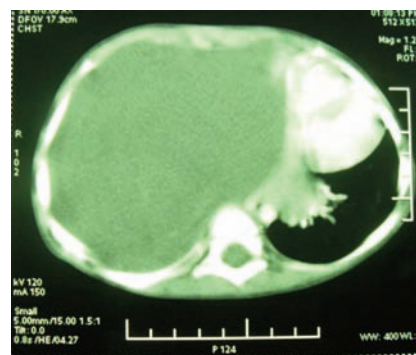


Figure 2. Computed tomography scan of chest shows large hypoattenuated mass in the right hemithorax.

relationship has been considered between lipoblastoma and fetal white adipose tissue.⁷

More than 90% of the reported cases have been in the children and infants less than 3 years of age with a male to female ratio of

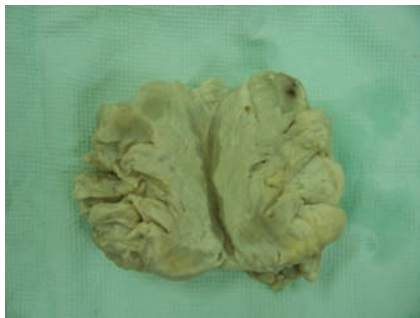


Figure 3. Gross specimen of the mass shows large white mass.

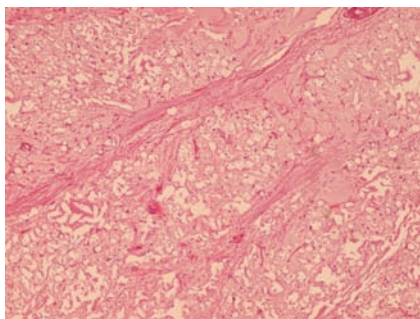


Figure 4. Microscopic section of the mass shows lobulated appearance and myxoid to fatty tissue with bland looking tumor cells and small vessels.



Figure 5. Postoperative chest X-ray.

about 3/1.⁸ Lipoblastoma occurs in the extremities in more than 80% of the patients, most commonly upper extremity such as axilla, shoulder, upper arm, elbow and hand. The second most common location has been reported in the neck and trunk.⁷ Intrathoracic location of lipoblastoma is extremely rare.⁹ Our case was a 15-month-old boy with a huge intrathoracic and mediastinal mass who presented with dyspnea. The diagnosis of lipoblastoma was made after surgical excision and pathologic evaluation. Radiographically by CT scan, lipoblastoma presents as a nonspecific mass with soft tissue density.⁹ It can mimic benign and malignant lipomatous tumors and hemangioma. It means that differentiation between the various adipose tissue tumors cannot be made by CT scan.¹⁰ MRI is the method of choice for preoperative diagnosis of lipoblastoma with a characteristic intermediate to high signal intensity on T1-weighted images according to the amount of immature fat.¹¹ In our case CT scan showed a large intrathoracic mass, but unfortunately MRI has not been done, so we didn't have preoperative diagnosis. In all of the previously reported cases of lipoblastoma, operative surgery and complete tumor excision has been performed with a clear plane of dissection from surrounding tissues.^{3,4,8} This was also true in our case, in which the tumor was easily detached from surrounding pericardium and pleura. The reported tumor size has been between 2 to 21 cm,¹² so our case was considered huge with a greatest diameter of 20 cm which has caused compression symptoms on heart and lung. Histologically, lipoblastoma shows a typical and characteristic morphology with lobular pattern and variable amount of myxoid stroma and white fat.¹ It is somehow similar to myxoid liposarcoma, but it differs by the absence of pleomorphism, atypia and hyperchromasia.³

Lipoblastoma is a rapidly growing tumor; the treatment of choice is surgical excision. After complete excision the prognosis is excellent with recurrence rate of less than 25%.¹² Our case was cured by complete excision to be followed by imaging for the possibility of recurrence. However no metastasis have been reported in this tumor.¹³ As a result, lipoblastoma should be considered in the differential diagnosis of rapidly growing soft fatty masses of children within the thorax and mediastinum.

References

1. Coffin CM, Lowichik A, Putnam AP. A clinicopathologic and Immunohistochemical analysis of 59 cases. *Am J Surg Pathol* 2009;33:17-5-12.
2. Pham NS, Poirer B, Fuller SC, et al. Pediatric lipoblastoma in the head and neck: A systematic review of 48 reported cases. *Int J of Pediatr Otorhinolaryngol* 2010;74:723-8.
3. Gulhan SS, Adams PY, Sarica EA, et al. Chest wall lipoblastoma in a seven-months-old girl: a case report. *J Pediatr Surg* 2004;39:1414-17.
4. Moaath A, Raed E, Mohammad R, Mohammad S. Lipoblastoma: a rare mediastinal tumor. *Ann Thoracic Surg* 2009;88:1695-7.
5. Jaffe RH. Recurrent lipomatous tumours of the groin: liposarcoma and lipoma pseudomyxomatodes. *Arch Pathol* 1926;1:381-7.
6. Chung EB, Enzinger FM. Benign lipoblastomatosis. An analysis of 35 cases. *Cancer* 1973;32:482-92.
7. Torre M, Borel C, Saitua F, et al. Lipoblastoma with unique localization requiring tracheal and esophageal resection. *J Pediatr Surg* 2010;45:E21-3.
8. Spinelli C, Costanzo S, Severi E, et al. A thoracic wall lipoblastoma in a 3-month-old infant. A case report and review of the literature. *J Pediatric Hematol Oncol* 2006;28:594-600.
9. Ko SF, Shieh CS, Shih TY, et al. Mediastinal lipoblastoma with intraspinal extension: MRI demonstration. *Magn Reson Imaging* 1998;16:445-8.
10. Chen CW, Chang WC, Lee HS, et al. MRI features of lipoblastoma: Differentiating from other palpable lipomatous tumor in pediatric patients. *Clin Imaging* 2010;34:453-7.
11. Castellote A, Vazquez E, Vera J, et al. *Radiographics* 1999;19:583-600.
12. Kanu A, Oermann CM, Malicki D, et al. Pulmonary lipoblastoma in an 18-month-old child: A Unique tumor in children. *Pediatr Pulmonology* 2002;34:150-4.
13. Stringel G, Shandling B, Mancner K. Lipoblastoma in infants and children. *J Pediatr Surg* 1982;17:277-80.