ISSN 2073-9990 East Cent. Afr. J. surg

Mediastinal Lipoblastoma: A Case Report and Review of literature

A. Tadesse[1], K. Philipos[2]
[1]Consultant pediatrics surgeon and assistant professor of surgery, Department of Surgery, School of Medicine, Addis Ababa University
[2]Assistant professor of surgery, Department of Surgery, School of Medicine, Addis Ababa University
Correspondence to: Amezene Tadesse, Email: amezenet@yahoo.com

Lipoblastomas are rare benign tumors that occurs in infancy and early childhood, accounting to less than 1% of all childhood neoplasms. These neoplasms have excellent prognosis for full recovery, they don't metastasize and are non-aggressive. They arise from the fetal-embryonal fat, lipoblasts[1]. Lipoblastomas occur most frequently in male children. About 70% of them arise in the limbs but they are also known to occur in the trunk (usually retroperitoneum), head or neck. Primary tumors located in the mediastinum and chest wall are rare[3] with 20 cases of Mediastinal lipoblastoma reported in English literature to date. We report a rare case of Mediastinal lipoblastoma in a 15 months old male child with the operative findings and subsequent outcome.

Case presentation

A 15 month old male child presented to our hospital with shortness of breath and recurrent cough of 5 months duration. He had no history of fever, weight loss or poor intake, no failure to thrive. Otherwise past medical history was unremarkable, no family history of malignancy. Pertinent physical finding was stridor with otherwise normal chest findings. PA and Lateral chest radiographs revealed a large posterior mediastinal well defined soft tissue mass displacing the trachea and carina anteriorly (Figure 1).

Chest CT scan revealed a well-defined hypodense mass in the posterior mediastinum compressing the trachea and extending into the middle mediastinum and the possibility of lipoma or a fatty lesion was entertained (Fig. 2). The child was subsequently operated with right posterolateral thoracotomy with intraoperative finding of a right sided posterior mediastinal fatty soft tissue mass which was lobulated and with cystic(mucoid containing) parts. The mass was sent for histopathological examination and was found to be consistent with lipoblastoma. Subsequently the child recovered fully and discharged improved. On subsequent follow up, the child continues to thrive normally with no chest symptoms and control chest X-rays revealed normal finding (Figure 3).

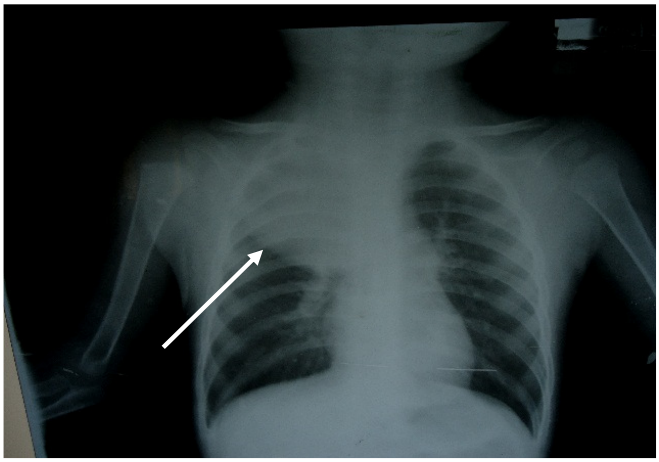

Figure 1. PA Chest X-ray Showing a Soft Tissue Mass Lesion in the Mediastinum (arrow)

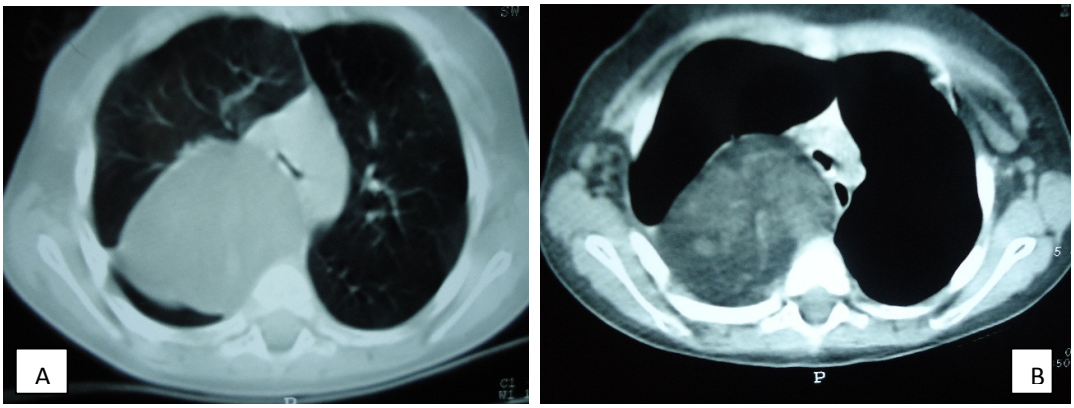

Figure 2. (A and B) Axial Chest CT scans revealed hypodense soft tissue density mass in the posterior mediastinum compressing the carina.

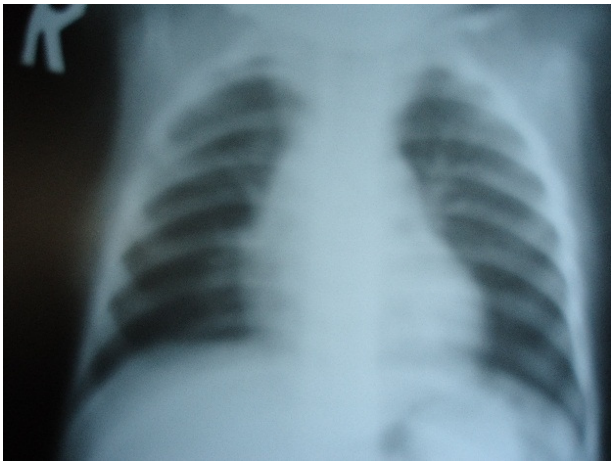

Figure 3. Follow up PA Chest X-ray, Normal Findings

Histopathological examination revealed well capsulated tissue containing regular lobules of fat cells which are uniform sized and have eccentric nucleus. There are large areas of myxoid degeneration. No necrosis or atypia seen.

Discussion

The term "lipoblastoma" was first used by Jaffe[6] in 1926 then confirmed by Vellios[2] in 1958 and has seen been used to indicate a rare tumor of infancy and early childhood arising from embryonal adipose tissue. Since then, approximately 200 cases have been reported in the English literature[7,8].

Vellios et al described in 1958 2 forms: a localized well-circumscribed lesion (lipoblastoma) that arises from the superficial fat tissue, or an uncapsulated, diffuse type (lipoblastomatosis) that grows from deeply situated adipose cells[2]. Nearly 90% are detected in children younger than 3 years[4] and 70% of them occur in the extremities[5], although several other sites have been described and occurrence in the trunk is very rare. The male: female sex ratio is 3:1. And the age of our male patient falls within this high risk group.

Definitive preoperative diagnosis is very difficult and most cases are preoperatively considered to be lipomas as is true with our case where the possibility of a mediastinal fatty lesion was entertained. Clinically, it is impossible to separate lipoblastomas from tumors of other histology

like lipomas and liposarcomas (especially the myxoid variant). This benign tumor shows more uniform growth with distinct lobulations, similar to the one found intra-operatively.
Histopathology and immunohistochemistry remains to be the gold standard in differentiating lipoblastoma from liposarcoma. Myxoid morphology in lipoblastoma is extremely uncommon [16, 17] and in such cases where histopathology is inconclusive, genetic rearrangement of the PLAG1 (pleomorphic adenoma gene 1) oncogene on chromosome 8q12 confirms the diagnosis of lipoblastoma. PLAG1 gene rearrangement has been detected in 70 % of lipoblastoma cases[18]. PLAG1 gene function is mainly in mitogenesis, proliferation, apoptosis and IGF-2 upregulation and is mainly expressed in humans in fetal tissues and in low levels postnatally[18,19]. Unfortunately our pathology unit lacks the facility for such genetic tests and we relied heavily on gross and microscopic findings consistent with lipoblastoma.

Lipoblastomas are typically painless and rapidly growing. The mass is easily detectable when it involves superficial tissues or whenever deep-seated tumors displace or compress surrounding organs. As in our case, the huge mediastinal mass compressed the carina with subsequent development of stridor and dyspnea. Symptoms vary depending on the site of the primary tumor. In the mediastinum, findings may include recurrent chest infection, cough, dyspnoea, or hypoxia[4] and even severe respiratory distress and pleural effusion (if ruptured)[10,11].

On PA chest x-ray, lipoblastoma appears as a non-specific soft tissue density mass [10] and on CT scanning, a fat containing soft tissue mass with septations and post contrast non-enhancement will be its characteristic features [12]. As previously mentioned the most important differential diagnoses are lipomas and liposarcomas where both may have similar clinical and radiologic features but in the case of liposarcoma age is most important factor as it rarely presents below the age of 10 years with peak incidence in the third decade of life and when it presents, especially the myxoid variant, it is almost impossible to differentiate histologically from lipoblastoma[13].

The preferred mode of treatment is surgical excision. Intraoperatively, a well localized mass of adipose tissue is found and can easily be excised completely most of the time and aggressive operative procedure is not advised, especially in infants and small children[14]. Same was the operative finding in our case where complete and safe excision was possible with the resultant excellent outcome. Mognato et al[15] even questioned whether surgery was necessary for such tumors as there are possibilities for spontaneous resolution including in those cases incompletely removed and even their evolution into mature lipomas but such scenarios needs further study and the obvious need for surgical excision together with the safe and complete excision in our case weighs greatly than the complications and risks associated with the procedure.

References

1. B-Kchaou I, Boccon-Gibodb L, Ducou Le Pointe H. Chest wall lipoblastoma in a 11-months-old girl. Arch Pediatr 2007; 14:915-6.
2. Vellios F, Baez J, ShumackerHb. Lipoblastomatosis: a tumor of fetal fat different from hibernoma; report of a case, with observations on the embryogenesis of human adipose tissue. Am J Pathol 1958:34(6):1149-59.http://www.ncbi.nlm.nib.gov/pubmed/13583102
3. Tahbaz O, BakhshayeshKaram M, Zahirifard S, et al. Mediastinallipoblastoma: report of a case. Iran J Radiol 2005;2(3-4):117-20.
4. Whyte AM, Powell N (1990) Case report: mediastinallipoblastoma in infancy. ClinRadiol 42:205

ISSN 2073-9990 East Cent. Afr. J. surg

5. Mahour GH, Bryan BJ, Isaacs H (1988) Lipoblastoma and lipoblastomatosis. A report of six cases. Surgery 104:577
6. Jaffe RH (1926) Recurrent lipomatous tumors of the groin: liposarcoma and lipoma pseudomyxomatodes. Arch Pathol 1:381-387
7. Abel RM, Bruan RT, Rafaat F, Haigh F, Sethia B, Parikh D (2003) Axillary Lipoblastoma: tumor recurrence in the right atrium. J PediatrSurg 38: 1246-1247
8. Mcvay MR, Keller JE, Wagner CW, Jackson RJ, Smith SD (2006) Surgical management of lipoblastoma. J Pediatr Surg 41: 1067-1071
9. Castellote A, Vazquez E, Vera J,Piqueras J, Lucaya J, Garcia-Pena P et al. Cervicothoracic lesions in infants and children. Radiographics.1999; 19(3); 583-600.
10. Cowling MG, Holmes SJ, Adam EJ. Benign chest wall lipoblastoma of infancy producing underlying bone enlargement. PediatrRadiol. 1995; 25(1):54-55.
11. Dishop MK, O`Connor WN, Abraham S, Cottrill CM. Primary cardiac lipoblastoma. PediatrDevPathol. 2001; 4(3):276-280.
12. Blak WC, Burke JW, Feldman PS, Johnson CM, Swanson S, CT appearance of cervical lipoblastoma. J Comput Assist Tomogr 1986; 10;696-698.
13. Mentzel T, Calonje E, Fletcher CD, Lipoblastoma and Lipoblastomatosis: a clinicopathological study of 14 cases. Histopathology.1993; 23(6):527-533.
14. A. Kucera, J. Snajdauf, M. Vyhnanek, J. Moravek, R. Kodet, E. Stejkalova, M. Dvorakova, Lipoblastoma in children: an analysis of 5 cases. ActaChirBelg, 2008, 108, 580-582.
15. Mognato G, Cecchetto G, Carli M, is surgical treatment of lipoblastoma always necessary? J Pediatr Surg. 2000. 35:1511-13.
16. Coffin CM, Lowichik A, Putnam A, Lipoblastoma- A clinicopathologic and immunohistochemical analysis of 59 cases. Am J SurgPathol. 2009; 33:705-12.
17. Farrugia MK, Fearane C, Benign lipoblastoma arising arising in the neck. PediatrSurg Int. 1998; 13:213-1.
18. Gisselsson D, Hibbard MK, Dal Cin P, Sciot R, His BL, KozakewichHp, et al PLAG1 gene alterations in Lipoblastoma. Am J Pathol. 2001; 159:955-62.