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CLINICAL CASE

Mediastinal lipoblastoma in paediatrics

Lipoblastoma mediastínico en pediatría

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What do we know about the subject matter of this study?

The lipoblastoma is a benign neoplasm of embryonic white adipose tissue and is almost exclusive to the pediatric age. It is infrequent to find it in the mediastinal region.

What does this study contribute to what is already known?

It reports more cases of a little-known pathology along with a brief review of the study and management of this entity.

Abstract

Objective: To describe two cases of mediastinal lipoblastoma, an infrequent and little-known pathology, which is extremely rare in the mediastinum, with no cases reported in our country. **Clinical Cases:** Two case reports. Both patients were boys younger than three years, in which a mediastinal mass was found incidentally on a chest x-ray. The study was complemented with a CT scan and with a thoracoscopic biopsy in one of the cases. Complete resection of the tumor was achieved in both patients through thoracotomy. One of the patients presented Claude Bernard Horner syndrome as a complication from surgery, which resolved spontaneously after two years and the second case had no complications. **Conclusion:** Mediastinal lipoblastoma is a very rare pathology. Descriptions found in existing literature are similar to the cases presented in this article. We can conclude that mediastinal lipoblastomas in pediatrics present a very similar pattern and presentation, having a good prognosis if complete resection is achieved. Also, it is essential to distinguish it from its differential diagnoses in order to rule out malignancy.

Keywords:
Lipoblastoma;
children;
mediastinum;
neoplasms

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Introduction

Lipoblastoma is a benign neoplasm of embryonic white adipose tissue, first described by Vellois et al in 1958. To date, there are only 14 published cases of mediastinal lipoblastoma in the databases PubMed, Scielo, and Lilacs (searched using the terms "Lipoblastoma" and "Mediastinum").

These neoplasms represent 6% of all adipose tissue tumors, which are classified as lipomas (64-90%), lipoblastomas (5-30%), liposarcomas (1-4%), and hibernomas (2%)[1]. They appear almost exclusively in pediatric age, 90% are diagnosed before the age of 3, and with a male:female ratio of 3:1[2-4]. The most frequent location of lipoblastoma is the trunk (48%), followed by limbs, head and neck, retroperitoneum, and mediastinum[1].

These tumors are characterized in the pathological anatomy for being a well-circumscribed lesion presenting a fibromembranous pseudocapsule with abundant myxoid matrix, composed of immature mesenchymal tissue with adipocytes in different stages of maturation. They are lobulated with internal septa and plexiform capillary network. Differing from other tumors by not having cellular atypia[5,6]. They are fast-growing tumors and, in most cases, do not infiltrate adjacent tissues, metastasize or present malignant degeneration.

The clinical picture may present respiratory symptoms or may be asymptomatic, depending on the size and location of the tumor[7]. The definitive diagnosis will be based on the anatomopathological study, however, we can suspect initially with a thorough anamnesis, physical examination, and imaging studies. It is essential the complete excision of the lesion, avoiding common complications of surgery such as bleeding, infections, injuries to nerve structures, among others. Despite it is a benign neoplasm, it can be locally invasive presenting a risk of recurrence if not completely excised[8].

It is important to discriminate lipoblastomatosis from a lipoblastoma due to its multifocal presentation and high morbidity when infiltrating adjacent tissues. Regarding mediastinal cases, they present infiltration into the epidural and intraspinal space, phrenic nerve, brachiocephalic vein, heart, and lungs. In other areas, they can also infiltrate soft tissues and muscle planes. Sometimes, the margin between the tumor and adjacent tissues may be unclear, therefore, it is essential to clearly determine the resection area before surgery, for proper surgical planning and thus avoid further complications without damaging adjacent structures[1].

The objective of this report is to describe a little know pathology that has a very low incidence and is even less frequent when located in the mediastinum, of which there are no described reports in our country. We report two clinical cases of mediastinal lipoblastoma in pediatric patients, including its presentation, management, and prognosis.

Clinical Cases

Case 1

A 2-year-old male patient, with no history of morbidity, weight loss or chronic respiratory symptoms, presented with symptoms of acute respiratory infection at the emergency department. A chest X-ray (figure 1) showed a 6-cm-diameter lesion on the right lung apex. The study was complemented with a CT scan which showed a 5cm septated lesion of adipose tissue in the right mediastinum. With a density lower than 0 Hounsfield Units (HU) defined as mediastinal lipoblastoma (figure 2).

Dissection of the lesion was performed through thoracoscopy before resection in the eighth right intercostal space via thoracotomy. The sample was sent for a deferred biopsy which was reported as a lipoblastoma. The patient developed transitory Horner Syndrome, which disappeared after 2 years of follow-up, without other complications.

Case 2

A 1-year-old male patient, with no history of morbidity, weight loss or chronic respiratory symptoms, presented with symptoms of acute respiratory infection at the emergency department. A chest X-ray showed a 13-cm-diameter mediastinal mass, which collapsed and displaced the left lung from the apex (figure 3).

The patient developed progressive respiratory insufficiency, thus needing mechanical ventilation. He was diagnosed with pneumonia secondary to respiratory syncytial virus. A CT scan was performed to complete the study, which showed a well defined hypovascular mass of 8 x 9 x 5 cm, with a density lower than 0 UH, in the upper left hemithorax that caused mass effect on the pulmonary parenchyma (figure 4). A thoracoscopic biopsy was performed before surgery which was reported as a low-grade myxoma. Laboratory tests showed alpha-fetoprotein and human chorionic gonadotropin within normal ranges.

In a second stage, the lesion was resected via thoracotomy with section of the sixth rib due to the large size of the mass, achieving a complete resection of it. The sample was sent to a deferred biopsy which reported a lipoblastoma. One month after surgery, the patient showed good response, without complications.

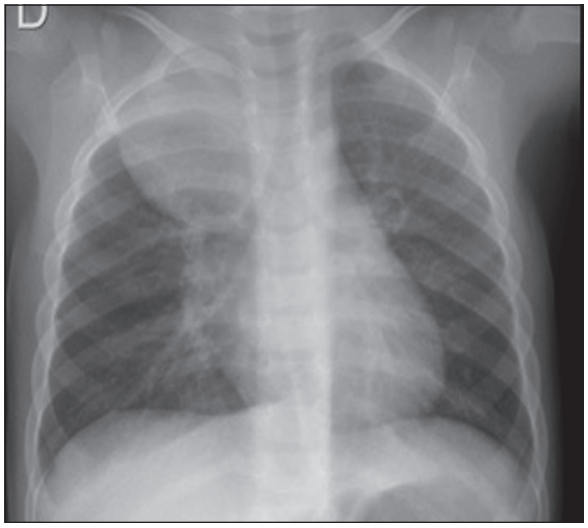

Figure 1. Chest x-ray. A lesion of 6 cm in diameter is observed in the vertex of the right lung.

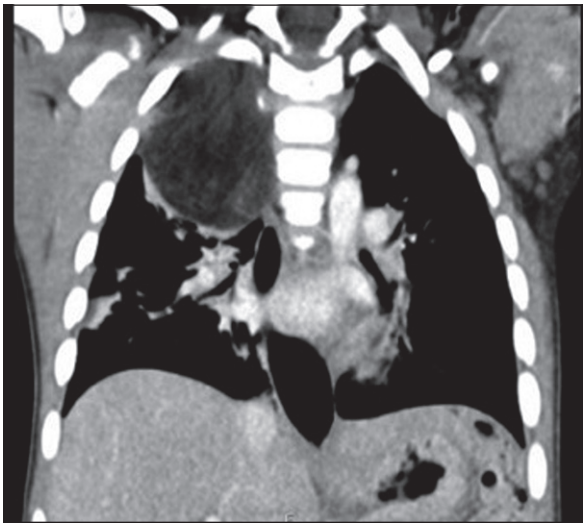

Figure 2. Chest Computed Tomography. A septated lesion of adipose tissue is observed in the right mediastinum of approximately 5 cm, with a density of less than 0 Hounsfield Units, reported as mediastinal lipoblastoma.

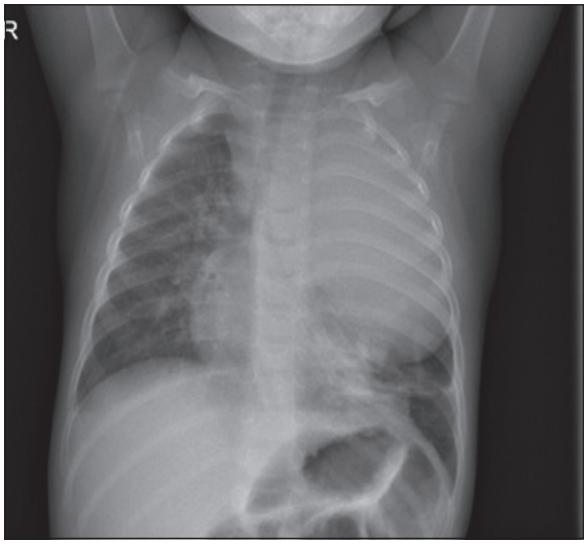

Figure 3. Chest x-ray. A mediastinal mass of approximately 13 cm in diameter is observed. It collapses and displaces the left lung to the contralateral side from the vertex.

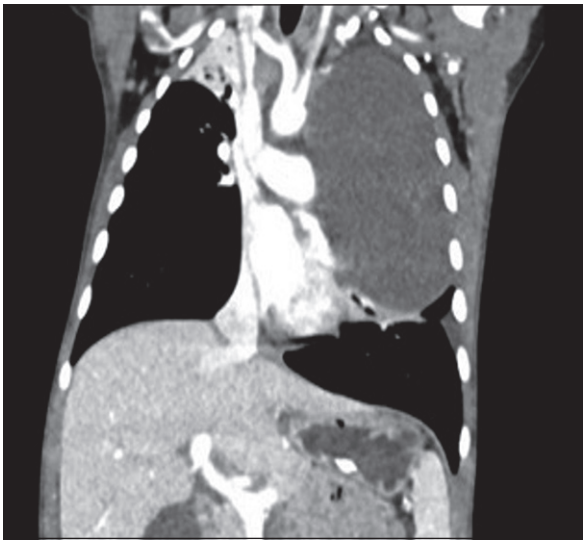

Figure 4. Chest Computed Tomography. A hypovascular mass of 8 x 9 x 5 cm is observed, with a density less than 0 HU, well defined in the left upper hemithorax that exerts a mass effect on the lung parenchyma.

Discussion

In the case of a mediastinal mass, the initial study should start with a thorough anamnesis and physical examination to guide the differential diagnosis. In most cases, lipoblastoma occurs in early childhood and is usually not related to other syndromes[1]. Lipoblastoma is extremely rare in children under the age of 10, unlike other lipomas, such as hibernoma, which occurs most often between the ages of 30 and 40, and liposarcoma, which appears most often over the age of 40[6,9,10].

The study should continue with imaging tests. It is difficult to distinguish on imaging between adipose tissue tumors (lipomas, liposarcomas, and lipoblastomas) since they share some similarities on the CT scan, such as their density and that they do not enhance with contrast material. A useful tool for differentiating lipoblastoma from liposarcoma is the absence of infiltration into adjacent structures, causing only mass effect[11,12].

Regarding the MRI, there is no consensus in the literature on its advantages over CT scans[13]. In our cases, a CT scan was requested to complement the study of the initial chest x-ray finding and the MRI study was not necessary as the CT scan provided the necessary preoperative information.

Ultrasound, although inferior to CT scan, can be used as a study for lipoblastoma. It shows hyperechoic tissue without calcifications which differentiates this tumor from a teratoma[13]. Although it is useful in the diagnosis of lipoblastoma, in our case, it was not helpful due to the mediastinal location.

Finally, needle biopsy does not provide a significant sample thus it is not recommended for the diagnosis of lipoblastoma. In our case, we can confirm this result since the previous thoracoscopic biopsy was not representative of the total lesion, thus it was reported as a low-grade myxoma when the definitive diagnosis was a lipoblastoma.

Regarding the cytogenetic study, this tumor can be associated with chromosomal translocation with deletion of the long arm of (8q 11-13). It has been described that 95% of myxoid liposarcomas have a chromosomal translocation (12;16) (q13;p11) which is also characteristic of this tumor[1]. Therefore, cytogenetic studies would be useful when there are doubts concerning the diagnostis, to differentiate lipoblastoma or lipoblastomatosis from liposarcoma[3]. In our case, the cytogenetic study was not necessary due to the high suspicion of benignity from the medical history, laboratory and imaging studies.

Unlike other tumors, lipoblastomas do not have elevated human chorionic gonadotropin or alpha-fetoprotein, which is a useful tool for differentiating them from their malignant differential diagnoses. In both cases, we used such tool as initial study before surgery, which was useful to recognize the benignity of the lesion. All these options can guide the lipoblastoma diagnosis, however, once the tumor was resected, the biopsy of the whole tumor shows the definitive diagnosis.

The management of lipoblastoma is the complete resection of the lesion, presenting a very good prognosis and low recurrence if the procedure results successfully. The decision of the surgical technique to use will depend on the location, size, and involvement with adjacent organs. The larger the size and the greater the involvement of adjacent organs, the more difficult the resection and has a worse prognosis. If possible, thoracoscopic dissection is recommended to achieve the least invasive surgery possible and then remove the tumor via thoracotomy if necessary. A recurrence of 14-25% has been described thus it requires follow-up, ideally every 6 months for 2 years[14]. This recurrence is mainly attributed to incomplete resection or infiltrating lesions.

Conclusion

Mediastinal lipoblastoma is a very rare entity, being one of the few benign tumors found in the mediastinum at the pediatric age. Its initial study should start with a CT scan but the definitive diagnosis is achieved by the pathological anatomy, where the biopsy of the whole piece is strictly necessary. The management of this tumor is its complete resection.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

Financial Disclosure

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References

1. Hicks J, Dilley A, Patel D, Barrish J, Zhu S, Brandt M. Lipoblastoma and Lipoblastomatosis in infants and childhood: Histopathologic, Ultrasound and Cytogenetic features. Ulrastructural Pathol. 2001;25:321-33.
2. Barros A, Catoni D, Araya J, Cancino M, Correa S, Rostion G. Caso Clínico: Lipoblastoma y Diagnóstico de Tumores de Partes Blandas. Rev. Ped. Elec. [en línea] 2006;3(2):18-22.
3. Hernández F, Nistal, Encinas JL, et al. Lipoblastoma: el menos conocido de los tumores. Cir Pediatr 2004;17:175-8.
4. Ozsen M, Yalcinkaya U, Yazici Z, Sarisozen MB. Lipomatous tumors in pediatric patients: a retropective analysis of 50 cases. Turk patoloji dergisi 2019. [Epub ahead of print].
5. Susam-Sen H, Yalcin B, Kutluk T. Lipoblastoma in children: A review of 12 cases. Pediatr Int. 2017;59(5):545-50.
6. Bruyeer E, Lemmerling M, Vander Poorten V, Sciot R, Hermans R. Paediatric lipoblastoma in the head and neck: three cases and review of literatura. Cancer Imaging. 2012;12:484-7.
7. Al-Mubarak G., Bryant A., Crawford J., et al. Surgical and Anesthetic Management of a Mediastinal Fatty Tumor: Lipoblastoma. Ann Thorac Surg. 2015; 100:e97-8.
8. Canonico F, Parassini M, Malaspina C. Nonmyxoid mediastinal lipoblastoma in a 2-year old girl: Case report with US, CT and MRI findings. Ultrasound Med Biol. 2011;14(1):14-7.
9. Collado L, Sierre S, Bosalec A, Lipsich J. Hibernoma: tumor retroperitoneal de grasa parda. Informe de un caso pediátrico. Arch Argent Pediatr 2011;109(6):e126-e129/e126.
10. Miranda DG, Gómez S, de la Torre D, Pérez A, Góngora J, Salas H. Liposarcoma mixoide en niños: Reporte de un caso. Rev Hosp Jua Mex 2006; 73(3):87-90.
11. Hanafiah M, Noryati M, Arni T. Mediastinal lipoblastoma: unexpected finding of a chest infection. BMJ 2013: bcr2013009879.
12. Kanu A, Oermann C, Malicki D, Wagner M, Langston C. Pulmonary lipoblastoma in a 18 month old child. A unique tumor in children. Pediatr Pulmonol 2002;34:150-4.
13. Geramizadeh B, Javadi F, Foroutan H. Intrathoracic lipoblastoma in a 15-month-old infant. Rare tumors. 2011;21;3(4):e51.
14. Ziegler A, Karplus G, Serour F, Peer M. Huge Mediastinal Lipoblastoma in a Nine Year-old Boy Successfully Removed Surgically. Heart, Lung and Circ 2015; 24(7):e101-3.