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INTERACTIVE CASE

A case of thoracic mass in an infant - What is the diagnosis?

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Pulmonary topography of a male patient, aged 1 year and 10 months, with a history of pneumonia since the first year of life (Figure 1 and 2). In the first episode, he presented pleural effusion and received in-hospital treatment. Tuberculosis screening had been negative. After discharge, he developed a productive cough. Computed tomography without contrast (CT) detected a voluminous subpleural cystic formation, with thick contents and septations inside it, located in the left hemithorax. Treatment was started with amoxicillin-clavulanate, during which the respiratory symptoms worsened. The patient was admitted and transferred to the pediatric UTI of Hospital Federal de Bonsucesso (HFB). A new CT indicated exploratory thoracotomy.

1- The main childhood lipomatous neoplasia is:

A Angiolipoma
B Lipoma
C Liposarcoma
D Hibernoma
E Lipoblastoma

Lipoma is the most common adipocytic tumor in children and adolescents; it corresponds to approximately two thirds of lipomatous neoplasms in the first twenty years of life. Sheybanii EF 2016.

Lipoblastomas are the second most common and are responsible for approximately 30% of adipocytic tumors in childhood. Navarro OM 2011.

Other adipocytic tumors include hibernoma, angiolipoma, diffuse lipomatosis, nerve lipomatosis, and liposarcoma.

2- Concerning Lipoblastomas, it can be said that:

A They are malignant tumors triggered by the proliferation of embryonic adipose cells.
B They are infiltrative tumors affecting deep soft tissues.
C Most lymphoblasts occur in the trunk and limbs.
D They mostly affect adolescents and young adults.
E They are often associated with syndromes.

- A lipoblastoma is a benign neoplasm resulting from the proliferation of embryonic adipocytes. It is most frequent among newborns and young children (usually under 3 years of age). (Kok, 2010)[1].
- It may be superficial and clearly circumscribed, or deep and diffused (lipoblastomatosis).
- The torso and limbs are the parts of the body that are most frequently affected, but retroperitoneal lipoblastoma, mesenteric lipoblastoma, genital lipoblastoma, mediastinal lipoblastoma, cervical lipoblastoma, lipoblastoma of the thoracic wall, lipoblastoma of the parietal pleura, and, very rarely, pulmonary intraparenchymal lipoblastoma have all been described Kanu A, Tracker MM, and Kanu A[2,3].

3- In relation to lipoblastomas, which statement is correct?

A They are tumors of small dimensions without mass effects.
B The diagnosis can be confirmed by computed tomography.

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Figure 1. Chest X-rays. Opacity in all left hemitorax and deviation of mediastinum on the left.

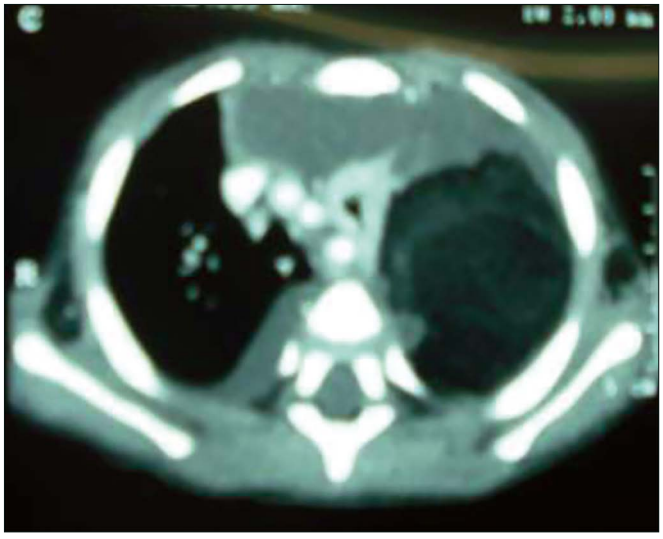


Figure 2. Chest tomography - tumor in the posterior mediastinum.

C Abnormalities on chromosome 11 have been associated with this type of neoplasia.
D They are more frequent in females.
E Although benign, they may invade locally.

- A lipoblastoma may become large in size, as in the case of a girl who had a retroperitoneal tumor weighing 1.9 kg and measuring 25 x 20 x 7 cm, thus causing a mass effect and dislocating the spleen and intestine. In this case, the kidney was rotated and dislocated to the median line. Kok.
- Most of the literature says that these tumors are more frequent in males, but in the studies by Kok and Telisinghe[1] and by Mahour et al.[4], neither sex was dominant, and in the study by Diley et al.[5], incidence was higher in females.
- Although benign, they may invade locally. Recurrence is also possible in 9% - 25% of cases.
- Computed tomography reveals the adipose nature of the tumor, but it does not allow to distinguish between lipoma, liposarcoma, and mixed-type liposarcoma. The diagnosis is histological in nature.
- In cases of lipoblastoma, abnormalities involving the rearrangement of the long arm of chromosome 8 and the PLAG1 gene have been described. In the case of myxoid liposarcoma, translocations occur in chromosomes 12 and 16. A genetic study may help to distinguish between cases in which a histopathological analysis cannot provide a diagnosis. Fallon SC[6].

4- In imaging studies, which of the conditions listed below can mimic primary pleural tumors?

A Extramedullary hematopoiesis
B Round atelectasis
C Extrapleural hematoma
D All of the above conditions
E None of the above conditions

From an imaging study perspective, primary parietal pleural tumors may be mimicked by the following conditions:

- Pleural effusion, loculated hemothorax and empyema
- Diffuse pleural thickening
- Round atelectasis
- Pleural plaques associated with exposure to asbestos
- Thoracic spasm
- Metastatic disease
- Extrapleural hematoma
- Extramedullary erythropoiesis

The patient underwent surgery to dissect the mass occupying all of the left hemithorax, (Figure 3). Histopathological examination revealed mature adipose tissue, compatible with lipoblastoma, and absence of malignancy (Figure 4). He presented excellent clinical evolution, and was discharged on the twentieth postoperative day, with a radiological image of pleural thickening and reduction in left pulmonary volume.

5- Mediastinum masses are causes of anterior, middle and posterior location:

A Parathyroid tumor, esophagus tumors and hiatal hernia.
B lymphoma, fibrosing mediastinitis and bronchogenic cyst.



Figure 3. Macrosopic feature of the cirurgical piece.

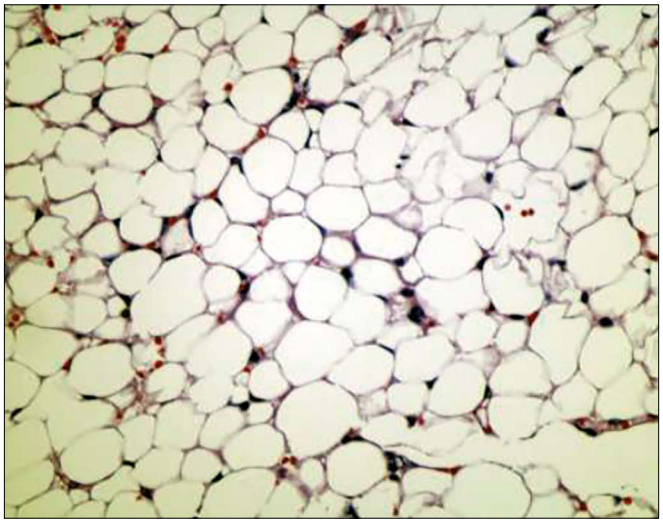


Figure 4. Histopathology revealing mature adipose tissue, compatible with lipoblastoma, and absence of malignancy.

C Pericardial cyst, teratoma, paravertebral abscess.
D Lymphoma, bronchogenic cyst, neurofibroma.
E Neurofibroma, bronchogenic cyst, esophageal duplication cyst.

- Knowledge of the mediastinal compartments is essential for establishing differential diagnoses in the mediastinumis. There are two types of mediastinal division. The first type divides the mediastinum into: superior (located above the fifth thoracic vertebra (T5); approximately at the level of the carina) and inferior (located below the T5).
- The other classification type, which is more commonly used, divides the mediastinum into: anterior (posterior border of the sternum to the posterior border of the heart); middle (posterior border of the heart to the anterior border of the spine); and posterior (starting at the anterior border of the spine).
- The main lesions found in the anterior mediastinum are referred to as "the 3 Ts of the anterior mediastinum:" Teratoma, Thymoma, and Terrible Lymphoma. Others include tumors of the parathyroid or ectopic thyroid, fibrosing mediastinitis, vascular aneurysms, cardiac tumors, Morgagni hernias, pericardial cysts, lipomas, and lymph node metastasis.
- In the middle mediastinum: tumors of the esophagus and megaesophagus, bronchogenic cysts, tracheal tumors, fibrosing mediastinitis, vascular aneurysms, lymphoma, hiatal gastric hernias, and lymph node metastasis.
- The main lesions found in the posterior mediastinum are neural neoplasms and paravertebral abscesses.

6- For the study of thoracic masses and nodules, from the radiological aspect we can correctly state that:

A Nodules are leased lesions, with speculated edges and diameter greater than 3.0 mm.
B Masses are opacities located in the mediastinum or lungs, with well-defined contours, and larger than 5.0 mm.
C Acinar pulmonary nodules are rounded opacities of 5 to 8 mm in diameter and typically multiple.
D Pulmonary nodules are rounded opacities with different border characteristics and diameters of less than 3 cm.
E Rib fractures and soft tissue lesions can simulate thoracic nodules and are called pseudonodules.
F Lung masses are totally solid, with a calcification indicating malignancy.

- A mass is a pulmonary, pleural, or mediastinal lesion observed in a chest x-ray as an opaque area greater than 3 cm in diameter, regardless of its density or the details of its borders. A mass is generally referred to in terms of solid opacity or a partially solid opacity. CT allows to determine size, location, attenuation, and other characteristics more accurately.
- A nodule is defined as any poorly- or well-defined opacity measuring up to 3 cm in diameter.
- Acinar nodules are pulmonary opacities that are approximately 5 to 8 mm in diameter and that presumably represent anatomical acini that became opaque as a result of consolidation. This classification is used only when there are numerous lesions.
- A pseudonodule is a lesion that mimics a pulmonary nodule, such as a rib fracture, a skin lesion, a device on the patient's body surface, anatomical variants, or areas of increased opacity.

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