CASE REPORT

Benign Fibrous Histiocytoma of the Lung: A Case Report

Histiocitoma Fibroso Benigno do Pulmão: Relato de um Caso

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ABSTRACT

A fibrous histiocytoma is a benign tumoral lesion generally localized in the dermis or the superficial subcutaneous tissue. Fibrous histiocytoma of the lung is a very rare tumor, with only a few cases in the literature.

We present a case of a 48-year-old man, asymptomatic, with a solitary pulmonary nodule with a high metabolism in PET/TC. Given the location and suspicious hypermetabolism in PET/TC, the patient underwent a wedge resection, with pathological examination concluding on a primary benign fibrous histiocytoma of the lung.

RESUMO

O histiocitoma fibroso é uma lesão tumoral benigna geralmente localizada na derme ou no tecido subcutâneo superficial. O histiocitoma fibroso do pulmão é um tumor muito raro, existindo poucos casos na literatura.

Apresentamos o caso de um homem de 48 anos, assintomático, com um nódulo pulmonar solitário de metabolismo elevado na PET/TC. Dada a localização e hipermetabolismo suspeito no PET/TC, o doente foi submetido a ressecção em cunha, cujo exame anatomopatológico conclui ser um histiocitoma fibroso benigno primário do pulmão.

Fibrous histiocytoma of the lung can be a difficult diagnosis, with a non-specific clinical history as well as radiologic findings. Hence, histopathological examination remains vital in establishing a definitive diagnosis. Prognosis is generally excellent, as these tumors usually have low recurrence rates after complete excision.

Keywords:
Histiocytoma, Benign Fibrous; Lung Neoplasms

O diagnóstico de histiocitoma fibroso do pulmão pode ser difícil, pois tem história clínica e achados radiológicos inespecíficos. Por conseguinte, o exame histopatológico continua a ser vital para estabelecer um diagnóstico definitivo. O prognóstico é geralmente excelente, uma vez que estes tumores costumam apresentar baixas taxas de recorrência após excisão completa.

Palavras-chave:
Histiocitoma Fibroso Benigno; Neoplasias do Pulmão

INTRODUCTION

Pulmonary nodules are often incidental findings in imaging studies and may pose a diagnostic challenge. Distinguishing benign fibrous histiocytoma from other pulmonary lesions, including malignant tumors, can be challenging based on clinical and radiological features alone.

As such, histopathological examination is crucial for accurate diagnosis. Benign fibrous histiocytoma (BFH) is a benign tumoral lesion consisting of fibroblastic and histiocytic cells, which is accompanied by varying numbers of inflammatory cells.

These tumors are usually localized in the dermis or the superficial subcutaneous tissue.[1] Generally, they present in the form of single, slow-growing nodules in the lower extremities.

There are only a few cases of primary fibrous histiocytoma of the lung described in adults.

BFH of the lung has been reported in a wide age range, but with most cases have been documented in the pediatric age group. There doesn't seem to be a strong predilection for gender, as cases have been reported in both males and females. Imaging studies such as chest X-rays or computed tomography (CT) scans often reveal a solitary pulmonary nodule. Many cases of BFH of the lung are asymptomatic and often discovered incidentally during imaging studies performed for unrelated reasons. The lack of symptoms may contribute to underreporting and underdiagnosis.

CASE REPORT

We report the case of a 48-year-old man, non-smoker, with a personal history of hypertension, hypercholesterolemia and focal epilepsy in the context of mesial temporal sclerosis.

During an investigation for palpitations, the patient performed a cardiac magnetic resonance imaging (MRI) that incidentally detected a pulmonary nodule. The patient denied respiratory symptoms, such as cough, sputum, wheezing, shortness of breath, as well as weight loss.

A thoracic CT scan with contrast was performed in June of 2023, which showed a well-circumcised solid nodule with 2.3 cm located in the anterior segment of the right upper lobe, which contacted the mediastinal pleura.

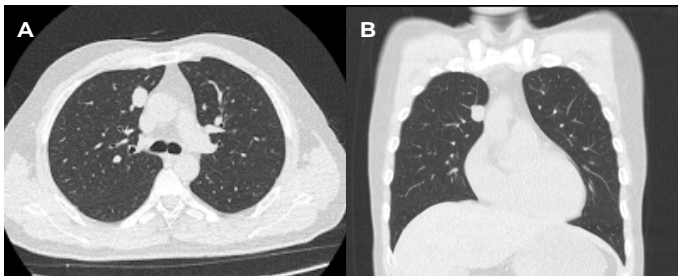

Figure 1 CT-thoracic scans (axial [**A**] and coronal [**B**] views) showing a peri-mediastinal nodular formation in the right upper lobe.

Blood biochemistry tests were normal and a PET/CT scan was performed to assist the solitary pulmonary nodule diagnosis, showing intense FDG involvement (SUV max: 14) localized to the pulmonary nodule.

Pulmonary function tests with plethysmography showed no alterations, with an FEV1 of 109%, DLCO-SB 67% and DLCO-VA 84%.

Given the location of the nodule, with a high metabolism in PET/CT, with no possibility for transthoracic biopsy nor endoscopic

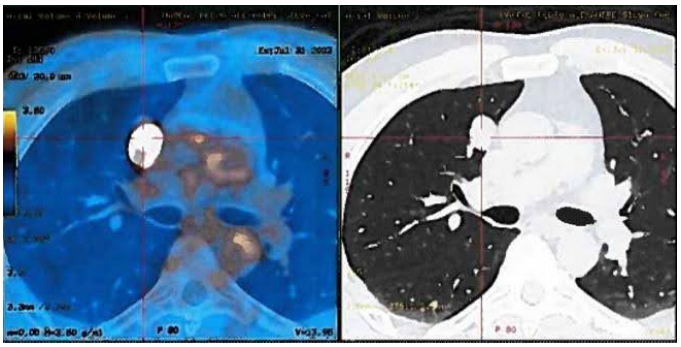

Figure 2 PET/CT showing a hypermetabolic nodular formation in the right upper lobe.

approach, the patient was submitted to an atypical pulmonary resection via video-assisted thoracic surgery (VATS), with no complications.

Microscopically the nodule corresponded to a fibrohistiocitary proliferation with fusiform cells (CD163+, AE1/AE3-, CAM5.2-, DES-, SMA-, CD34-, S100-, HMB45-, EMA- e ALK1-), without atypia, a chronic inflammatory infiltrate and abundant foamy cells. A morphology, as well as an immunophenotypic profile, that was compatible with a benign fibrous histiocytoma of the lung.

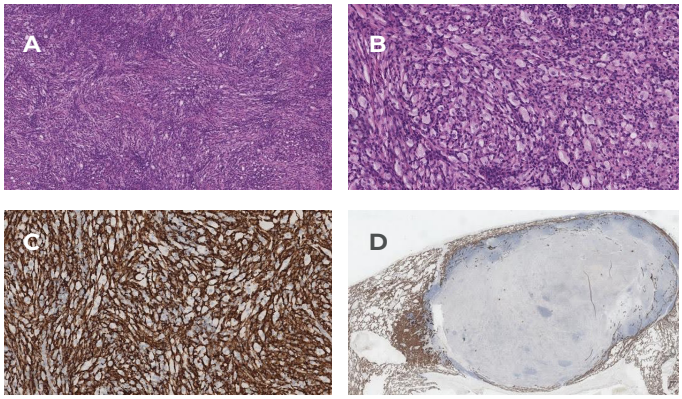

Figure 3 Microscopic features include a fibrohistiocytic proliferation with a swirled pattern of bland spindle cells, a chronic inflammatory infiltrate (**A** – HE X80) with abundant foamy cells (**B** – HE X200). Immunohistochemistry for CD163 (**C** – X200) was diffusely and strongly positive, but negative for cytokeratin AE1/AE3 (**D** – X40).

The patient was later evaluated at a Dermatology consult, which confirmed no skin lesions that could be compatible with cutaneous histiocytomas.

DISCUSSION

Fibrous histiocytoma usually occurs in the soft tissues, tendons and joints of the upper and lower extremities.

Fibrous histiocytoma or fibroxanthoma of the lung consists of an extremely rare tumor in adults. The available literature regarding fibrous histiocytoma of the lung includes, in its majority, case reports or small case series, predominantly from the last century.[2–5] Although they compose one of the most common primary lung tumors in children under the age of 16, they are a rare entity in adults, representing 1% of all surgically resected lung lesions.[6]

The pathologic hallmark of these lesions is a proliferation of spindle cells with accompanying inflammatory infiltrate of plasma cells, lymphocytes and eosinophils. However, it is still unclear whether these lesions represent a primary inflammatory process or a low-grade malignancy with prominent inflammatory response.

Clinical history and radiological diagnosis are often non-specific. This type of lesion is usually noticed during the routine examination of asymptomatic patients.[7] Symptoms that may emerge include cough, hemoptysis, chest pain and fever. The appearance of benign histiocytomas of the lung may not be specific, as radiological features may overlap with other benign or malignant conditions, such as hamartomas, carcinoid tumors as well as multiple infectious diseases. Typically, it presents as a solitary pulmonary nodule, well-circumscribed or lobulated, showing a low homogeneous attenuation in CT. Cystic changes or cavitation were observed in a few cases.[8] In addition, despite being a benign lesion, it can involve activity in the PET-CT, as shown by other cases in the literature.[7] In some cases, only surgery can provide a definite diagnosis.

It is important to note that to obtain an accurate diagnosis, alongside careful and personalized management, it is required a multidisciplinary approach, including pulmonologists, radiologists, thoracic surgeons, as well as pathologists.

Complete surgical resection is the treatment of choice, with complete tumor excision and tumor size ≤ 3 cm being factors associated with decreased risk of recurrence.[8,9]

The natural history of this entity is variable, with lesions typically remaining stable or growing slowly. Long-term follow-up is recommended given the possibility of recurrence and the fact that some lesions become locally invasive.

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RR: First draft, image collection and literature review.
CM: Critical and final review of the work.
IB: Critical and final review of the work.
RC: Image collection critical and final review of the work.
Ri R: Image collection critical and final review of the work.
MTA: First review and critical and final review of the work.
All authors approved the final version to be published.

Declaração de Contribuição:

RR: *Primeiro rascunho, recolha de imagens e revisão da literatura.*
CM: *Revisão crítica e final do trabalho.*
IB: *Revisão crítica e final do trabalho.*
RC: *Recolha Imagens revisão crítica e final do trabalho*
Ri R: *Recolha Imagens revisão crítica e final do trabalho.*
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