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Case report

Diffuse vascular hamartoma with malignant behavior

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ABSTRACT

Pulmonary hamartomas are benign lesions, usually asymptomatic and incidentally discovered on a routine chest radiograph; occasionally, however, this benign lesion may cause life threatening symptoms due to it's location and diffuse vascular involvement. We report the case of a 27 year-old male, non-smoker, who presented with dyspnea, cough, hemoptysis and weight loss. He was found to have a mass in the right hilar region which also involved the right main bronchus, pulmonary artery and esophagus. Surgical biopsy of the lesion led to the diagnosis of diffuse vascular hamartoma. Although it was a benign lesion, due to the size and location, surgical removal was not possible and patient died 10 years after being diagnosed with the condition.

1. Introduction

A hamartoma is a benign tumor characterized by “normal” tissues with a disorganized growth. Pulmonary hamartomas (PH) were first described by Albrecht in 1904 [1]. The lesion is usually characterized by the growth of fat, connective tissue and cartilage tissues within the pulmonary parenchyma (80%) or the endobronchial tree (10–20%) [2]. Parenchymal lesions are usually an incidental finding on a routine chest X-Ray. The endobronchial tumors usually present with new onset of respiratory symptoms, which are most commonly, recurrent chest infections dyspnea or hemoptysis. Pulmonary hamartoma are the most common form of benign lung tumors with an incidence of approximately 0.025% of all lung tumors [2,3]. The lesions usually appear in the periphery of the lung (hilar location is rare). The difference between hamartomas and normal tissue is that hamartomas grow in a disorganized mass, although most hamartomas grow slowly, at a rate similar to normal tissues. The pathogenesis of PH remains unclear. Hamartomas are usually divided into: either chondromatous pulmonary hamartoma or a vascular hamartoma; the former being the most common type. The chondromatous hamartomas are formed by mature lobular cartilage and less frequently adipous and fibromixoid tissues. They may exhibit calcified foci. On the other hand vascular hamartomas represent abnormal growth of vessels mixed with pulmonary parenchyma, similar to abnormal vascular growth in other organs [4].

We present a case of a vascular hamartoma with a malignant behavior, involving vessels and the esophagus.

1.1. Case report

A 27-year-old male, non-smoker presented with hemoptysis, dyspnea, fever and thoracic pain of 8 weeks duration. Associated symptoms included persistent and productive cough (mucoid sputum) and progressive weight loss, which he could not quantify.

On physical examination bilateral crackles involving the lung bases were evident. Chest radiography revealed elevated right hemidiaphragm and right perihilar opacity.

Lung Computed Tomography Scan showed a mass in the right hilar region, with multiple calcifications, about 6 cm in greatest transverse axis, causing extrinsic compression of the right main bronchus, carina and the proximal third of the left main bronchus. It also involved the right pulmonary artery and displaced the left atrium, the esophagus and the proximal end of the superior vena cava. Hilar and subcarinal lymphadenopathy with calcifications was also present There was volume reduction of the right lung due to elevation of the diaphragm (Fig. 1).

Flexible bronchoscopy revealed marked inflammation and blunting of the carina, along with narrowing of both the main bronchi as well as trunchus intermedius (Fig. 2). Endobbronchial biopsy and transbronchial needle aspiration revealed intense

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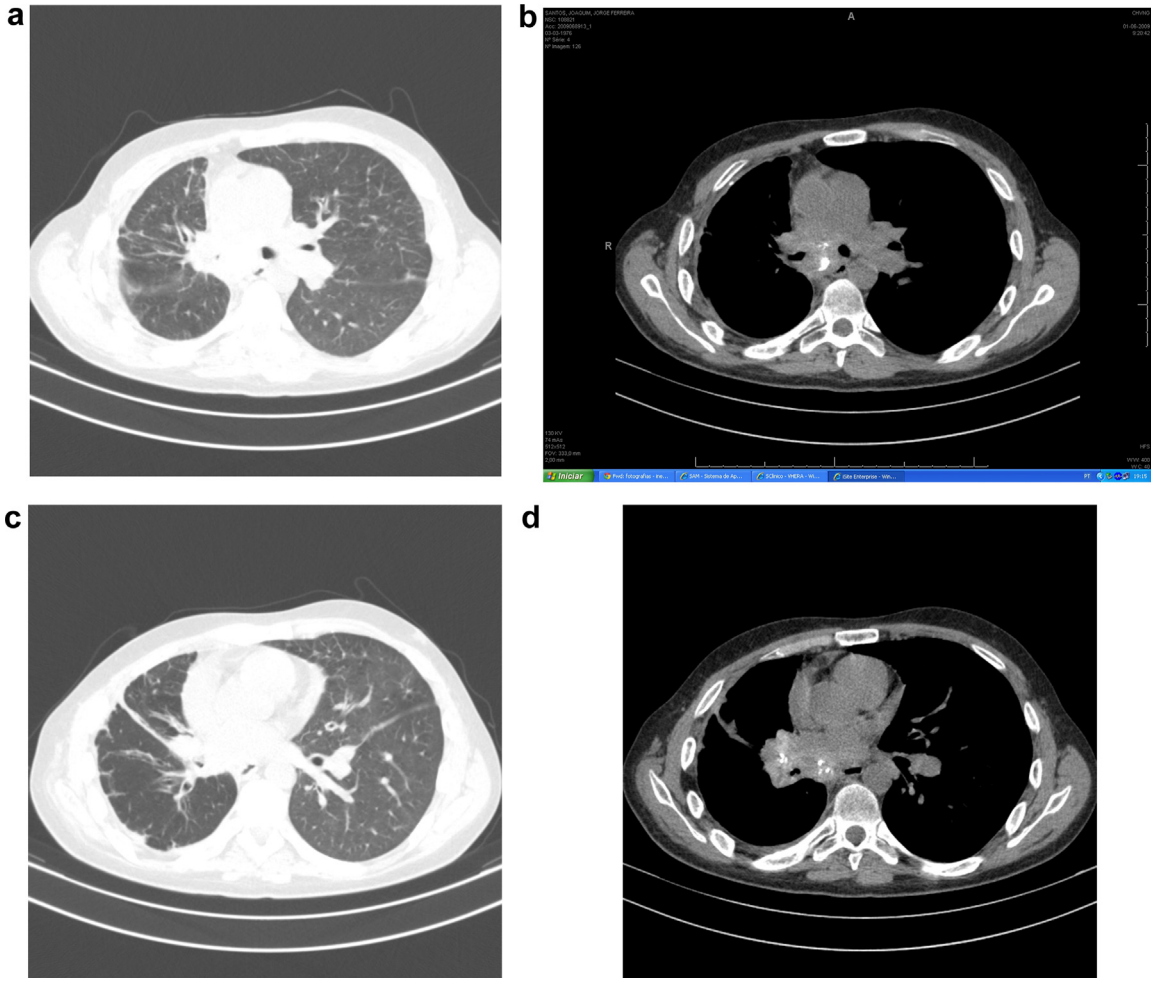

Fig. 1. a,b,c: Chest CT with mediastinal mass on right hilar region with right main bronchus caliber reduction, d: Right main bronchus caliber reduction and right pleural thickening.

chronic inflammatory process. No signs of malignant cells were found in bronchial lavage. Ziehl-Neelson, Giemsa and Gram stain did not reveal any micro-organisms.

The patient was subjected to transthoracic needle aspiration guided by the computed tomography. There were no signs of malignancy.

A right thoracotomy, a lung biopsy and decortication of the pleural process were performed due to the important thickening of pleura. Gross examination of the specimen revealed thickening of the vessel walls and pleura with chronic inflammatory changes. Histological examination of the biopsy revealed - pulmonary parenchyma with large, irregular vessels with thickened heterogenic, irregular walls related to the hamartomatous nature of the disease (Fig. 3a-b) and pleural thickening with chronic inflammatory infiltrate and fibrosis. Diagnosis of vascular hamartoma was established.

Total surgical removal of the lesion was not possible due to the vascular invasion of the pulmonary artery and esophagus. The patient underwent radiation therapy, with stabilization of the lesion size. He had several episodes of hemoptysis, associated with progressive exertional dyspnea and developed respiratory insufficiency. Flexible bronchoscopies were performed trying to achieve control of hemoptyses with argon plasma coagulation on several occasions. Patient refused to undergo heart-lung transplantation.

Ten years after being diagnosed with the condition the patient died of the chronic illness and worsening respiratory insufficiency.

2. Discussion

Pulmonary hamartomas are usually found in adults with a peak incidence in the sixth decade of life with a male preponderance; male: female ratio being 2:1 to 3:1 [5]. These benign lesions are often asymptomatic and are typically discovered as an incidental coin lesion on a routine chest film. Pulmonary hamartomas characteristically appear as well-defined, solitary pulmonary nodules. Radiologically, hamartomas account for 7%–14% of pulmonary coin lesions [6–8]. PH can occur in all parts of lung, but most often, they are found in the periphery and rarely near the hilar regions. Our patient had an atypical radiologic presentation along with histological findings of a mainly vascular component; the latter also being less frequent than chondromatous type [5].

Today, despite the advances in medical therapy pulmonary resection remains the curative treatment for patients with pulmonary hamartoma. Controversy, however, still exists related to the indication and timing of the surgery [5]. Since most pulmonary hamartomas are nonexpanding or slowly growing neoplasms, some authors believe that surgery is necessary only when expansion is recorded in young or middle aged patients or accompanying by

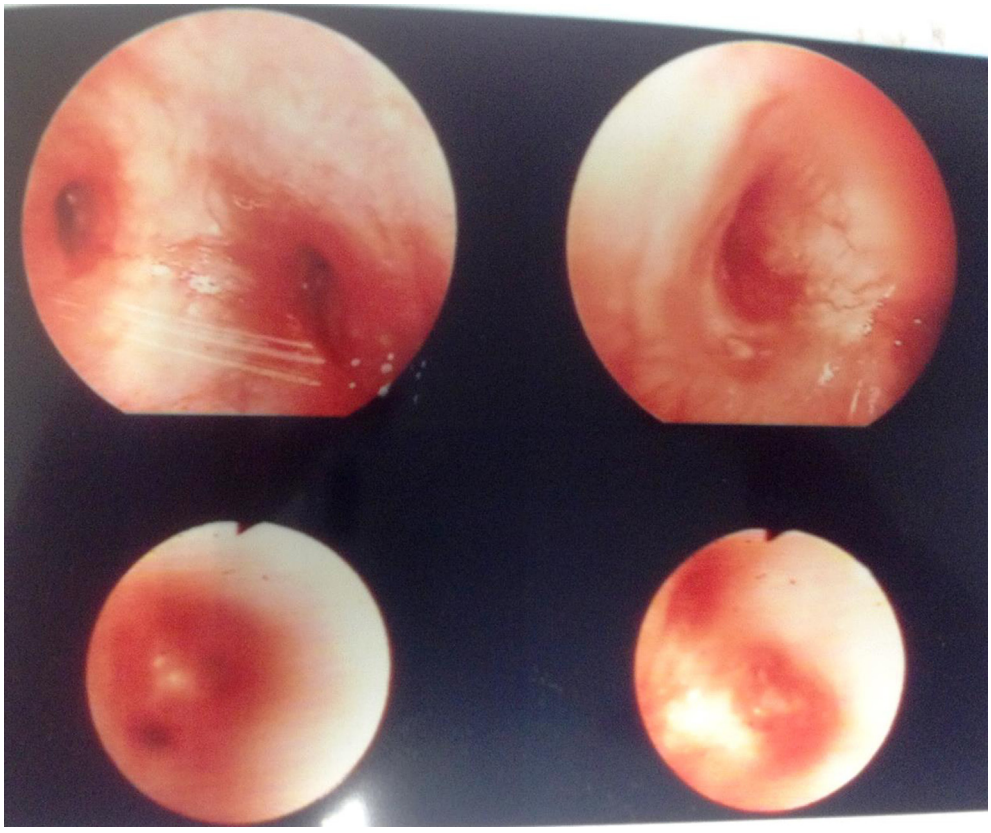

Fig. 2. Blunting of the Carina with extrinsic compression.

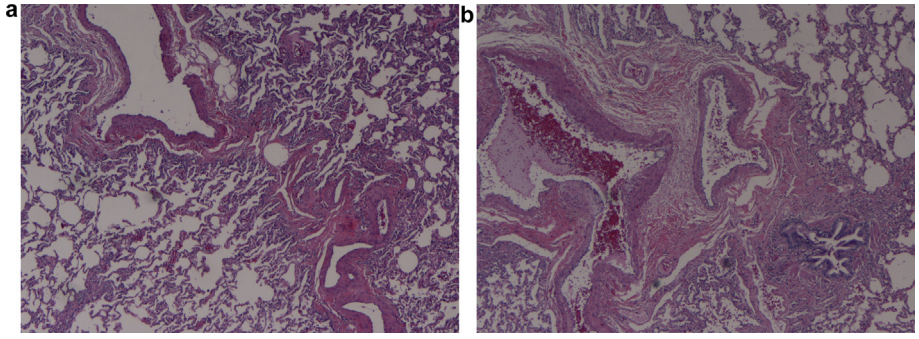

Fig. 3. Pulmonary parenchyma with large vessels with thickened heterogenic walls and irregular form related to the hamartomatous nature of the disease. (a): Pulmonary parenchyma with large vessels. (b): Large vessels with thickened wall.

obvious pulmonary symptoms. In spite of the benign nature of the disease, in this case it behaved as a malignant one, by invading the right pulmonary artery and esophagus, to which surgery and lung transplant were not treatment options [9], making this case a very rare one with no similar prior descriptions in the English literature.

Conflict of interest

We declare no conflicts of interest.

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