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Case Report

Extrapulmonary Hamartoma with Hemothorax: A Case Report

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Abstract

Pulmonary hamartoma is the most common benign tumor of the lung. However, its extrapulmonary manifestation is rare, with only 7 other cases found in the literature. We describe a 72 year-old patient who developed progressive dyspnea with unilateral pleural effusion on the left, as evidenced on chest radiography and computed tomography. Videothoracoscopy was indicated and a hemothorax and an extrapulmonary lesion with 2 pedicled (one ruptured) was found. Pleural effusion was treated and the tumor was resected, with relief of the patient's symptoms and preventing its recurrence. Subsequently, in anatomopathological analysis, it was found to be an hamartoma.

Keywords: Hamartoma; Hemothorax; Videothoracoscopy; Neoplastic; Thoracic Surgery

Introduction

Hamartomas are Mesenchymal tumors and commonly consist of respiratory epithelium mixed with cartilage, glands, fibrous tissue, blood vessels, calcifications and fat in varying concentrations [1,2], the latter being, when present, a pathognomonic landmark of hamartomas [3], also called hamartomas benign mesenchymal, adenochondroma or pulmonary fibroadenoma [1]. Macroscopically hamartomas are a firm white or gray multilobulated mass that easily detaches from the surrounding parenchyma, of cartilaginous consistency with occasional fragments of calcifications or bones [2]. Despite its benign characteristics and in most cases growth of up to 3,2 millimeters per year, it can be confused with malignant nodulations in the images, causing a challenge to differentiate them [4-6]. Despite presenting continuous growth, it usually occurs slowly and typical hamartomas usually double in size after two years of the disease [7] and even has a risk of malignancy, but it is extremely rare [8].

Hamartoma is the most common benign tumor [9,10], equivalent to about 75% of cases [11,12] and about 8% of all lung cancers in general [13]. This disease has a prevalence of 0,3% in the general population [14], with greater involvement in male individuals in a relation of up to 4:1 [1] and from the sixth decade of life [11]. They are typically characterized as single, well-defined nodules less than 4 centimeters in diameter [1,10] and located in the pulmonary parenchyma itself or, less commonly, in the tracheobronchial tree (in up to 20% of cases) [8,10,15], which may be polypoid especially when endobronchial [11]. However, its manifestation outside these places, classified as extrapulmonary, is rare and with few literary descriptions.

Few atypical presentations of hamartoma have been described in the literature, such as the tumor penetrating the visceral pleura adhering to the mediastinal [10,16], located in the posterior mediastinum [17], presenting in the pulmonary hilum [18], with pleural dissemination [6,19] or even developing outside of the visceral pleura [12]. This study aims to describe a case of extrapulmonary hamartoma with an atypical presentation in parietal pleura, manifesting by pleural effusion and dyspnea.

Materials and Methods

We used data available in the patient's medical record after hospital discharge through retrospective analysis with the authorization of a clarification term. The project was approved by the Brazilian Ethics Committee under number 33339620.7.0000.0055.

Case Report and Discussion

Patient JRM, male, 72 years old, with no history of smoking, started to have dyspnea for about 15 days with progressive worsening associated with dependent ventilatory chest pain. He subsequently had unmeasured fever and denied wight loss. Upon admission, physical examination revealed tachypnea (respiratory rate of 25 incursions per minute), dullness to percussion on the basis of left hemithorax with abolished breath sounds in this region and creations in the middle third of the left lung.

This was followed by a diagnostic based on the suspicion of pleural effusion to be clarified, and a chest X-ray was requested, which proved fluid in the left hemithorax (Figure 1). At this time, the laboratory showed leukocytes of 10,200 with 8% rods, PCR of 187,6 and hyponatremia (Na = 128). The next step was a computed tomography scan of the chest, which showed pulmonary consolidation in the region of the left lower lobar bronchus and confirmed the left pleural effusion, therefore, bronchoscopy was indicated. Bronchoscopy showed only clear mucous secretion in a bilateral bronchial tree.

Thus, videothoracoscopy on the left was indicated, with the finding of a massive frankly hematic pleural effusion (hemothorax) and an oval lesion measuring 6,5 x 5,2 centimeters with 1,0 centimeters of average thickness in the parietal pleura, dark, with two vascular pedicles, one of then is ruptured (Figure 1), being resected (Figure 2) and sent for anatomopathological analysis, verifying that it is a hamartoma (Figure 3).

Figure 1: Hamartoma located in the parietal pleura with hemothorax caused by the breached vascular pedicle in videothoracoscopy.

Figure 2: Hamartoma after surgery on surgical table.

Figure 3: Absence of atypia or malignity, presence of vascular component with extensive hemorrhage and presence of cystic areas, mature cartilage, with ossification and mature adiposus tissue compatible with pleuro-pulmonary hamartoma.

The etiology of hamartomas is uncertain, but it is accepted that some chronic inflammatory agents may be one of the possible factors involved in aetiogenesis [20] associated with some abnormalities found in karyotypes that revealed chromosomal changes in 6p21 and 14q24 [14].

The suspicion of hamartoma tends to occasionally occur mainly through imaging tests performed in search of some other pathology, since in most patients they are located in the pulmonary periphery and are therefore asymptomatic [2,6,10,16]. However, some cases may have different symptoms, especially in atypical locations. Endobronchial hamartomas can cause some degree of airway obstruction in which they are located and cause obstructive symptoms such as chronic cough [11,15].

In the present case, the patient had atypical location besides also atypical symptoms and his diagnosis was not easily predictable and was not found in the mains suspicions of differential diagnoses. The diagnosis was made by cytological study since histopathological and ultrastructural studies are not of high value [2]. Treatment is usually conservative with serial imaging exams, except in the impossibility of excluding the malignancy, a situation in which a surgical approach is indicated. After surgery, sarcomatous transformation or disease recurrence is extremely rare [2,16].

Some other few cases had atypical presentation sites and have their particularities [6,10,12,16-19]. In the patient described, we excluded the possibility of involvement of the tracheobronchial tree by means of flexible bronchoscopy before proceeding with surgical therapy. This approach was necessary because the patient was symptomatic due to the tumor.

Conclusion

The case described, in addition to having extremely rare neoplastic location, presented a very atypical clinical picture that differs from the vast majority of other cases and is important enough to indicate surgical resection and improve the patient's quality of life. In addition, as it is a non-specific condition, it was only possible to complete the diagnosis through the surgery itself with referral of the piece for anatomopathological study.

Conflict of Interest

We declare that there is no conflict of interest in publishing the research.

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