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Case Report

Calcifying Fibrous Tumor within Lung Parenchyma: Case Report

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Abstract

Calcifying fibrous tumor is a rare benign neoplasm, with few cases reported within lung parenchyma. Due to its rarity and lack of specification on radiological exams, it is important to distinguish it from others thoracic tumors. This case reported is about a 67-year-old woman with an incidental finding on chest tomography of a well-defined mass within lung parenchyma. The patient was submitted to surgery and the diagnosis of a calcifying fibrous tumor was established by histology.

Keywords: Calcifying fibrous tumor; Pulmonary; Lung parenchyma

Background

Calcifying Fibrous Tumor (CFT) is defined as a benign tumor first described in soft tissue in 1988 by Rosenthal and Abdul-Karim as Childhood fibrous tumor with psammoma bodies [1,2]. It is frequently localized in the gastrointestinal tract but it can also be observed in different sites and rarely in lung parenchyma. It has a wide age range, varying from weeks to 84 years, with a female predominance [3]. Considering that CFT is a rare tumor, this report presents a case with aim to discuss the possible differential diagnosis.

Case Presentation

Sixty-seven years old, woman who had been submitted to a chest tomography due to an episode of cough and fever was treated with antibiotics and resolution of the symptoms. In tomography, it was observed a heterogeneous mass measuring 4.1 × 3.0 cm in the middle region of the right hemithorax within lung parenchyma and close to the pleural surface. The lesion was well defined and had multiple calcifications inside (Figure 1). The patient was submitted to a wedge resection and the specimen was sent to pathological analysis. Grossly the tumor was well circumscribed but without a capsule, and composed by a white firm mass with calcification. Microscopic examination revealed a hypocellular fibrous tumor with hyaline collagen. There were numerous psammoma bodies and dystrophic calcification, as well as mild lymphoplasmacytic chronic inflammatory infiltrate (Figure 2). Immunohistochemical staining was negative for STAT6, BcL2, ALK, β-catenin and focal positivity for CD34.

Discussion

Usually, the lesions in CFT are solitary, as observed in this case report; however, it can also be multiple. The most common location is in the gastrointestinal tract, followed by pleura, neck, mesentery, mediastinium, peritoneum, omentum, soft tissues and adrenal gland. Rare locations have been reported such as lung, pericardium, spleen, liver, breast, gallbladder, spermatic cord, tongue and fallopian tube [3]. The symptoms are variable because the localization of the tumor is diverse. CTF of the thorax patients can be asymptomatic, as an incidental finding, present with chest pain, or dry cough [1]. The radiological appearance is of multiple or single well-defined lesion with areas of increased attenuation attributed to calcification [1], as noted in this case reported. In the largest literature review, the most common way to diagnose and exclude other conditions has been through tissue specimen [3]. Macroscopically, the tumor is well circumscribed, unencapsulated with a solid and firm texture. Histologically, the tumor is hypocellular without atypia and composed by dense hyaline collagen, mild mononuclear inflammatory infiltrate and dystrophic calcification and/or psammoma bodies. The morphological characteristics are sufficient for diagnosis but ancillary studies can be used to exclude differential diagnosis. By immunohistochemistry, the neoplastic cells may be CD34, vimentin and factor XIIIa positive. To differentiate from a solitary fibrous tumor the CFT cells are negative for BCL2, STAT6, CD99. On the other hand, there is not expression of β-catenin in CFT, which are usually positive in desmoid fibromatosis and inflammatory myofibroblastic tumor [1]. In addition, these

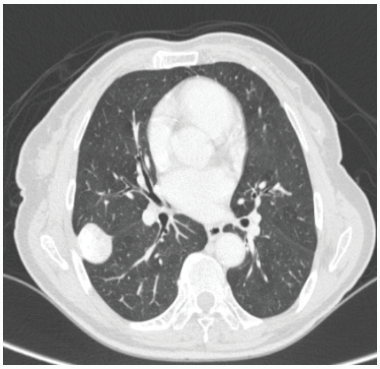

Figure 1: Computed tomographic scans reveal a well-defined mass in the lung parenchyma.

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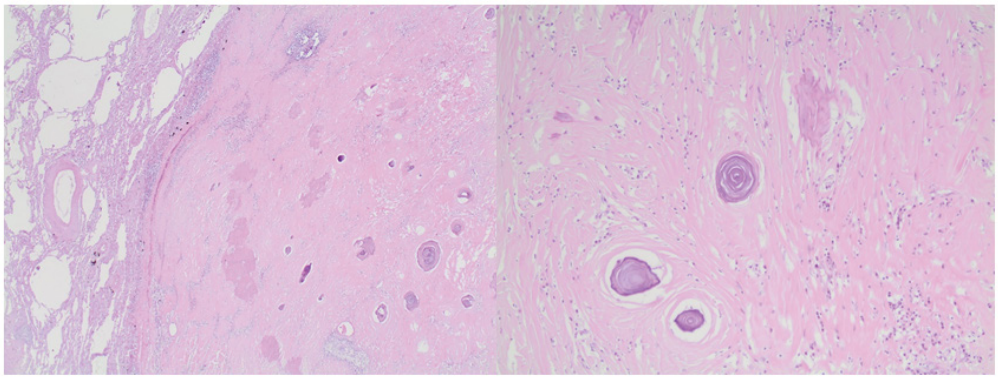

Figure 2: Well defined tumor, unencapsulated in the lung parenchyma, composed by hyaline dense fibrous tissue with frequent psammoma bodies and calcification, with mild lymphoplasmacytic cells. A) HE 20x and B) HE 100x.

tumors are more cellular than CFT and lacks exuberant calcifications. The etiology remains unknown but some authors concern it originated as trauma response [3]. Deleterious mutations in ZN717, FRG1 and CDC27 and loss of 6p22.2 has also been reported [1]. Furthermore, some authors have hypothesized that these lesions are an IgG4-related disease, given that the plasma cells were reported to be IgG4 positive by immunohistochemistry, has dense fibrosis and elevated serum IgG4, although these findings are still controversial [4]. The main treatment for this condition is surgical excision with complete resection and the survival rate is estimated in 100%, with no death attributed to CFT in the literature.

Conclusion

CFT in the lung parenchyma is a rare begin lesion and should be considerate in the differential diagnosis on image and histopathological exams.

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