

Case Report

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Calcifying Fibrous Tumor of Lung: Report of a Case and Review of the Literature

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Abstract

Calcifying fibrous tumor (CFT) is a rare, benign, fibrous lesion typically found in subcutaneous and deep soft tissues of the extremities, trunk and neck of children, teenagers and young adults. Some cases have been described in the pleura and mediastinum. In contrast, CFTs of the lung are extremely rare and only 3 cases have been previously reported in the English literature. We report a giant CFT of the left lung, involving the hilum and treated with a left intrapericardial pneumonectomy in a 31-year old man. To the best of our knowledge, at this time only four pulmonary CFT cases, including our case, have been reported in literature. Our case is the widest thoracic CFT lesion described in the literature. This report highlights the clinical pathologic characteristics and differential diagnosis of this unusual pulmonary entity.

Introduction

Calcifying fibrous tumor is a benign mesenchymal solid neoplasm originally described as a soft tissue tumor in children [1]. CFT has been reported across a variety of different anatomic sites, such as the gastrointestinal tract, pleura, neck, axilla, scrotum, groin, and solid organs [2]. Amongst these, pulmonary lesions are very uncommon and only 3 previous cases have been described in this location [3-5]. CFTs are composed of scarce myofibroblasts in a hyalinized fibrosclerotic stroma accompanied by an infiltration of inflammatory cells such as plasma cells, macrophages, and lymphocytes. Psammomatous and dystrophic calcification are also present throughout the lesion.

The aetiology of these lesions is unknown and for a long time there was disagreement even about the inflammatory or neoplastic nature of these tumors [6-8]. Actually, CFT is mostly considered a true neoplasm. Molecular data and its tendency to local recurrence supports this hypothesis.

Herein, we describe the clinical, radiological, morphological and immunohistochemical features of a CFT of the lung in a young man.

Case presentation

A 31 year old man was referred to our hospital with the presumptive diagnosis of a wide neoplastic lesion of the left lung, causing dyspnea and chest pain. Routine laboratory data presented no alterations or were within normal limits. The patient had no other past medical history. The computed tomography (CT) revealed a large lesion of the lower left lobe, involving the main pulmonary artery and both the pulmonary veins at their origin from the left atrium. The lesion was also adherent to pericardium and descending aorta (Figure 1). Bronchoscopy was completely negative and an attempt to get a diagnosis with a percutaneous needle biopsy was unsuccessful (adequate but not diagnostic sample). At the left sided thoracotomy, we found a giant lesion at the left pulmonary hilum, involving all its main structures (left main bron-

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chus, pulmonary artery and veins at the origin). Repeated frozen sections showed a spindle cells proliferation without features of malignancy. The pleura was completely free from disease. Due to the dimension and the location of the tumor, an intrapericardial pneumonectomy was necessary to achieve a complete resection. The post-operative period was uneventful.

On gross examination, the lesion measured 14 X 10 X 8 cm, was lobulated, well circumscribed and firm but unencapsulated. On cut surface the tumor appeared white to tan to grey and the calcifications sometimes imparts a gritty texture (Figure 2). Conventional hematoxylin and eosin (HE) stained slides showed a hypocellular proliferation of relatively small, inconspicuous, mature fibrocytes set in a densely collagenous background. Dystrophic or psammomatous calcifications and a variable inflammatory infiltrate, composed chiefly of lymphocytes and plasma cells, were present. Lymphoid aggregates with active germinal centers were also observed. No atypical features were identified; tumor necrosis, cellular anaplasia or mitoses were absent (Figure 3). Immunohistochemically, the spindle-shaped cells were positive only for vimentin and factor XIIIa, negative for pan-cytokeratin, EMA, CD34, desmin, smooth muscle actin, CD99, ALK1, and S100 protein (Figure 4).

Morphological and immunohistochemical features were consistent with the diagnosis of calcifying fibrous tumor.

At the 4 years follow-up, the patient is disease free with a good quality of life.

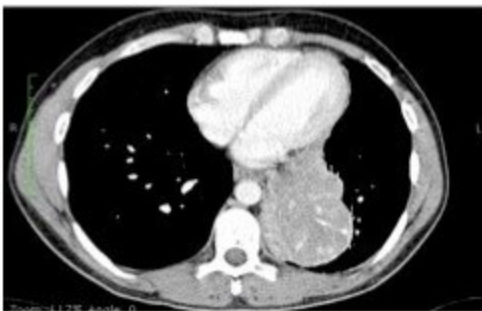


Figure 1: CT scan of the chest showing a sharply circumscribed mass within the lower left lobe involving the main pulmonary artery and both the pulmonary veins at their origin from the left atrium.

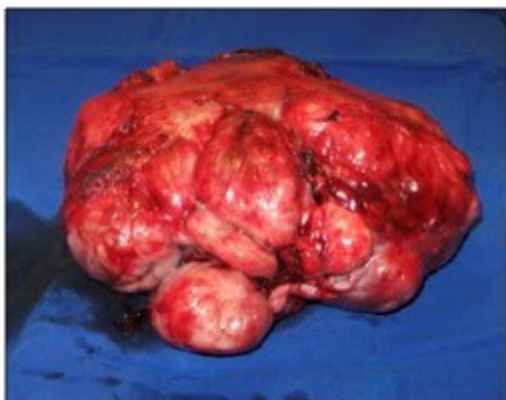


Figure 2: The excised specimen, measuring 14 X 10 X 8 cm was submitted for histology. It is composed by a solid, lobulated mass white to tan to grey in its gross appearance.

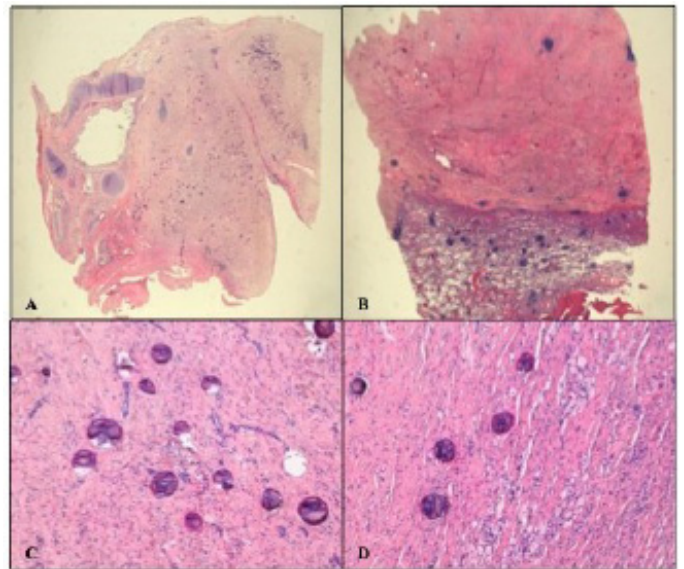


Figure 3: Low power photomicrograph showed a well circumscribed fibrous proliferation within the lung parenchyma and close to the bronchus. (A, B: HE, original magnification 2,5x). High power photomicrograph showed a hypocellular spindle cells proliferation in a dense hyalinised collagen background with dystrophic or psammomatous calcifications and a variable inflammatory infiltrate (C, D: HE; original magnification 40x).

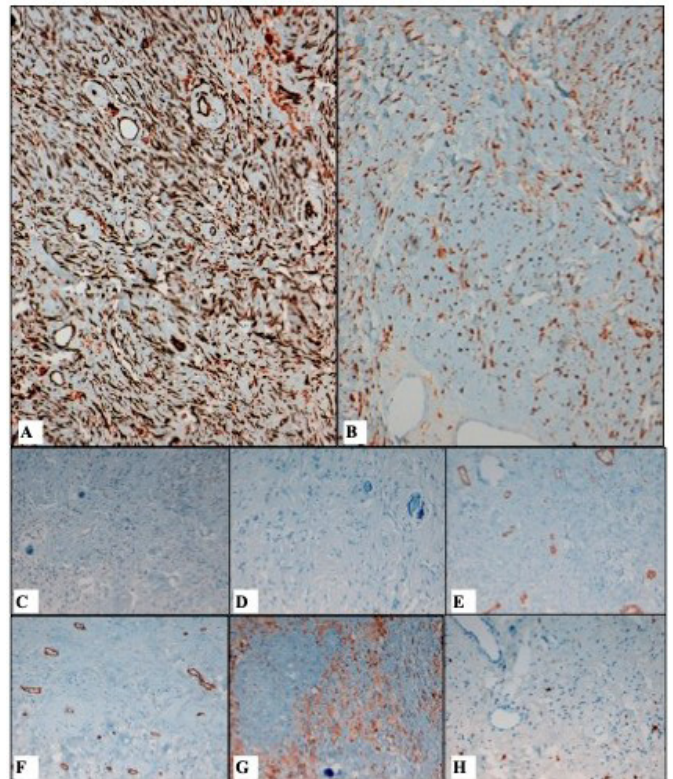


Figure 4: Immunohistological studies showed positive staining for vimentin (A; original magnification 40x) and Factor XIIIa (B; original magnification 40x). Negative staining were demonstrated for cytokeratin AE1/AE3 (C; original magnification 40x), ALK1 (D; original magnification 40x), smooth muscle actin (E; original magnification 40x), CD34 (F; original magnification 40x), CD68 (G; original magnification 40x) and, CD117 (H, original magnification 40x).

Discussion

Calcifying fibrous tumor was described for the first time in 1988 by Rosenthal and Abdul-Karim as a “childhood fibrous tumor with psammoma bodies” [1]. These lesions were subsequently renamed by Fetsch et al, in 1993, in “calcifying fibrous pseudotumor” [9]. However, due to a local recurrence rate of approximately 10%, these lesions were renamed CFTs in 2002 by the World Health Organization (WHO) classification of tumors of soft tissue and bone [10].

CFT is a rare benign mesenchymal tumor commonly characterised histologically by hyalinized collagenous fibrous tissue, with

bland spindle cells, psammomatous or dystrophic calcifications, and focal lymphoplasmacytic infiltrate. CFT is usually located in soft tissues of the extremities and trunk, other locations are rare and anecdotally reported. Its occurrence in the thoracic cavity is unusual, and even less frequently is its localization to the lung. Our review of the published English literature showed only 3 previous cases of CFT located in the lung. Peaches et al. in 2003 reported the first CFT with pulmonary localization.5 Subsequently, Soyer et al. in 2004 and Ozkan et al. in 2014 reported two more cases [3,4] to the best of our knowledge we report not only the fourth case of a lung CFT and the widest intrathoracic CFT lesion described in the literature [2] (Table 1).

Table 1:

Case	References	Age	Sex	Location	Size	Treatment	Follow-up
1	Peachell et al	31	M	Right hilum	2,7 cm	Lobectomy	NA
2	Soyer et al	7	M	Right lower lobe	5 cm	Lobectomy	1 week
3	Ozkan et al	64	M	Left hilar area	3,5 cm	Lobectomy and lymph node dissection	1 year
4	Present case	31	M	Left lung	14 cm	Pneumonectomy	4 years

The aetiology of this tumor is still unclear, but various pathogenic factors have been suggested to be involved. These include infection (Epstein-Barr virus), abnormal immunological reactions (Castleman’s Disease), or trauma. Indeed, CFT was initially thought to represent a reactive process resulting from abnormal tissue healing. Instead, later studies suggested a true neoplasm nature. Recently, it has been postulated that CFT may represent a sclerosing end-stage of an inflammatory myofibroblastic tumor (IMT). Tomassen et al. showed by methylation profiling that CFT and IMT share the same epigenetic profile but ALK1, a consistent marker of IMT, has been found only exceptionally in CFT [11]. Thus, the possibility that CFT is a distinctive lesion with unique clinical and biological significance is still valid.

The most important differential diagnosis of CFT includes other spindle cell tumors or pseudotumor conditions. These include: nodular fasciitis, fibroma of tendon sheath, desmoid-type fibromatosis, calcifying aponeurotic fibroma, schwannoma, IgG4-related disease, inflammatory myofibroblastic tumor, synovial sarcoma, solitary fibrous tumor, sclerosing leiomyoma, leiomyosarcoma, and fibrosarcoma. Metastatic lung tumors with calcifications, such as carcinoma of the ovary, breast, and thyroid, should also be included in the differential diagnosis. However, the histological features and immunohistochemical staining are almost always sufficiently distinct to allow easy segregation from a variety of reactive or neoplastic entities that enter the differential diagnosis. Dystrophic calcifications and psammomatous bodies are specific to CFT, which distinguish the tumor from many pseudotumor conditions. Absence of cellular atypia, storiform pattern, frequent mitosis, and bizarre tumor cells are the characteristics that distinguish CFT from soft tissue sarcomas such as leiomyosarcoma, fibrosarcoma and malignant fibrous histiocytoma.

Concerning the management of CFT a complete resection is the treatment of choice. The local recurrence rate varies from 17% to 30% in different series. Thus, the inclusion of a margin of normal tissue and a periodic follow-up would seem optimal to minimize this possibility.

With respect to the pre-operative diagnostic tools, percutaneous needle biopsy has been described mainly in chest wall and pleural lesions, but with disappointing results; our report confirms this datum.

Conclusions

CFT of the lung is extremely rare, and only 4 cases, including the present case, have been described in the literature. Although rare, CFT must be included in the differential diagnoses of lung lesions. The most important diagnostic aid is to bear this entity in mind and focus the attention on the histological criteria for differential diagnosis. CFT has shown an excellent prognosis. The conservative excision is almost always sufficient to cure the patient. However, local recurrence may occur. Thus, clinical follow-up is mandatory.

Conflict of interest: No potential conflict of interest relevant to this article was reported.

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