

Calcifying fibrous pseudotumor of lungs

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Abstract: Calcified fibrous pseudotumors can generally be detected as solitary masses in various regions of the body, and were first described in 1988. In this case report, we discuss an adult patient whose tumor was localized in the lung, which has not been reported in the literature before.

Key words: Pseudotumor, lung, calcifying, fibrous

1. Introduction

Calcifying fibrous pseudotumor (CFP) is a rare condition. It is seen as soft tissue pathology, and the lesions frequently observed solitarily are characterized by hyalinized collagen, psammomatous or dystrophic calcification, and lymphoplasmacytic infiltration. It occurs most commonly in childhood and young adulthood (more in women). The present study presents the findings of a lung-located CFP case, which is the second case reported in the literature.

2. Case report

A 64-year-old asymptomatic male presented for a routine health checkup; a mass was observed in the left hilar area on PA chest radiography. A thorax CT examination showed the lesion to be a hypodense mass 29 mm in diameter, located in the lower lobe of the left lung, which did not allow contrast enhancement and caused suspension in the segmental arteries (Figure 1). FDG involvement of the mass defined in a PET/CT examination was determined as SUVmax 4.61 and pathologic FDG involvement was not observed in any area except for the mass (Figure 2). The fiberoptic bronchoscopic evaluation did not determine any pathology in the tracheobronchial system; a fine-needle aspiration biopsy of the mass indicated a tumor of mesenchymal origin. Due to the location of the mass adjacent to vascular structures in the lower lobe hilus, the patient underwent left thoracotomy, lower lobectomy, and lymph node dissection. The patient was discharged on the fifth postoperative day without developing any complications. Macroscopic examination of the mass

showed a white 35-mm tumoral mass of firm consistency located in the parenchyma adjacent to the bronchus. Cross sections prepared from the mass included spindle cells that were well demarcated, with pulmonary parenchyma infiltrating the bronchus wall and a tumoral structure consisting of psammoma-like calcification (Figures 3A–3C). Immunohistochemical examination showed diffuse vimentin and focal EMA positivity in tumor cells; CD34, desmin, actin, pancreatin, and calretinin were negative. Trichrome dye showed the tumor was rich in collagen bundles. Amyloid staining showed no amyloids. In line with these findings, the case was diagnosed as calcifying fibrous pseudotumor. No metastatic focus was observed in the mediastinal lymph nodes. No clinical or radiological pathology was determined during postoperative control examinations, and the patient remained in good health during the 1 year of postoperative follow-up.

3. Discussion

Rosenthal and Abdul-Karim first characterized childhood fibrous tumor with psammoma bodies in 1988 in 2 pediatric cases; in 1993, a further 10 cases were identified by Fetsch et al. and were termed calcifying fibrous pseudotumor (1). In 1999, the World Health Organization categorized CFP as a soft tissue tumor.

CFP is a rare condition. The lesions observed as soft tissue pathology are characterized with hyalinized collagen, psammomatous or dystrophic calcification, and lymphoplasmacytic infiltration (1). As determined in our case, CFP is mostly observed as a solitary lesion. It is most

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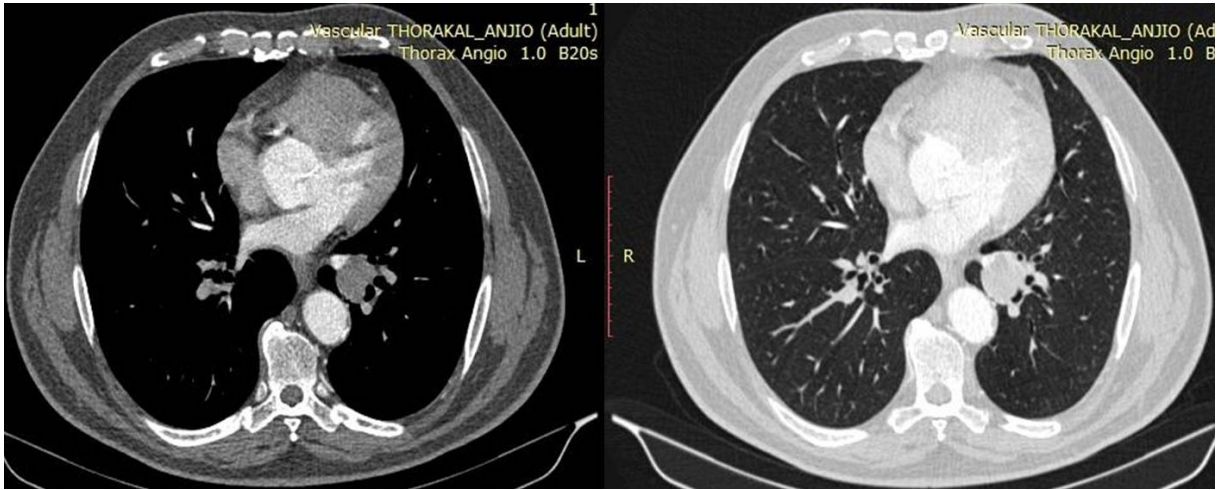


Figure 1. Findings of thorax CT examination.



Figure 2. Findings of PET/CT examination.

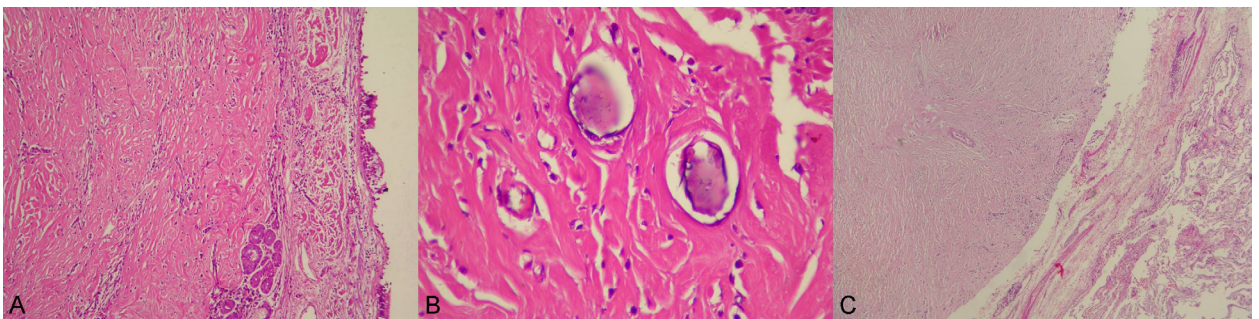


Figure 3. A. Adjacency of the tumor with bronchial mucosa (HEX200), B. psammoma-like calcification in dense fibrous stroma (HEX400), C. adjacency of the tumor with pulmonary parenchyma (HEX40).

common during childhood and early adulthood (more in women) (2).

The literature reports different locations for the tumor, i.e. peritoneum, extremities, pleura, mediastinum, dorsum, inguinal region, scrotum, thorax wall, cervix, axilla, heart, velum, and lungs (3,4). Our literature survey identified one pediatric patient with a tumor located in the lung (4). Ours is the second case of lung-located CFP in the literature, and the first report of lung-located CFP in an adult.

The differential diagnosis of the tumor includes thymic epithelial tumor, solitary fibrous tumor, soft tissue

ectopic calcification, osteochondroma, and inflammatory myofibroblastic tumor (5). The absence of cellular atypia, storiform pattern, frequent mitosis, and the characteristic features of tumor cells differentiate the tumor from other soft tissue sarcomas such as leiomyosarcoma, fibrosarcoma, and malign fibrous histiocytoma. Despite the absence of any case reporting metastasis, there are cases of recurrent diagnosis due to failure to completely excise the mass (1). There are no definitive clinical or laboratory findings to differentiate CFP from other pulmonary tumors, including tumor indicators and imaging techniques. Complete surgical excision of the lesion provides the cure.

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