

Calcifying Fibrous Tumor of the Pleura: A Rare Case with an Unusual Presentation on CT and MRI¹

흉막에서 발생한 석회화 섬유성 종양: 증례 보고¹

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Calcifying fibrous tumors (CFTs) are rare benign mesenchymal tumors consisting of hyalinized collagenous fibrotic tissue with a lymphoplasmacytic infiltrate and dystrophic calcifications. Radiographic features have seldom been described, and there are no reports describing magnetic resonance imaging (MRI) findings. Here, we report a pleural CFT in a 47-year-old woman. The tumor mimicked an intrapulmonary lesion on initial computed tomography scans but migrated inferiorly and presented as an extra-pulmonary lesion on MRI. The tumor showed iso-signal intensity on T1-weighted images (WIs), low signal intensity on T2WIs, and slight rim enhancement on enhanced T1WIs.

Index terms

Calcifying Fibrous Tumor
Pleura
CT
MR

INTRODUCTION

Calcifying fibrous tumors (CFTs) are rare benign tumors of uncertain etiology. These lesions consist of hyalinized collagenous fibrotic tissue interspersed with lymphoplasmacytic infiltrate and extensive dystrophic calcifications mimicking psammomatous features (1). CFT was first introduced as a “childhood fibrous tumor with psammoma bodies” by Rosenthal and Abdul-Karin in 1988 and was renamed “calcifying fibrous pseudotumor” by Fetsch et al. (2) in 1993. According to the World Health Organization classification, “calcifying fibrous tumor” is the term currently used (3). CFT of the pleura is a very rare entity first reported by Pinkard et al. (1) in 1996. To date, 15 cases of CFT of the pleura have been reported. The present report focuses on radiographic features of CFT of the pleura including magnetic resonance imaging (MRI) findings not reported previously.

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This study was approved by the Institutional Review Board of our hospital. The requirement for patient informed consent was waived due to the retrospective nature of the study. Patient records and information were anonymized and de-identified prior to analysis.

CASE REPORT

A 47-year-old woman presented with an abnormal chest X-ray taken for a health exam. She was a non-smoker, had no history of asbestos exposure, and did not have any respiratory complaints. All laboratory findings were normal. There was an approximately 4.5 cm, oval-shaped, well-circumscribed mass in the periphery of the left lower lung zone on a plain film (Fig. 1). An axial computed tomography (CT) scan (Fig. 2A, B) revealed a 4.5 cm sized, well-defined, lobulated, and partially calcified lesion locat-

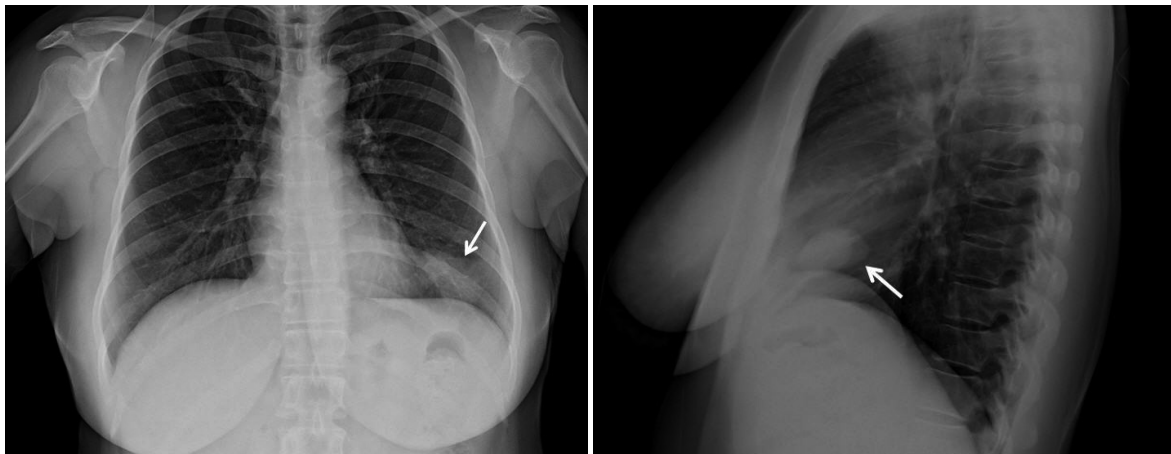


Fig. 1. Plain chest X-ray of the calcifying fibrous tumor of the pleura. Chest PA (A) and lateral view (B) show a 3 × 4.5 cm, oval-shaped, well-circumscribed mass-like lesion (arrow) in the left lower lung periphery.

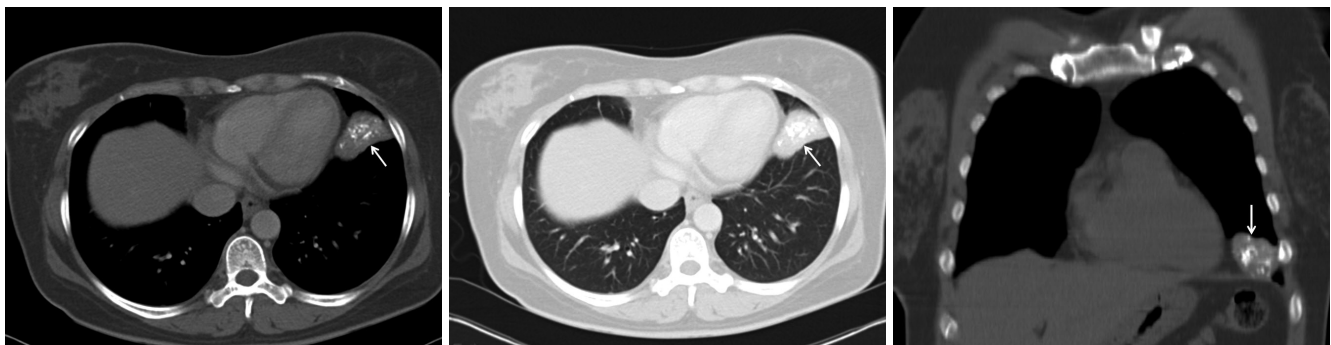


Fig. 2. High-resolution CT image of the calcifying fibrous tumor of the pleura. Axial CT scan on mediastinal (A), lung window setting (B), and coronal view mediastinal setting (C) show a 2.5 × 4.5 cm well-circumscribed, lobulated mass (arrow) with coarse calcification, located above the diaphragm in the lingular segment of the left upper lobe.

ed in the lingular segment of the left lung. The solid mass seemed to be an intrapulmonary lesion due to its acute angle relationship with adjacent structures (Fig. 2C). However, the mass seemed to have migrated inferiorly on coronal MRI and appeared to be located in the extra-pulmonary space between the left lower lung and the diaphragm (Fig. 3A). The mass was well-circumscribed, surrounded by the pleura, and had an obtuse relationship with the chest wall. The lesion showed iso- to intermediate signal intensity on T1-weighted images (T1WIs) (Fig. 3B) and low signal intensity on T2-weighted images (T2WIs) (Fig. 3C). No signal change was detected on fat saturation T2WIs. Slight peripheral rim enhancement was observed after gadolinium administration (Fig. 3D).

A complete resection was performed via video-assisted thoracoscopic surgery (VATS). The mass was located on the left anterior chest wall and was 6 cm in longest diameter, well demarcat-

ed, and firm (Fig. 4A). The cut surface of the mass revealed a solid, white, fibrous matrix without hemorrhage or necrosis (Fig. 4B). Fibrotic tissue with dystrophic calcification was noted on a microscopic evaluation (Fig. 4C). Immunohistochemical staining was positive for vimentin (Fig. 4D) and CD99 but negative for smooth muscle actin and CD34. The final pathological diagnosis was CFT of the pleura. The patient underwent a follow-up CT scan after 6 months, and there was no evidence of recurrence.

DISCUSSION

CFT of the pleura is a very rare disease; we found only 15 cases reported in the literature. Among these prior cases, 10 were women (67%; age, 23–52 years), and the age range for the men was 34–54 years. Symptoms included chest discomfort or pain

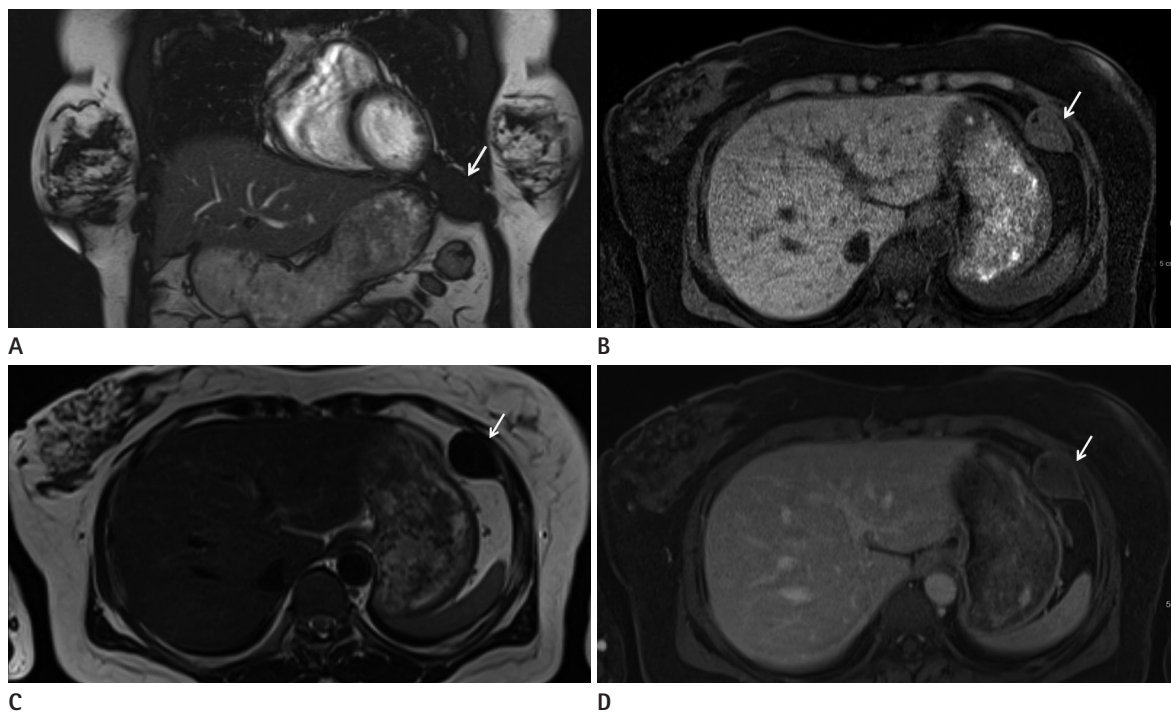


Fig. 3. MR images of the calcifying fibrous tumor of the pleura. The ultrafast spine echo Half-Fourier-acquisition single-shot turbo spine-echo coronal image **(A)** shows an inferiorly migrated mass located in the extraparenchymal space between the left diaphragm and left lung. The mass (arrow) shows iso- to intermediate signal intensity on the T1-weighted image **(B)** and low signal intensity on the T2-weighted image **(C)**. Slight peripheral rim enhancement was observed on the gadolinium-enhanced T1-weighted image **(D)**.

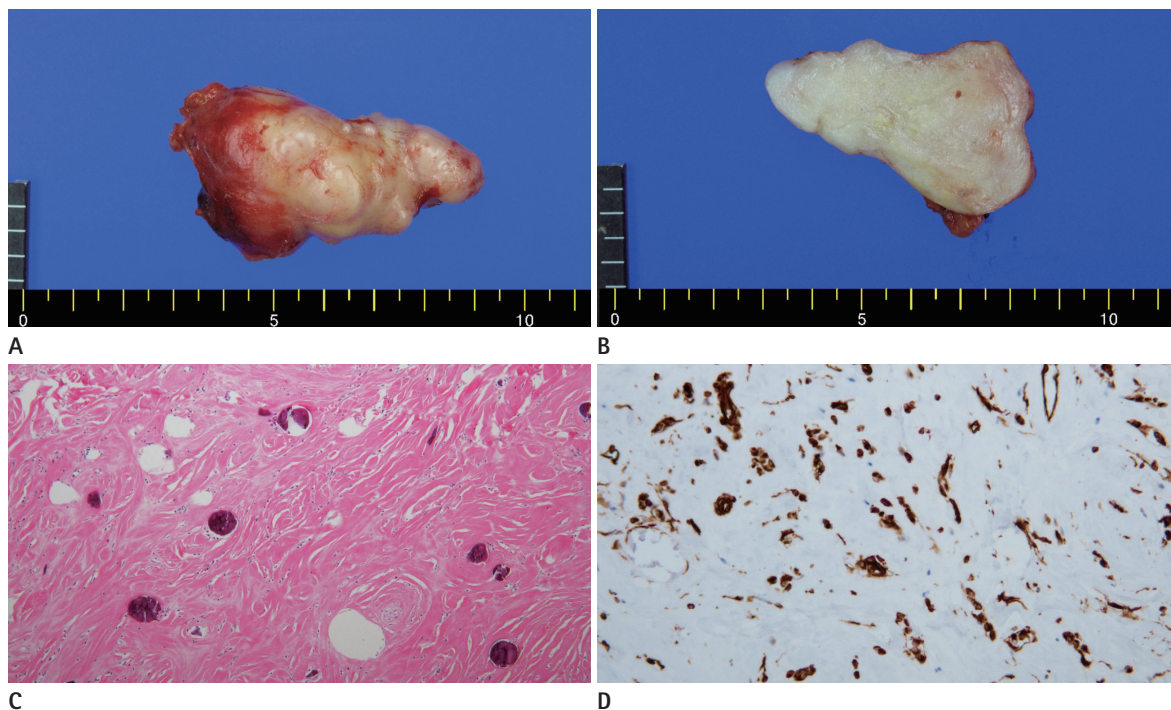


Fig. 4. Gross images and histology of the specimen. The mass measured up to 6 cm, well demarcated, firm and lobulated **(A)**. Cut surfaces of the mass revealed a solid, white, fibrous matrix without hemorrhage or necrosis **(B)**. On microscopic exam, fibrotic tissue with dystrophic calcification was observed [hematoxylin and eosin stain ($\times 100$)] **(C)** with vimentin positive on immunohistochemical staining ($\times 200$) **(D)**.

(40%), and asymptomatic cases were found incidentally after radiographic exams. More than half (67%) of these cases had numerous small, nodular lesions that were not visible on CT but were seen preoperatively and were confused with malignant lesions, such as mesothelioma (4) or metastatic lesions (3), particularly in patients with a history of asbestosis or cancer.

Most cases showed well marginated, heterogeneous, pleural-based extra-parenchymal masses on CT, with an obtuse angle relationship to the chest wall, mediastinum, or diaphragm and were usually located in the lower part of the thorax. All of the cases showed varying degrees of calcification in the center of the mass. Isaka et al. (5) reported ¹⁸F-fluorodeoxyglucose positron emission tomography-CT findings with a maximum standardized uptake value of 2.0, which is low and indicative of a benign tumor.

No prior reports have described MRI findings for CFT of the pleura. In our case, T1WIs showed an iso-signal to skeletal muscles and low signal intensity on T2WI, which may have been related to the hypocellularity and scattered calcification of the tumor. However, unlike the intense enhancement after the gadolinium injection observed in other benign fibrous tumors of the pleura, our case showed only mild peripheral enhancement. The central portion of the mass consisted mainly of collagen and calcium and lacked a cellular component. A few inflammatory cells and spindle cells were seen at the periphery of the mass under a microscope, which may explain the minimal peripheral enhancement and no enhancement in the central portion.

Histological findings of CFT include unencapsulated, densely hyalinized collagen bundles with fibrotic proliferation and calcifications with laminated features mimicking psammoma bodies. Our case showed typical calcifications with lymphocyte and plasma cell infiltration. Sparse spindle cells with no atypia were found. Immunolabeling is not mandatory for the diagnosis (6), but vimentin staining was positive in spindle cells. CD34 staining in a submesothelial fibroblast-originating tumor is usually negative in CFTs and positive in localized fibrous tumors of the pleura. However, some cases of CFT of the pleura have stained positive for CD34 (3, 7), suggesting a common origin for CFT and solitary fibrous tumors of the pleura. Unlike hypercellular solitary fibrous tumors, which lack calcification, CFTs are hypocellular with dystrophic or laminated calcification. In our case, the central portion of the mass mainly consisted of collagen and calcium and lacked a cellular component, whereas a few inflam-

matory cells and spindle cells were seen on the periphery of the mass. The lesion seemed to be intrapulmonary on the CT scans. The change in mass location was explained by respiration or a change in patient position. Ferretti et al. (8) reported that large tumors of the pleura can be difficult to localize because they often form acute angles that mimic intrapulmonary masses. Many prior cases of CFT of the pleura have revealed tumor pedicles that connect the tumor to the pleura (1, 5, 9).

The current mainstay treatment for CFT of the pleura is total excision of the mass via VATS or open surgery. Somatic soft tissue CFTs have a low frequency of local recurrence with a good prognosis (3). No evidence of recurrence 6–12 months after resection has been reported in previous cases of CFT of the pleura, but long-term observations are required.

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흉막에서 발생한 석회화 섬유성 종양: 증례 보고¹

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석회화 섬유성 종양은 섬유성 조직, 림프구형질세포, 이형성 석회화 등으로 이루어진 드문 간엽성 양성 종양이다. 영상소견에 대한 보고는 많지 않으며, 자기공명영상소견은 아직 보고된 바가 없다. 이에 47세 여환에서 발생한 흉막 기원 석회화 섬유성 종양의 영상소견을 보고하고자 한다. 이 종양은 CT에서 폐실질 내의 병변으로 보였으나, 이후 시행한 MRI에서는 폐실질 외, 흉막기원의 종양으로 관찰되었다. T1-weighted image (이하 WI)에서는 근육과 비슷한 신호강도를, T2WI에서는 낮은 신호강도를 보였으며, 조영 후 T1WI에서 종양의 가쪽으로 약간의 조영증강을 보였다.

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