Abstract: Calcifying fibrous pseudotumor (CFP) of the pleura is a rare benign lesion. Eleven cases of CFP of the pleura have been reported, and multiple nodules were present in seven cases. We report one additional case of multiple CFP occurring in the right pleura. The patient was 35-year-old asymptomatic male who presented at our hospital with incidentally found multiple pleural nodules. Diagnosis was made by video-assisted thoracoscopic surgery and complete resection of all disseminated lesions was possible with followed thoracotomy. Although multiple lesions may prevent the complete resection and CFP of the pleura is considered as benign lesion, complete surgical resection of all lesions seems to be the best therapy for CFP of the pleura to reduce additional dissemination and local recurrence.

Key Words: Tumor, Benign, Calcifying fibrous pseudotumor, Pleura.

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Calcifying fibrous pseudotumor (CFP) was originally reported by Rosenthal and Abdul-Karim as a childhood fibrous tumor with psammoma bodies in 1988. In 1993, the term of CFP was first adopted by Fetsch et al. with an analysis of 10 cases. CFP is a rare benign soft tissue lesion that primarily affects children and young adults and most commonly arises in the extremities. Since three cases of CFP, in which lesion involved the pleura, were first described by Pinkard et al. in 1996, 11 cases of CFP of the pleura have been reported in the literature.1 We present a patient who showed multiple disseminated CFP of the pleura which were completely extirpated.

CASE REPORT

A 35-year-old man was referred to our hospital for further evaluation of a pleural nodule which was incidentally found in a chest radiograph during his medical check-up for employment (Figure 1). He had no symptom and specific medical history except appendectomy 4 years previously. Chest computed tomography (CT) confirmed multiple pleural nodules of the right chest (Figure 2). Video-assisted thoracoscopic surgery was performed for histologic examination to exclude malignancies. During the operation, we could find multiple small nodules on the surface of lung, diaphragm, and parietal pleura. We performed wedge resection of the right lower lobe that contained three nodules and local excision of a single soft tissue mass located in the parietal pleura. The lesion of the parietal pleura was the largest of all nodules. Since intraoperative rapid pathologic diagnosis of the frozen section revealed chronic inflammatory cell infiltration with no malignant findings, thus we performed no additional resection. Several weeks later, the final pathologic diagnosis was proven to CFP. We decided to perform resection of the remained nodules using thoracotomy for complete resection due to lack of knowledge about the prognosis of multiple presentation of tumor. Totally, 16 nodules were resected including previous operation. At second operation, six nodules were resected from the right lower lobe. Two small nodules were resected from the middle lobe and the inferior pulmonary ligament. Remaining four nodules attached to the thoracic diaphragm were resected including the diaphragm and the resected diaphragm was repaired.

All pulmonary lesions were located on the visceral surface of the lung and thus multiple wedge resections for complete resection were possible. The resected lesions were well limited but nonencapsulated pale gray rubbery masses ranged 0.4 to 3.0 cm in longest dimension. Microscopic examination by hematoxylin-eosin stain confirmed the pathologic diagnosis of CFP. The lesion mostly composed of dense hyalinized collagenous tissue containing lymphoplasmacytic infiltrates and dystrophic calcifications (Figure 3). Immunohistochemical studies showed positive findings for vimentin, smooth-muscle actin and CD34, but negative findings for desmin and anaplastic lymphoma kinase. The patient’s postoperative recovery was uneventful and no evidence of local recurrence or distant metastasis has been found in the 16 months since his second operation.

DISCUSSION

CFP is a extremely rare benign pleural lesion, but easily recognizable by its unique histopathologic features characterized by a dense hyalinized collagenous tissue interspersed with benign spindle cells, lymphoplasmacytic infiltrate, and, particularly, psammomatous and/or dystrophic calcifications. Grossly, CFP of the pleura shows well-circumscribed but unencapsulated, solid, or firm with a uniform gray-white fibrous cut appearance without hemorrhage or necrosis and lesions are pleural-based.
The multiple CFP of the pleura has the possibility of confusion with malignant pleural fibrous neoplasm including desmoplastic mesothelioma or disseminated metastatic sarcoma. Desmoplastic mesothelioma shows a storiform collagen pattern, collagen necrosis, bland acellular collagen, and focal cytologic features of malignancy with positive findings for cytokeratin and obviously invasive. In disseminated metastatic fibrous sarcoma with pleural calcification can be confused radiologically and, thus histologic confirmation is necessary. These include synovial sarcoma, giant cell tumor of the bone and carcinomas of the colon, ovary, breast and thyroid. To exclude these malignancies, either partial or complete resection of the lesions may be required.

Mito et al. suggested that CFP of the pleura affects slightly older individual (mean age of 34 years) than CFP of the somatic soft tissue (mean age of 14.5 years). In addition to their case, we reviewed more three cases and our present case. Of all the 12 patients, only 4 patients had solitary tumor, and remained 8 patients showed multiple or disseminated lesions. In the setting of multiple or disseminated CFP, lesions usually seemed as small multiple disseminated nodules with a relatively large main mass. These findings suggest the possibility of pleural dissemination from a primary tumor. In these cases, small multiple nodules could not be detected in the preoperative CT scan. And preoperative CT scan of our patient also did not show all small nodules. Eight patients

FIGURE 1. Preoperative chest radiograph reveals a pleural nodule in the right lower chest (black arrow).

FIGURE 2. Serial views of computed tomographic scan reveal multiple pleural nodules in the right chest.

FIGURE 3. Microscopic findings show dense interwoven collagen bundles with scattered spindle shaped cells, psammomatous calcifications (A) (hematoxylin and eosin, ×100), and lymphoplasmacytic infiltrate (B) (hematoxylin and eosin, ×100). Immunohistochemistry reveals that the most spindle cells are positive for vimentin (C) (×200).
were female. It also suggests slight female predilection of this tumor.\textsuperscript{1}

Although local recurrence has been reported in a few cases of CFP, many authors recommend that local excision is an adequate therapy for CFP of the pleura, as it is for CFP of the somatic soft tissue.\textsuperscript{3,5,6} And the recurrence of disease or distant metastasis of CFP of the pleura was not documented so far. In the two cases reported by Hainaut et al. and Shibata et al., the multitude of pleural lesions prevented complete excision.\textsuperscript{1} But the long-term prognosis remains undetermined. Thus, we believe that a complete resection of all lesions is the best way to treat this rare tumor even in the multiple lesions and good prognosis can be expected with complete resection. We performed complete resection of all 16 nodules and long-term follow-up is necessary to confirm the value of the complete resection in the setting of multiple disseminated nodules.

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**REFERENCES**