Multiple Calcifying Fibrous Pseudotumor of the Bilateral Pleura

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Calcifying fibrous pseudotumor is a rare lesion characterized histologically by hypocellular hyalinized collagenous tissue with calcifications and patchy lymphocytes infiltration. Occurring most often in children and young adults, calcifying fibrous pseudotumor is a clinically benign lesion that can form over a broad anatomic distribution, including in subcutaneous and deep soft tissues, but is rarely found in the pleura. The cause and mechanisms of pathogenesis of calcifying fibrous pseudotumor are unknown. In this article, we describe a case of a 44-year-old woman with multiple calcifying fibrous pseudotumor disseminated in the bilateral pleura that was pathologically diagnosed. We discuss the differential diagnosis with other benign or malignant soft tissue diseases and also review the recent literature on this rare benign entity. Complete resection of all disseminated lesions was possible with followed thoracotomy. Although multiple lesions may prevent the complete resection and calcifying fibrous pseudotumor of the pleura is considered as benign lesion, complete surgical resection of all lesions seems to be the best therapy for calcifying fibrous pseudotumor of the pleura to reduce additional dissemination and local recurrence.

Key words: calcifying fibrous pseudotumor – bilateral pleura – resection

INTRODUCTION

Calcifying fibrous pseudotumor (CFP) is a rare soft tissue lesion that has been documented in recent 10 years. Since the clinical feature of CFP is unique and rare, it is important to be well aware of diagnosis, treatment and prognosis of CFP. To our knowledge, only 12 cases of CFP of the pleura have been reported in the literature (1,2). However, all the cases of CFP are described in which the lesion involved the unilateral pleura. We would like to add one case of multiple CFP disseminated in the bilateral pleura that was pathologically diagnosed.

CASE REPORT

A 44-year-old woman was referred to our hospital for further evaluation of pleural nodules that were incidentally found in a chest radiograph during her medical examination. She presented with non-specific chest pain. Chest computed tomographic (CT) scan showed multiple pleural nodules in the bilateral thorax. No hilar or mediastinal lymphadenecstasy was seen (Fig. 1). Family history and past medical history were unremarkable. A tentative diagnosis of diffuse mesothelioma was made. Right video-assisted thoracoscopic surgery was performed for histological examination. During the operation, we found that multiple pale gray nodules of diverse sizes disseminated throughout the chest wall. We performed local excision of the largest pleural nodule measured about 2.5 cm located in the parietal pleura for intraoperative rapid pathologic examination. Since rapid pathologic diagnosis of the frozen section revealed hyalinized fibrosclerotic nodular lesions with microlcalifications with no malignant findings, we decided to convert to open surgery. Thoracotomy was performed, and all remaining nodules were extirpated completely. A subsequent left thoracotomy was undertaken with excision of all left-sided nodules 1 month later. The follow-up CT scan has shown no new lesion at 12 months (Fig. 2).

On postoperative gross examination, all nodules were well circumscribed, non-encapsulated and solid with a uniform white fibrous cut appearance without any hemorrhage or necrosis (Fig. 3). Histologically, the nodules were composed
Figure 1. (A) Pre-operative computed tomographic scan showed multiple pleural nodules in the bilateral thorax. (B) No hilar or mediastinal lymphadenectomy was seen.

Figure 2. Hematoxylin and eosin, ×200. (A) Calcifications, (B) hyalinized degeneration collagen fiber, (C) collagen fiber comprising fibrocytes and (D) varying amount of inflammatory cells infiltration.
chiefly of hypocellular hyalinized fibrosclerotic tissue with scattered cytologically bland spindle cells, scant inflammatory cells, mainly lymphocytes infiltration and small calcifications. The surface of the nodules was occasionally covered by a single layer of mesothelial cells. Asbestos bodies were not detected in or around the nodules. Taken together, a pathologic diagnosis of multiple CFP was made.

DISCUSSION

CFP was first reported as a ‘childhood fibrous tumor with psammoma bodies’ in two girls by Rosenthal and Abtul-Karin (3) in 1988. In 1993, Fetsch et al. (4) described 10 adult cases and renamed the entity ‘calcifying fibrous pseudotumor’ for the histological character of CFP is hypocellular hyalinized collagenous tissue with calcifications and/or psammomatous but not limited to the psammoma bodies in the lesion (6). Non-thoracic cases are predominantly reported in children or young adults with a mean age of 14.5 years (3,4). The pleural cases occur mainly in females aged from 23 to 54 years with a median age of 38.7 years (1). Many case reports of multiple intrathoracic CFP are from Asian countries such as Japan, Korea and India, with our patient being of Chinese origin. Of the patients with pleural CFP, some complained of chest pain and/or non-productive cough, whereas a few were asymptomatic.

With respect to the number of nodules, multiple or disseminated lesions involving the unilateral pleura were present in eight patients and solitary nodules were present in four patients. In our patients, multiple CFP disseminating in the bilateral pleura was pathologically diagnosed. In the setting of multiple CFP, lesions usually appeared (7); and hypocellular lesion prominently comprising fibrocytes or myofibroblasts intermixed with varying amount of inflammatory cells infiltration.

The pathogenesis of CFP remains unknown to us nowadays. Fetsch et al. (4) thought that CFP was a distinctive form of fibrous pseudotumor, some others considered CFP as a reactive process to inflammation or tumor-like lesions and still others presumed CFP was the end stage of inflammatory myofibroblastic tumors. Although Hill compared histological and immunohistochemical features between seven cases of CFP and seven cases of inflammatory myofibroblastic tumors, they did not illuminate that they were the same illness.

Since CFP has been reported in recent years and is extremely rare, recognition of the pathologic features of CFP is important because of the possibility of confusing it intraoperatively with some benign or malignant soft tissue diseases such as calcified granuloma, calcified pleural plaque, chronic fibrous pleuritis and mesothelioma of pleura. Calcified granuloma generally presents pathologic changes of granuloma formation with the character of well-demarcated nodular hyperplasia composed by effused monocytes and macrophages. Langhans giant cells may be found in the lesion. Calcified pleural plaque disperses in

Table 1. Calcifying fibrous pseudotumor (CFP) and other similar diseases differential diagnosis

<table>
<thead>
<tr>
<th>Disease name</th>
<th>Fibrosis and hyalinosis</th>
<th>Calcification</th>
<th>Cell infiltrating</th>
</tr>
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<tbody>
<tr>
<td>CFP</td>
<td>+</td>
<td>+</td>
<td>Hypocellular, varying inflammatory cells infiltration</td>
</tr>
<tr>
<td>Calcified granuloma</td>
<td>+, nodosity</td>
<td>+</td>
<td>Granulomatous lesion</td>
</tr>
<tr>
<td>Chronic fibrous pleurisy</td>
<td>+</td>
<td>+/-</td>
<td>Diffusing pleural thickening and plaque fibrosis</td>
</tr>
<tr>
<td>Mesothelioma of pleura</td>
<td>+</td>
<td>+</td>
<td>Infiltrative growth, abundant cells with certain of nuclear atypia</td>
</tr>
</tbody>
</table>
thickened partial pleural with inequality of sizes and often exists in the bilateral thoracic cavity. Calcified plaque is large and has broad distribution with irregular shape but distinct boundary. The plaque is smooth and ivory white, consisting of uniform and intensive-layered collagen fiber in parallel alignment or interweaving reticulum. There are usually no inflammatory cells or blood vessels in lesion sites. The patients have history of exposure to asbestos. Chronic fibrous pleuritis demonstrate fibrous exudation on the surface of the pleura and infiltration of neutrophil chronic inflammatory cells. It shows clear zone phenomenon of vertically aligned capillaries to pleura with more cells adjacent to pleura surface and less cells away from the surface. There are fibrous thickening and adhesions in advanced stage of disease. There are spindle-shaped cells or fibroblast in benign mesothelioma of the pleura. Condensing collagen fibers are in stagger arrangement or network structure. The tumor shows non-infiltrative growth in fat or muscle of chest wall or lung tissue. In malignant mesothelioma of pleura cells present nuclear atypia and nucleolus and cytoplasm is acidophilia. The tumor shows infiltrative growth in fat or muscle of chest wall or lung tissue. There is no zone phenomenon or obviously hyperplastic capillaries Table 1).

CFP is a slow growing benign lesion and not considered as true neoplasm. Although local excision is performed in a few cases of CFP, the recurrence of disease or distant metastasis of CFP of the pleura was not documented so far. Some authors suggest CFP may recur if lesions are inadequately excised (1,5). Moreover, the patient presented with nonspecific chest pain in the studied case. Thus, we believe that a complete resection of all lesions is the best way to treat this rare tumor.

Conflict of interest statement
None declared

References