Korean J Thorac Cardiovasc Surg 2011;44:318-320 □ Case Report □
ISSN: 2233-601X (Print) ISSN: 2093-6516 (Online) DOI:10.5090/kjtcs.2011.44.4.318

Calcifying Fibrous Pseudotumor of the Anterior Mediastinum

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Calcifying fibrous pseudotumors are rare soft-tissue lesions pathologically characterized by hyalinized collagen, psammomatous or dystrophic calcification, and lympho-plasmacyte infiltration. They are clinically benign with an extremely low rate of recurrence and complete surgical resection is known to be the treatment of choice. We performed the resection of a calcifying fibrous pseudotumors in the anterior mediastinum without complications.

Key words: 1. Mediastinal neoplasm
2. Pseudotumor

CASE REPORT

A fifty-one-year-old man was referred to our department because of a bulging contour in the right paratracheal area on chest x-ray, which was identified during a routine medical checkup (Fig. 1A). He did not complain of any respiratory symptoms and denied fatigue, weight loss, weakness, or regurgitation after swallowing. He was a non-smoker and had a history of cholecystectomy fifteen years before presentation. Physical examination revealed nothing specific. The complete blood count (CBC) and chemistry were within normal limits and abdominal ultrasound showed small cysts in the liver and kidney. There were no lesions in the abdomen suspected of malignancies. Chest CT a showed well-defined calcified mass in the right anterior mediastinum with its largest diameter at 3.3 cm (Fig. 1B). Based on the principles of anterior mediastinal mass treatment, we planned complete excision via sternotomy. Under general anesthesia, a median sternotomy was performed. The mass was hard in consistency and completely distinctive from the thymus and mediastinal fat tissue (Fig. 2). It was easily dissected from the surrounding structure, but firmly adhered to the brachiocephalic vein. The adhered part of brachiocephalic vein was resected en bloc with the mass and primary repair of the vein was then performed. Pathologic findings revealed a 4.2×2.7×2.1 cm sized well-circumscribed mass, which was not encapsulated and had a white to yellow cut surface (Fig. 3A). Microscopically, it consisted of abundant hyalinized collagen and psammomatous calcification (Fig 3B). Immunohistochemically, the spindle cells of this mass did not show cytoplasmic stain with antibodies against actin, desmin, or CD 34. The patient did not have any postoperative complications and was discharged on the fifth day postoperatively. During 11 months of follow-up, he has been doing well without any evidence of recurrence.

DISCUSSION

A calcifying fibrous pseudotumor (CFP) is a rare benign fibrous lesion characterized by dense hyalinized collagenous tissue in the sparse spindle cells, psammomatous or dystro-

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Received: March 2, 2011, Revised: March 21, 2011, Accepted: April 1, 2011
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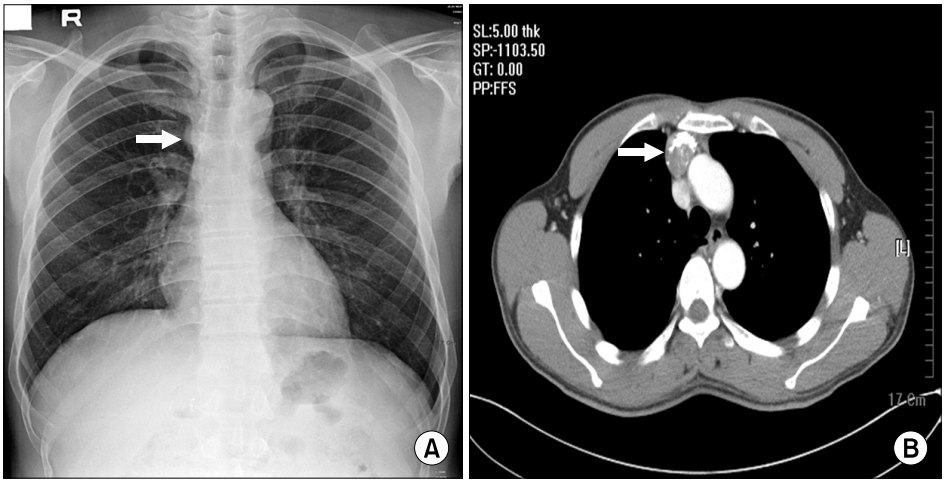

Fig. 1. Bulging contour in right paratracheal area on chest PA (A) and calcified anterior mediastinal mass (B).

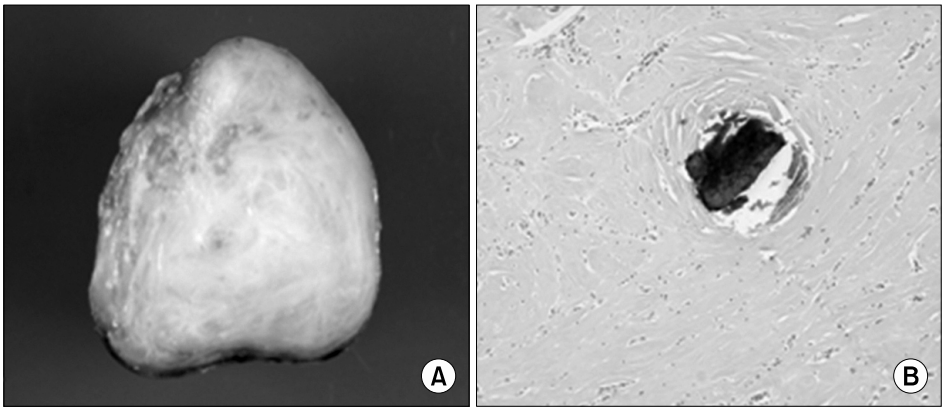

Fig. 3. The mass is well circumscribed and firm and has a gray-white fibrous appearance (A). The photomicrograph of the mass shows a uniform dense collagenous matrix with a psammomatous calcification (B) (H&E, ×100).

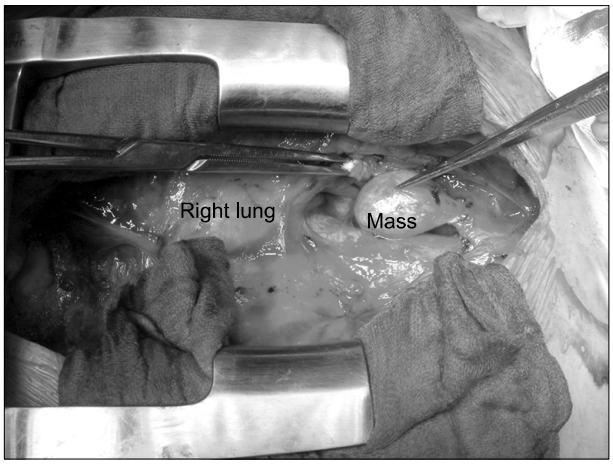

Fig. 2. Intraoperative photograph shows a well-circumscribed mass in the anterior mediastinum.

phic calcifications, and a variable quantity of lymphoplasmacytic infiltrates [1]. Such lesions were initially reported as childhood fibrous pseudotumors with psammomatous bodies [2] and thereafter named CFP by Fetsch et al. in 1993 [1]. Most of the reported cases were in children and young adults (ranging from 1 to 33 years old) and occurred in the soft tissue of the neck, trunk, or extremities. CFP has also occurred in the pleura as multiple nodules, in the mediastinum [3], or even in the myocardium [4]. CFP usually does not cause systemic symptoms, but it can cause specific local symptoms such as thoracic outlet syndrome according to its location [5]. This case did not show any systemic or localized symptoms and occurred in a middle-aged man. The fact that CFP was found in a man in his fifties is quite unusual, but it is difficult to presume the natural disease course of this lesion because an old chest PA was not available.

The differential diagnosis includes thymic epithelial tumor, solitary fibrous tumor, ectopic calcification of soft tissue, osteochondroma, and inflammatory myofibroblastic tumor (IMT).

The pathologic features are the key to differential diagnosis and those of CFP are well established. The most prominent feature is psammomatous or dystrophic calcification. Immunohistochemically, CFP shows only weak or absent expression of CD 34 and nonspecific expression of alpha smooth muscle actin or desmin [6]. This case showed no distinctive expression of CD 34, actin, or desmin. Although once CFP was considered to be a sclerosing end stage inflammatory myofibroblastic tumor, it is now well recognized that CFP and IMT have their own distinct histologic, immunohistochemical, and electron microscopic features [6]. For example, anaplastic lymphoma kinase-1, one of the distinguishing features of IMT, is not common in CFP [7].

Complete surgical excision is the definitive treatment, but in some cases it is not possible due to its location, such as encasement of the anterior descending coronary artery or infiltration into the myocardium [8]. If any CFP lesions in specific locations cause compressive or constrictive symptoms and prevent complete resection, partial resection or marsupialization should be performed for the relief of symptoms. Though recurrence is rarely known [1], regular follow-up is obligatory.

We report a case of CFP in the anterior mediastinum of a middle-aged man. Complete excision was the treatment of choice for CFP due to its benign nature, good prognosis, and rare recurrence.

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