

Ectopic Cystic Thymoma Associated with Raynaud's Phenomenon

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Mediastinal cystic tumors are well-margined round lesions that comprise 12% to 18% of all mediastinal masses. These lesions include a variety of diseases with overlapping radiologic appearances and variable prognoses. Pathological examinations are almost always required for differential diagnosis. We encountered a case of anterior mediastinal tumor discovered in the process of investigation of Raynaud's phenomenon. Taking into account the tumor location, a pericardial cyst was initially suspected. However, the tumor was surgically resected and histopathological examinations demonstrated thymus-like tissue in the cyst walls. Raynaud's phenomenon greatly improved after surgery. These findings suggested that cystic thymoma originated from ectopic thymic tissue and is accompanied by paraneoplastic syndrome. (Ann Thorac Cardiovasc Surg 2007; 13: 118–21)

Key words: mediastinal tumor, cyst, thymoma, Raynaud's phenomenon

Introduction

Mediastinal cysts show various characteristics, and represent 12% to 18% of all primary mediastinal tumors. Due to the similarity in imaging appearances, we sometimes have difficulty with differential diagnosis without pathological examination. We report on a case of cystic thymoma that simulated a pericardial cyst.

Case Report

A previously healthy 37-year-old man presented with a two-month history of Raynaud's phenomenon. He had normal findings on physical examination, 12-lead electrocardiography, and blood examination was normal except for a higher value of serological anti-nuclear factor (titers >1/1,280). A Chest x-ray showed a smoothly mar-

gined density in the region of the right cardiophrenic angle (Fig. 1A). A contrast-enhanced computed tomography (CT) scan demonstrated a huge bilocular mass adjacent to the right atrium with partial contrast enhancement in the right anterior mediastinum (Fig. 1B). Neither pleural nor pericardial effusion was evident.

Magnetic resonance imaging (MRI) demonstrated a 10×7.6 cm oval-shaped mass with a high signal intensity of the mass contents on the T1-weighted image (Fig. 2A). An echocardiogram showed the heterogeneous structure with right atrial compression, and was suggestive of a pericardial cyst (Fig. 2B).

We could not detect any cause of Raynaud's phenomenon but the possibility arose of paraneoplastic syndrome. Therefore, the patient underwent operative exploration. Macroscopically the smoothly surfaced mass was extrapericardial, situated in the right paracardiac area. Then it was resected through the midclavicular discission in the fourth intercostal space under a video-assisted thoracic procedure. Careful observation in the anterior-superior mediastinum after removal of the tumor showed the existence of the atrophic thymus-like organ without any direct continuity to the resected mass. The loculated mass (10×11×9 cm) was encapsulated by a focally calcified capsule and contained a yellowish fat-like substance (Fig.

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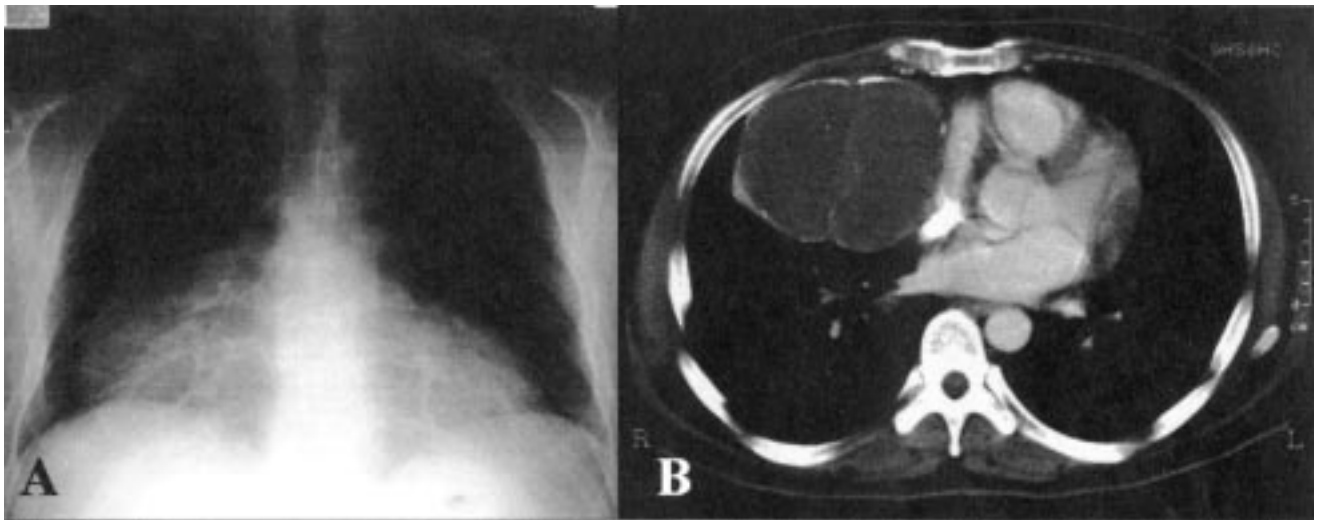


Fig. 1.

A: Posteroanterior chest radiogram demonstrated a large dense mass in the region of the right cardiophrenic angle.

B: Contrast-enhanced chest CT scan showed a large loculated tumor encapsulated by a focally enhanced wall in the right anterior mediastinum adjacent to the right atrium.

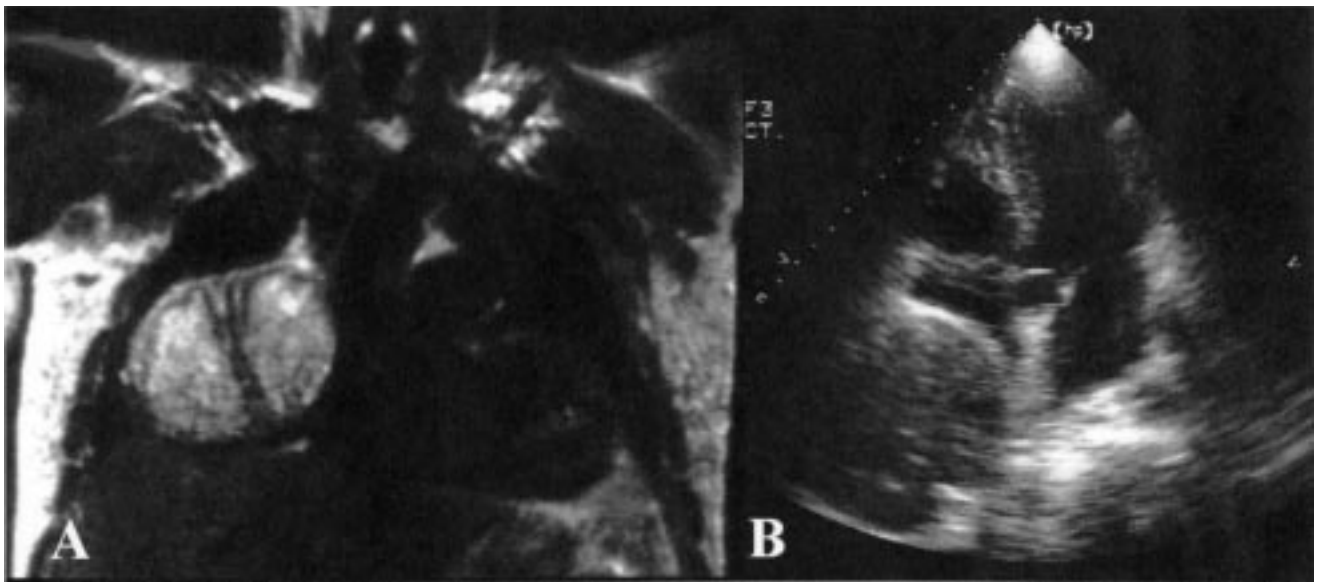


Fig. 2.

A: An approximately 10×7.6 cm smoothly marginated mass was detected by chest MRI in the coronal plane. The contents of the mass demonstrated heterogeneously high signal intensity on T1-weighted image.

B: Echocardiogram showed a 9.2×8.5 cm tumor pressing against the right atrium.

3A). Histopathological examinations showed markedly thickened capsular connective tissue with occasional foci of lymphocytes. Hassall corpuscles were also focally noted (Fig. 3B). Hemorrhage and necrotic material were seen in the lesion without any evidence of malignancy. The diagnosis was of ectopic cystic thymoma.

The patient was relieved of Raynaud's phenomenon

by the third post-operative day and serological anti-nuclear factor value rapidly normalized. There has been no sign of recurrence for 1 year after surgery.

Discussion

Mediastinal tumors represent a wide variety of disease

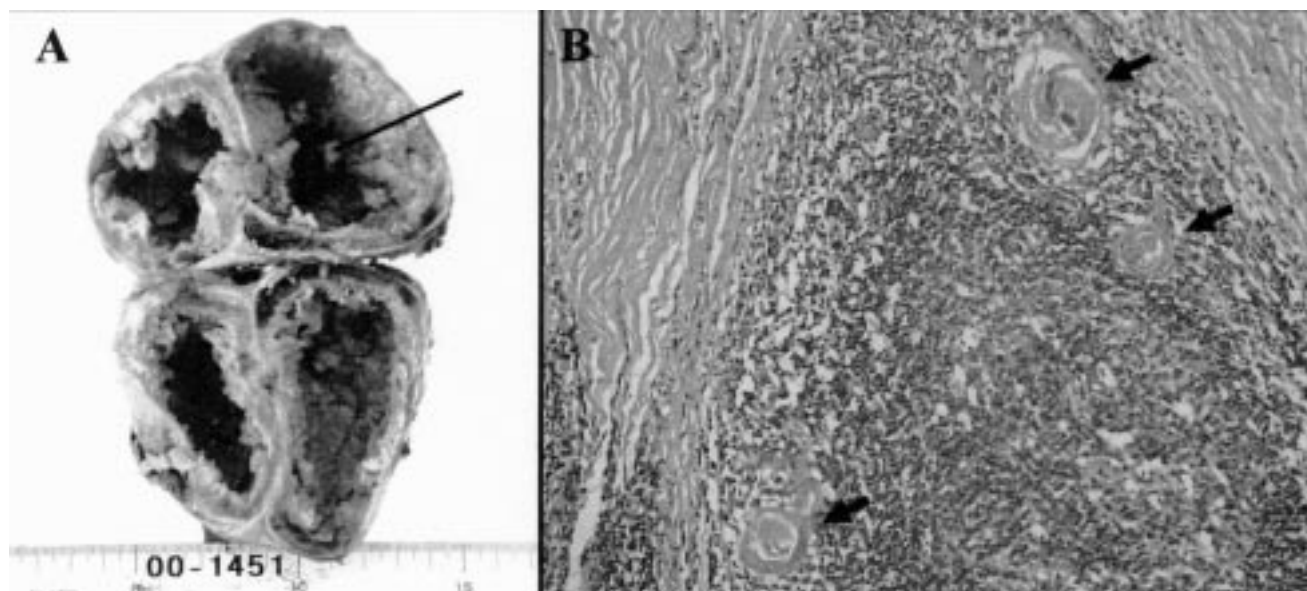


Fig. 3.
A: Cut surface of the resected specimen showed bilocular cysts containing a yellowish fat-like substance. Walls of the cyst were focally calcified and partially growing inward toward the cavity. One representative section (bar) was obtained for histopathological examinations.
B: Microscopic image of the section demonstrated markedly thickened capsular connective tissue with occasional foci of lymphocytes and Hassall corpuscles (arrows). There was no evidence of malignancy.

states. Information about the location and content of the mass is useful for differential diagnosis, and pathological examination is almost always required.¹⁾ We encountered a case of cystic anterior mediastinal tumor. Pure cystic masses are generally benign, but degeneration of the cystic masses sometimes alter their original appearances, which may promote misdiagnosis.²⁾

In our case, a well-margined spherical mass at the right cardiophrenic angle seen on chest radiogram was initially suspected to be a pericardial cyst. An echocardiogram was also suggestive of the diagnosis. However, the tumor lacked the typical characteristics of a pericardial cyst, such as epithelial lining of the cavity wall and water-like fluid contents.³⁾

Thymoma is the most common primary tumor in the anterior mediastinum. It is characterized by predominantly cystic formation and is called cystic thymoma, which should be discerned from non-neoplastic congenital thymic cyst.⁴⁾ Suster et al. reported that the features of cystic thymoma were: solid expanses within the cyst walls; perivascular spaces and foci of medullary differentiation; absence of an epithelial lining of the cysts.⁵⁾ At first, it was difficult for us to make an accurate diagnosis on this case because of the extensive necrotic lesions. However,

we managed to find thymus-like tissues including Hassall corpuscles in the cyst walls. The lack of both epithelial lining in the cyst walls and typical water-attenuation masses with imperceptible walls suggested that thymic cyst could be excluded from the differential diagnosis.

Thymoma is usually found in the normal location of the thymus in the anterosuperior mediastinum. However, ectopic genesis of thymoma has been rarely reported at many other sites including pericardium, associated with thymic rests in these tissues.⁶⁾ The thymic gland is embryologically derived from the third, and to a lesser extent, the fourth pharyngeal pouches. Congenital thymic cysts arise from embryonic tissue of the thymopharyngeal duct, and are found anywhere along the embryologic course of the thymic gland. The third and fourth pharyngeal pouches are located adjacent to the pericardial sac, and sometimes thymic remnants may migrate to the middle and lower mediastinum, where pericardial cysts are usually found.⁷⁾

Almost 40% of patients with thymoma seem to have an autoimmune- paraneoplastic syndrome.⁷⁾ Myasthenia gravis is the most common of these syndromes. In this case, Raynaud's phenomenon disappeared with a normalization of serological anti-nuclear factor value after total

removal of the tumor. We believe the symptoms could be explained by parathymic syndrome, although any truly causative link of thymoma to Raynaud's phenomenon is difficult to confirm.⁸⁾

We have reported on a case of ectopic cystic thymoma simulating a pericardial cyst and associated with Raynaud's phenomenon. Anterior mediastinal neoplasms constitute a varied group of tumors and are occasionally visible as cystic lesions. They sometimes have similar radiological image, which may mislead regarding the differential diagnosis. However, the clinical history and pathological examinations in addition to the imaging appearances facilitate accurate diagnosis and adequate treatment for these patients.

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