Inflammatory Myofibroblastic Tumor Mimicking Invasive Thymoma: a case report

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Inflammatory myofibroblastic tumor is an unusual disease that is most commonly seen in the lung and orbit and extremely rare in the mediastinum. As far as we know, there have been only two case reports of mediastinal inflammatory myofibroblastic tumor occurring in patients over 40 years old. In literature, we found no such case of mediastinal inflammatory myofibroblastic tumor that can present as invasive thymoma on the basis of the image pattern. Here, we report 57-year-old woman with anterior mediastinal inflammatory myofibroblastic tumor, which shows local invasion to the pericardium and central necrosis on chest CT scan. These uncommon image patterns have not been documented in literature and present diagnostic challenges.

CASE REPORT

The 57-year-old women in her usual health status came to our outpatient department presenting with left chest pain for two months. There was no fever nor abnormal breath sound on physical examination. However, her chest plain film showed a bulging soft tissue mass in the left hilar region (Fig. 1). So she was admitted for further evaluation.

The blood test revealed normal finding except high platelet level (522000/µL). CT scan was then performed (Fig. 2a, 2b). The tumor had no calcification and was located at the anterior mediastinum on non-enhanced image. After contrast administration, this tumor showed moderate homogenous enhancement with central necrosis. In addition, this tumor was tightly adjacent to the pericardium associated with pericardial effusion. Left-sided pleural effusion was also seen. The above imaging findings were highly suggestive of invasive thymoma with possible pericardium invasion. Other differential diagnoses include lymphoma, Castleman disease and other mesenchymal tumor of mediastinum.

The patient had sternotomy and pericardiectomy for tumor excision. A huge soft and yellowish mass about 9.5 × 6 × 3cm was found at anterior medias-
tinum adhesive to peripheral tissue (Fig. 3). There was also 100c.c. yellowish pericardial effusion and 200c.c. bloody pleural effusion. The tumor easily bleeds during operation. The pathologic findings showed fibrovascular tissue heavily infiltrated with a mixture of acute and chronic inflammatory cells on hematoxylin and eosin stain (HE stain) (Fig. 4a). Inflammatory cells including plasma cells, neutrophils, lymphocytes and histiocytes were confirmed by CD68 immunostain (Fig. 4b). Spindle cell proliferation and scattered large mononuclear and multinucleated cells were observed. Some were positive for smooth muscle actin and all were negative for ALK, CD30, CD15 and S-100 protein (Fig. 4c, 4d). The above findings were suggestive of inflammatory myofibroblastic tumor. The pericardium showed only reactive change microscopically. The patient has been well without tumor recurrence or chest pain one year after surgery.

**DISCUSSION**

Inflammatory myofibroblastic tumor is an uncommon, quasineoplastic lesion that was first observed in the lung and described by Brunn in 1939, and was named as inflammatory pseudotumor by Umiker et al in 1954 [1, 3]. It has been described by many other different names such as xanthomatous pseudotumor or plasma cell granuloma (especially found in the heart) because of its complexity and variable histologic characteristics and behavior [1].
To the best of our knowledge, inflammatory myofibroblastic tumor most commonly involves the lung and the orbit, and it usually occurs in children and young adult [1, 2, 5]. Besides, it is also the most common primary tumor of the lung in childhood [4]. Most of the patients are asymptomatic, but some may seek medical help because of cough, dyspnea, chest pain, or hemoptysis. Also, some of the patients may have an elevated erythrocyte sedimentation rate (ESR), mild anemia, or thrombocytosis [5]. The cause of inflammatory myofibroblastic tumor is still unknown [1, 6].

The histologic characteristics may vary from benign to malignant. And three main histologic patterns have been typically described: 1. Scarce, loosely arrayed spindle cells with a myxoid, vascular, and inflammatory proliferation resembling granulation tissue or nodular fasciitis. 2. Abundant, compact spindle cells with inflammation resembling fibrous histiocytoma or fibromatosis. 3. Sparse,

Figure 3. The post-operative finding shows a 9.5 × 6 × 3cm mass at anterior mediastinum with adhesion to peripheral tissue.

Figure 4. a. Histopathologic findings show fibrovascular tissue heavily infiltrated with a mixture of acute and chronic inflammatory cells. (HE stain 200X) b. Inflammatory cells including plasma cells, neutrophils, lymphocytes and histiocytes were confirmed (CD68 100X) c. Some of the spindle cells are positive for smooth muscle actin (Actin 100X) d. ALK immunostain showed all negative. CD30, CD15 and S-100 protein (not shown) also showed all negative.
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dense plate-like collagen with entrapped inflammatory cells resembling a desmoids tumor or scar [5, 6].

Though lung and orbit are the most common organs involved in this disease, almost every site in the body has been reported to have been affected. Initial image examination may occasionally show pulmonary inflammatory myofibroblastic tumor with mediastinum invasion [3]. In fact, the mediastinum is rarely a primary location of inflammatory myofibroblastic tumor, with the occurrence rate of 1.1-5.5% in several large studies [2, 5]. After reviewing previous studies, we can find only two cases of mediastinal inflammatory myofibroblastic tumor that occurred in patients over 40 years old [2]. Although there is a case report that inflammatory myofibroblastic tumor in the mediastinum presenting as superior vena cava syndrome on CT image and the other case report about pulmonary inflammatory myofibroblastic tumor with mediastinal invasion mimicking thymoma, there has been as yet no report that primary anterior mediastinal inflammatory myofibroblastic tumor can present itself as an invasive thymoma on image study [1, 3, 7, 8].

There are many differential diagnoses of anterior mediastinal tumor, such as thymic tumor, lymphoma, Castleman disease, germ cell tumor, or intrathoracic thyroid goiter. According to previous study [9], thymic tumors such as invasive thymoma or thymic carcinoma often show more heterogeneous enhancement on CT image study. Some may show pleural or pericardial implants or invasion. As anterior mediastinal lymphoma, we may see more prominent mediastinal lymphadenopathy. The main tumor may demonstrate homogenous attenuation with little if any enhancement. In addition, the hallmark of Castleman disease is strongly enhancing lymph nodes. As compared with our case, this tumor has no significant image character and is similar to invasive thymoma in imaging characteristics due to highly suspicion of pericardium invasion. But other mesenchymal tumor of mediastinum may also have the same image presentation. It is very difficult to make a definite diagnosis on the basis of CT imaging findings. In such case, surgical approach and pathologic diagnosis play very important roles.

Here, we report a case of anterior mediastinal inflammatory myofibroblastic tumor in a 57-year-old female patient, who showed an uncommon image pattern mimicking an invasive thymoma. This image presentation has not been described previously. As compared with previous study and our case, we could know that mediastinal inflammatory myofibroblastic tumor can present as superior vena cava syndrome, pericardium invasion, or tumor necrosis [1, 3, 5, 7, 8]. But these image findings are not specific so as to facilitate a definite diagnosis. Further surgical and pathologic diagnoses are very important for such a case.

**REFERENCES**

炎性肌纖維性腫瘤極相似侵犯性胸腺瘤：病例報告

黃維迪 許清寅 楊斐適

馬偕紀念醫院 放射線科

炎性肌纖維性腫瘤是一種少見的腫瘤，最常好發於肺部與眼框，卻極少出現在縱膈腔。就目前為止，超過四十歲的病人合併有縱膈腔炎性肌纖維性腫瘤只有兩個病例報告。而文獻記載中並沒有任何紀錄關於炎性肌纖維性腫瘤可在影像上相似侵犯性胸腺瘤。我們報告一個五十七歲女性在前縱膈腔長出炎性肌纖維性腫瘤，並表現出心包膜侵犯，中心性壞死等類似侵犯性胸腺瘤的少見影像表現。進一步的確定診斷需要手術與病理報告證據。