

Case Report

A Rare Case of Inflammatory Myofibroblastic Tumor of the Diaphragmatic Parietal Pleura with Dissemination

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Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm that occurs at different sites in the body. Pleural IMT in particular is especially rare. IMTs infrequently tend to have malignancy. We report a rare case of advanced diaphragmatic parietal pleural IMT with dissemination. A 30-year-old woman complained of right upper abdominal pain. Computed tomography showed a large lobulated mass over the right diaphragm, but no disseminated nodules were noted. Intraoperatively, we found the primary tumor arising from the diaphragmatic parietal pleura and a dozen disseminated nodules, and we removed them completely. The histopathological and immunohistochemical diagnosis was IMT.

Key words: inflammatory myofibroblastic tumor, pleura, dissemination

Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm that has been reported to occur in nearly every site in the body, including the lung, bladder, spleen, breast, pancreas, liver, colon, prostate, peripheral nerves, soft tissue and orbit. In the chest site, IMT commonly arises in the lung nodules. Pulmonary IMTs were reported to occupy 0.04% of resected lung nodules [1, 2]. A pleural IMT of the diaphragm or chest wall is a rare clinical entity compared to pulmonary IMT [3-5]. IMT has been considered a primary immunologic lesion, fibrogenetic disorder, or a specific reaction to infectious agents, trauma, or adjacent necrosis or neoplasm [2]. Chromosomal translocation involving the anaplastic lymphoma kinase (ALK) gene was present in 50% of IMTs, and approximately half of IMTs may be malignant [6]. Histologically, IMT belongs to a subgroup

in the broad category of inflammatory pseudotumors and consists of a variable mixture of collagen, inflammatory cells and usually cytologically bland spindle cells showing myofibroblastic differentiation [7]. IMT is an intermediate malignant soft tissue tumor because it can be infrequently confused with a malignant tumor to invasive locally, recur or metastasize [3][8]. Here we report a rare case of advanced diaphragmatic parietal pleural IMT with dissemination.

Case Report

A 30-year-old woman complained of right upper abdominal pain and sought medical attention at a nearby outpatient clinic. Chest X-ray showed blunting of the right costophrenic angle, and abdominal computed tomography (CT) revealed a 7.8 cm well-defined low attenuation lesion over the right diaphragm. She

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was subsequently referred to our hospital.

Her past history included appendectomy at the age of 14. Her family history was unremarkable. She had been smoking half a pack of cigarettes per day for 14 years upon visiting our hospital.

She had experienced pain in the right upper abdomen when coughing over the last few months. No



Fig. 1 Contrast-enhanced computed tomography showed a 7.8×3 cm heterogeneous lobulated mass over the right diaphragm. It showed early phase heterogeneous and delayed enhancement.

other abnormalities were observed on a chest and abdominal physical examination. Her complete blood count was within the normal range. Her levels of blood urea nitrogen, creatinine, liver function, C-reactive protein (CRP) and electrolytes were within normal limits.

Contrast-enhanced CT showed a 7.8×3 cm heterogeneous lobulated mass over the right diaphragm with early phase heterogeneous and delayed enhancement (Fig. 1). Very slight pleural effusion but no disseminated nodules were noted. The maximum standardized uptake value on fluorodeoxyglucose positron emission tomography in the tumor lesion was 2.6. In the tumor, T1-weighted magnetic resonance imaging (MRI) showed isointense lesions, and T2-weighted MRI showed mixed isointense and mild hypointense lesions. We preoperatively diagnosed solitary fibrous tumor (SFT) arising from the pleura of the diaphragm or chest wall.

The patient underwent video-assisted thoracic surgery for the purpose of diagnosis and treatment. Intraoperatively, the primary tumor, with a stalk, was seen arising from the diaphragmatic parietal pleura and was flexible on the diaphragm without invasion. A dozen disseminated 3–15 mm nodules were found nearby on the parietal pleura of the chest wall and diaphragm, but no pleural effusion was observed (Fig. 2). The primary tumor and all visible disseminated nodules were completely removed.

The histological findings on hematoxylin and eosin

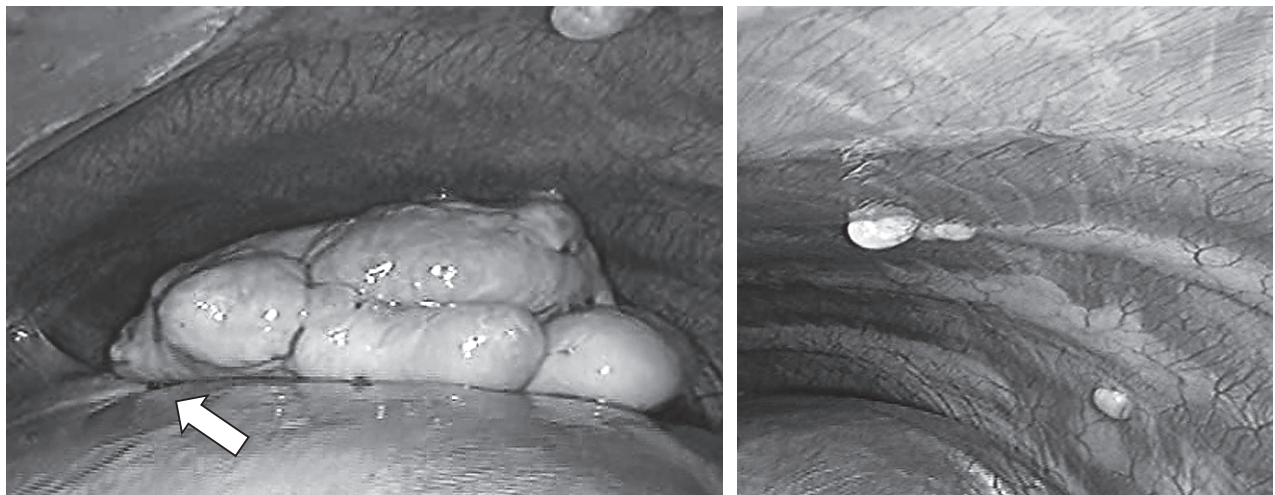


Fig. 2 The primary tumor arose from the diaphragmatic parietal pleura with a stalk (white arrow). Then, a dozen disseminated 3–15 mm nodules were found on nearby parietal pleura on the chest wall and diaphragm.

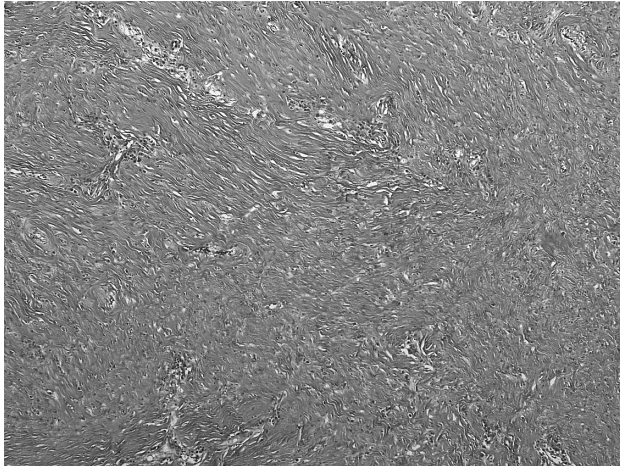


Fig. 3 The hematoxylin and eosin histology suggested the proliferation of spindle cells (fibroblasts and myofibroblasts) and monomorphic cells (macrophages, lymphocytes and plasma cells).

staining suggested proliferation of spindle cells (fibroblasts and myofibroblasts) and round cells (macrophages, lymphocytes and plasma cells) (Fig. 3). However, no evidence of malignancy, nuclear pleomorphism, or necrosis was found. Occasional mitoses, with 2 mitoses/50 high-power fields, were observed. In immunohistochemistry, the tumor was negative for cytokeratin AE1/3, CD117, smooth muscle cell actin (SMA) and S100. It was positive for calponin and HHF-35, and slight staining for desmin and CD34 was seen. The percentage of MIB-1 staining in the tumor was less than 1%, indicating low proliferative capacity. ALK was negative. There were no signs of a specific infectious disease such as tuberculosis. We finally diagnosed IMT after the histopathological and immunohistochemical analyses. The primary tumor arising from the diaphragmatic parietal pleura and the disseminated nodules showed the same pathological results.

We did not perform adjuvant chemotherapy or radiotherapy. The patient has remained asymptomatic and free from recurrence for 1 year after the operation.

Discussion

Previous studies have reported that most IMTs manifest as solitary masses in the lung parenchyma [1, 2]. In the chest, IMTs arising from extrapulmonary sites such as the pleura, diaphragm and chest

wall are rare and have appeared in only a few reports [3-5]. IMT can occur commonly in patients under 40 years of age, and our patient was young, at age 30 [2]. The findings of clinical examination and laboratory tests tend to be nonspecific or unremarkable [5]. The nonspecific imaging characteristics of IMT make it difficult to reach a preoperative diagnosis [9, 10]. We made a preoperative diagnosis of SFT based on the imaging findings and the frequency of occurrence in the chest wall or in pleural tumors.

Histologically, IMT is a distinctive neoplasm characterized by the presence of myofibroblasts and chronic inflammatory cells; the histopathological findings in this case were consistent with IMT [7]. In immunohistochemistry, positive results for SMA (86%) were frequently seen in a previous report [11]. The tumor was negative for SMA in the present case, but it was positive for calponin, HHF-35 and desmin, which were myofibroblast markers same as SMA. According to these results and other negative stains such as cytokeratin AE1/3, CD117 and S100, the pathological diagnosis was IMT.

Most IMTs manifest as solitary masses with discrete margins and have good prognoses if completely resected, because recurrence after complete resection is rare [3]. However, patients with incompletely resected or recurrent IMTs have been reported to have poor prognoses, because no effective treatment for this disease has been established [3]. In this case, the primary tumor and all visible disseminated nodules were completely removed, but visually unidentified disseminated nodules might have been missed. We did not perform adjuvant chemotherapy or radiotherapy because they have not been established as efficient treatments for IMT [5]. Anticancer drug combinations of paclitaxel and carboplatin or celecoxib and methotrexate have been reported to be helpful for advanced IMTs that are unresectable, show invasiveness, or have metastasized [12, 13]. It has been reported that non-steroidal anti-inflammatory drugs and steroids are also therapeutic [14]. Crizotinib, an ALK inhibitor, has been effective in some cases where ALK is positive on immunohistochemistry [15]. However, the results of these treatments have been widely variable, ranging from ineffective to complete regression.

We reported here a rare case of advanced diaphragmatic parietal pleural IMT with dissemination.

The patient has remained asymptomatic and free from recurrence for one year after resection, but careful observation is required.

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