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CASE REPORT

Primary inflammatory myofibroblastic tumor of the diaphragm: Report of a case

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Abstract

We report a case of primary inflammatory myofibroblastic tumor of the diaphragm in a 64-year-old man. The patient was hospitalized for a computed tomography (CT)-detected large tissue mass at the left lower lung field. Complete tumor excision followed by pathological investigation was performed. Microscopically, the tumor showed staggered arrangements of spindle myoepithelial cells, lymphoblastic and eosinophil cells. Immunohistochemically, the proliferating spindle cells showed positive staining for smooth muscle actin, vimentin, CD68 and Desmin, but negative for cytokeratin, leukocyte common antigen, epithelial membrane antigen, and S-100. This is the first reported inflammatory myofibroblastic tumor of the diaphragm found in an adult. The postoperative course was uneventful and the patient had no recurrence 18 months after surgery.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a well-described disease and can occur in various organs and tissues such as the lung, skin, soft tissues, breast, gastrointestinal tract, pancreas, oral cavity, bone, and central nervous system.[1–4] Various synonyms like inflammatory pseudotumor, plasma cell granuloma, pseudosarcomatous myofibroblastic proliferation, plasma cell pseudotumor, inflammatory myofibrohistiocytic proliferation, atypical myofibroblastic tumor and post operative spindle nodule, are interchangeably used to describe this unusual pathological entity.[5] In the 2002 World Health Organization classification of soft tissue tumors, this neoplasm was renamed to IMT.[6]

Diaphragm IMT is extremely rare. Hoer *et al*. report the first case in a five-year-old boy.[7] Here we report the second case, but the first diaphragm IMT in an adult, based on clinical and imaging findings, tumorectomy, and histopathological examination.

Case report

A 64-year-old male complained of discomfort mainly under the xiphoid, for one month, accompanied by poor appetite, hiccup, nausea and vomiting, and shortness of breath. There was no past history of exposure to tuberculosis. Physical examination revealed flatness of percussion in the axillary line and posterior axillary line of the left lower lung, but no obvious positive signs in the other regions.

A large tissue mass was observed, via chest radiograph, at the left lower lung field. Computed tomography (CT) demonstrated a large solid mass (9.4 cm × 7.6 cm) on the left chest wall that broke into the thoracic cavity (Fig. 1). After posterolateral thoracotomy, the tumor was found partially projecting into the abdominal cavity and adhering to the left lower lobe of the lung. The tumor was completely excised and parts of the diaphragm and left lower lobe of the lung were resected.

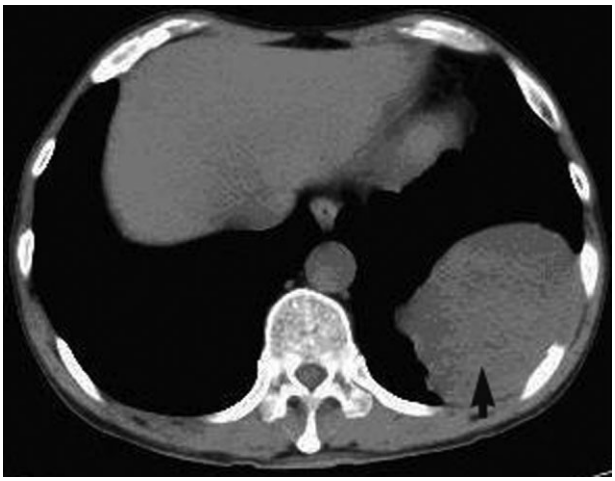

Figure 1 Computed tomography (CT) showed that a large solid-tissue mass had broken into the thoracic cavity.

By gross examination, the tumor was gray-pink with an irregular border and smooth capsule and 10.5 cm × 6 cm × 8 cm in size (Fig. 2a). The inner side of the tumor was gray-yellow in color without any necrosis (Fig. 2a). Microscopically, the tumor showed staggered arrangements of spindle myoepithelial cells, lymphoblastic and eosinophil cells (Fig. 2b). By immunohistochemistry, the proliferating spindle cells showed positive staining for smooth muscle actin (SMA, Fig 2c), vimentin (Vim, Fig 2d), CD68 (Fig. 2e) and Desmin (Fig 2f), but negative for cytokeratin (CK), leukocyte common antigen (LCA), epithelial membrane antigen (EMA) and S-100. Pathological diagnosis was inflammatory myofibroblastoma of the left diaphragm (invading partial lung tissue). The patient recovered without complications and was discharged and did not experience recurrence during the 18 months of follow-up.

Discussion

IMT has been increasingly reported involving various organs, however, diaphragm involvement of IMT is still rare. Although most reports suggest an inflammatory pathogenesis with an exaggerated response to tissue damage and subsequent pseudotumor formation, there are reports suggesting a true neoplastic nature with clonal chromosomal changes in chromosome.[8] Almost 50% of IMTs demonstrate anaplastic lymphoma kinase (ALK) rearrangements by fluorescence in situ hybridization (FISH), and several fusion partners have been identified, including TPM3 at 1p23, ATIC at 2q35, CLTC at 17q23, CARS at 11p15, RANBP2 at 2q13 and SEC31L1 at 4q21. The TMP3-ALK, ATIC-ALK and CLTC-ALK have been reported in IMTs which represent translocation-derived of chimeric tyrosine kinases. When the RANBP2-ALK gene fusion type was established, the tumor demonstrated clinical aggressiveness, in comparison to other gene fusion types.

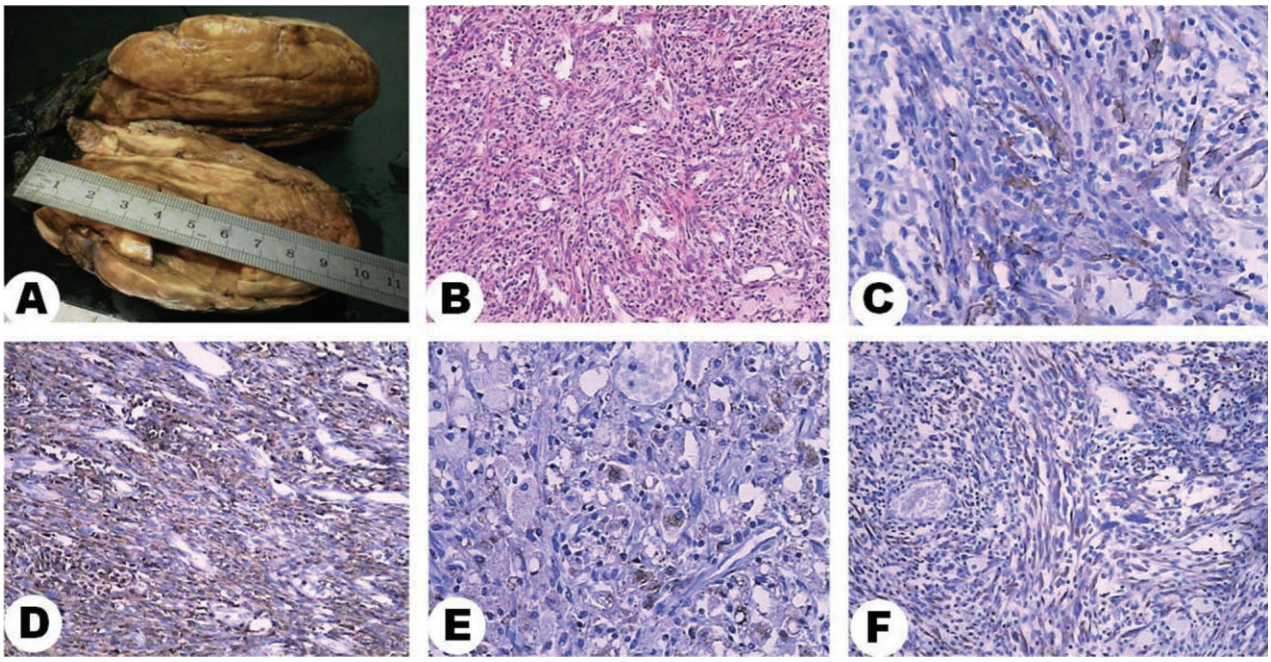

Figure 2 (**a**) The tumor measured 10.5 cm × 6 cm × 8 cm. (**b**) Hematoxylin–eosin (HE) staining of the spindle-cell tumor with admixed inflammatory cells (×100). Immunohistochemical stains showed that the proliferating spindle cells expressed (**c**) smooth muscle actin (SMA), (**d**) vimentin (Vim), (**e**) Desmin, (**f**) CD68.

Patients with IMT present nonspecific clinical features including fever, weight loss, night sweats, pain, malaise or site specific symptoms.[9] Sometimes abnormal laboratory parameters can be found, such as elevated erythrocyte sedimentation rate, leukocytosis, thrombocytosis, microcytic anemia, and/or polyclonal hypergammaglobulinaemia.[10] Our case indicates that diaphragm IMT is likely to present some nonspecific respiratory symptoms, such as shortness of breath and dyspnea, and digestive symptoms, such as poor appetite, nausea and vomiting, which are different from IMT in other sites.

The presenting symptoms of diaphragm IMT are similar to that of other primary tumors of the diaphragm (e.g. diaphragmatic cysts, lipomas, rhabdomyosarcoma, and fibrosarcoma). Therefore, it can be difficult to clinically, radiologically, and intraoperatively, differentiate IMT from other primary tumors of the diaphragm.[9] The diagnosis of IMT is based on the histological features of spindle myoepithelial cell proliferation, lymphocytic, and inflammatory infiltration, after exclusion of any causes, such as fungi and bacteria infection and trauma. Some immune-markers, such as ALK vimentin, actin, and CD68, are positive in 25% of cases.[4] ALK expression is usually found in younger patients. There are implications, but no clear-cut relationship has been established between ALK expression and prognosis. In our case, the tumor was positive for Vim, actin, CD68, SMA, Des and CD34, but negative for CK, LCA, EMA and S-100.

Although various non surgical treatment options have been proposed for IMTs, including systemic corticosteroid, immunosuppressive drugs, antibiotics, and radiation, with variable success, and some spontaneous regression has been reported,[11–13] excision remains the optimal measure in curing IMT because of the emergent requirement of relieving the mass effect; it is generally curative when total removal is performed.[14] In our case, the whole mass, including the invaded lung and diaphragm, were all resected.

In some sites, the behavior of IMTs are more aggressive, constituting a true neoplastic process because of the potential for local recurrence, development of multifocal tumors, infiltrative local growth, and even distant metastasis.[15] Therefore, postoperative follow-up of patients is critical. The recurrence rate varies depending by the anatomical site, from 2% for tumors confined to the lung, to 25% for extrapulmonary lesions. Moreover, distant metastasis of IMT is rare, occurring in <5% of cases reported. Extrapulmonary IMTs seem to exhibit a more favorable clinical course and treatment outcome, evolving with lower rates of recurrence, malignant transformation, metastasis, and mortality.[16] Hoer *et al.* considered the prognosis of IMT after adequate and complete removal as excellent in the first case of IMT of the diaphragm.[7] The final histologic diagnosis of our case was IMT invading partial lung tissue. The respiratory and digestive symptoms were relieved spontaneously after the excision of the tumor. There has been no evidence of tumor recurrence postoperatively during the 18 months of follow-up, without any adjuvant therapy.

Disclosure

No authors report any conflict of interest.

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