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Inflammatory myofibroblastic tumor in the mediastinum mimicking a malignant tumor

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT), also called "inflammatory pseudotumor", is a rare benign tumor composed of spindle cells with a variable infiltrate of inflammatory cells and fibrous tissue. There have been many reported cases of IMT in every organ system; however, IMT in the mediastinum is rare. We report a rare, proven case of spontaneous regression of IMT in the right cardiophrenic angle. The diagnosis was confirmed by histopathology, and the observation of serial ultrasonographic and computed tomography images over the course of three months revealed a picture consistent with spontaneous regression of the inflammatory pseudotumor.

Key words: • *inflammatory pseudotumor* • *spontaneous neoplasm regression* • *mediastinum*

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Inflammatory myofibroblastic tumor (IMT) is a benign lesion consisting of myofibroblastic spindle cells with an inflammatory infiltrate. IMT can develop at any location; however, its occurrence in the mediastinum is rare. We report an IMT in the mediastinum, for which the imaging results resemble those of an invasive malignant tumor arising from the mediastinal mesenchymal tissue. The final diagnosis was confirmed by histopathological findings of a ultrasonography-guided needle core biopsy. Findings on serial images by computed tomography (CT) over a four-month period were consistent with spontaneous regression of the IMT.

Case report

A 72-year-old man presented with upper abdominal discomfort and loss of appetite. On admission, his laboratory tests revealed a serum C-reactive protein concentration of 11.1 mg/dL; however, other laboratory data, including the serum white blood cell (WBC) count, were normal, and the patient was afebrile. Contrast-enhanced computed tomography (CT) revealed a heterogeneously enhanced mass with an irregular margin in the right cardiophrenic angle (Fig. 1). Ultrasonography (US) also showed a heterogeneously low-echoic structure (Fig. 2). On both CT and US images, the mass was adjacent to the right side of the pericardium and the superior aspect of the dome of the right hemidiaphragm. A small pericardial effusion was also noted. The initial diagnosis was a malignant tumor invading the pericardium and diaphragm which appeared to be inoperable.

A follow-up CT obtained two weeks after admission, however, showed a slight reduction in the volume of the mass (Fig. 3a). The patient was followed closely while receiving antibiotics during a one-month hospitalization, and was discharged thereafter. CT obtained one month after discharge (Fig. 3b) revealed an additional decrease in the volume of the mass. The mass was undetectable on a CT scan three months after discharge (Fig. 3c), thus suggesting a natural course of spontaneous regression of IMT over a four-month period.

US-guided biopsy was performed after the first follow-up CT, and histopathology disclosed a benign lesion composed of spindle cells with an inflammatory infiltrate consisting of lymphocytes, histiocytes, and plasma cells, and fibrous tissue, suggestive of IMT (Fig. 4).

Discussion

IMT is a rare benign entity of unknown etiology, consisting of a mass composed of spindle cells mixed with an infiltrate of variable numbers of inflammatory cells including lymphocytes, plasma cells, immunoblasts, and histiocytes, and fibrous tissue. The most frequently reported site of IMT is in the lung, while it occurs less commonly in the liver,

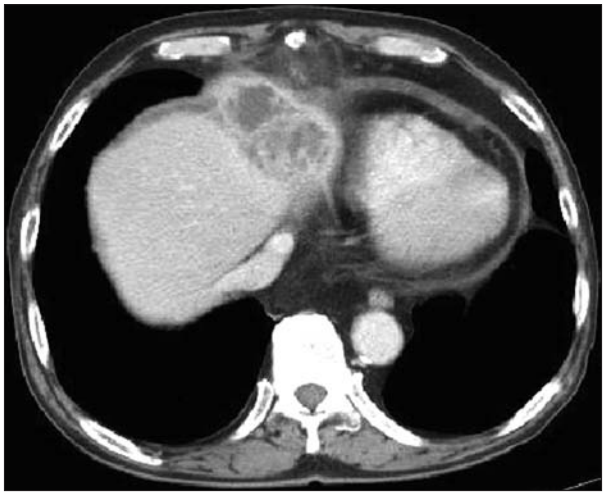


Figure 1. Contrast-enhanced axial CT image shows a heterogeneously enhanced mass with an irregular margin in the right cardiophrenic angle, immediately adjacent to the right aspect of the pericardium and the superior aspect of the dome of the right hemidiaphragm. It was thought to be an invasive malignancy. A small pericardial effusion is also seen.

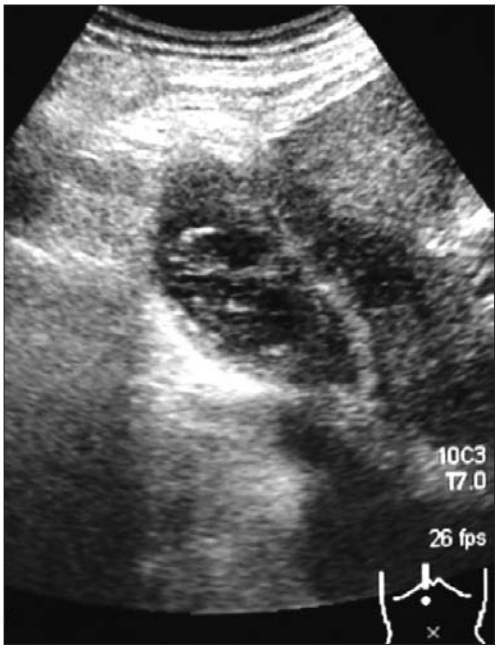
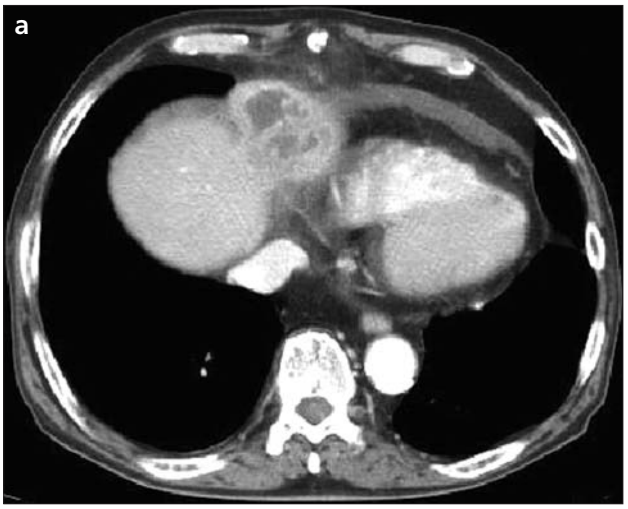
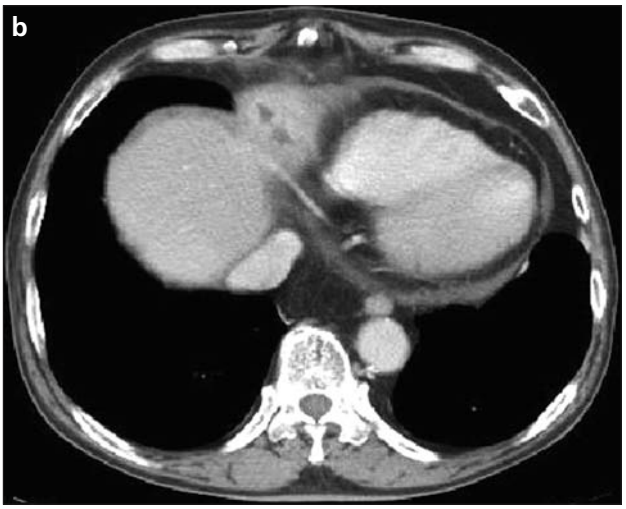


Figure 2. Longitudinal US image shows a heterogeneously hypoechoic mass lesion adherent to the diaphragm, superior to the liver.





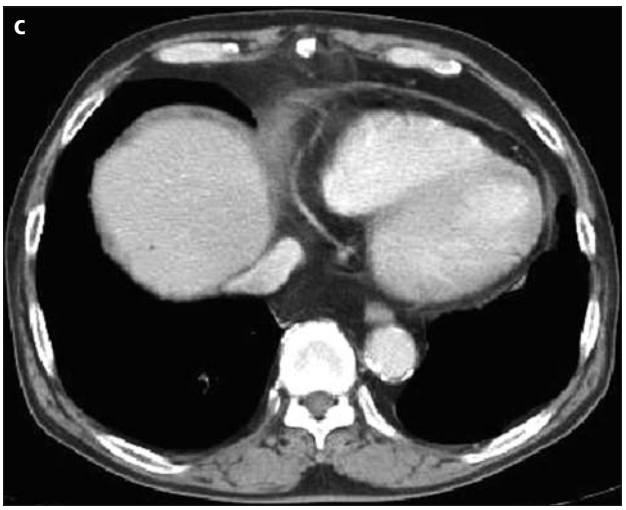


Figure 3. a–c. Contrast-enhanced axial CT two weeks after admission **(a)** demonstrates slightly reduced tumor volume. Contrast-enhanced axial CT one month after discharge **(b)** shows further decrease in tumor volume. On a follow-up contrast-enhanced axial CT three months after discharge **(c)**, the mass was not detectable.

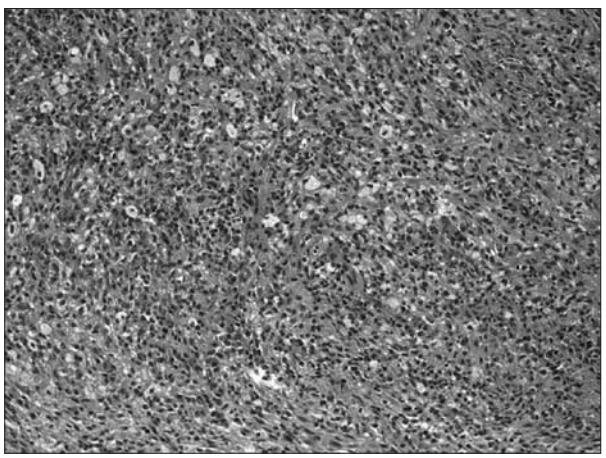


Figure 4. Photomicrograph demonstrating spindle cells and inflammatory cells including lymphocytes, histiocytes, and plasma cells, and a moderate amount of collagen, consistent with an inflammatory myofibroblastic tumor.

spleen, stomach, orbit, urinary bladder (1), and rarely, the mediastinum, which accounts for about ten cases to date (2–6) based upon a MEDLINE search from 1966 to 2006.

It remains difficult to distinguish IMT from a malignant tumor on the basis of imaging. As was seen in our case, CT images of IMT mimic those of an invasive malignant tumor with an infiltrative margin. The heterogeneous enhancement with contrast normally seen on a CT scan reflects the histopathology of the tumor, which is composed of spindle cells, a variable inflammatory infiltrate, and fibrous tissue. This enhancement is nonspecifically variable, depending on the balance between the cellular component and the fibrous tissue. In addition, an infiltrative peritumoral margin on imaging reflects the inflammatory characteristics of this unencapsulated tumor. As a result, in most cases, a definitive diagnosis is made based on the histopathological findings from either a resected tumor or a needle biopsy.

The clinical presentation of IMT may not be sufficient to make an accurate diagnosis. IMT is often accompanied by elevated serum C-reactive protein and/or an increased WBC count, reflecting the inflammatory characteristics of this tumor; however, in malignancy, a nonspecific inflammatory reaction is often suggested by laboratory tests.

According to our MEDLINE search, spontaneous regression of IMT has been reported sporadically in lung, liver, orbit, urinary bladder, and elsewhere. In addition, there have been various reports of spontaneous regression of malignant tumors (e.g., hepatocellular carcinoma, renal cell carcinoma, lung carcinoma). However, reduction in tumor size is often more reflective of the inflammatory characteristics rather than of the malignancy per se. The present case is of interest because the volume of the tumor was found to have decreased spontaneously and dramatically on serial CT scans.

In summary, radiologists should be aware of IMT in the mediastinum mimicking an invasive malignancy. In addition, when a tumor resembling malignancy is seen in any location with an inflammatory reaction, a careful evaluation of serial CT scans could be an important technique for making an accurate diagnosis. If a spontaneous but slight regression is noted, then IMT should be considered in the differential diagnosis. In order to avoid unnecessary invasive surgery, it is important for physicians to identify IMTs, which sometimes can regress spontaneously.

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