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CASE REPORT

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Inflammatory Myofibroblastic Tumor of the Lung: Unusual Imaging Findings of Three Cases

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Summary

Background: Inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor, is a benign disorder composed of fibrous tissues, myofibroblasts and inflammatory cell proliferation with obscure etiology. Although it is the most common lung tumor in children, it is seen rarely in adults constituting less than 1% of adult lung tumors.

Case Reports: In this report, we present different and rare CT manifestations of three adult patients with lung IMT.

Conclusions: In conclusion, IMT is a rare lung tumor in adults and may simulate malignancy. It should be considered in the differential diagnosis when a large mass with lobulated contour or harboring coarse calcification is observed.

MeSH Keywords: Lung Neoplasms • Multidetector Computed Tomography • Plasma Cell Granuloma, Pulmonary

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Backlground

Inflammatory myofibroblastic tumor (IMT) is a rare tumor with obscure etiopathogenesis in which different inflammatory cells and myofibroblastic spindle cells are seen histologically. First defined by Umiker and Iverson in 1954 [1,2], it is the most common lung tumor in children and adolescents. The condition is rare in adults constituting 0.7% of all adult lung tumors [3]. Clinical and radiological findings of IMT are generally nonspecific and differentiation from malignant lung tumors remains a challenge. Here, we reported different CT features of three adult patients with IMT.

Case Report

Case 1

A 69-year-old non-smoker female patient presented to our hospital with a one-month history of dyspnea, chest pain and dry cough. She denied weight loss or fever. Laboratory investigations revealed an elevated white blood count (15.4 K/uL, reference value 4–11 K/uL), elevated lactate dehydrogenase (388 U/L, reference value 135–215 U/L), high erythrocyte sedimentation rate (51 mm/hour) and increased C-reactive protein (7 U/mL, reference value 0–0.5 mg/dL). Physical examination was unremarkable. Chest x-ray revealed a lobulated peripheral mass in the right upper and mid lung zone. Contrast-enhanced chest CT showed a pleural-based lobulated mass in the anterior segment of the right upper lobe. The mass measured 4×3×3 cm, displayed central cavitation, and extended towards the right hilum along with the right upper lobe anterior segmental bronchus (Figure 1). Histological examination of core needle biopsy was consistent with an IMT with organizing pneumonia pattern. Wedge resection was performed, and there was no recurrence 9 months after surgery.

Case 2

A 52-year-old male patient was admitted to our institution with complaints of dyspnea and chest pain that were increasing with effort. His complaints had been present for the last 3 months and become more severe recently. The patient had a 20 pack-year smoking history. He denied weight loss or fever. Physical examination was unremarkable. Laboratory investigations revealed mildly elevated white blood count (11.3 K/uL, reference value 4–11 K/uL) and lactate dehydrogenase 258 U/L (reference value 135–215 U/L). Chest x-ray revealed high attenuation right paracardiac

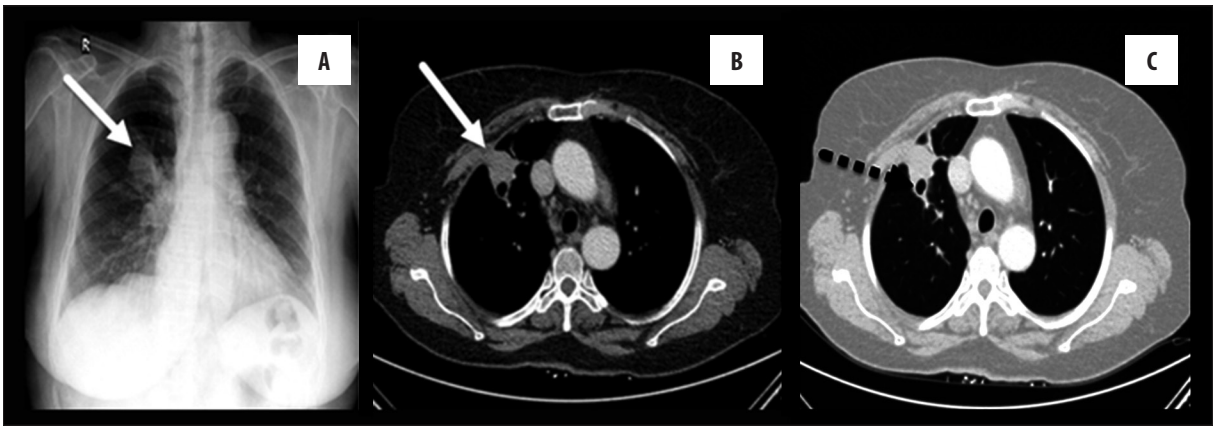

Figure 1. (**A**) Chest x-ray shows opacity with a lobulated contour in the upper mid zone (white arrow); (**B, C**) contrast enhanced axial CT scan of the thorax demonstrating a pleura-based lobulated mass (white arrow) in the anterior segment of the right upper lung lobe with central cavitation (black dashed arrow).

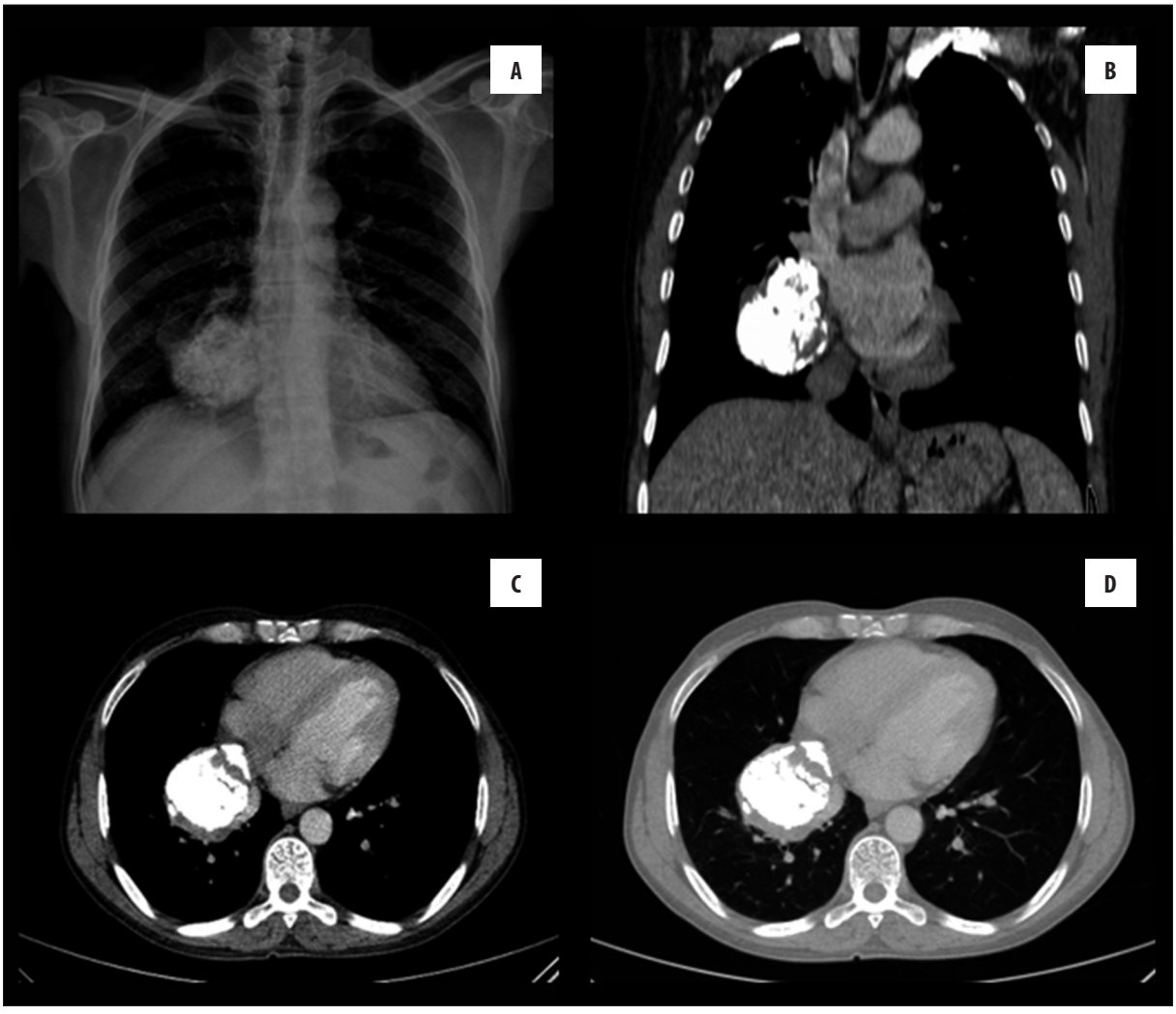

Figure 2. (**A**) Chest x-ray shows lobulated high density opacity in the paracardiac location of the mid zone of the right lung, (**B**) contrast enhanced coronal reformatted and (**C, D**) contrast enhanced axial CT scan of the thorax shows dense coarse inner calcifications leading to the right lower lobe from right hilum.

opacity with well-defined lobulated contours. Contrast-enhanced chest CT examination showed a 7×7×6-cm lesion with dense coarse inner calcifications located in the right inferior pulmonary ligament. The mass displaced the right inferior pulmonary vein backward without invasion into broncho-vascular structures (Figure 2). Right lower

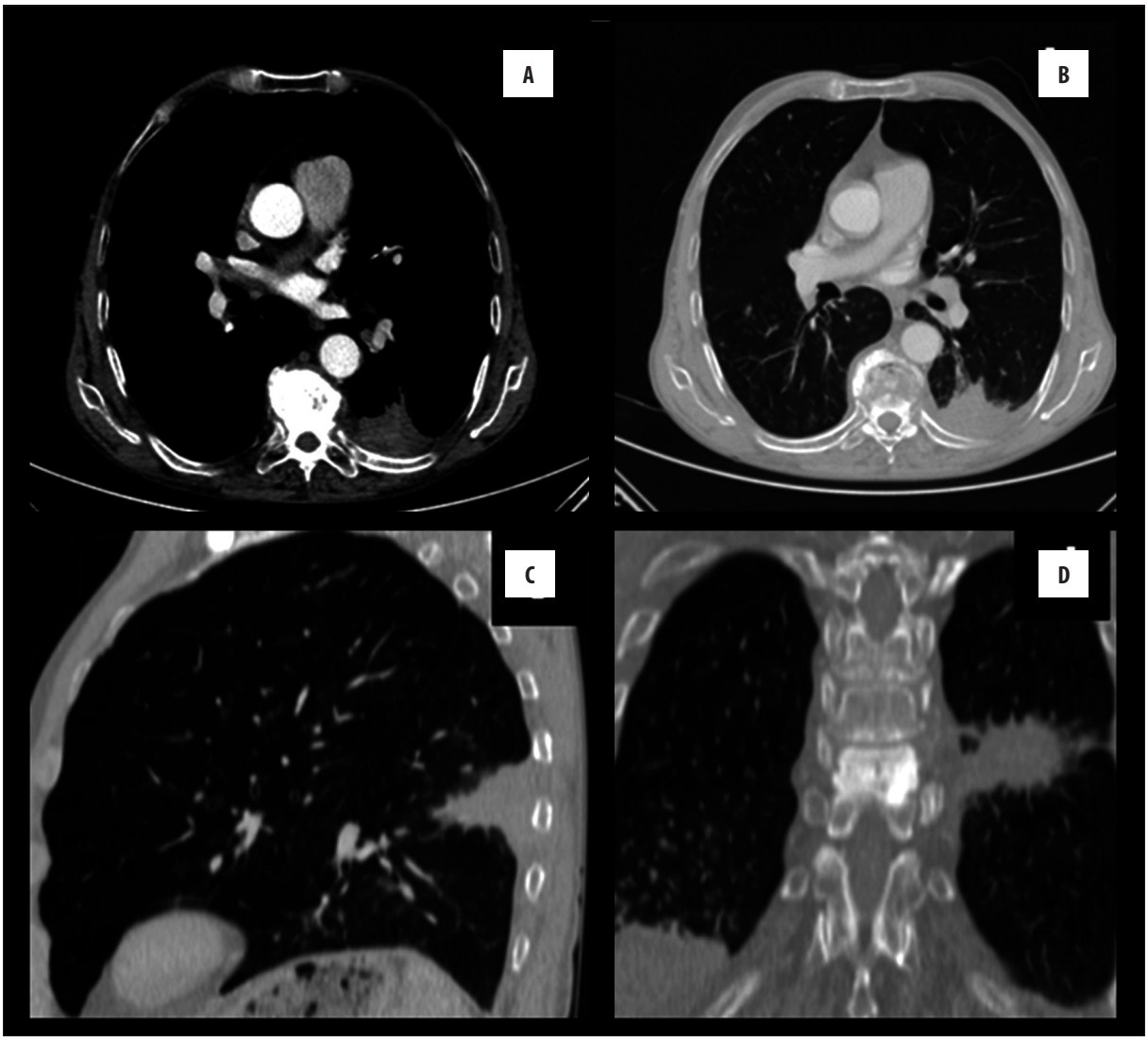

Figure 3. (**A, B**) Contrast enhanced axial CT scan of the thorax, (**C**) sagital reformatted and (**D**) coronal reformatted CT scan of the thorax shows pleura-based consolidated lesion with spiculated contours in the superior segment of the right lower lung lobe.

lobectomy was performed and IMT with a fibrous histiocytic pattern was diagnosed pathologically.

Case 3

A 62-year-old male suffering from chronic obstructive pulmonary disease (COPD) for 9 years developed an incidental mass on chest x-ray during a routine follow-up. The patient had a 24–pack-year smoking history. He denied weight loss, sputum or fever. His physical examination was unremarkable. Laboratory investigations revealed mildly elevated white blood count (11.3 K/uL, reference value 4–11 K/uL) and high erythrocyte sedimentation rate (88 mm/hour). Contrast-enhanced chest CT showed a 4×4×5-cm pleura-based consolidative lesion with spiculated contours in the superior segment of the right lower lung lobe. Subsegmental atelectasis was present around the lesion (Figure 3). Patient underwent a core needle biopsy to exclude malignancy, and the lesion was diagnosed as an IMT with organizing pneumonia pattern pathologically. After biopsy the patient was lost to further follow-up.

Discussion

Inflammatory myofibroblastic tumor is an idiopathic benign condition composed of fibrous tissues, myofibroblasts and inflammatory infiltration, predominantly histiocytes and plasma cells. They can involve many anatomic sites and have propensity to be locally aggressive and multifocal [4]. Due to the complexity and variable histological characteristics and behavior of the condition, numerous synonymous terms such as inflammatory pseudotumor, plasma cell granuloma, xanthogranuloma, fibrous histiocytoma, xanthoma and fibroxanthoma have been used to describe this entity. IMT can simulate a malignant process both clinically and radiologically.

IMT is the most common primary lung tumor in children under 16 years of age. Most of the reported cases are under 40 years of age [1]. Being a very rare lung tumor in adults, it constitutes only 0.7% of all lung tumors [5]. Its incidence was reported to be 0.04% [6]. All our cases were older than 50 years of age and the lesions mimicked malignancy in all cases.

Patients with pulmonary IMT are frequently asymptomatic as in our third case which was detected incidentally during a routine examination. Nonspecific symptoms such as dyspnea and weakness were reported rarely as in our first two cases. Other complaints include cough (10%), chest pain (11%), hemoptysis (9%), and fever (7%) [7]. Although most of the reported cases were male, the disease does not have gender predilection [8]. Laboratory tests are usually nonspecific, but increased levels of CRP and/or elevated erythrocyte sedimentation rate have been described in about 50% of IMT cases, as were the case in our patients.

Radiologically, IMT is frequently seen as a peripherally located solitary nodule or mass in the lower lobes. It may have lobulated contours and show varying degrees of heterogeneous enhancement on CT [6]. It can also occur in the hilum of the lungs and may contain focal calcification or rarely manifest as a consolidative mass. Although not a usual finding, cavitation may also be observed within a mass or consolidation [9-11]. Different forms of calcifications (e.g. peripheral, punctate or fine) are seen in 15% of IMTs. However, calcification is very rare in adult IMTs [12]. We observed three different manifestations of IMT, namely cavitation in case 1, densely calcified perihilar mass in case 2, and consolidative mass lesion in case 3. To the best of our knowledge, no previous case of entirely calcified IMT located in the inferior pulmonary ligament similar to case 2 in this report has been reported. Pulmonary IMTs may cause atelectasis due to bronchial compression, or they may rarely occur as multiple lesions [13]. On magnetic resonance imaging, IMTs were reported to have heterogeneous signal characteristics with a little higher signal than the muscle on T1- and T2-weighted images and show heterogeneous contrast enhancement as seen on CT [14].

The differential diagnosis of IMT includes primary lung cancer, metastasis, hamartoma, chondroma or pulmonary granuloma. Endobronchial lesions may also be confused with carcinoid tumors if extensive contrast enhancement is observed [1]. Although local invasive forms of IMT have been reported, complete resection is the recommended treatment method. Recurrence has been reported rarely in incomplete resection [8].

Inflammatory myofibroblastic tumor can be divided into three histologic types; organizing pneumonia, lymphohistiocytic or fibrous histiocytic pattern. Fibrous histiocytic pattern is the most common form, characterized by spindle-shaped myofibroblasts arranged in whorls and lymphohistiocytic pattern is the least common form characterized by mixture of lymphocytes and plasma cells. Organizing pneumonia pattern is characterized by airways filled with plump fibroblasts, histiocytes, mononuclear cells and fibroblasts [9]. We observed fibrous histiocytic pattern in our case 2, and organizing pneumonia pattern in case 1 and 3.

Conclusions

In conclusion, IMT is a rare lung tumor in adults and may simulate malignancy. Having no specific clinical or imaging findings, it should be considered in the differential diagnosis when a large mass with lobulated contour or harboring coarse calcification is observed. The presence of elevated acute phase reactants and lack of hilar or mediastinal lymphadenopathy strengthen the diagnosis. Surgical resection is the recommended treatment in lung IMTs with favorable prognosis in completely resected cases.

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