

Inflammatory pseudotumor of lung, a case report and review of literature

SB PATEL, DM SHAH, KG GOSWAMI, SR SHAH, PM SHAH

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Introduction:

Inflammatory pseudotumor of lung is a rare benign lung tumor of uncertain etiology, occurring more commonly in the younger patients. They are also known as 'plasma cell granuloma', 'histiocytoma', and 'fibroxanthoma' depending on the predominant cell type. Although benign, they have the capacity for local invasion, rapid growth and sarcomatous transformation. Some cases have shown regression either spontaneously or after corticosteroid therapy. This case is being reported because of its rarity and typical presentation.

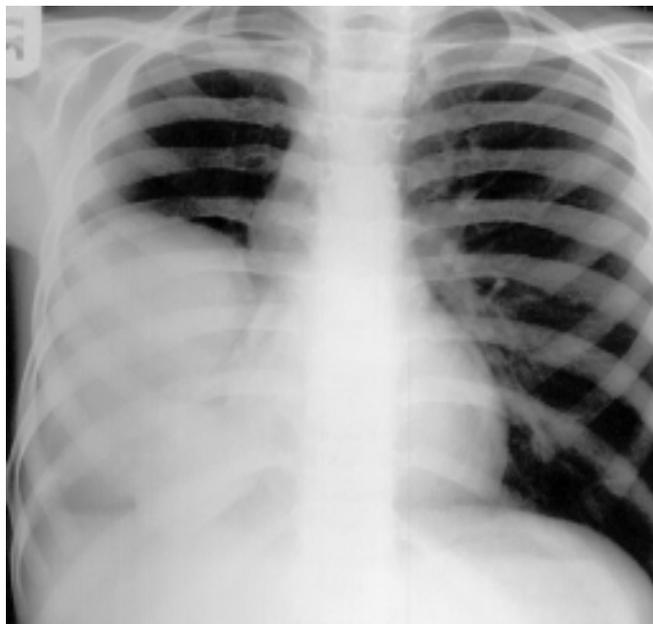


Figure 1. Chest roentgenogram postero-anterolateral view showing lobulated soft tissue mass without cavitation in right mid-lower zone.

Case Report:

A 14 years male presented with history of cough, common cold and repeated pneumonitis since last one year. Chest

AP and Lateral roentgenogram were taken outside (fig. 1 & 2). They showed presence of sharply marginated lobulated soft tissue opacity in right mid and lower zones without internal cavitation or internal air-bronchogram. Considering it to be a mass lesion, patient was advised CT scan thorax. CT scan thorax was also done outside. Noncontrast CT scan thorax showed presence of a large well defined sharply marginated lobulated solid hypodense lesion in right lower lobe (fig. 3). The lesion was abutting pleura. There was presence of internal calcification within the mass. No evidence of pleural effusion was seen. Contrast enhanced CT scan showed mild homogenous enhancement of the mass (fig 4). Considering it to be a mass lesion, patient was operated outside. Right pneumonectomy was done. Outside histopathology report showed no evidence of malignancy with presence of inflammatory cells. Then patient was referred to our institute along with all investigation reports and histopathology slides. All roentgenograms and CT scan thorax were reviewed. Histopathology slides were also reviewed and a diagnosis of inflammatory pseudotumor of lung was made. As final diagnosis came at our institute, and the disease itself is very rare, we like to report this case; even though patient was investigated and operated outside our institute.

Discussion:

The varying terminology used to describe the inflammatory pseudotumor of lung, like plasma cell granuloma, inflammatory myofibroblastic tumor, histiocytoma, fibroxanthoma, xanthogranuloma, plasma cell-histiocytoma complex, have caused a great deal of confusion in understanding the natural history of the disease. Umiker and Iverson preferred to include all these lesions under the designation of inflammatory pseudotumor since morphologically these simulate a tumor, histologically these are composed of inflammatory cells and show complete maturity of fibroblastic

From the Department of Radio-diagnosis, Gujarat Cancer and Research Institute, Asarwa, AHMEDABAD - 380016

Request for Reprints: Dr. Dipal M. Shah MD, DMRE, Junior Lecturer, Department of Radio-diagnosis, Gujarat Cancer and Research Institute, Asarwa, AHMEDABAD - 380016

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component with striking lack of mitosis.



Figure 2. Chest roentgenogram right lateral view showing lobulated soft tissue mass situated in lower zone, posteriorly.

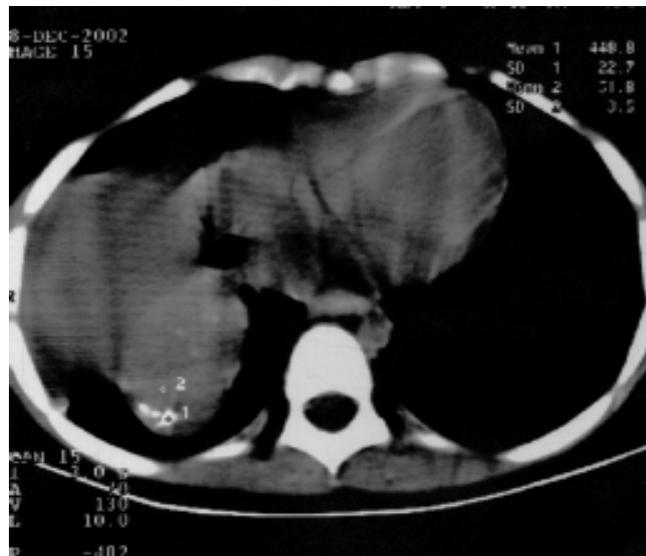


Figure 3. Noncontrast CT scan thorax showing lobulated hypodense soft tissue mass in right lower lobe with presence of internal calcification.

It is a non-neoplastic process characterized by unregulated growth of inflammatory cells. The cause of this inflammatory response remains unknown and various theories attribute it to a metabolic disturbance, organized cellular growth developing in association with pulmonary infection, viral origin or antigen-antibody interaction in relation to an agent, which is no longer identifiable (1).

The role of antibiotics has been speculated upon. However, neither bacteria nor fungi have been grown in tissue cultures in any of the resected specimens but a history of prior infection could be obtained in about one third of patients (1).



Figure 4. Contrast enhanced CT scan thorax showing mild homogenous enhancement of the lobulated mass in right lower lobe.

Grossly, they are round, well circumscribed but unencapsulated intrapulmonary masses. Microscopically they are circumscribed tumefactive lesions, composed of a variable mixture of collagen, inflammatory cells and benign mesenchymal cells including spindled myofibroblasts, fibroblasts, plasma cells, lymphocytes, foam cells, giant cells and macrophages.

Inflammatory pseudotumor of lung is benign tumor of childhood. The reported incidence in various reports varies from 0.04 % to 0.7 % of lung masses (1). Though usually reported in pediatric age group, they may occur at any age. It is the most common primary lung mass seen in children. There is no sex predilection (2).

The symptomatology of the tumor is generally in the form of cough, chest pain, dyspnoea and haemoptysis and is generally nonspecific. Fever and clubbing have been observed particularly in younger patients and generally disappear after resection of the lesion (1). The laboratory data are not of much help in diagnosis of inflammatory pseudotumor.

The most common radiographic presentation is that of a solitary well-circumscribed round or oval pulmonary mass (3). This is seen in over 50 % of patients. Approximately 25 % of patients will present with a coin lesion (4). The tumor size may range from 0.5 to 36 cm (5,6). The pulmonary mass can grow quite large, occupying entire lung.

Calcification is not infrequent in inflammatory pseudotumor of lung. The nature of calcification in inflammatory pseudotumor is most commonly described as mottled with calcific flecks (7) and is thought more often to be fine in nature rather than coarse. Dense calcification has been reported (8). Calcification in inflammatory pseudotumor is more common in children than in adults in cases reported in literature. Bahadori and Liebow reported 40 cases of inflammatory pseudotumor out of which 16 were children and 24 were adults (7). Calcification was present in 2 cases, both of them were children. Monzon et al; in reporting 28 children with inflammatory pseudotumor, described calcification in 4 children (6). Berardi and colleagues reviewed 181 of inflammatory pseudotumor and found 8 instances of calcification. Out of 8 cases, 5 cases of calcification occurred in children (5). Microscopic evidence of calcification in inflammatory pseudotumor is more commonly appreciated than radiographic evidence of calcification.

A small group of patients may present with ill-defined areas of pneumonic consolidation or atelectasis. In most cases, the lesion is usually intraparenchymal with secondary compression and entrapment of bronchi occurring with growth (4). Occasionally, endobronchial and endotracheal lesions may cause post obstructive pneumonia or atelectasis (9). Rarely, multiple lesions are present. Central cavitation is a very unusual feature. Hilar lymphadenopathy is rare and pleural effusion has not been reported to occur with this etiology (4). Inflammatory pseudotumor can manifest as a mediastinal mass (10). Aggressive features of inflammatory pseudotumor have been described, including vertebral destruction (11), vascular invasion (12), recurrence (13) and metastatic spread (14).

There is considerable diversity of appearance of inflammatory pseudotumor on CT in cases that have been studied so far. CT scan most commonly demonstrates a mass of heterogeneous attenuation. Variable enhancement patterns have been described on contrast enhanced CT scans, including nonenhancement, mild enhancement, heterogeneous enhancement and peripheral enhancement. Variable calcification patterns have been described on CT scans including punctate, dense, flocculent and curvilinear (2).

A study of MRI findings in five patients of inflammatory pseudotumor showed heterogeneous lesion with signal intensity slightly greater than skeletal muscles on T1-weighted images and high signal intensity on T2-weighted images. Diffuse heterogeneous enhancement of the lesion was noted on gadolinium-enhanced images (2).

The radiological differential diagnosis includes primary neoplasm, metastasis, hamartoma, chondroma,

sclerosing hemangioma, pulmonary granuloma. Endobronchial inflammatory pseudotumor may mimic carcinoid (2). Resection of the lesion is treatment of choice. However, non-surgical treatment like radiotherapy and steroids have been employed in the setting of incomplete surgical resection, tumor recurrence and patient being unfit for surgery (1).

In conclusion, inflammatory pseudotumor of lung is a relatively uncommon lesion, which is typically solitary, peripheral, sharply circumscribed, lobulated mass. Most patients are symptomatic at diagnosis and may have history of prior lower respiratory tract infection. The radiographic and CT appearance of inflammatory pseudotumor is quite variable and nonspecific. Therefore, when clinical presentation of pulmonary neoplasm or infection is atypical or problematic, it is important for radiologists to consider the diagnosis of inflammatory pseudotumor to be aware of the variety of its different radiographic and CT manifestations, and to proffer it as a diagnostic possibility.

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