

Inflammatory Myofibroblastic tumor of lung (Pseudotumor of the lung)

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Introduction

Inflammatory pseudotumor first observed in the lung and described by Brunn in 1939, was so named by Umiker et al in 1954 because of its propensity to clinically and radiologically mimic a malignant process (1,2). An inflammatory pseudo tumor (IPT), known as a plasma cell granuloma, is a relatively uncommon non-neoplastic tumor-like process with an unidentified etiology that usually occurs in children and young people. Inflammatory pseudo tumor is a rare disease in the adults (3,4). Patients with IPT are usually asymptomatic, with a solitary pulmonary nodule or mass detected on routine chest roentgenogram. IPT can behave as a malignant tumor both clinically and radiologically. Cough, fever, dyspnea, and hemoptysis are the usual presenting symptoms. We report a case of lung involvement with radiologic and pathologic correlations in an elderly patient who presented with cough with mucoid expectoration, chest pain, anorexia and malaise of 6 weeks duration. Initial radiological appearance with pathological correlation proved the presence of pseudo tumor in our patient who was then put on treatment. The lesion regressed on that side and developed recurrence on the opposite side. This makes this case interesting with bilateral pseudotumor with recurrence and good response to treatment in adult age group.

Case report:

75 year old lady was referred to chest outpatient department with complaints of cough with mucoid expectoration, left sided dull aching chest pain, anorexia and malaise of 6 weeks duration. She had no other underlying systemic disease or respiratory symptoms in the past. On examination patient was anaemic. Respiratory examination revealed an impaired percussion note in the left upper lobe area with crackles.

Frontal chest radiograph revealed a homogenous opacity with ill defined margins in the left upper lung fields (Figure 1). Computerized tomogram of the chest showed presence of a homogenous solid density mass lesion with defined borders in left upper lobe (Figure-2). Hematological examination showed hemoglobin of 9 gm%, leukocyte count of 22600 cells/ mm³ with predominant neutrophils (88%) and an ESR of 120 mm/ hr. Sputum samples for Gram stain and pyogenic culture, Ziehl-Neelsen smear staining for acid fast bacilli & three cytology specimens for malignant cells were negative. A CT guided FNA and biopsy was carried out. FNAC was inconclusive but biopsy showed proliferation of cellular fibrotic tissue with entrapped flattened alveolar spaces and focal collection of plasma cells and lymphocytes (Figure-3). At places the infiltrate was seen to diffusely involve the tissue with the fibrotic bands also infiltrating the skeletal muscle fibers (Figure-4). These features were suggestive of an inflammatory myofibroblastic tumour.

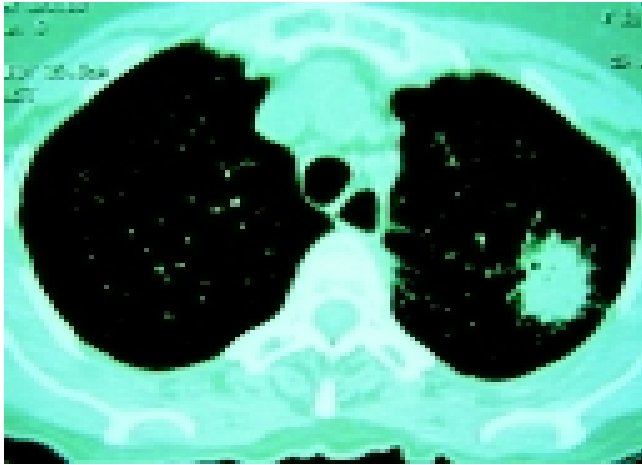


Picture 1 - Frontal chest radiograph (Premedication)
Showing a homogenous opacity with ill defined margins in the left upper lung fields

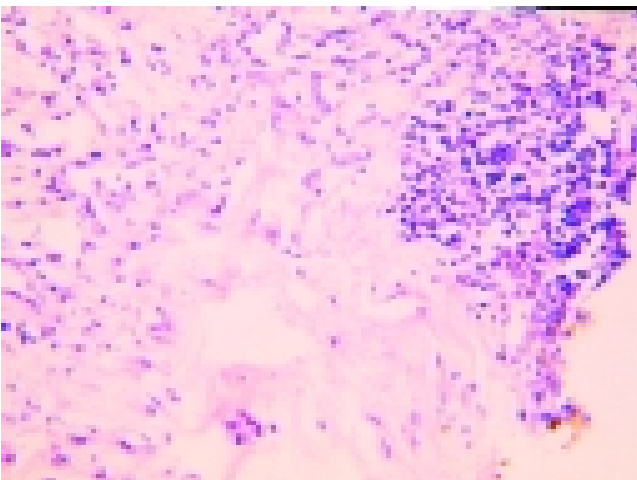
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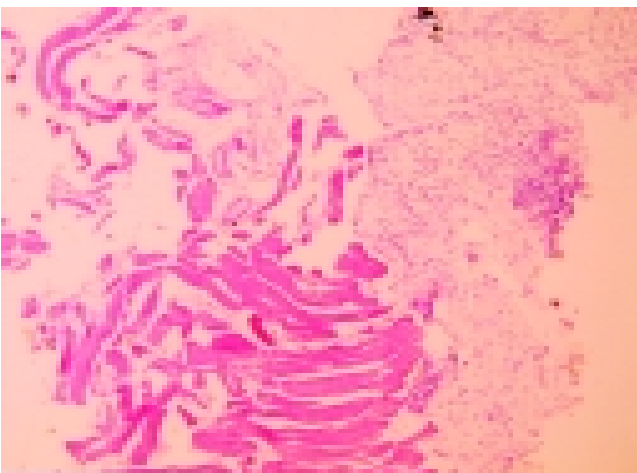
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Picture 2 - Axial CT (Premedication)
Showing homogenous solid density mass lesion with defined borders in left upper lobe



Picture 3 - Histopatology slide
Showing proliferation of cellular fibrotic tissue with entrapped flattened alveolar spaces and focal collection of plasma cells and lymphocytes



Picture 4 - Histopatology slide
Showing that the infiltrate was diffusely involving the tissue with the fibrotic bands & infiltrating the skeletal muscle

Bronchoscopy study showed mucosal inflammation of left upper lobe bronchial segments and the washings showed heavy growth of *Pseudomonas* by culture method. Patient was treated with anti *Pseudomonas* antibiotics as per the sensitivity pattern along with systemic steroids. Surgery was not considered in view of her advanced age.

On treatment after 10 days there was a radiological resolution of the left upper lobe lesion but a new triangular shaped lesion appeared in the right mid zone (Figure-5). Patient was continued on antibiotics with steroids for 3 weeks duration with symptomatic improvement. A repeat CT scan performed at the end of a month showed near complete resolution of both the left upper mass lesion and right middle lobe lesion with minimal residual scarring. Repeat complete blood count showed a leukocyte count of 11000 cells/ mm³ with ESR of 48 mm/ hr. Subsequent follow ups over a period of 3 months have not shown any clinical and radiological signs of recurrence of the lesion.



Picture 5 - Frontal chest radiograph (Postmedication)
Showing complete resolution of the left upper lobe lesion but a new triangular shaped lesion appeared in the right mid zone

Discussion

Inflammatory pseudotumor has been described in literature by various names in various locations as plasma cell granuloma (heart), inflammatory myofibroblastic tumor (lung), inflammatory myofibrohistiocytic proliferation, histiocytoma, xanthoma, fibroxanthoma, xanthogranuloma, fibrous xanthoma, plasma cell-histiocytoma complex (lung), plasmocytoma, solitary mast cell granuloma, inflammatory fibrosarcoma (bladder) (1). Inflammatory pseudotumor, also known as plasma cell granuloma and inflammatory myofibroblastic tumor, is a rare non-neoplastic lesion in adults consisting mainly of spindle-shaped mesenchymal and inflammatory cells

(1,5,6). Inflammatory pseudotumor most commonly involves the lung and the orbit, but it has been reported to occur in nearly every site in the body, from the central nervous system to the gastrointestinal tract (1). Because inflammatory pseudotumors mimic malignant tumors both clinically and radiologically, the radiologist should be familiar with this entity and help avoid unnecessary radical surgery when ever possible.

Inflammatory pseudotumor has been reported to occur in nearly every site in the body, from the central nervous system to the gastrointestinal tract. The extrapulmonary sites in which pseudotumor has been recorded are in the orbit, stomach, testis, esophagus, liver, spleen, pancreas, kidney, adrenal gland, retroperitoneum, diaphragm, mesentery, bladder, heart, thyroid, tonsil, fourth ventricle, spinal cord meninges, central nervous system, maxillary sinus, nasal cavity, nasopharynx, larynx, trachea (1,3,12).

The underlying cause and pathogenesis remain uncertain and various theories have been put forwarded by various authors. The commonly believed ones are following trauma or surgery, immune-autoimmune mechanism and secondary to infection (1). In certain cases it is thought to result from inflammation following minor trauma or surgery or to be associated with other malignancy (1,3,4). The ones secondary to infection are found in association with the organism include mycobacteria associated with spindle cell tumor; Epstein-Barr virus found in splenic and nodal pseudotumors; actinomycetes and nocardiae found in hepatic and pulmonary pseudotumors, respectively; and mycoplasma in pulmonary pseudotumors (1,7). In the present case possibility of pseudomonas induced pulmonary pseudotumor was considered because of the evidence of Pseudomonas growth in bronchial washings.

There have been reports of patients under 16 years of age developing inflammatory pseudo tumors that most frequently present as primary tumor-like lesions of the lung (5). It was reported that the incidence of the development of such tumors was only 23 of 56,400 cases that underwent thoracic surgery (0.04%) (5). Pseudo tumors accounted for only 0.7% of cases of lung and bronchogenic tumors (5). Though it is generally acknowledged that inflammatory pseudo tumors involve a non-neoplastic process characterized by unregulated growth of inflammatory cells, the existence of genuine involvement of neighbouring structures or its rapid recurrence in this case may appear incongruous and raises the possibility of an inflammatory pseudo tumor being a neoplasm. Some cases may be related to an infectious process (5).

Many patients are asymptomatic and are diagnosed incidentally on routine chest radiography. A history of lower respiratory tract infection is present in 18 to 30% of cases (8). Inflammatory pseudo tumor may have aggressive

features and they may encase bronchi or invade mediastinal structures, chest wall, and diaphragm. In such instances cough, fever, dyspnea, pulmonary infection and hemoptysis are the usual presenting manifestations. Multiple recurrences and metastatic spread can also be seen (8).

Macroscopically inflammatory pseudo tumors are well-circumscribed, unencapsulated, white-yellow masses of firm consistency (8). On microscopic examination based on the predominant histopathologic features, the lesions can be divided into three histologic types: (a) organizing pneumonia pattern, characterized by airways filled with plump fibroblasts and foamy histiocytes and parenchyma replaced with a mixture of histiocytes, mononuclear cells, and fibroblasts; (b) fibrous histiocytic pattern, which is the most common, and is characterized by spindle-shaped myofibroblasts arranged in whorls; and (c) lymphohistiocytic pattern, which is the least common (1,9,14). The histological picture consisting of polymorph with mature plasma cells, lymphocytes, vacuolized histiocytes, fibroblasts and other elements of the reticulo-endothelial system has been named plasma cell granuloma of the lung (10). Once the myofibroblast was identified and its function in tissue repair was established, this cell type was found in a variety of soft tissue lesions from nodular fasciitis to malignant fibrous histiocytoma. The myofibroblast was eventually recognized as the principal cell type in the inflammatory pseudotumor, which provided the opportunity to redesignate this tumor as inflammatory myofibroblastic tumor (IMT) (11).

Radiological features of inflammatory pseudotumor are variable and nonspecific. Although many pseudo tumors can be diagnosed presumptively on chest radiographs, a CT scan of the thorax may be necessary for a definitive diagnosis. The diagnosis of a pulmonary pseudotumor should be considered any time a lenticular opacity is identified superimposed on the central portion of the lung on a chest radiograph (13). CT features of pulmonary inflammatory pseudotumor has been found to be typically a solitary, peripheral, sharply circumscribed mass with an anatomic bias for the lower lobes (9,15). Local invasion and primary involvement of the mediastinum and hilar structures are unusual manifestations (15). CT calcifications within the lesion occur more frequently in children than in adults (9,14). They are useful for differential diagnosis if present, but they are usually non-specific in shape and configuration (16). If present calcification pattern ranges from an amorphous, mixed, or fine fleck-like pattern to heavy mineralization (1,17). Atelectasis and pleural effusion may occur (13,15). Cavitations and lymphadenopathy are very rare.

MRI shows intermediate signal intensity on T1WI and high signal intensity on T2WI. Dystrophic calcification within the lesion is not rare and is seen more frequently

in children than in adults. Multiple lung lesions, pneumonic consolidation, atelectasis, hilar masses, and cavitation are unusual findings (18). CT and MRI are helpful for demonstrating the extent of the potentially aggressive disease. Lung inflammatory myofibroblastic tumor is an uncommon cause of solitary lung nodule (0.7 % of lung tumors) (19). Central airway, vascular, esophageal, diaphragmatic, and chest wall invasion can be delineated by these modalities.

Treatment of the inflammatory pseudotumor can be diverse from antibiotics, corticosteroids & radiation to surgical treatment (8,20). Complete surgical resection, if possible, is the treatment of choice for most inflammatory pseudotumors (1,8,20). Corticosteroids response is often unpredictable in cases on pulmonary inflammatory pseudotumor. Early and complete surgical resection of the inflammatory pseudotumor provides cure and prevents local recurrence (8,20).

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