

**Case Report**

Disease Control of Pulmonary Inflammatory Pseudotumor by Multidisciplinary Approach

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Abstract

Introduction: Inflammatory pseudotumor is a rare tumor of the lung, which is usually misdiagnosed as malignancy. Most of early cases are treated with complete excision, but advanced cases: radiation therapy, steroids and chemotherapy will be a part of treatment plan. Multidisciplinary approach can save many cases of inflammatory pseudotumor and other rare diseases.

Case Report: we reporting a case of advanced inflammatory pseudotumor with had a long disease control with multidisciplinary approach in the form of chemotherapy, steroids, radiation therapy and surgery.

Conclusion: Advanced IP of the lung is rare disease with unknown etiology, where multidisciplinary team is needed for diagnosis and treatment: maximum safe debulking, chemotherapy, radiation therapy and steroids can be the suitable combined treatment for many advanced cases

Keywords: advanced pulmonary inflammatory pseudotumor; multidisciplinary approach; disease control

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Consent: We confirm that the patient has given the informed consent for the case report to be published.

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Introduction

Inflammatory pseudo tumor (IP) was described as clinically invasive histologically benign mass lesion of unknown etiology; it has a variety of histologic presentations and has been recognized under various names like plasma cell granuloma, inflammatory myofibroblastic tumor, histiocytoma complex, xanthomatous pseudotumor, fibrous xanthoma, and inflammatory myofibrohistiocytic proliferation [1]. It occurs mainly in the lung, but can arise in mesentery, bone, omentum, with less than 5% occur in head and neck [1].

IP of the lung was reported to have frequency of 0.04-0.07 of all respiratory tumors [2], most cases were less than 49 years old with non-specific symptoms and frequently misdiagnosed with other commoner lesions in the lung [3]. Cefolio et al. [4] reported that the tumors could be divided into the two types, based on the presence or absence of local invasion of vessels. The first type is the noninvasive type, which is asymptomatic and the second type is the invasive type, which is symptomatic.

Most successful therapy for IP of the lung through reported cases is surgery; complete resection usually has less frequency of recurrence than partial resection [3, 5]. Some cases of IP are Un-resectable, and they were treated by steroids, chemotherapy or radiotherapy [6].

We used multidisciplinary approach for treatment of such rare condition of advanced IP of the lung to reach the best results for disease control.

Case report

Female patient 35 years old presented on April 2012 by cough and chest pain not responding to treatment, computed tomography (CT) chest was showing right lower lung solid soft tissue mass and mediastinal lymph node enlargement.

The patient had four trials of open and CT guided biopsy with misdiagnosis as lung fibrosis, or other conditions and she had a history multiple courses of antibiotics, steroids with no response.

She presented in Nasser institute adult oncology unit after last open biopsy showing inflammatory pseudo tumor on January 2014, and new CT was done showing large RT lung lesion invading mediastinum and RT lung collapse (Figure 1).

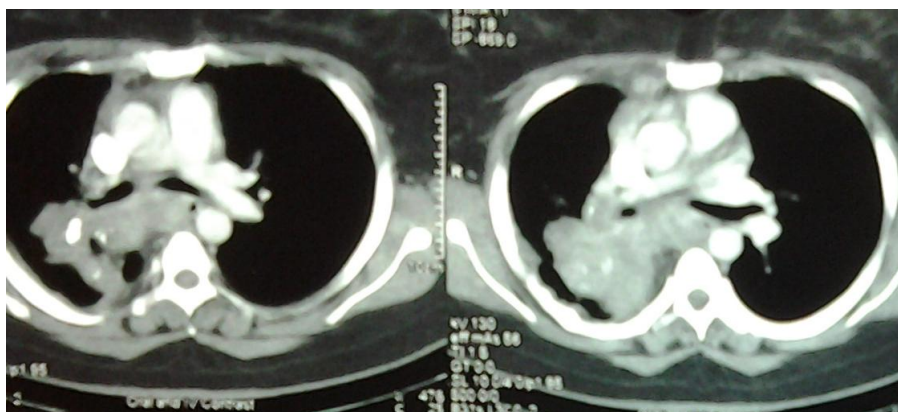


Figure 1 CT chest on January 2014 before Radiotherapy and Chemotherapy

Treatment

The right lung lesion was Un-resectable as consulted by chest surgical and oncology committee, and for that we decided to begin treatment by high dose steroids and RTH for tumor downsizing.

Radiotherapy

3D conformal radiation therapy for the 30 Gy 15 fractions in 3 weeks 15MEV,

Steroids

The patient had 16 mg dexamethasone/ day intramuscular for one week, followed by 8 mg/day next week, then 4 mg/day third week then interruption gradually.

CT after one month of radiotherapy was showing disease reduction in size (Figure 2) which was associated by improvement of patient symptoms and signs.



Figure 2 CT after 3D conformal radiotherapy and steroids and before chemotherapy

Chemotherapy

After consultation of oncology and chest surgery committee, surgeon decided that the tumor is still not resectable and more down staging was still needed, so our committee decided to begin chemotherapy in the form of: cyclophosphamide 750 mg/m²/21days for three cycles, which was well tolerated by the patient and then she was referred to chest surgery after new CT which was showing further decrease in size of tumor (Figure 3).



Figure 3 CT after 3 cycles of cyclophosphamide and before surgery

Surgery:

The patient was operated many times for biopsy before chemotherapy and radiotherapy; but this time was for radical resection after chemotherapy and radiotherapy, where there was tense adhesion caused by previous thoracotomy, so meticulous dissection of the lesion of the right lung was done where we observed the following:

- 1- Lesion involve the whole right lung where pneumonectomy can be the ideal decision
- 2- Lesion was hard, gritty sensation, originating from parenchymal lung tissues, the remaining lung tissues was not healthy.
- 3- Radical removal of the mass was not completed because there were un-resectable adhesions to the wall of LT atrium and intra-cardiac extension of the lesions through the pulmonary veins. This was confirmed by opening the pericardium for trial of complete resection
- 4- maximum debulking of the tumor was done.

Histopathological picture post surgery:

Grossly the specimen received as a fragmented vaguely well-defined mass measuring 5X3 cm. Cut section is firm to hard, nodular and heterogeneous, some areas are whitish, yellowish and black (Figure 4).

Histologic picture revealed collapsed lung parenchyma including a non-capsulated nodule composed of variable thickness collagenous bands with hyalinosis interrupted by slit like spaces, blood vessels and patches of coarse calcification. There were dense mononuclear lymphocytic inflammatory infiltrate rich in plasma cells (Figure 5).

Immunohistochemical staining for CD3 and CD20 showed polyclonal lymphocytic infiltrate excluding the possibility of non-Hodgkin's lymphoma. Immuno-staining for both CD15 and CD30 are negative excluding the possibility of Hodgkins lymphoma (Figure 5).

Histological examination and immunohistochemical staining confirmed the final diagnosis of inflammatory pseudotumor.

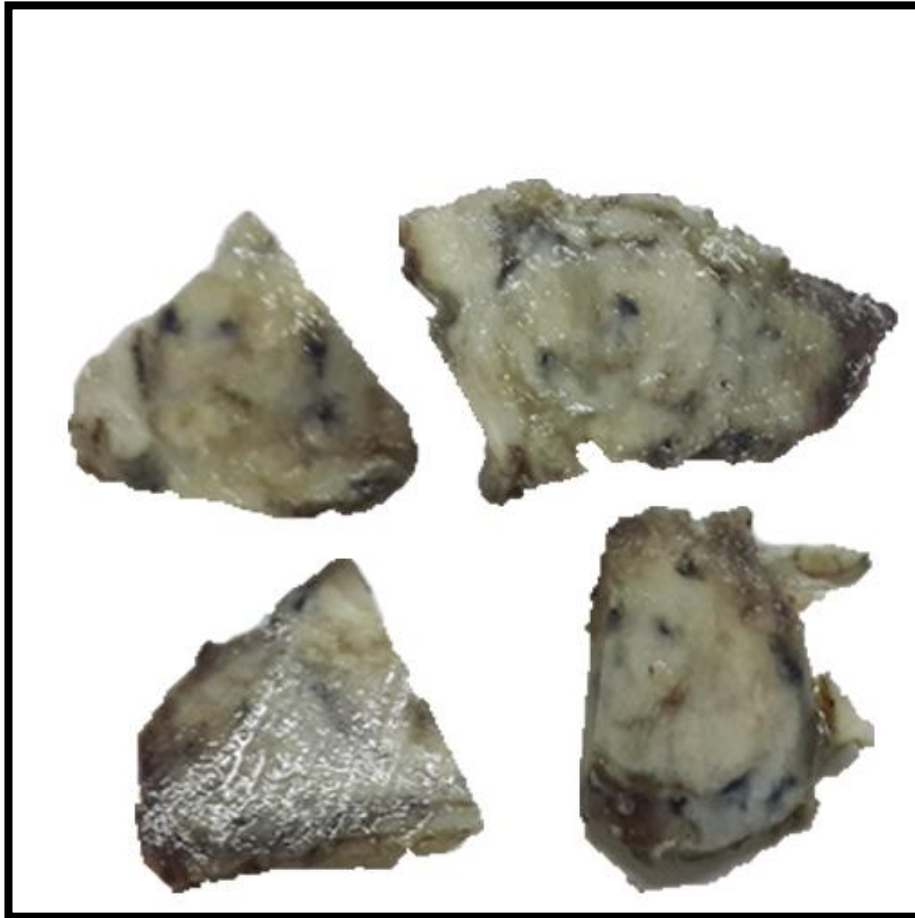


Figure 4 Gross picture of vaguely nodular mass with heterogeneous cut surface.

Follow up

The patient had a long period of recovery, and her chest symptoms were decreased, follow up CT was showing large residual tumor, so our committee decided to begin chemotherapy in the form of COP: cyclophosphamide 750 mg/m² D1/21 days, vincristine 2 mg D1/ 21 days and prednisolone 60 mg/day from D1 to D5/ 21 days.

The patient had 3 cycles and disease stabilized.

Now she's on maintenance steroids and still on follow up till December 2014.

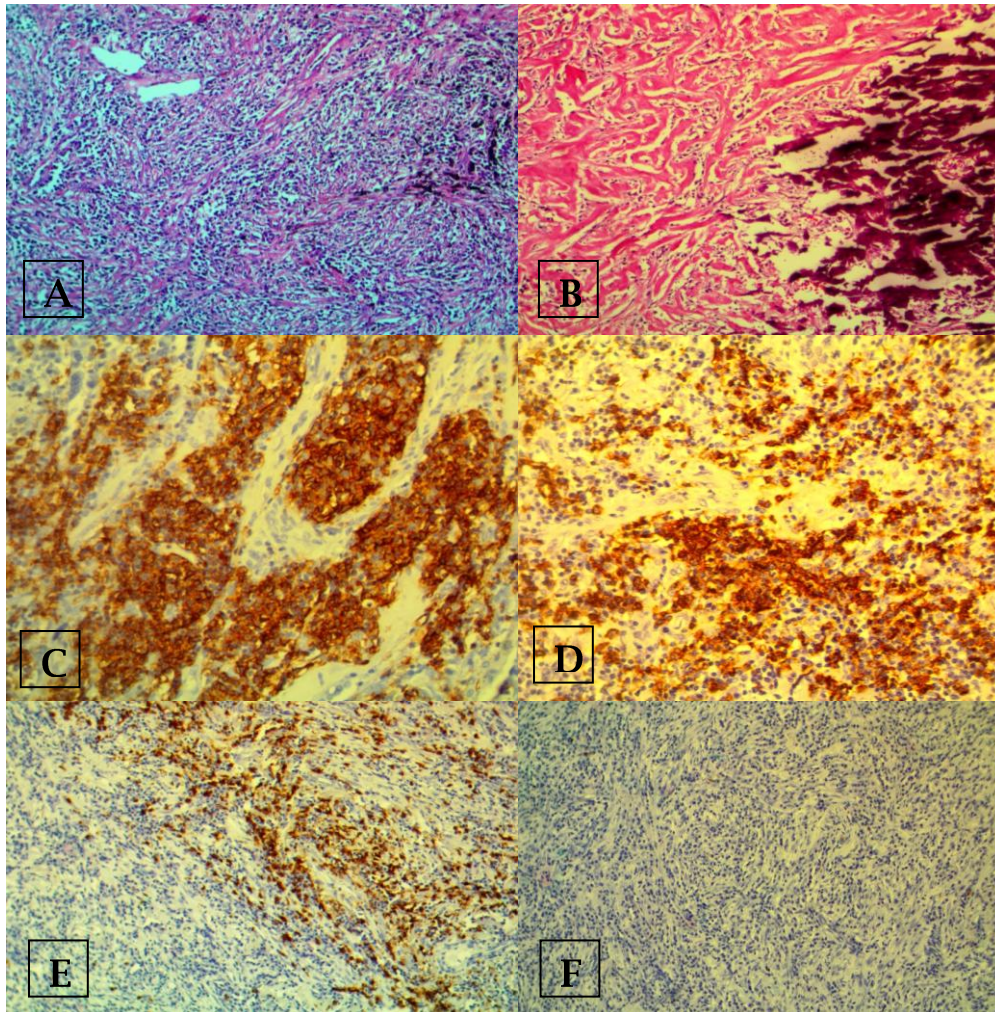


Figure 5 A- Histopathologic picture showed mononuclear lymphocytic infiltrate rich in plasma cells separated by collagenous stroma (Hx&E, X100). B- Areas of hyalinosis and calcification (Hx&E, X100). C&D- Immunostaining for CD3 and CD20 showed polyclonal lymphocytic infiltrate (X200). E- Immunostaining for CD15 revealed scattered positive monocytic cells (X200). F- Immunostaining for CD 30 was negative (X100).

Discussion

IP most commonly involves the lung and orbit, but it has been described in almost any location, more frequently seen in the lower lobe of the right lung, and forms a solitary, oval, and well defined lobulated mass that is peripherally located. A mass lesion is located peripherally in 87% and centrally in 6%, and the lesions can present radiologically as multiple nodular (5%), pleural based, cavitary lesions (5%), or can present with lobar atelectasis (8%) or hilar lymphadenopathy (5%) [7].

The etiology of pulmonary IP is still unknown, but immunological and infectious causes have been postulated [7].

The clinical presentation of the tumor can be variable, ranging from asymptomatic (70 -78%) to symptoms like cough, hemoptysis, chest pain, dyspnea, fever, etc., Radiology of the chest is helpful in localizing the position which involve to segment or lobe, and it's usually solitary [4].

Most reported cases of inflammatory pseudo tumor were treated by complete or partial surgical resection, with or without steroids. Most early cases of pulmonary IP which was treated with bronchoscopy resection had small size of tumor ranging from 1-2 cm [9]. Partial resection of IP was done in large tumors [5] with the use of radiotherapy and steroids to get the best tumor control.

Cyclophosphamide was used in some recurrent cases of IP in combination with steroids and radiotherapy with good results of treatment [10], but in our advanced case of pulmonary inflammatory pseudo tumor, disease control occurred after multidisciplinary approach: cyclophosphamide, steroid, radiotherapy and maximum debulking surgery. Close follow up in such a case with late diagnosis of IP is very important.

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