How pseudo is an inflammatory pseudotumor?

Dilip Gude,  
Ramasubba Rayudu,  
Dharam Bansal

Departments of Internal Medicine, 
Radiology, Pulmonology and 
Critical Care, Medwin Hospital, 
Nampally, Hyderabad, 
Andhra Pradesh, India

Address for correspondence:  
Dr. Dilip Gude,  
AMC, 3rd Floor, Medwin 
Hospital, Chirag Ali lane, 
Nampally, Hyderabad - 500 001, 
Andhra Pradesh, India.  
E-mail: letsgo.dilip@gmail.com

INTRODUCTION

Inflammatory pseudotumors (IPTs) are rare, well- 
circumscribed, unencapsulated, quasi-neoplastic tumors of 
unregulated growth of inflammatory cells, first recognized 
by Umiker and Iverson.[1] Various terms have been used to 
describe them based on the predominance of the particular 
type of cytology, such as plasma cell granuloma, plasma cell 
pseudotumor, inflammatory myofibroblastic proliferation, 
omentum-mesenteric hamartoma, histiocytoma and 
fi broxanthoma. It can roughly be categorized into an 
organizing pneumonia pattern, a fibrous histiocytic 
pattern (most common) and a lymphohistiocytic pattern 
(least common).[2] It is not yet established if these lesions 
represent a primary inflammatory process or a low-grade 
malignancy with a prominent inflammatory response.

CASE REPORT

We report a case of a 33-year-old male who presented with 
a history of cough with productive sputum, shortness of 
breath, recurrent fever (low grade), generalized weakness, 
anorexia and weight loss of 6 months duration. He also had 
an episode of hemoptysis. He was a non-smoker and worked 
as a clerical assistant at an office (no occupational exposure 
to potential allergens or toxic agents). On examination, he 
was thin built, and had pallor and clubbing. An impaired 
percussion note and decreased breath sounds were found 
on the left. Lab investigations revealed high erythrocyte 
sedimentation rate (ESR; 55 mm/hour), anemia (Hb 
8 g/dl), and thrombocytosis (580,000). X-ray chest revealed 
a large lobulated soft tissue mass lesion overlying the left 
cardiac border, situated in anterior and middle mediastinum 
[Figure 1]. Contrast-enhanced computed tomography 
(CT) thorax showed a hypodense mass lesion, measuring 
8.4 × 6.1 cm, adjacent to left hilum without any evidence 
of calcification or contrast enhancement [Figure 2]. 
Radiological differential diagnosis was that of hydatid cyst, 
empyema or neoplasm. Fine needle aspiration cytology 
(FNAC) was done which was inconclusive (no malignant 
cells). Patient was taken up for thoracotomy and a large 
mass, measuring 8 and 10 cm, arising from the lower lobe 
of left lung adherent to pericardium and diaphragm without 
a definite plane of cleavage was found. The necrotic mass 
was excised. Grossly, the specimen was lobular with a 
rubbery tan-yellow cut surface. Histopathology showed 
bland spindle-/stellate-shaped myofibroblasts loosely 
arranged in a myxoid stroma with scattered plasma cells and 
inflammatory cells [Figure 3]. There were varying mitoses, 
prominent lymphoplasmacytic infiltrate and a few germinal 
centers. The immune profile of the specimen was positive 
for smooth muscle actin, strongly positive for vimentin, 
negative for desmin and equivocal for epithelial membrane 
antigen. Final diagnosis of IPT of the left lung was made. 
The patient had been doing well and a follow-up of 2 years

A B S T R A C T

Inflammatory pseudotumor (IPT) is a rare lesion of unclear etiology reported in various organs. Although mostly benign, these tumors may pose a therapeutic challenge in cases of recurrence. We report the case of a young male who presented with a clinical and radiological picture suggestive of a malignancy in the thorax and upon evaluation was noted to have IPT of the lung. Complete surgical resection was done with no evidence of tumor recurrence. We review the literature and discern the epidemiological, clinical, pathophysiological and management aspects of IPTs.

Key words: Inflammatory pseudotumor, lung, malignancy, recurrence
showed no evidence of tumor recurrence. There was a feeling of well-being with acceptable weight gain (5 kg), and no further cough, shortness of breath or fever was reported.

**DISCUSSION**

Though IPTs of the lung are rare (0.04–0.7% incidence of all lung masses[3]), they pose a diagnostic and/or therapeutic challenge to the managing team of clinicians, pathologists, and surgeons. IPT is a distinctive pseudosarcomatous lesion occurring primarily in the viscera and soft tissues. It is seen mostly in children and young adults (half of the patients are less than 40 years[4]) with an equal sex distribution. Although initially reported and is more common in the lung, it has a propensity to involve 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, nasopharynx, larynx and trachea.[5] Two other notable cases of IPT have been reported from India, a 12-year-old presenting with massive hemoptysis[6] and a 3.5-year-old initially misdiagnosed to have a neuroblastoma.[7] Another 78-year-old developed a metachronous lung IPT, 7 years after having an orbital IPT.[8] WHO has recently classified the lesions into myxoid vascular, compact spindle cell, and hypocellular fibrous patterns with varying degrees of overlap within a lesion. Occasionally, IPTs are known to calcify, cavitate, invade the mediastinum or hilum, or present with pleural effusion. About 10% of the lesions are endobronchial. Clinically, most patients are asymptomatic, with a solitary nodule or mass detected by routine chest roentgenogram. Systemic symptoms may predominate in some, and recurrent pneumonia, collapse, pleural effusion, and a variety of mediastinal compression symptoms are not uncommon. Lab investigations may show elevated ESR, anemia, thrombocytosis, and hypergammaglobulinemia, which characteristically resolve after the lesion is excised.[8] Some of the postulated pathophysiological mechanisms include trauma or surgery, immune–autoimmune mechanism, associated with other malignancy or secondary to infection. Specific infectious agents have been incriminated, such as mycobacteria in association with spindle cell tumor, and nocardiae and mycoplasma in pulmonary pseudotumors.[9] The finding of IgG-predominant, polyclonal plasma cells and the development of about one-third of lesions after an infection further reinforces IPT being a reactive inflammatory process.

Proliferation of spindle cells, arranged in short fascicles with a focal storiform (whorled or cartwheel-like) architecture, associated with a variably dense polymorphic
infiltrate of mononuclear inflammatory cells is the most consistent histopathologic finding.[9] Cytologic atypia, ganglion-like cells, P53 expression and DNA aneuploidy along with focal and/or vascular invasion and nuclear pleomorphism are suggestive of more malignant behavior and a worse prognosis.[10]

IPTs have been shown to radiologically and clinically mimic a malignant process. Chest radiograph may show a lenticular opacity superimposed on the central portion of the lung, while CT typically shows a solitary, peripheral, sharply circumscribed mass with an anatomic bias for the lower lobes. Magnetic resonance imaging (MRI; intermediate signal intensity on T1WI and high signal intensity on T2WI) may help in delineating the extent of contiguous spread.

Complete surgical resection (wedge resection preferred over lobectomy or pneumonectomy), either by video-assisted thoracotomy or by open thoracotomy (if larger lesions and invasive nature), is the definitive treatment. If the patients are poor surgical candidates or have multiple nodules or unresectable disease, nonsurgical modalities like treatment with glucocorticoids, radiotherapy, and chemotherapy may be tried. Complete tumor excision and tumor size ≤3 cm are factors associated with a decreased risk of recurrence.

ACKNOWLEDGMENT

We thank the Departments of Internal Medicine, Radiology and Pulmonology/Critical Care for their perpetual support.

REFERENCES


How to cite this article: Gude D, Rayudu R, Bansal D. How pseudo is an inflammatory pseudotumor?. Indian J Med Paediatr Oncol 2011;32:204-6

Source of Support: Nil, Conflict of Interest: None declared.

Author Help: Reference checking facility

The manuscript system (www.journalonweb.com) allows the authors to check and verify the accuracy and style of references. The tool checks the references with PubMed as per a predefined style. Authors are encouraged to use this facility, before submitting articles to the journal.

- The style as well as bibliographic elements should be 100% accurate, to help get the references verified from the system. Even a single spelling error or addition of issue number/month of publication will lead to an error when verifying the reference.
- Example of a correct style
- Only the references from journals indexed in PubMed will be checked.
- Enter each reference in new line, without a serial number.
- Add up to a maximum of 15 references at a time.
- If the reference is correct for its bibliographic elements and punctuations, it will be shown as CORRECT and a link to the correct article in PubMed will be given.
- If any of the bibliographic elements are missing, incorrect or extra (such as issue number), it will be shown as INCORRECT and link to possible articles in PubMed will be given.