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Inflammatory myofibroblastic tumor of the lung in an adolescent boy

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INFLAMMATORY MYOFIBROBLASTIC TUMOR OF THE LUNG IN AN ADOLESCENT BOY

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ABSTRACT

Inflammatory myofibroblastic tumors of the lung are rare, here, an unusal case is described. A 14-year-old boy presented with a history of weight loss and clubbing and was found to have a solitary circumscribed mass in right lower lobe treated with lobectomy. This case indicates the need for early and complete removal of the inflammatory myofibroblastic tumor of the lung.

KEY WORDS: *Pseudotumor. Spindle cells. Lobectomy.*

INTRODUCTION

Inflammatory pseudotumors (IPTs), inflammatory myofibroblastic tumors or plasma cell granulomas are benign, essentially non-neoplastic, tumor-like masses which are inflammatory in origin.[1] Although they have been widely reported under different names (e.g., postinflammatory tumors, histiocytoma, xanthoma, fibroxanthoma, xanthogranuloma, plasma cell tumor), their first detailed description was made in 1973.[2] We present a case that illustrates the diversity of clinical presentations and unpredictability of the natural course of IPTs.

CASE REPORT

A 14-year-old physically fit boy, who was a non-smoker, presented with history of weight loss for 4 months. He had been well until 4 months back when he noticed some weight loss over the course of two years. He did not have any other complaints or insignificant past medical and drug histories. His physical examination was unremarkable except of clubbing of the fingers. Chest radiograph (Figure 1) showed a well circumscribed 5 cm right lower lobe mass. Computed tomographic scan (Figure 2) showed a circumscribed sharply defined mass in the right lower lobe located in the peripheral portion of the lung away from the main bronchus. It was a solid lesion with the differential possibility of slowly growing neoplasm or hamartoma. In view of the uncertain diagnosis and prognosis, triple-regimen antibiotics was started. Repeated radiological examination showed no change in the mass size. A right thoracotomy and right lower lobectomy were performed. Histologic examination revealed proliferating spindle cells consistent with the features of inflammatory pseudotumor of the lung (Figure 3). It was further studied by immunohistochemistry which showed strong reactivity of spindle cells of the lesion for smooth muscle actin. He remained well with no tumor recurrence in his follow-up visits.

Figure 1: Chest X-ray showed a rounded mass in the right lower lobe mass.

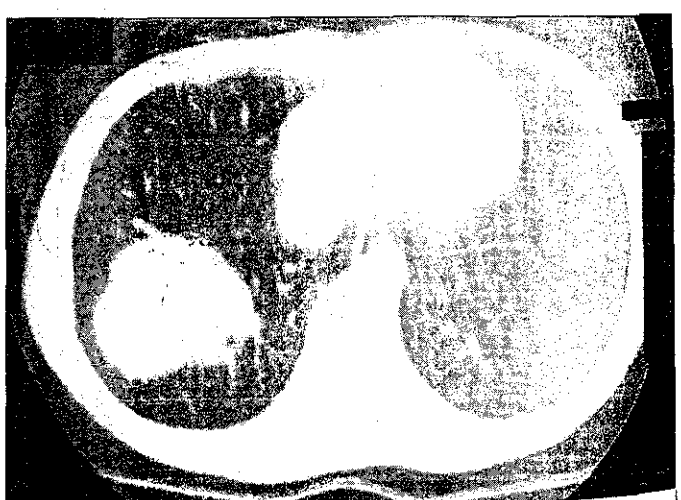

Figure 2: Computed tomographic scan showed a rounded right lower lobe mass treated by right lower lobectomy.

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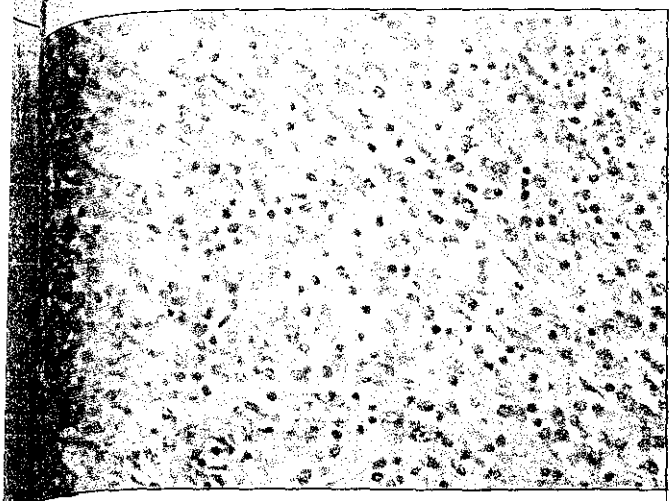

Figure 3: Photomicrograph shows an inflammatory myofibroblastic tumor. The lesion is composed of spindle cells arranged in interlacing fascicles, with admixed diverse inflammatory cells. The spindle cells have cellular atypia and no mitotic activity and the inflammatory cells are mature. (Hematoxylin-eosin, original magnification 400X).

DISCUSSION

Inflammatory pseudotumors of the lung are rare, with no predisposition to sex or race. More than half of the patients are less than 40 years of age and 15% are between the ages of 1 and 10 years. Their etiology and pathogenesis remain uncertain, although prior lung infection was recorded in one-third of the patients.[1, 2] Suggestions include immune mediated response and organizing intraalveolar pneumonia with progression to other histopathologic types.[1]

Although IPTs are regarded as inflammatory or reactive lesions rather than neoplasms, they may have features such as angioinvasion, local recurrence, distant metastases and neoplastic clonal changes.[3-5]

Macroscopically, IPTs are well circumscribed, non-encapsulated, firm, white yellow masses. Most are parenchymal but some are endobronchial and may cause obstruction. Less than 5% invade the mediastinum and chest wall.[1,2,6] Microscopically, the lesions consist of variable mixtures of fibroblasts and granulation tissue, fibrous tissue and inflammatory cells including lymphocytes, histiocytes, giant cells, macrophages, neutrophils, eosinophils and typically large number of plasma cells.[1,2] Immunohistochemistry has demonstrated the polyclonal nature of the plasma cells with immunoglobulin G predominance.[5]

Many patients are asymptomatic and the IPTs are discovered by incidental findings on examination of radiographs; if the patients are symptomatic, cough, hemoptysis, shortness of breath, clubbing, chest pain and arthralgia may be noted.[1]

Radiologically IPTs typically present as solitary circumscribed masses. Multiple lung masses, pneumonic consolidation, atelectasis, hilar masses and cavitation are quite unusual.[1,2,6] Percutaneous fine needle aspiration biopsy is considered insufficient for diagnosis and frozen section is also subject to errors.[1] Early and complete surgical resection of the IPTs remains the best treatment option to exclude malignancy and to achieve cure. Delay in diagnosis and treatment may increase considerably the magnitude of the surgical intervention required.

Non-surgical treatment modalities including radiotherapy, chemotherapy and steroids may have a place in the setting of incomplete surgical resection, multifocal disease, postoperative tumor recurrence or contraindication to lung resection.[7, 8]

The natural history of IPTs is unpredictable. Although spontaneous regression may occur, local expansion may cause significant morbidity and occasionally death.[2]

Although the most common picture of the IPTs is one of an asymptomatic, well-circumscribed lung mass that may mimic cancer, clinicians need to bear in mind their diverse clinical presentations. Surgical excision is usually indicated to reach a firm diagnosis and to attempt cure.

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