INFLAMMATORY PSEUDOTUMORS OF THE LUNG

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Abstract:
Inflammatory pseudotumors (IPTs) are benign, tumorlike masses, which are inflammatory in origin. These pseudotumors of the lung are rare and their etiology and pathogenesis remain an enigma. Surgical resection is recommended as the treatment of choice for pulmonary pseudotumors. Complete resection leads to excellent survival.

Key Words: Pseudotumors, Inflammatory, Myofibroblastic, Plasma Cell

Inflammatory pseudotumors (IPTs) or plasma cell granulomas are benign, essentially nonneoplastic, tumorlike masses, inflammatory in origin. Although they have been widely reported under different names (eg, postinflammatory tumors, histiocytoma, xanthoma, fibroxanthoma, xanthogranuloma, plasma cell tumor), their first detailed description was made in 1973.[1] Surgical resection is recommended as the treatment of choice. Complete resection leads to excellent survival.

Inflammatory pseudotumors of the lung are rare, with no predisposition to sex or race. Its incidence is reported to be 0.04-1% of all the tumors of the lung.[1] 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.[2]

Although IPTs are regarded as inflammatory or reactive lesions rather than neoplasms, they may have features such as angioinvasion, local recurrence, distant metastases and cytogenetic clonal changes.[3-5] The notion of inflammatory myofibroblastic tumor being a reactive lesion or a neoplasm had been controversial, it has been recently thought of as a neoplasm because of clonal chromosomal abnormalities, chromosomal rearrangements involving the ALK receptor tyrosine kinase locus region or DNA aneuploidy.[11] It grows locally and slowly so taking into account these histopathologic and biological findings, it may be regarded as low grade malignancy or benign tumor.

Macroscopically, IPTs are well circumscribed, nonencapsulated, 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, neutorphils, eosinophils and typically large numbers of plasma cells.[1,2] Immunohistochemistry has demonstrated the polyclonal nature of the plasma cells with immunoglobulin G predominance (Fig.1)[5]

In 1990, Pettinato and colleagues[2] referred to this entity as inflammatory myofibroblastic tumor because the bulk of the lesion invariably consisted of not specific inflammatory cells, but proliferative myofibroblasts and fibroblasts. Most of the spindle cells were myofibroblasts, which showed immunohistochemical staining for vimentin and smooth muscle actin.

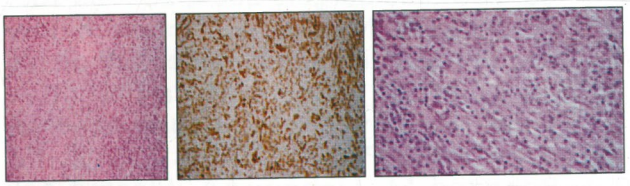

Fig.1. Photomicrographs showed inflammatory myofibroblastic tumor. The lesion is composed of spindle cells arranged in interlacing fascicles with admixed diverse inflammatory cells. Immunohistochemically, (in the middle) the spindle cells of the lesion show diffuse and strong reactivity for smooth muscle actin (immunohistochemistry for smooth muscle actin).

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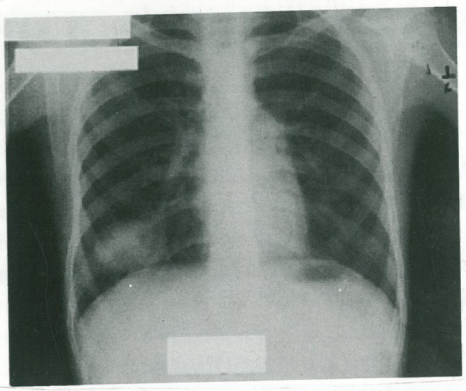

Figure 2. Chest-X-ray of a 12 yr old baoy showing Right Lower Lobe Pseudotumor

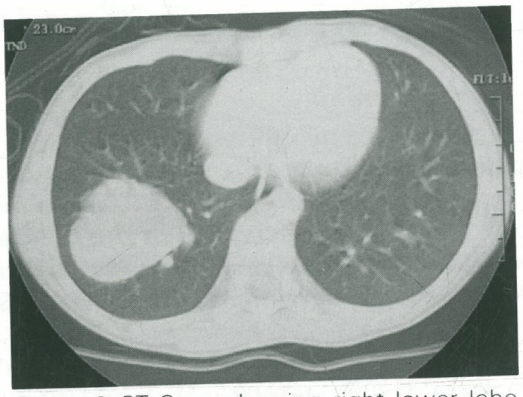

Figure 3 CT Scan showing right lower lobe Pseudotumor

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.(Figure 2, and 3)

Multiple lung masses, pneumonic consolidation, atelectasis, hilar masses and cavitation are unusual.[1, 2] Percutaneous fine needle aspiration biopsy is considered insufficient for diagnosis and frozen section is also subject to errors. [2]

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. The surgical resection usually requires a lobectomy but sometimes pneumonectomy may be required in very large pseudotumors. Wedge resection or segmental resection in patients with marginal lung functions should also be considered.

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. [1]

CONCLUSION:

Although the most common picture of the inflammatory pseudotumors 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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