Pulmonary Myofibroblastic Pseudotumor: A Rare Surgical Pathology

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Abstract

Pulmonary myofibroblastic pseudotumors are rare surgical pathologies, of unknown origin, with benign behavior and a good prognosis if completely resected. We present a 21-year-old male with a solitary pulmonary nodule found during a routine chest X-ray with CT scan confirmation. After a 16-month follow-up, the nodule increased in size and the patient developed mild dyspnea. He underwent an elective left postero-lateral thoracotomy and excision of the mass with an upper lobectomy. Pathologic studies revealed a pulmonary myofibroblastic pseudotumor.

Key words
Inflammatory myofibroblastic pseudotumor · lung · nodule

Bibliography

Thorac Cardiov Surg 2006; 54: 430 – 432

DOI 10.1055/s-2006-924088 · ISSN 0171-6425

Abbreviations

IMP	Inflammatory myofibroblastic pseudotumor
CT	Computed tomography

Introduction

Myofibroblastic pseudotumors of the lung are a rare surgical pathology. The pathogenesis of this tumor is related to an exaggerated inflammatory response to tissue injury of unknown cause [1]. The main cells recognized in this pathology are myofibroblasts, giving the name to this lesion. These tumors displace surrounding anatomic structures leading to organic dysfunction by local compression [2]. They exhibit a benign behavior and have a good prognosis if completely resected.

Case Report

A 21-year-old healthy male came to our hospital for a routine medical check-up. Standard chest X-ray revealed a 2.5 cm, well defined nodule in the left para-hilar region (Fig. **1 a**). Computed tomography (CT) scan showed a tumor without calcifications. No mediastinal lymph nodes were seen (Fig. **1 b**). The patient remained asymptomatic over a 13-month period of clinical and X-ray follow-up. Then he developed new onset exertional dyspnea. During the following 3 months, under X-ray evaluation, the nodule underwent an increase in size of up to 3.2 cm at its largest dimension (Fig. **1 c**). Surgery was advised. A left posterolateral thoracotomy was carried out and excision of the mass was completed successfully. Due to the hilar location of the nodule, dissection was difficult and a complete upper lobectomy was performed. The lower lobe and the parietal pleura were normal; lymph nodes at the left inferior pulmonary ligament and the aorto-pulmonary window were slightly enlarged and were resected. Pathologic studies revealed a myofibroblastic pseudotumor of the lung with plasma cell infiltration (Fig. **2 a** and **b**).

The patient's hospital course was uneventful and he was discharged home 10 days after surgery. At the six and twelve month follow-up, the patient was asymptomatic, doing well and with no evidence of recurrence on chest X-rays or CT scan.

Discussion

The inclusion of inflammatory myofibroblastic pseudotumors in the World Health Organization classification of soft tissue tumors was made in 1994. They were described as tumors composed of myofibroblastic differentiated cells, usually accompanied by numerous plasma cells and lymphocytes [2].

Pseudotumors have been found in almost every organ and system, in all ages and races. If located in the lungs, they are detected mostly because of systemic signs and symptoms such as fever, anemia, thrombocytosis, and other specific symptoms like cough, hemoptysis, chest pain, upper respiratory tract infections [3, 4]. In our patient, the diagnosis of the solitary pulmonary nodule was incidental and after 13 months of observation he only presented with mild exertional dyspnea.

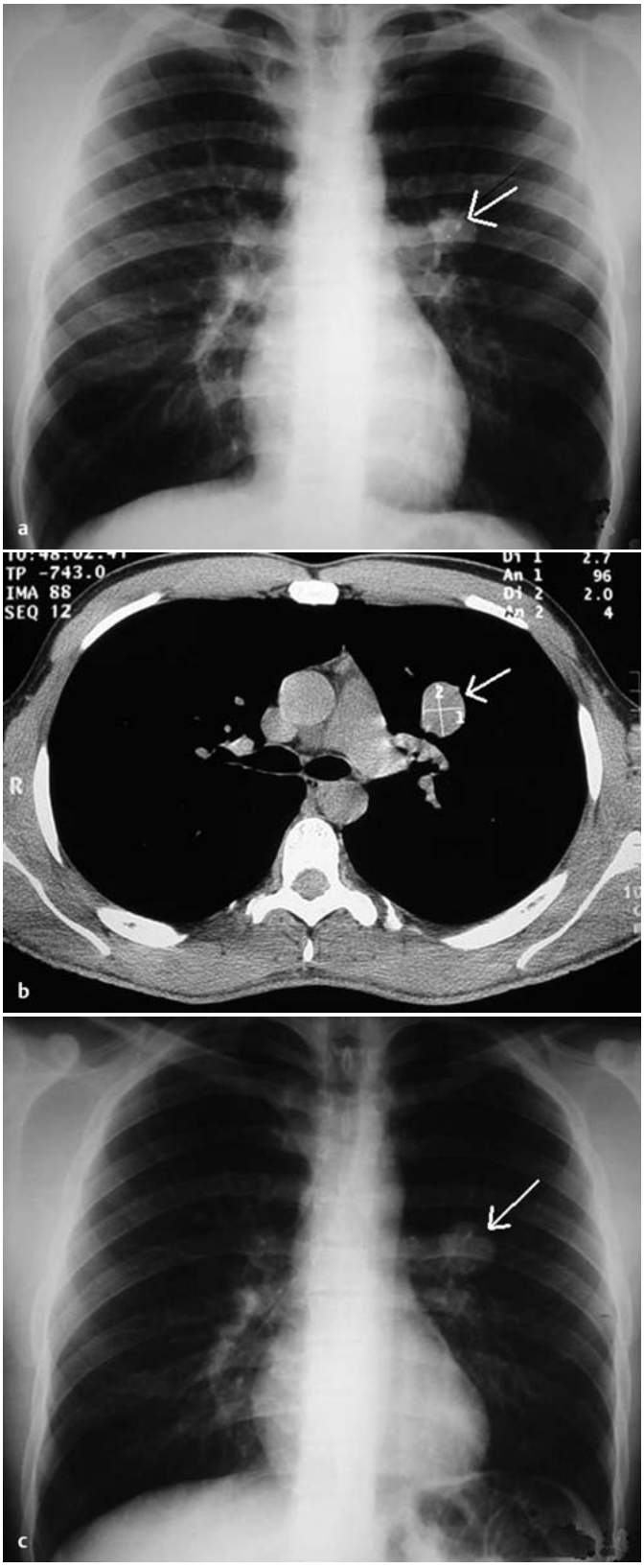

Fig. **1 a** to **c** **a** Solitary pulmonary nodule in the left lung (arrow). **b** CT evidence of a nodule in the left pulmonary hilum (arrow). **c** Follow up chest X-rays that show enlargement of the left pulmonary nodule (arrow).

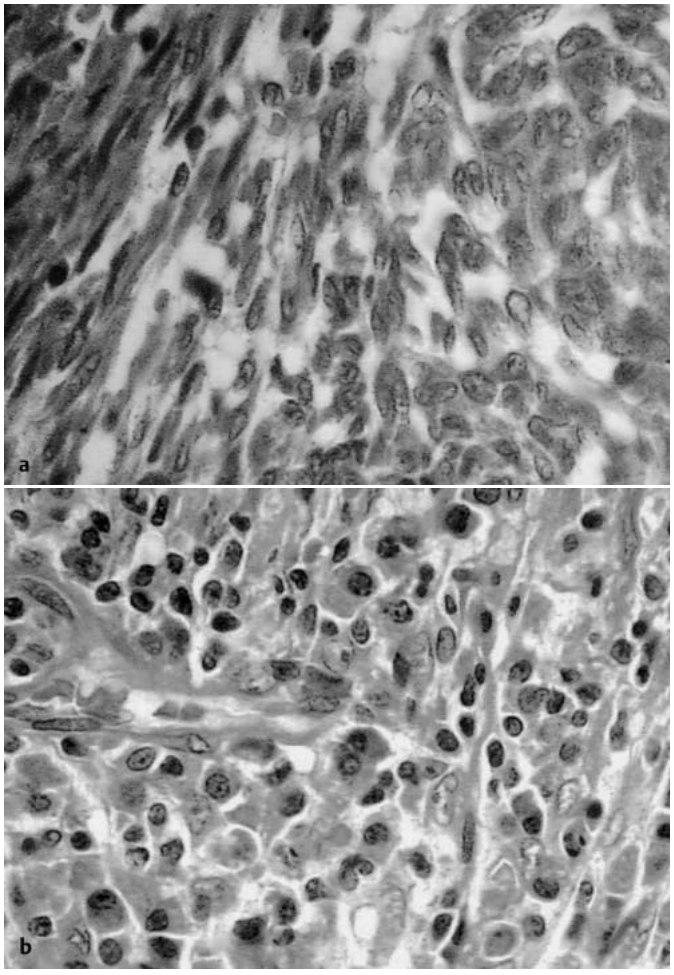

Fig. **2a** and **b** **a** Characteristic myofibroblasts in nodule tissue. **b** Multiple plasma cells in tumoral tissue.

Pseudotumors commonly vary in size; reports in the literature describe lesions from 1 to 40 cm [3, 5, 6]. Their surface after they are cut is gray-white and firm, with a fascicular pattern, or can be soft with a myxoid appearance, occasionally with calcifications [3]. They are rare tumors that present as solitary masses consisting of inflammatory and mesenchymal cells [5]. Their true incidence is not known because of the different names used to describe them [4]. A report from the Mayo Clinic and Mayo Foundation showed a 0.04% incidence of inflammatory pseudotumors of the lung in all thoracic procedures performed between 1946 and 1993 [6]; this series included 56 400 cases.

There have been attempts to classify these tumors into two types according to their clinical presentation. Noninvasive inflammatory pseudotumors, which occur mainly in asymptomatic patients, are characterized by small nodules which do not invade surrounding structures and which can be completely resected. The second type includes tumors that invade the lung, mediastinum or thoracic wall; these require extensive surgery and patients are mostly symptomatic [6].

On X-ray examination, there are no specific signs for an IMP, they are usually visible as a solitary circular nodule within the parenchyma. CT scan helps to define the nodule and its relations [4].

Precise diagnosis usually is not made prior to surgery and commonly presents a dilemma to the surgeon during operation [6]. The diagnosis requires thoracotomy, since bronchoscopy is usually negative and fine needle aspiration is frequently non-diagnostic [5]. Frozen transoperative studies should be interpreted with caution because of IMP's resemblance to sarcomatoid carcinoma of the lung [3] or plasmacytomas.

Whenever possible, total resection should be done, because there is a direct relationship between the degree of resection and the chance of recurrence [4]. We were able to accomplish this resection in our patient. The entity is associated with excellent prognosis. In a review of the literature, there is consensus that the treatment of choice is surgical resection and 10-year recurrence is almost nonexistent.

Conclusions

IMP is a very rare disease, most frequently localized in the lungs. These cases require significant pulmonary resection and are associated with excellent prognosis and survival.

Acknowledgement

We wish to thank Magdalena Gomez M.D. for her assistance in the preparation of this manuscript.

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Received October 24, 2005

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