

CASE REPORT

A rare case of inflammatory pseudotumour of the bronchus, occurring in an achondroplastic woman

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ABSTRACT: An inflammatory pseudotumour of the lung in an achondroplastic woman is described. The occurrence of both conditions in the same patient probably represents a coincidental finding. However, recurrent pulmonary infections, as frequently seen in achondroplasia, may be relevant, because a history of antecedent infection is noted in one third of cases of inflammatory pseudotumours. Despite extensive debridement of the pseudotumour by neodymium yttrium aluminium garnet (NdYAG) laser therapy, early recurrence occurred.

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Inflammatory pseudotumours of the lung are rare in adults. They pose a problem in differentiating between a benign and a malignant process. On the basis of a case report, the clinical, radiographic and pathological features of pseudotumours of the lung are described.

Case history

In February 1991, a female aged 54 yrs, who had been well until two weeks earlier, was admitted to hospital when she developed fever followed by a productive cough and dyspnoea.

On physical examination the patient was an overweight, achondroplastic, mentally-retarded female. Her temperature was 38°C, pulse 96 beats·min⁻¹ and respiration rate 18 breaths·min⁻¹. Auscultation revealed absence of lung sounds over the left hemithorax. Heart and abdominal examination were normal. An X-ray film of the chest revealed atelectasis of the left lung with shifting of the mediastinum. Computerized tomography of the thorax confirmed the complete collapse of the left lung. No pathological lymph nodes were visualized.

Laboratory examination showed an elevated sedimentation rate and white blood cell count (14.3×10^9 cells·l⁻¹). Renal and liver function tests, as well as tumour markers, were normal. No infectious cause could be found with appropriate bacteriological and serological techniques. Neither echography of the abdomen nor bone scintigraphy revealed any evidence of malignant disease. Fibreoptic bronchoscopy disclosed a gray endobronchial mass originating from the left lower lobe and protrud-

ing into the left main bronchus. As a first biopsy yielded insufficient material, the procedure was repeated with the rigid bronchoscope. Microscopic examination of this biopsy specimen showed an inflammatory pseudotumour (fig. 1). Microscopic investigation confirmed the benign character of the tumour, that was composed of cellular connective tissue with many capillaries. There were numerous slender mesenchymal cells with small ovoid nuclei and a fine homogeneously dispersed chromatin. Mitotic figures were rare. A dense diffuse inflammatory infiltrate, consisting of lymphocytes, numerous plasma cells, histiocytes, foam cells and polymorphonuclear granulocytes, was present. Because of the endobronchial growth, the respiratory insufficiency and mental status

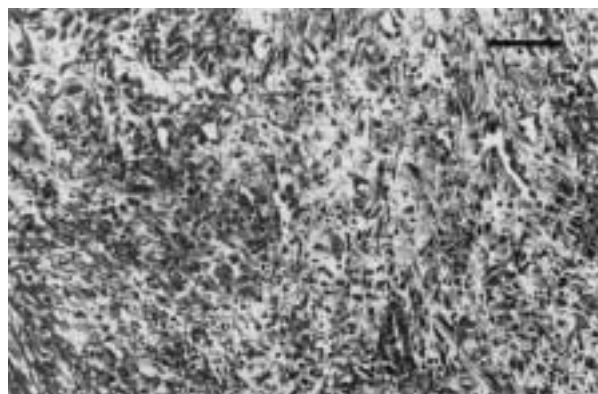


Fig. 1. – Intrabronchial inflammatory pseudotumour consisting of highly vascularized connective tissue intermingled with numerous inflammatory cells. (Masson's trichrome stain; bar = 100µm).

of the patient, neodymium yttrium aluminium garnet (Nd-YAG) laser therapy with additional debridement was preferred to surgical excision. At discharge, chest radiography showed complete re-expansion of the left lung.

The patient was well until August 1991, when respiratory insufficiency developed again, due to tumour growth in the left main bronchus and lower trachea, with ensuing complete atelectasis of the left lung. Several laser treatment sessions with additional debridement under general anaesthesia were necessary to resect the polypoid mass. Pathological examination of these specimens confirmed the initial diagnosis. During the last session, severe haemorrhage occurred, requiring endoscopic haemostasis and precluding further procedures.

Four weeks later, the patient developed a paraplegia. Spinal magnetic resonance imaging (MRI) scan (fig. 2) revealed vertebral and medullar compression at the Th 10 level. Some contiguity with the tumour in the collapsed left lung was suspected. As the paraplegia was already well-established, no surgical treatment was performed, and the patient died because of urosepsis in December 1991.

Autopsy confirmed the diagnosis of inflammatory pseudotumour of the bronchus, originating from the left lower lobe (fig. 3), without infiltration of mediastinum or spine. No malignant pathology could be demonstrated as the cause of the thoracic medullar compression. Degeneration of the intervertebral cartilaginous disc with protrusion of its core into the spinal canal, frequently encountered in achondroplasia, was responsible for the patient's paraplegia.



Fig. 2. – Spinal magnetic resonance imaging (MRI) scan showing vertebral and medullar compression at the Th 10 level.

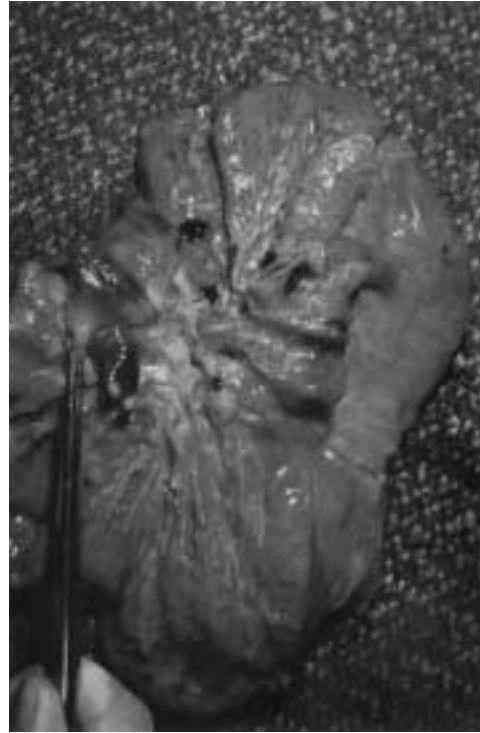


Fig. 3. – Endobronchial inflammatory pseudotumour originating from the left lower lobe and protruding into the left main bronchus.

Discussion

Lung, mediastinum and, rarely, lymph nodes can be the seat of so-called inflammatory pseudotumours. These lesions behave as reactive processes. They may be multifocal or recurrent, thereby mimicking neoplasms. Because of the presence of plasma cells and fat-laden mononuclear cells (foam cells) within these lesions, they are also called histiocytoma, fibroxanthoma, xanthogranuloma or plasma cell granuloma.

Depending on the predominant cell types, inflammatory pseudotumours are divided into three groups: the organizing pneumonia type with predominant fibroblast-like spindle cells; the fibrous histiocytoma type; and the lymphoplasmacytic type [1]. Endobronchial growth is more often seen in the fibrous histiocytoma type, as was the case in our patient.

GOLBERT and PLETNEV [2] reported an incidence of 0.7% inflammatory pseudotumour among 1,075 pulmonary and bronchial tumours that they encountered. Approximately 60% of these tumours occur in patients less than 30 yrs of age [3]. They are the most common primary tumour-like lesions of the lung in children under 16 yrs of age.

Inflammatory pseudotumours usually present as solitary, circumscribed masses. They are firm and white, yellow or gray on section. Although they are usually found in the lung and major bronchi, they have also been reported in the liver, stomach, kidney, retroperitoneum, peritoneal cavity, spinal cord meninges, posterior cranial fossa [4] and the bladder [5]. The lesions vary from 1 to 12 cm and grow slowly. Endobronchial growth is predominant in 12%, leading to symptoms of cough and

haemoptysis [6]. The lesion is sometimes found incidentally. Atelectasis is the presenting finding in children [2]. Although a history of recent acute respiratory illness, occurring in one third of the patients, suggests that the process represents a stage in the resolution of acute pneumonia, successful culture of the organism is unlikely, because the organism has been eradicated by normal host defences long before radiographic detection of the lesion [1]. As achondroplastic patients develop recurrent pulmonary infections, due to thoracic cage deformity produced by abnormal rib development [7], an antecedent pulmonary infection could be associated with the pathogenesis of this tumour in this particular patient. Nevertheless, we were not able to trace this in the patient's history.

If the patient's general condition is good, the appropriate therapy, when anatomically possible, is surgery with wedge resection, lobectomy or pneumonectomy [8]. Multiple bilateral nodules pose a more difficult problem. Resection and histological diagnosis of one of the nodules and observation of the other may be considered in some cases, especially if the patient is a poor operative risk [9]. Adjuvant therapeutic modalities include radiation therapy, steroid therapy [10] and antibiotics. Despite an extensive literature search, we found no report on Nd YAG laser treatment of an inflammatory pseudotumour. The reported case should, therefore, be regarded as the first.

Death associated with inflammatory pseudotumour is unusual. The prognosis in the vast majority is good, even after local resection, and despite occasional limited extension into the mediastinum.

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