Inflammatory myofibroblastic tumour of the lung in a five-year-old girl

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Abstract

The inflammatory myofibroblastic tumour of the lung is considered a rare diagnosis of lung masses. We report the case of a five-year-old girl who presented with recurrent pyrexia, dry cough, and shortness of breath. Chest X-ray and computed tomography showed a total atelectasis of the left lower lobe and a segmental atelectasis of the left upper lobe. The mass was removed in toto, histopathology revealed the diagnosis of an inflammatory myofibroblastic tumour of the lung. The patient is without any signs of relapse 30 months after surgery.

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1. Introduction

Inflammatory myofibroblastic tumours belong to the group of soft tissue tumours and have also been named plasma cell granuloma, inflammatory pseudotumour, xanthoma xanthomatosus pseudotumour, fibrous histiocytoma or histiocytoma, mast cell tumour or mast cell granuloma. The aetiology is still not completely understood. These tumours are considered as rare lesions in the differential diagnosis of lung masses, first described by Brunn in 1939 [1]. In children, the inflammatory myofibroblastic tumour is the most common primary lung mass [2, 3]. Thirty-five percent of inflammatory myofibroblastic tumours have occurred in children under 15 years [2]. The treatment of choice for inflammatory myofibroblastic tumour of the lung is, for diagnostic and therapeutic reasons, complete resection [4, 5].

We report a case of a five-year-old girl who presented with atelectasis due to an unknown lung mass and review the literature related to inflammatory myofibroblastic tumour of the lung.

2. Case report

A five-year-old girl presented with a 9-month history of recurrent respiratory infections with a dry cough which had been treated with antibiotics. Additional symptoms were pyrexia, weight loss, night sweat and shortness of breath. The girl presented in a reduced general condition with a progressive cough. She was referred with suspicion of foreign body aspiration to our institution. Chest X-ray showed a total atelectasis of the left lower lobe and segmental atelectasis of the left upper lobe with compensatory overinflation of the contralateral lung. Using computed tomography (CT), a lesion of diameter of 7×14 mm in the left main bronchus with bronchial obstruction and atelectasis of the left lung, mediastinal shift to the left, and compensatory overinflation of the right lung was seen (Fig. 1). Bronchoscopy revealed a total obstruction of the left main bronchus by a nodular structure covered with mucosa. The blood examination showed signs of an inflammation: anaemia (Hb 115 g/l), leukocytosis (24.6×10⁹/l) without leftward shift, elevated erythrocyte sediment reaction (ESR) (63 mm/h) and a CRP with 32 mg/l.

A thoracotomy was performed, and a parenchymal sparing endobronchial resection of the tumour was performed via bronchotomy. The mass was completely removed. Histopathological examination revealed the diagnosis of an intrabronchial, inflammatory pseudotumour of the lung, of the fibrohistiocytoma type (Fig. 2). The postoperative course was uneventful. Chest X-ray and bronchoscopy showed a complete re-expansion of the left lung. The patient was discharged in good general condition without any signs of relapse 30 months after surgery (clinical examination, CT, bronchoscopy).

3. Discussion

Inflammatory myofibroblastic tumours of the lung mimic malignant neoplasms clinically and radiographically [6]. These tumours only rarely appear to be locally invasive [6] or to recur rapidly [5]. Furthermore, malignant variants are discussed. Pathophysiologically, an unregulated proliferative...
tion of inflammatory cells seems to be involved [7]. It has been hypothesized that an initial infection may lead to an inflammatory myofibroblastic tumour and the occurrence of microorganisms such as Mycoplasma, Nocardia, Actinomyces [7], Epstein-Barr [8] and human herpes virus have been described. The incidence reported in the literature is <1% of all lung masses [5, 7] but inflammatory myofibroblastic tumours account for as many as 56% of benign pulmonary tumours in children. Inflammatory myofibroblastic tumours occur at any age. In children, they account for most common primary lung masses [2, 3]. Two-hundred and thirty cases of primary lung tumours in children have been reviewed by Hartmann and Shochat, 78 (34%) were benign and 56% of these were classified as inflammatory myofibroblastic tumours [3]. Bahadori and Liebow reported in their study that more than one-third of the patients were <20 years, and a quarter were 10 years or younger, the youngest patient was 13 months old. Male and female subjects seem to be affected equally. In 24 of the 40 patients, the lesion was discovered during a routine examination, and there was no history of previous or present illness [2].

The fact that about 50% of the patients are asymptomatic holds the risk of false diagnosis. Other patients are afflicted with cough, haemoptysis, chest pain or dyspnoea, and sometimes additional systemic features like low grade pyrexia, weight loss, microcytic hypochromic anaemia, polycyonal hyperglobulinaemia and a raised ESR, which are possibly caused by interleukins produced by the lesion.

Five-year survival in case of inflammatory myofibroblastic tumours is 91.3%, but the recurrence rate after resection is 4% and appears in locations of incomplete resection [7]. That is why the treatment of choice for diagnostic and therapeutic reasons is a complete resection [4, 5]. Laser can be applied successfully, but there are also reports about recurrent relapse of the tumour which finally ended in surgical intervention [9]. In the present case, this uncertainty was the reason for the decision for direct surgical resection.

4. Conclusion

Inflammatory myofibroblastic tumours are rare benign lesions that can occur in any age group, but are commonly found in patients who are <40 years of age [4]. Surgery remains the treatment of choice both for diagnostic and therapeutic reasons, even if the inflammatory myofibroblastic tumour may be diagnosed by fine-needle aspiration in a few cases [10].

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References

eComment: Malignant behavior of inflammatory pseudotumors

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Inflammatory myofibroblastic tumor of the lung is rare and its incidence is reported to be 0.04–1% of all pulmonary tumors. We have read with great interest the article by Ochs et al. concerning such a type of tumor [1]. Although these lesions can grow at a wide variety of other sites, they usually arise within the lung.

The aim of this brief comment is to highlight the risk of malignant transformation of these tumors. Sarcomatous transformation is described in the literature and can occur with both pulmonary and extrapulmonary inflammatory pseudotumors. Spencer reported a sarcomatous transformation of two inflammatory pseudotumors of the lung of the 27 observed [2]. Coffin and colleagues reported a sarcomatous transformation in 2 of 84 extrapulmonary inflammatory pseudotumors [3]. Donner and associates reported one case of inflammatory pseudotumor arising in the soft tissue of the forearm and progressing into sarcoma after five recurrences [4]. The possibility of a sarcomatous transformation should, therefore, be taken into account in patients with inflammatory pseudotumor. Caution is needed, however, because inflammatory pseudotumors may resemble low-grade sarcomas from a histologic point of view.

The treatment of choice of inflammatory pseudotumor of the lung is surgery. Wedge resection, if radical, is suitable for curative purposes. When it is not technically feasible, the lesion is removed with major resection (lobectomy or pneumonectomy) [5]. In some cases, neighboring anatomic structures (chest wall, diaphragm) also need to be excised. Long-term follow-up is imperative to detect recurrence.

References