Endobronchial Inflammatory Pseudotumor of the Lung as a Cause of Atelectasis

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Abstract
Inflammatory pseudotumor is the most frequent primary lung tumor in children under the age of 16. Inflammatory pseudotumor of the lung is usually peripherally located, but lesions may rarely be endobronchial. We report here the case of a five-year-old girl with unresolving pneumonia and atelectasis at the left lower lobe. Radiologically, atelectasis due to bronchial obstruction was considered, and bronchoscopic and histopathological investigations confirmed the definite diagnosis.

Keywords: atelectasis, lung neoplasm, inflammatory pseudotumor, children

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INTRODUCTION

When atelectasis is radiologically determined in children with recurrent or unresolving pneumonia, the initial diagnosis to consider is foreign body aspiration. Benign or malignant tumoral lesions are rare in childhood as causes of obstructive atelectasis [1,2]. Inflammatory pseudotumor (IPT) is the most frequent primary lung tumor in children under the age of 16 [3]. A case report with endobronchial IPT is presented.

CASE REPORT

A five-year-old girl was consulted at our outpatient clinic with a seven-month history of unresolving pneumonia.

The patient presented to a health care facility with persisting symptoms such as cough and recurrent lower respiratory tract infections. Although she had received antibiotic therapy she had persisting symptoms over a four-month period, resulting in hospitalization three months prior to presentation, with pneumonia and atelectasis at the basal zones of the left side of the thorax. Her symptoms improved with a 15-day course of antibiotics for pneumonia. Atelectasis due to foreign body aspiration was excluded with bronchoscopy and the patient was discharged. But 10 days ago when the patient's symptoms recurred as high fever and cough, she was sent to our department for investigation of the etiology of atelectasis and performance of a fiberoptic bronchoscopy.

It was learned that her mother had two pregnancies and two deliveries; our patient was born after a normal spontaneous vaginal delivery (40 weeks, 3500 g). Her mother reported smoking 3 cigarettes/day during the pregnancy. The elder sister of the case was 12 years of age and healthy. No remarkable features were noted in her family medical history other than tuberculosis in her father's uncle.

Physical examination revealed weight 16 kg (10-25%), height 104 cm (10-25%), heart rate 80/min, respiratory rate 20/min, and blood pressure 90/70 mmHg.

The auscultation of the lung fields revealed crepitant rale in left upper zone, left middle and lower zones, and the respiratory sounds were diminished in left lower hemithorax. Her complete blood count showed a white blood cell count of 12800 with 72% neutrophils. C-reactive protein (CRP) was 3.4 mg/dl. Biochemical tests were in normal ranges. On the work-up chest radiograph, left lower lobe infiltration and left mediastinal shift were observed (Figure 1). Computerized tomography (CT) scan showed pneumonic consolidation in the left lung, lower lobe atelectasis and mediastinal shift due to volume decrease (Figure 2).

Flexible fiberoptic bronchoscopy was performed to exclude associated malformations of the airways. Bronchoscopically, an endobronchial lesion was found in the left lower lobe bronchus. Although the lesion was thought to be an organized foreign body, biopsy was performed to exclude the possibility of a polyp or a tumoral lesion. Pathological examination of the biopsy revealed the lesion was compatible with a reactive-natured lesion. Because defi-

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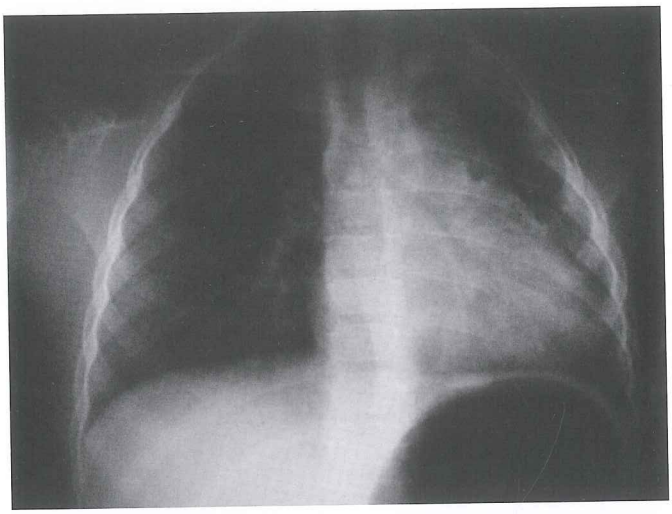

Figure 1. On chest radiograph, left lower lobe infiltration and left mediastinal shift were observed.

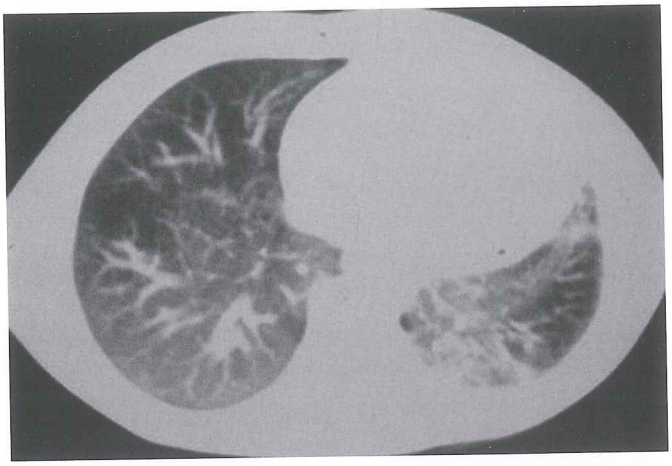

Figure 2. Computerized tomography scan showed pneumonic consolidation in the left lung, lower lobe atelectasis and mediastinal shift due to volume decrease.

nite diagnosis could only be provided by examination of the whole lesion, rigid bronchoscopy was performed and the lesion was excised. Microscopic examination revealed a tumor with spindle cells just under the epithelium and invasion of the deep tissue by inflammatory cells including plasma cells. These findings were typical of an inflammatory myofibroblastic tumor (Figure 3). Ten days after the bronchoscopy, there was significant improvement in atelectasis radiologically (Figure 4).

DISCUSSION

Initially described in 1937, IPT, inflammatory myofibroblastic tumor or plasma cell granuloma are synonymous terms for an inflammatory solid tumor that contains spindle cells, myofibroblasts, plasma cells, and histocytes [4]. Inflammatory pseudotumor of the lung (IPL) is by far the most common benign tumor of the lung in children, accounting for 84% of the cases. Childhood lesions account for 43% of all IPL cases. In the pediatric age group, IPL is commonly seen in older children, but has also been reported in children as young as 12 months of age [5]. Girls and boys are affected equally [6].

Although the lung is the best known and most common site, IPT occurs in diverse extrapulmonary locations, such as the skin, soft tissues, larynx, liver, mesentery, heart, spleen, stomach, small intestine, brain, spinal cord, kidney, renal pelvis, urinary bladder, major salivary glands, pancreas, thyroid, lymph nodes, breast and orbita [5,6,7,8]. Coffin et al. [8] reported their experience of 84 cases, and the sites of involvement included abdomen, retroperitoneum or pelvis (61 cases), upper respiratory tract (12 cases), trunk (8 cases), and extremities (3 cases).

Children present with fever in 22% of cases, cough in 20%, chest pain in 11%, hemoptysis in 9%, or pneumonia in 8% [5]. Our case presented with unresolving pneumonia. The etiology of unresolving pneumonia was obstructive atelectasis due to endobronchial IPT. Kim et al. [9] reported a series of 28 cases, and asymptomatic cases accounted for 11.1%; the locations of the lesions were parenchymal (85.7%), endobronchial (10.7%), and endotracheal (3.6%).

Although IPT is the most frequent primary lung tumor in children under the age of 16, clinical history and radiological diagnosis are often non-specific, making the diagnosis difficult. Usually surgery can provide a definite diagnosis. On chest X-ray, a solitary, well-demarcated, sometimes lobulated coin lesion in the periphery of the lung, frequently in the lower lobes, is the most typical image. In 15% of the cases, calcifications are detected, and in less than 10% ipsilateral effusion and in less than 7% hilar and mediastinal adenopathies are seen. Constriction and tapering of the vessels and bronchi, also present in our case, develop in 8-14% of cases [10].

IPT is a non-neoplastic reactive inflammatory condition. The concept of a benign lesion has been recently challenged by both clinical demonstration of recurrence and cytogenetic evidence of acquired clonal chromosomal abnormality [11,12]. Therefore, excisional surgery is the treatment of choice for ITL [13,14]. Such excisions, both diagnostic and curative, carry minimal risks and avoid unnecessary mutilation. Closely monitored follow-up is mandatory, as the natural history of this lesion is not yet well understood [15,16]. After the extirpation of the lesion, our case improved both clinically and radiologically and is being followed.

This case was presented in order to emphasize that performance of flexible bronchoscopy is important in chil-

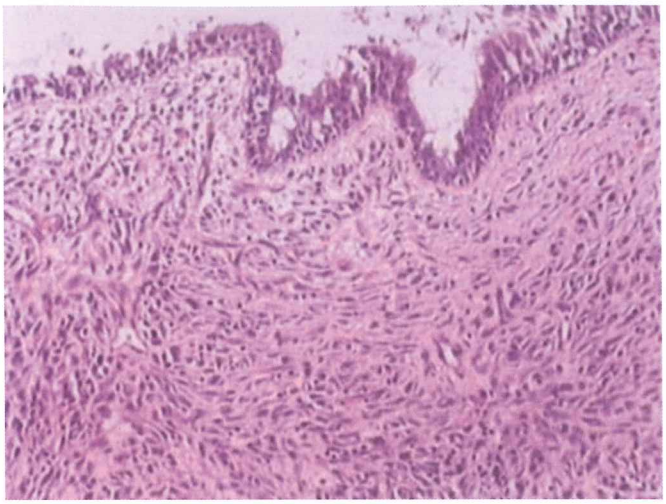

Figure 3. Microscopic examination revealed a tumor with spindle cells just under the epithelium and invasion of the deep tissue by inflammatory cells including plasma cells.

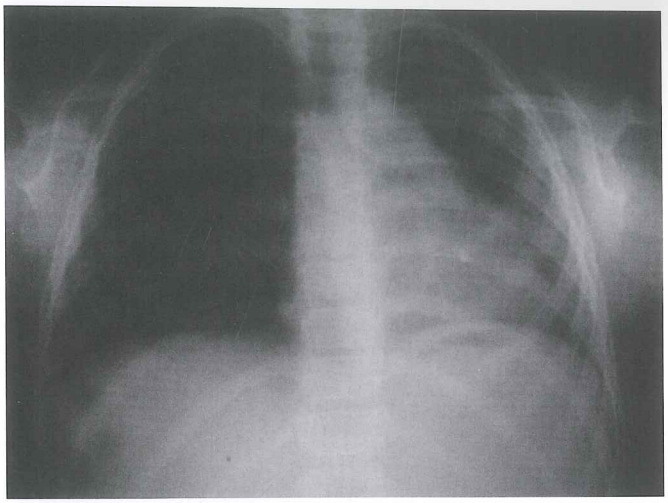

Figure 4. Ten days after the bronchoscopy, significant improvement in atelectasis was demonstrated radiologically.

dren as well as adults, and to remind physicians that inflammatory pseudotumor, which is a rare endobronchial tumoral lesion, should be considered in the differential diagnosis for atelectasis.

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