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### Reference

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# Inflammatory myofibroblastic tumor of the lung: a rare cause of atelectasis in children

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**Abstract** Although rare, inflammatory myofibroblastic tumor is the most common primary lung mass in children. We report the case of an 11-year-old boy investigated for persistent cough and dyspnea with complete left lung atelectasis mimicking pneumonia. CT and MRI showed an endobronchial mass of the left main bronchus. The boy underwent endoscopic resection of the tumor and histology was in favor of an inflammatory myofibroblastic tumor of the lung. This diagnosis should be suspected in children with recurrent pneumonia. The prognosis is good after complete resection.

**Keywords** Inflammatory myofibroblastic tumor · Atelectasis · Children

## Introduction

Inflammatory myofibroblastic tumor (IMT) of the lung is a rare benign tumor first described by Brunn in 1939, as cited by

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Jeba et al. [1]. It belongs to the group of soft-tissue tumors and is also called inflammatory pseudotumor, fibrous histiocytoma, plasma cell granuloma, plasmocytoma, solitary mast cell tumor, pseudoneoplastic pneumonia or fibroxanthoma.

IMT can involve any anatomical location, but the lung is the most commonly affected site in children as well as in adults. The other anatomical locations are head and neck, mesentery, liver and bladder. In children, IMT is the most common primary lung mass that can behave like malignant tumors. Clinical presentation and radiologic findings are variable and not specific. Diagnostic confirmation is based on histological examination.

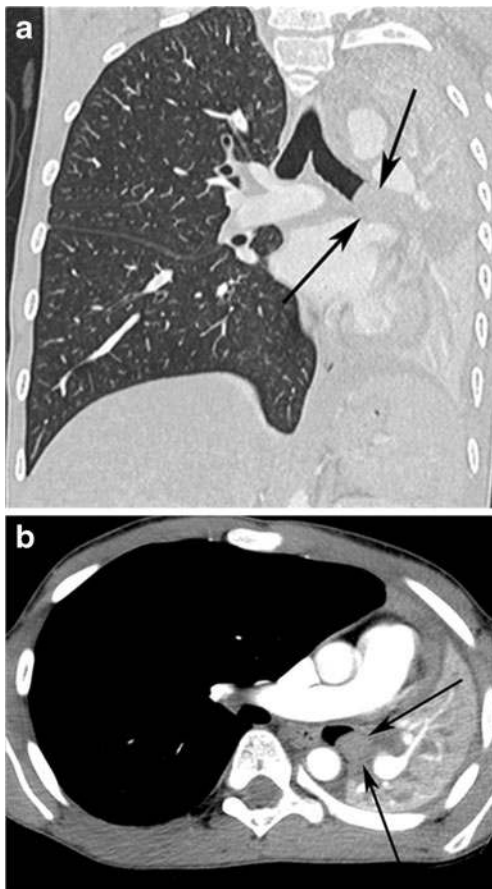
## Case report

An 11-year-old otherwise healthy boy had a 2-month history of dyspnea and coughs and was treated for asthma with no clinical improvement. One month after the onset of respiratory symptoms, he developed fever and the cough became persistent. Chest radiograph showed left lower lobe atelectasis. The diagnosis of pneumonia was established and antibiotherapy prescribed. One month later, he was referred to our emergency department because of unsatisfactory clinical course with complete left lung radiologic atelectasis (Fig. 1). White blood cell count, blood chemistry and CRP level were unremarkable. Tuberculin skin test was negative. Chest US showed pulmonary consolidation without significant pleural effusion. Chest CT was performed on a 16-slice machine (GE LightSpeed, Waukesha, WI, USA) after intravenous 2 ml/kg injection of a 300 mgI/ml contrast medium (CTDI of 6.44 mGy). Besides total atelectasis of the left lung with mediastinal shift to the left and compensatory overinflation of the right lung, CT showed a well-delineated homogeneous mass of 1.8×1.2×1.7 cm in the left main bronchus (Fig. 2). This lesion completely obstructed the

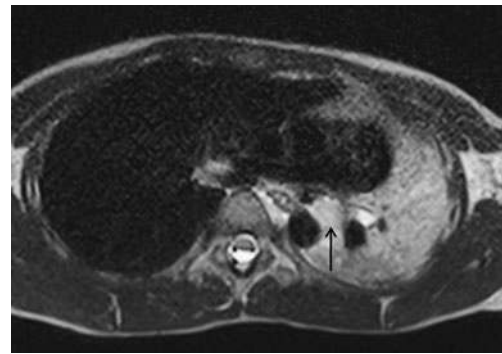


**Fig. 1** The chest radiograph shows complete left lung atelectasis with compensatory overinflation of the right lung

main bronchus and was not enhanced by contrast material. Chest MRI was performed on a 1.5-T (Magnetom Avanto, Siemens, Erlangen, Germany) and imaging protocol

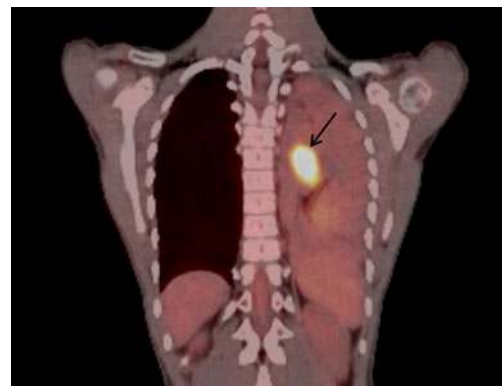


**Fig. 2** Contrast-enhanced and reformatted coronal (a) and axial (b) CT images show an obstructive homogeneous, well-circumscribed and non-enhanced mass in the left main bronchus (arrows)



**Fig. 3** Axial T2-weighted MR image shows a well-delineated mass (arrow) with high signal intensity in the left main bronchus and atelectasis of the left lung

included Balanced Steady-State Free Precession (bSSFP), coronal T1-weighted turbo spin-echo sequences (TR/TE 641/22 ms), axial and coronal T2-weighted turbo spin echo (TR/TE 1,000/80 ms) and fat-suppressed T1-weighted spin-echo sequences after intravenous administration of gadobutrol (Gadovist, Bayer, Zurich, Switzerland) at a dose of 0.1 mmol/kg body weight. MRI confirmed the presence of a homogeneous mass in the left main bronchus with high signal intensity on T2- and T1-weighted images with no significant contrast enhancement (Fig. 3). Whole-body MRI was also performed and the protocol included 3-D inversion recovery (inversion time=160 ms) T2-weighted turbo spin-echo (SPACE, TR/TE 2,000/151 ms, isotropic voxel dimensions, 1.4 mm), coronal T1-weighted spin-echo sequences (TR/TE 550/11) and diffusion. No other lesion was found. On [F-18]-fluoro-2-deoxyglucose (FDG) PET/CT (Biograph 16; Siemens, Erlangen, Germany), this mass showed increased metabolism (SUV max 21.8) (Fig. 4). No other hypermetabolic lesions were found on PET/CT. Lung volumes were markedly decreased with a restrictive syndrome (total lung capacity 55% of predicted) on spirometric studies. Bronchoscopy visualized the endobronchial tumor, which was obstructing incompletely the distal end of the left main bronchus (Fig. 5). The polypoid shape



**Fig. 4** Fused coronal PET/CT image demonstrates intense 18F-FDG uptake in the pulmonary mass (arrow)



**Fig. 5** Endoscopic view of the left main bronchus shows an obstructive endobronchial mass

of the tumor allowed total endoscopic resection with a diathermic snare combined with argon plasma coagulation and no complications. Histological analysis revealed a mesenchymal tumor more commonly called inflammatory myofibroblastic tumor. Cytogenetic analysis confirmed this diagnosis with positive characteristic rearrangement at 2p23.9 of the gene coding for the anaplastic lymphoma kinase receptor tyrosine kinase (found in 50% of IMT cases). Chest radiographs after resection showed complete resolution of the atelectasis. Spirometry showed normal total lung capacity, and repeated bronchoscopies at 1-month and 3-month follow-ups were normal.

## Discussion

Most of the causes of atelectasis in childhood are related to airway obstruction. Intra- and extrabronchial obstruction may be exogenous due to foreign body, or to mucous plugs. Endobronchial tumors are a rare cause of atelectasis.

IMT is a rare lesion; it belongs to the group of soft-tissue tumors and has also been called inflammatory pseudotumor, fibrous histiocytoma, plasma cell granuloma, plasmocytoma, solitary mast cell tumor, pseudoneoplastic pneumonia, and fibroxanthoma. This is probably due to its predominant cellular component. It can involve any anatomical location; however, the lung is the most commonly affected site [2]. Other frequent locations include the bladder, mesentery, omentum, retroperitoneum, pelvis, gastrointestinal tract, liver, spleen, kidney and pancreas. Some tumors can be multifocal and involve several organs. The true incidence of IMT of the lung is difficult to establish because of various nomenclatures used. However, it is relatively rare among all primary lung neoplasms, accounting for only 0.04%–0.7% of them [3]. According to many series,

IMT is the most common primary childhood lung tumor, with an incidence ranging from 12% to 57% [2]. There is a near equal male/female occurrence [2, 4].

Pathophysiologically, an unregulated proliferation of inflammatory cells seems to be present [3]. The cause of this inflammatory response remains unknown. It has been hypothesized that an initial infection leads to IMT, and about 30% of patients have had a history of respiratory infection. In a case report Fiocchi et al. [5] described a direct progression from infectious disease to an IMT. Inflammatory lesions are thought to occur locally as a result of excessive response to tissue damage [6]. Others believe it to be a real neoplasm induced by genic reorganizations on the chromosome 23 [7].

Most frequent symptoms are cough, hemoptysis, chest pain, dyspnea and fever. It may also be asymptomatic [7].

Radiologic findings are variable and unspecific. The most common radiologic presentation is a solitary peripheral well-circumscribed lung mass, with variable size between 1.5 cm and 14 cm according to a series of 28 cases reported by Kim et al. [8]. The endobronchial polypoid forms arising in the trachea or the bronchus are the rarest but have also been reported, as in our case. They can show atelectasis and obstructive pneumonia and can spread into the mediastinum.

Contrast-enhanced CT or MRI is necessary to view the endobronchial mass that is not visible on the chest radiograph. CT and MRI allow a better characterization and delineation of the tumor. Contrast-enhanced CT shows a homogeneous or heterogeneous attenuated mass. Intralesional calcifications are possible but rare, and are shown more frequently in pediatric cases [4, 7]. IMTs are also known to rarely form a cavity. In such cases, MRI shows intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Absent or delayed enhancement with contrast injection on CT or MRI is described in the literature. Takayama et al. [4] reported two cases of IMT that were intra-pulmonary well circumscribed masses. Dynamic contrast enhancement CT in both cases showed delayed enhancement. They speculate that the delayed enhancement could be attributed to abundant fibrous tissues such as myofibroblasts [4]. High fixation of 18F-FDG of IMT on PET has been described [7] and may be used for assessment of multifocal forms.

Concerning transbronchial biopsy and transthoracic needle biopsy, Cerfolio et al. [2] believe that they often lead to confusion between IMT and fibrohistiocytic neoplasms, nodular sclerosing Hodgkin lymphoma, primary lung cancer and mediastinal fibrosis. Indeed, histological findings of a fragment cannot exclude a neoplasm with surrounding inflammation. Complete resection for both diagnosis and treatment is mandatory.

While inflammatory myofibroblastic tumor is a rare endobronchial tumor, it should be known as a possible cause of atelectasis in children and distinguished from a malignancy. The treatment is surgical, and a favorable prognosis relies on complete resection.

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