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CASE REPORT

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# Inflammatory myofibroblastic tumor of the lung: presentations of an uncommon finding

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## Abstract

*Inflammatory myofibroblastic tumor (IMT) is a rare occurrence most commonly affecting the pediatric population. It is considered a diagnostic challenge because of nonspecific clinical symptoms and radiologic presentations. IMT is a soft-tissue tumor that can occur throughout the body but is most commonly found in the lung. This article discusses the case of a 10-year-old girl who presented to our hospital with abnormal chest radiograph. Imaging revealed a heterogenous mass in the superior aspect of the left lower lobe. It was found incidentally when the patient presented to the emergency department. Surgical resection proved a diagnostic and therapeutic measure confirming diagnosis of IMT. The tumor has potential to cause illness either by its rare malignant potential or by mass effect. Because lesions are often asymptomatic with a predilection for pediatric patients, cases describing appropriate management are essential. IMT is a rare tumor of borderline malignant potential. IMT of the lung may invade hilar structures, the mediastinum, and the pleura. Lesions range in diameter from 1.5 to 14 cm. Etiology of the tumor is unclear but may include immune response to infection, trauma, or surgery. Affected patients may present with cough and shortness of breath but are often asymptomatic. Histology is unique but not specific. Immunohistochemistry can differentiate the tumor, by staining for overexpression of the anaplastic lymphoma kinase oncogene. In conclusion, surgical resection is the preferred diagnostic and therapeutic regimen for IMT. The majority of IMTs follow a benign course, but exceptions qualify as low-grade malignancies. As a result, it is important to identify, treat, and maintain vigilance. This case encompasses presentation, diagnosis, and treatment of pulmonary IMT and highlights best practices in addressing IMT for adolescent patients.*

**Keywords:** *inflammatory myofibroblastic tumor, lung neoplasm, plasma cell granuloma, pediatric, pulmonary nodule, thoracotomy*

## Introduction

This case describes the presentation of a rare lung tumor and best practices of how to treat it. Inflammatory myofibroblastic tumor (IMT) is a tumor of borderline malignant potential more common in patients younger than 40 years, especially in children and young adults [1]. Diagnosis of the tumor is most accurately achieved by observing its histology. As a result, IMT is best diagnosed by surgical resection, which is also the most effective therapeutic measure. This case report presents clinical, radiologic, and pathologic features of pulmonary IMT as addressed at an academic children's hospital. This paper has been reported in line with SCARE criteria [2].

## Case report

A 10-year-old girl presented to the emergency department after experiencing severe left shoulder pain after hitting her clavicle while completing a pull up at school. A chest radiograph was performed at an outside hospital and was suspicious for hilar enlargement (Figure 1A). The patient was referred to pulmonology, who recommended follow-up chest radiograph in 6 weeks. At time of follow up, radiology confirmed that the left perihilar opacity appeared to have increased in size compared with about 6 weeks prior (Figure 1B). Physical exam was unremarkable at both visits. The patient did not complain of shortness of breath and was able to participate actively in athletic competition with her peers. Complete pulmonary function testing showed normal spirometry but increased airway resistance and mild air trapping that could suggest mild obstructive lung disease. Additionally, her diffusion was mildly elevated corrected for alveolar volume. This was initially interpreted as incidental and not clinically significant. However, a tumor with increased vascularity or increased metabolic demand could falsely increase the uptake of carbon monoxide.

Although the appearance of the lesion could have represented atelectasis or pneumonia, enhanced computed tomography (CT) of the chest was recommended based on interval enlargement of the lesion. The CT exam revealed a 2.7 x 2.7 x 2.3 cm lobulated, heterogenous mass along the major fissure, primarily within the superior segment of the left lower lobe lung but extending into the upper lobe (Figures 2 and 3). Differential diagnosis based on imaging included IMT, hamartoma, carcinoid, chondroma, or other sarcoma. The lesion was thought to be benign but likely locally aggressive as it extended from the lower lobe into upper lobe. Core biopsy was thought to be risky because of proximity to pulmonary artery. After review of CT results, differential diagnoses, and therapeutic

options, it was determined that surgery should be performed for excisional biopsy.

During surgery, a left posterior lateral thoracotomy incision was made, and a muscle-sparing approach to the chest wall was employed at the left fifth intercostal space. A firm, round 4 cm mass was identified in the superior portion of the left lower lobe extending across the fissure and into a small portion of the left upper lobe. There were no other palpable masses and there were not identifiably enlarged lymph nodes. The lateral portion of the oblique fissure was exposed, vessels were ligated, and the tumor was removed via segmentectomy. Segmental lower lobe resection and wedge upper lobe resection were performed to resect the mass with 2 cm margins.

Upon excision, the specimen was sent fresh to surgical pathology. Fluorescent in situ hybridization results were negative for rearrangement of the anaplastic lymphoma kinase (ALK) gene at 2p23 locus. From these findings, the lesion was diagnosed as IMT. After pathologic sectioning and analysis, the lesion was diagnosed as IMT (Figure 4).

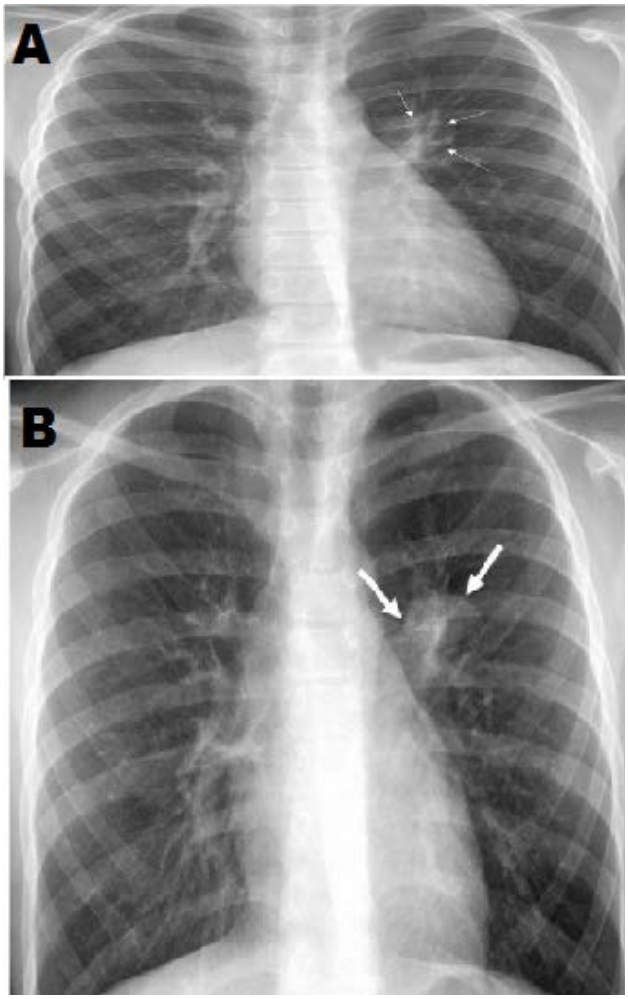


**Figure 1:** *(A) This posteroanterior chest radiograph shows hazy opacity projecting around the left hilum (small arrows). The chest is otherwise normal. (B) A second chest radiograph performed 6 weeks later again demonstrates the lesion. The margins of the lesion are now well defined (large arrows). The differential diagnosis includes mass, bulky lymphadenopathy, or vascular lesion. Round pneumonia is not considered because of the clinical history. Chest CT with contrast was advised.*

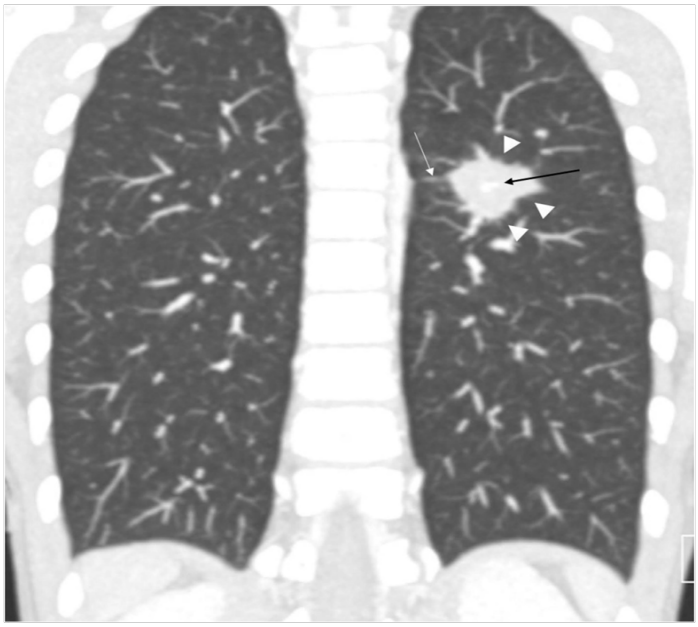

***Figure 2:*** *This 2-mm thick coronal reformatted image from the contrast-enhanced CT of the chest shows a solid mass with a central, coarse calcification (black arrow). The margins of the lesion are speculated (arrowheads), a finding that suggests infiltration into adjacent lung parenchyma. This finding can be seen with aggressive or infectious processes. There is stranding medial to the lesion (white arrow), a finding that can indicate lymphatic obstruction or discoid atelectasis.*

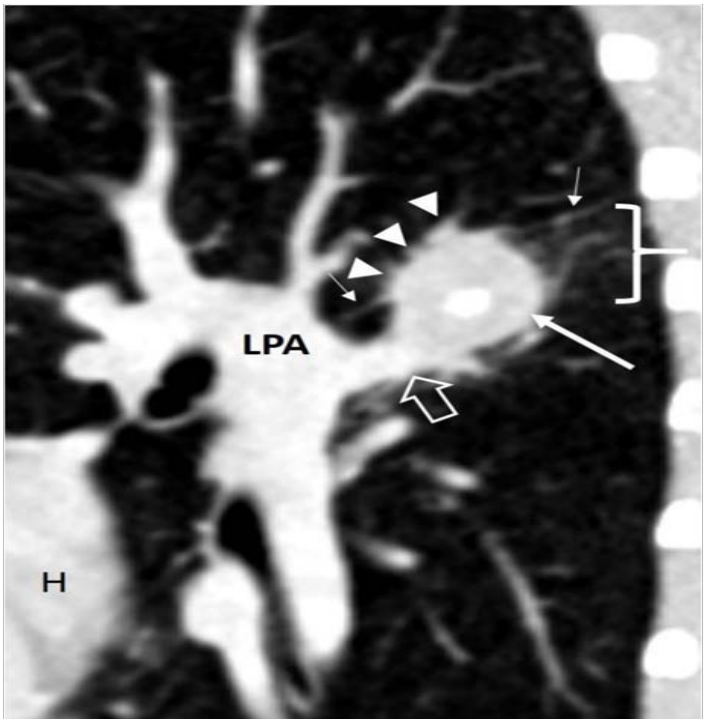


***Figure 3:*** *This 2 mm thick sagittal reformatted image from the contrast-enhanced CT of the chest again demonstrates the mass (large arrow) within the superior segment of the left lower lobe abutting the major fissure (small arrows). Spiculation of the margin of the mass (arrowheads) seen cranial to the major fissure identifies extension of the lesion across the fissure into the upper lobe. The mass is contiguous with the posterior margin of the superior segmental branch of the left lower lobar pulmonary artery (LPA). Ground glass opacity (bracket) posterior to the mass is most likely the result of volume loss due to small airway compromise. The differential diagnosis includes inflammatory myofibroblastictumor, hamartoma, carcinoid, or other sarcoma. H = heart.*

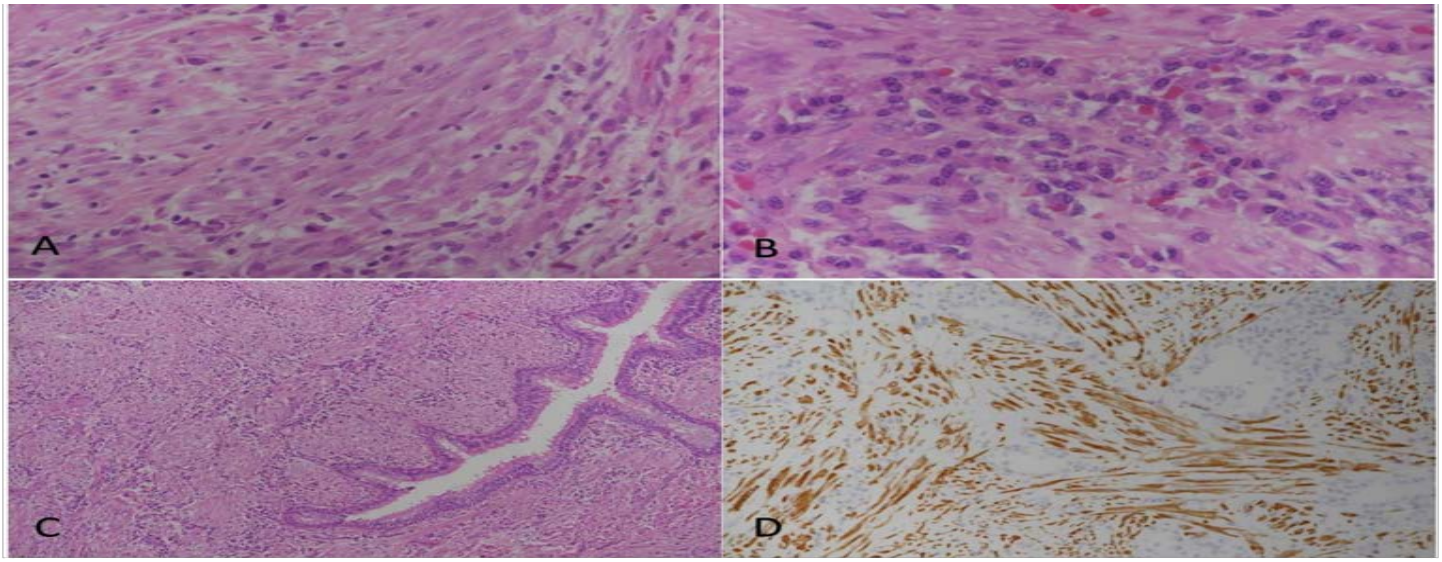


***Figure 4:*** *Photomicrographs of the inflammatory myofibroblastictumor showing interlacing fascicles of spindle cells with monomorphic nuclei containing small inconspicuous nucleoli with open chromatin pattern (A), numerous plasma cells infiltrating the proliferating spindle cells--no mitotic figures, atypical nuclei, or foci of necrosis were found (B), an incorporated non-neoplastic bronchiole, foci of lymphoid clusters, dystrophic calcification, and collagen deposition were also present, borders of the lesion were infiltrative and, in places, surrounding entrapped pulmonary blood vessels and bronchioles (C), and anaplastic lymphoma kinase-1 immunohistochemical reactivity in the lesional cells (D).*

## Discussion and conclusion

Inflammatory myofibroblastic tumor is still considered a tumor of borderline malignant potential because of its possibility of recurrence, tendency to be locally invasive, and history of malignant transformation. Recurrence is recorded in approximately 25% of patients related to the location, resectability, and multinodularity of the tumor [3]. Recurrent tumors occur in 4% of cases after incomplete resection, which is more frequent in patients with tumors that infiltrate more sensitive areas such as the hilum [4,5]. While metastasis is rare, it is a possibility and occurs in 5% of cases [3,5,6]. As a result, surgical resection was the best option in this case and should be considered in all presentations of pediatric inflammatory myofibroblastic tumor.

Inflammatory myofibroblastic tumor is reported to affect males and females equally. There is a slight predominance among children and young adults, although the tumor can occur throughout life [5]. The average age of patients with IMT is 10 years. Almost half of patients with IMT are asymptomatic with tumors incidentally diagnosed on radiology [7]. This was true in the case of our 10-year-old patient whose tumor was discovered on chest radiograph taken upon visiting the emergency department at an outside institution for complaint of unrelated pain. The case highlights the importance of remaining vigilant in all patient populations and pursuing a comprehensive work up of suspicious findings.

Clinical symptoms of IMT vary based on location of the tumor. Lung IMT most commonly presents with cough, shortness of breath, chest pain, and hemoptysis. Many times IMT is asymptomatic leading to its tendency toward incidental discovery. In the case presented, physical exam was unremarkable and history did not indicate clinical symptoms. The tumor was found incidentally. The course of treatment exemplifies appropriately aggressive follow up and emphasizes the importance of thorough examination in the event of a suspicious mass. Despite its relatively benign presentation, the possibility for more aggressive growth and metastasis in IMT and other differential diagnoses is cause for a clinical course similar to the one described in this case.

Computed tomography with contrast is a preferred means of imaging IMT as it provides enhancement of tissue as well as vascular behavior and regional extension. Inflammatory myofibroblastic tumor appears as a heterogenous nodule or mass and may be accompanied by focal atelectasis, pleural effusion, or vascularization. The present case showed focal areas of atelectasis around the tumor. Intralesional calcifications are commonly seen in children and exhibit a range of patterns such as amorphous, mixed, or fine fleck-line [7,8]. Calcification in this case was seen on CT and confirmed by pathology.

On gross examination, the tumor is a well-circumscribed mass rather than an infiltrative process. It has a tan, white, or yellow color to its whorled fleshy or myxoid cut surface [3,9]. Grittiness may be felt on sectioning because of the microcalcifications.

Histologic characteristics, together with the immunohistochemical staining profile of the tumor, favor the diagnosis of IMT and differentiate it from other neoplastic processes. Inflammatory myofibroblastic tumor displays a characteristic combination of bland-appearing myofibroblastic spindle cells with variable inflammatory cells in the background. The inflammatory infiltrate is often composed of a large number of lymphocytes, plasma cells, eosinophils, and neutrophils [4,10].Histology of the tumor in this case confirmed its diagnosis as IMT.

Histology of IMT is unique but is not entirely specific to the diagnosis. Immunohistochemical stains may be useful in identifying the tumor. Overall, the spindle cells of the tumor have characteristics of myofibroblasts causing positive results for vimentin in addition to muscle-specific proteins such as muscle-specific actin and smooth muscle actin in a majority of cases [4,10]. A common feature of IMT is overexpression of ALK protein, a protein tyrosine kinase receptor encoded on the second chromosome at 2p23 [6]. Inflammatory myofibroblastic tumor is positive for ALK rearrangement and overexpression in over half of cases [4]. In one study of 36 IMT cases conducted by Saab et al. [9], 64% of tumors

| Study | Age | Gender | Localization | Size (mm) | Follow-up period | ALK translocation or over expression |
|---|---|---|---|---|---|---|
| Carvalho (11) | 12 y | M | middle right lung | 6.0 x 2.5 x 1.7 cm | 6 mo | negative |
| Carvalho (11) | 39 y | M | upper right lung | -- | -- | positive |
| Degheili (12) | 43 y | F | upper left lung | 3.0 x 3.0 cm | 10 years | negative |
| Genchellac (7) | 4 y | F | inferior left lung | 4.9 x 5.5 x 6.2 cm | 2 years | -- |
| Hammas (5) | 3 y | -- | lower left lung | 8.5 cm | -- | positive |
| Tsuchiya (13) | 12 mo | M | right upper lung | 0.17 cm | -- | positive |
| Tsuchiya (13) | 8 mo | M | right middle lung | 0.24 cm | -- | positive |
| Zennaro (14) | 26 y | F | middle right lung | 3.0 cm | 12 mo | -- |

***Table 1:*** *Reported cases of pulmonary inflammatory myofibroblastictumor.ALK: Anaplastic Lymphoma Kinase.*

were ALK positive. The tumor in this case was negative for ALK rearrangement.

The features discussed above are characteristics common to IMT. Past case reports have described a collection of tumors that display these identifying features to varying degrees. Because presentation is variable, it is helpful to examine the body of literature available on the topic (Table 1). This summary of published reports of IMT found in the lung exemplifies the diverse collection of common features displayed by this lesion.

Preferred treatment of IMT is total surgical excision as a diagnostic and therapeutic regimen. Depending on the quality of the surgical resection, this tumor may have a 5-year survival rate of 91.3% [5].Although surgical resection in children is challenging, complete removal of the tumor is important in promoting good prognosis [11-15]. The surgical procedure in this case provided complete resection of the tumor and portends a good prognosis for the patient involved. The patient has proceeded through the post-operative course with no complications and will return in one year for follow-up imaging.

In conclusion, inflammatory myofibroblastic tumor is a rare tumor of borderline malignant potential most commonly found in the lung in children with a mean age of 10 years. Surgical resection by an experienced surgeon is the preferred diagnostic and therapeutic regimen. The tumor is often discovered incidentally, and often characterized by nonspecific clinical and radiological presentation. Diagnosis is confirmed by histology and immunohistochemistry. The majority of IMTs follow a benign course, but exceptions qualify as low-grade malignancies. As a result, it is important to identify, treat, and maintain vigilance.

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