Radiology Case Reports 20 (2025) 2468–2471

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Insights into inflammatory myofibroblastic tumor: A complex and challenging diagnosis ☆,☆☆

*Hajra Arshad, MD**, *Charles K. Crawford, BS*, *Elliot K. Fishman, MD*

The Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University, Baltimore, MD, USA

ARTICLE INFO

Article history:
Received 23 January 2025
Accepted 4 February 2025

Keywords:
Inflammatory myofibroblastic tumor
Computed tomography (CT)
Positron emission tomography (PET)

ABSTRACT

Inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor or plasma cell granuloma, is an uncommon soft tissue tumor of mesenchymal origin with low to intermediate malignant potential. IMTs are most commonly located in the lung, abdomen/pelvis, or retroperitoneum, primarily affecting children and young adults. Although metastasis is rare, IMTs can recur after surgical resection, as oftentimes resection is limited due to infiltration with surrounding tissue. Here, we present a challenging case of IMT in a 22-year-old male who presented with chest pain and shortness of breath. Extensive diagnostic workup, including multiple biopsies and imaging studies, raised suspicion of malignancy, though initial biopsies failed to confirm a diagnosis. This case highlights the diagnostic difficulties and the importance of a coordinated approach in the diagnosis and management of IMT.

Introduction

Inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor or plasma cell granuloma, is a rare soft tissue tumor of mesenchymal origin. It most commonly affects children and young adults with a median age of 9 occurring commonly in the lungs, abdomen/pelvis, or retroperitoneum [1]. Histologically, they consist of myofibroblastic spindle cells and an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils [2]. They are considered to be intermediate malignant capacity with frequent recurrences and rare metastasis [3]. The mainstay of treatment is surgical resection [4]. Due to its rarity and complex nature, it is a challenging diagnosis and requires a coordinated review of radiology and histopathology. We present a challenging case of IMT in the chest, which was diagnosed after multiple biopsies and resections. It is important to report such cases to add to available literature regarding the complexity of such cases.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

☆☆ Acknowledgments: There was no funding associated with this report.

* Corresponding author.
E-mail address: harshad1@jh.edu (H. Arshad).

https://doi.org/10.1016/j.radcr.2025.02.005

Case presentation

A young 22-year-old male, training in the Marines, with a family history of Brugada syndrome, presented with an episode of chest pain and shortness of breath during training. On further workup in the hospital, a large, lobulated, calcified mass was found in the left apical chest extending from apex to hilum with confluent adenopathy, described in Fig. 1. Fluorodeoxyglucose (FDG)-positron emission tomography (PET) scan was performed, which illustrated heterogenous FDG-avidity in the mass (Fig. 2). He underwent several biopsies which were inconclusive making the case very challenging to diagnose.

An initial biopsy from an outside institution suggested spindle cell sarcoma, but reviewing the material at our hospital did not reveal any evidence of malignancy.

Then, he underwent left-sided video-assisted thoracic surgery (VATS) with a biopsy of the thoracic mass. Intraoperatively, a large mass was found attached to the upper chest extending between lobes via the fissure and obscuring the posterior mediastinum. Numerous tumor biopsies were obtained and sent for pathology. After extensive testing, pathologic examination of the specimens did not reveal evidence of neoplastic tissue. There was dense inflammatory tissue with lymphoplasmacytic infiltration without identifiable malignancy. Given the large, calcified mass, the pathology was unsatisfying, and there was some uncertainty about whether the sampled (outer) portion of the tumor was in fact representative of the central calcified portion of the tumor. Therefore, Interventional Radiology (IR) - guided biopsy was recommended to target the center of the lesion.

The patient underwent IR-guided biopsy which revealed a dense fibroinflammatory infiltrate similar to the previous biopsy, and focal calcifications which were not present in the previous one, however, it was still insufficient for a definitive diagnosis. However, a repeat CT scan reported interval growth of the mass. An extensive review of the correlation between imaging, pathology and overall history still showed concern for malignancy, thus surgical resection of the mass was planned.

An initial attempt at resection through thoracotomy was complicated by an episode of ventricular fibrillation, leading to abortion of the procedure. Return of spontaneous circulation was achieved with chest compressions, 2 doses of epinephrine and 3 shocks. Later on, he finally underwent resection of the left mediastinal tumor and left upper lobectomy of the lung, which removed almost half of the tumor. The final diagnosis on pathology was a fibro-inflammatory process with dense fi-

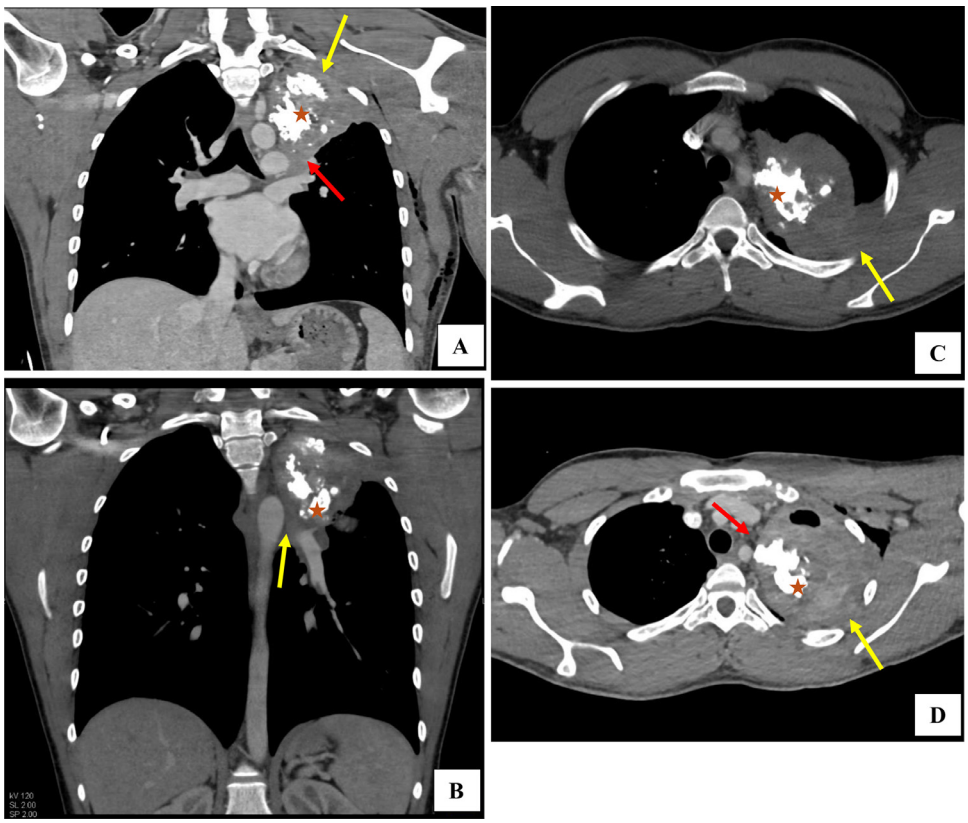

Fig. 1 – A 22-year-old male presenting with chest pain and shortness of breath during training. Coronal (A and B) and axial (C and D) CT scan shows a large lobulated soft tissue mass with coarse calcifications (orange star) in the left lung apex. The mass extends to the apical (A – yellow arrow) and posterior chest wall (C, D – yellow arrow), without invasion or significant mass effect on surrounding structures. Inferiorly, it extends to the left hilum and along the oblique fissure (A – red arrow). There is preserved fat plane between the mass and the aortic arch (B – yellow arrow). Left hilar lymphadenopathy is also seen (D – red arrow).

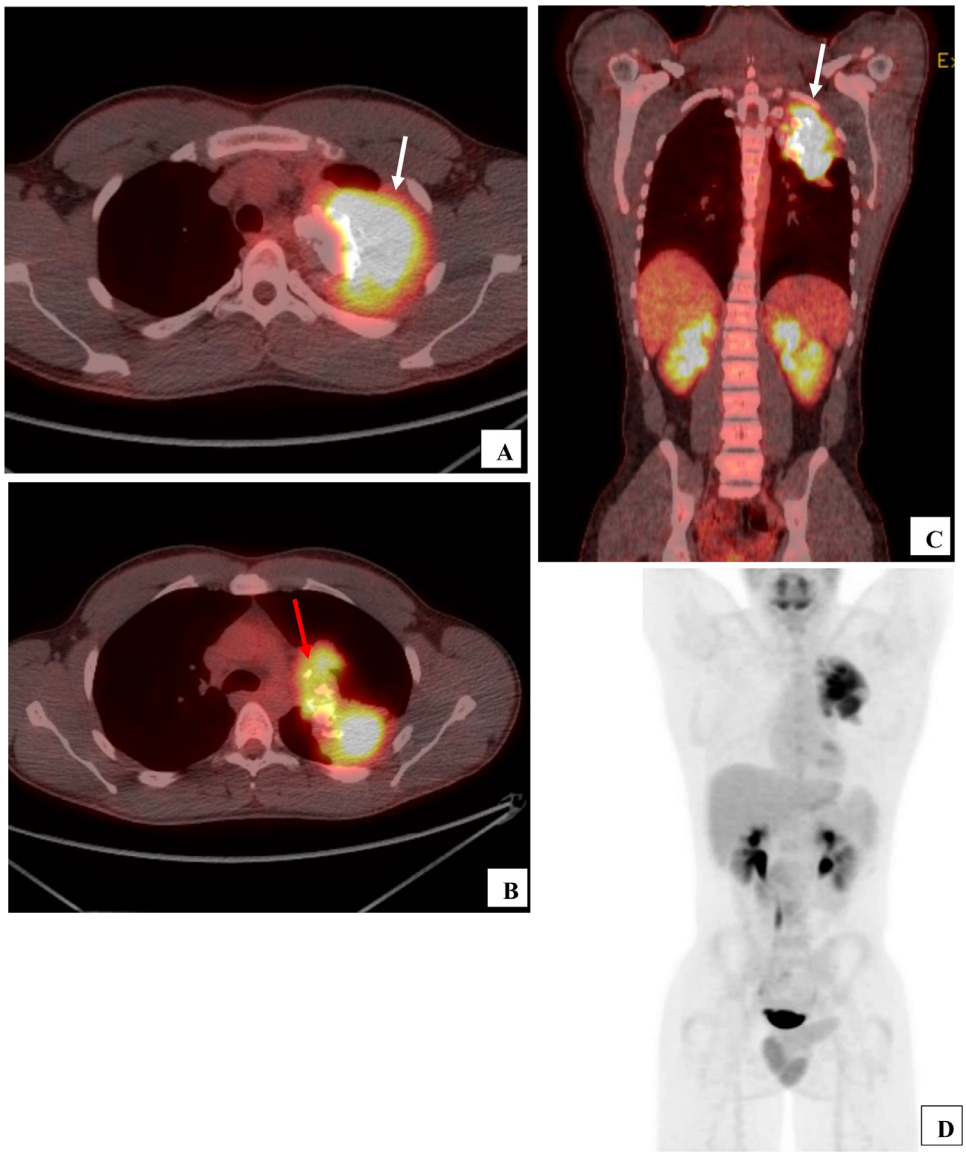

Fig. 2 – A 22-year-old male with a left chest apical mass, suspicion of a malignant process. Fluorodeoxyglucose (FDG)-positron emission tomography (PET) scan demonstrates a heterogeneously FDG avid partially calcified irregular mass in the upper lobe of the left lung (A, C – white arrow), confluent with hypermetabolic ipsilateral hilar lymphadenopathy (B – red arrow). No additional suspicious FDG avid lymph nodes or distant metastasis were identified (D).

brosis and calcifications, best regarded as an unusual (old) inflammatory myofibroblastic tumor (IMT). Fluorescence In Situ Hybridization (FISH) for ALK and ROS1 rearrangement were within normal limits.

The patient was kept on regular follow-ups with chest CT scan to monitor the residual mass size which has remained stable.

Discussion

Inflammatory myofibroblastic tumor (IMT) presenting in the first and second decade of life is a rare tumor with intermediate malignant potential [1,3]. It has a variable clinical and radiologic presentation, mimicking malignant tumors. The lung is the most common location for IMT. Clinically, lung IMT presents with symptoms of cough, chest pain, dyspnea, hemoptysis, and sometimes unspecified inflammatory symptoms of fever, malaise and weight loss may be present [5]. They may exert mass effect on surrounding structures as they grow larger.

On chest CT imaging, it typically presents as a well-defined, lobulated, or round solitary peripheral pulmonary nodule or mass. In some cases, it may appear as an area of consolidation or a nodule with ill-defined margins or spiculations, imitating lung cancer. The internal architecture can be homogeneous or heterogeneous, often exhibiting features such as hemorrhage, necrosis, and calcification, with variable degrees of contrast enhancement due to the presence of fibrosis [5,6].

Our patient had an ill-defined large left hilar lobulated soft tissue mass with coarse central calcifications along with hilar lymphadenopathy, suggestive of a malignant process.

Histopathological findings demonstrate dense proliferation of various spindle-shaped cells (myofibroblasts) with collagenous stroma and inflammatory cell infiltration, mainly plasma cells and lymphocytes [7]. Interestingly, our patient showed a bland myofibroblastic spindle cell proliferation eroding into the lung along with a dense lymphoplasmacytic inflammatory component in a severely sclerotic and hyalinized background stroma with scattered calcification, reflecting an old, and fibrotic IMT.

A study reported that approximately two-thirds of IMTs have anaplastic lymphoma kinase (ALK) and ROS1 gene rearrangements, which play a crucial role in both their pathogenesis and diagnosis [8]. ALK is a receptor tyrosine kinase, which becomes oncogenic due to gene fusion leading to overexpression of ALK protein, which is commonly used as a diagnostic and prognostic marker. They also identified a case RET mutation in a lung IMT. It has also been noted in the literature that ALK-positive cases are associated with improved outcomes, whereas ALK-negative IMTs tend to exhibit more aggressive behavior, and a higher incidence of metastasis compared to their ALK-positive counterparts [9,10]. Despite repeat testing on multiple biopsy samples, our patient did not test positive for ALK or ROS1 gene rearrangements.

The principal treatment is surgical resection with a favorable outcome, however complete resection might not be possible due to adhesions to surrounding structures. A tumor recurrence rate of 20% has been reported in the literature with positive surgical margins identified as a significant risk factor for tumor recurrence and mortality [11]. CT scans play a crucial role in monitoring IMT progression and postsurgical follow-up by providing detailed visualization of tumor size, internal structure, and enhancement patterns, allowing for precise quantitative assessment of changes over time. Regular CT follow-ups, especially in the first few years after treatment, are essential for detecting local recurrence, evaluating treatment efficacy, and guiding long-term management strategies for IMT patients.

Conclusion

Inflammatory myofibroblastic tumors (IMTs) are rare, intermediate malignancy tumors with variable clinical and radiological presentations, often mimicking more aggressive malignancies. Due to their rarity, IMTs are often challenging to diagnose, requiring a combination of radiological imaging, histopathology, and deep analysis of clinical history. Surgical resection is the primary treatment, and while recurrence rates are low, close postoperative serial surveillance is essential, especially within the first few years. This case emphasizes the importance of a multidisciplinary approach in diagnosing and managing IMTs and contributes valuable insight into the complexities surrounding these tumors. Further studies are necessary to better understand their pathophysiology and refine diagnostic and treatment strategies.

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for participation in research, which includes the permission to use data collected in future research projects such as the presented case details and images used in this manuscript.

REFERENCES

[1] Cantera JE, Alfaro MP, Rafart DC, Zalazar R, Muruzabal MM, Barquín PG, Pérez IV. Inflammatory myofibroblastic tumours: a pictorial review. Insights Imag 2015;6:85–96.

[2] Mehta B, Mascarenhas L, Zhou S, Wang L, Venkatramani R. Inflammatory myofibroblastic tumors in childhood. Pediatr Hematol Oncol 2013;30(7):640–5. doi:10.3109/08880018.2013.816810.

[3] Kube S, Vokuhl C, Dantonello T, Scheer M, Hallmen E, Feuchtgruber S, et al. Inflammatory myofibroblastic tumors—A retrospective analysis of the Cooperative Weichteilsarkom Studiengruppe. Pediatr Blood Cancer 2018;65(6):e27012. doi:10.1002/pbc.27012.

[4] Kube S, Vokuhl C, Dantonello T, Scheer M, Hallmen E, Feuchtgruber S, et al. Inflammatory myofibroblastic tumors—A retrospective analysis of the Cooperative Weichteilsarkom Studiengruppe. Pediatr Blood Cancer 2018;65(6):e27012.

[5] Raitio A, Losty PD. Treatment and outcomes in pediatric inflammatory myofibroblastic tumors–A systematic review of published studies. Eur J Surg Oncol 2024;3:108388. doi:10.1016/j.ejso.2024.108388.

[6] Fletcher CD. Pathology and genetics of tumors of soft tissue and bone. In: Classification of Tumors, 4. World Health Organization; 2002. p. 35–46.

[7] Coffin CM, Hornick JL, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J Surg Pathol 2007;31(4):509–20.

[8] Antonescu CR, Suurmeijer AJ, Zhang L, Sung YS, Jungbluth AA, Travis WD, et al. Molecular characterization of inflammatory myofibroblastic tumors with frequent ALK and ROS1 gene fusions and rare novel RET rearrangement. Am J surg Pathol 2015;39(7):957–67.

[9] Furuya K, Yasumori K, Takeo S, Sakino I, Uesugi N, Momosaki S, et al. Lung CT: part 1, mimickers of lung cancer??? Spectrum of CT findings with pathologic correlation. Am J Roentgenol 2012;199(4):W454–63. doi:10.2214/AJR.10.7262.

[10] Gleason BC, Hornick. JL Inflammatory myofibroblastic tumours: where are we now? J Clin Patholo 2008;61(4):428–37.

[11] Wang Z, Zhao X, Li K, Yao W, Dong K, Xiao X, et al. Analysis of clinical features and outcomes for inflammatory myofibroblastic tumors in China: 11 years of experience at a single center. Pediatr Surg Int 2016;32:239–43.