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CASE REPORT

A case of Pulmonary Inflammatory Fibroblastic Tumor with Clinical, Radiological, Histopathological Features and 2-year follow-up Results and Review of the Literature

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SUMMARY

Inflammatory myofibroblastic tumor (IMT) is a mesenchymal neoplasm. So in any organ, IMT has the potential of development. The most common sites that it arises are the lungs. It can occur in any age, but predominantly in children and adolescents. The etiology is not known. It is poorly understood on genetic and molecular level either. Clinical symptoms and radiological features are nonspecific in pulmonary IMT, can imitate lung cancer or tuberculosis. Diagnosis is based on histopathologic or immunohistochemical evaluation. The biological behavior is highly unpredictable, rarely metastase, frequently re-occure. Therapeutic approach rely mainly on complete surgical resection although there is no guideline for the treatment or follow-up. Here in, we report a pulmonary IMT case with clinical, radiological, histopathological features, and 2 year follow up results after complete surgical resection.

Keywords: Inflammatory pseudotumor; lung neoplasms; pulmonary inflammatory myofibroblastic tumor (pulmonary IMT); plasma cell granuloma; tuberculosis; xanthogranuloma.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare soft tissue tumor that arises from the mesenchyme. IMT was previously described as a benign, reactive lesion consisting of myofibroblasts and inflammatory cells. However, based on publications about metastasis and recurrence ability, the idea that IMT was a benign and reactive lesion was replaced by a neoplasm with a low malignancy potential. The most common site that IMT arises is the lung, which is called pulmonary IMT, but we should keep in mind that IMT may arise from any part of the body that has mesenchymal tissue. The other common sites are small and large intestines, mesentery, mediastinum, retroperitoneum, omentum and diaphragm.[1,2]

Pulmonary IMT is the most common neoplasm of childhood pulmonary neoplasms, but in adults, it is very rare. Most patients are asymptomatic, while some patients have nonspecific symptoms, such as cough, chest pain, hemoptysis, dyspnea, fever and fatigue. [3-5] The most common radiographic finding is the solitary pulmonary nodule, which is hypermetabolic in PET CT. Therefore, it is impossible to differentiate pulmonary IMT from lung carcinoma radiologically. Histopathological examination is required for the definitive diagnosis. The recommended treatment is complete surgical resection. Despite complete surgical resection, recurrence is still possible with a 2% rate. As the recurrence may develop many years after surgery, long-term follow-up is required. Here, we present a

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case with pulmonary IMT with her clinical, radiological features and 2-year follow-up results.

Case Report

A 63-year-old female patient with no respiratory complaints admitted to our outpatient clinic with a pulmonary nodule incidentally detected by thorax computed tomography (CT), which was ordered for the evaluation of her thyroid disease. She had allergic asthma and was on regular inhaler therapy. Thorax CT revealed two paravertebral nodules, one, 20x15 mm in diameter, in the left upper lobe apicoposterior segment and the other one, 7 mm in short axis, in the right lower lobe superior segment (Fig. 1). They were both hypermetabolic (left upper lobe nodule SUVmax: 13.7, right lower lobe nodule SUVmax: 2.7) on PET CT (Fig. 2). Upon this, sampling of both nodules by video-assisted thoracoscopic lung wedge biopsy was recommended. She preferred to be operated at another center. After the operation, she admitted to our department back with left upper lobe nodule wedge sample. The right one was sampled. In the evaluation of the material in the pathology department of our hospital, it was reported that it was an inflammatory myofibroblastic tumor and necrotic granuloma with a negative surgical margin (Figs. 3a, 3b, 3c). Tissue Mycobacterium tuberculosis PCR negative, but due to the high incidence in our country, upon necrotic granuloma results, we offered the patient to take antituberculosis treatment for six months. The treatment was completed without any complications. The patient was followed for recurrence of IMT after the surgery. We

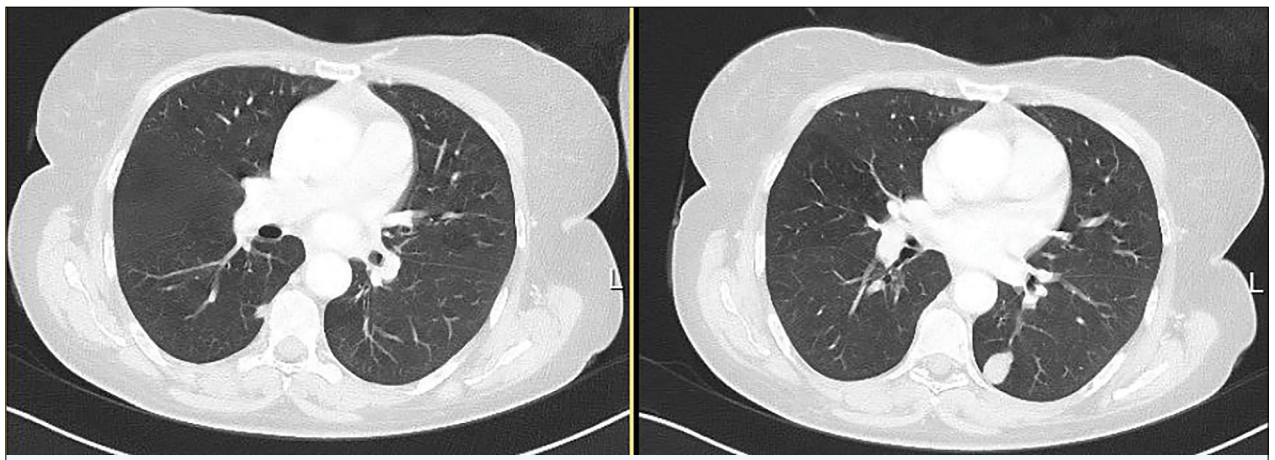

Fig. 1. Two paravertebral pulmonary nodules with a smooth margin, one 20x15 mm in diameter, in the left upper lobe apicoposterior segment, other 7 mm in short axis, in the right lower lobe superior segment in thorax CT.

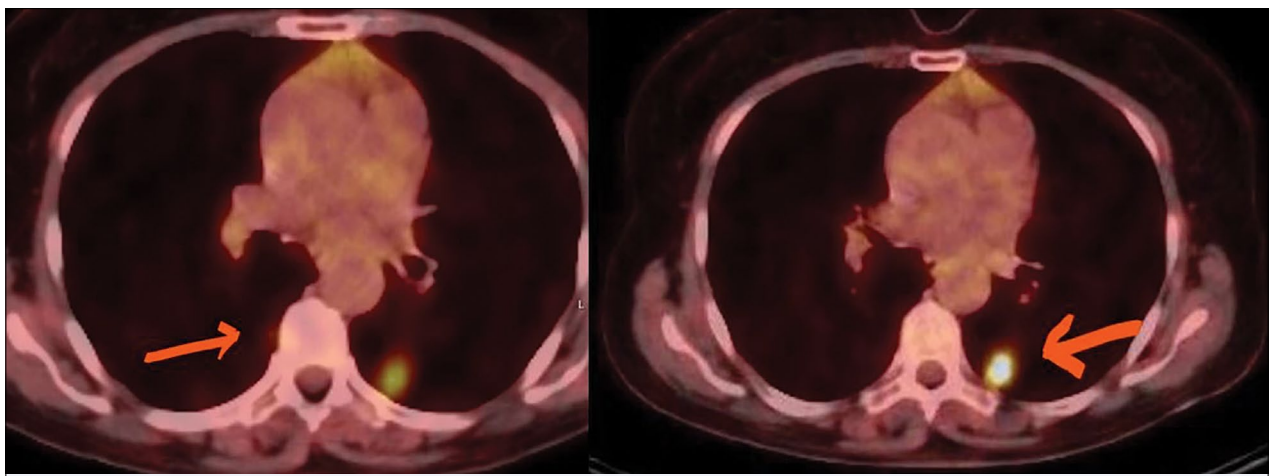

Fig. 2. Both of the nodules are hypermetabolic (left upper lobe nodule SUVmax: 13.7, right lower lobe nodule SUVmax: 2.7) on PET CT.

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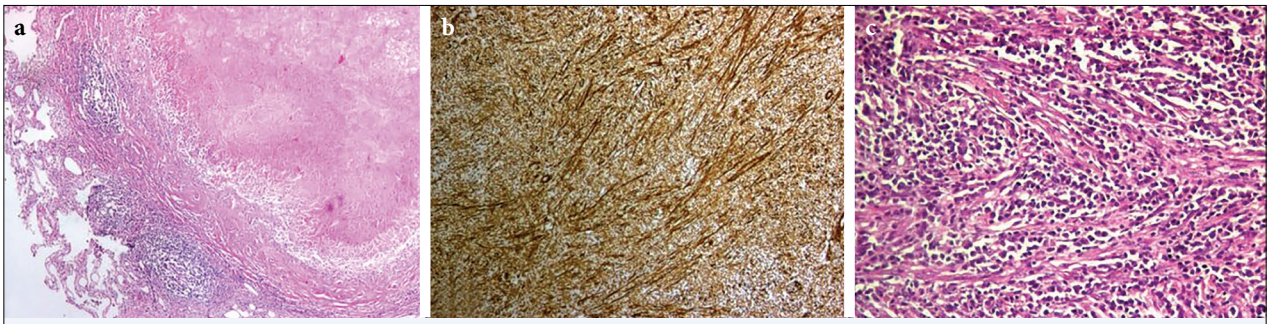

Fig. 3. Necrotic granuloma in the left upper lobe apical frozen material. (b) Spindle cells stained positively with SMA, immunohistochemically. (c) The present findings were interpreted as an inflammatory myofibroblastic tumor. ALK and Kaldesmon were negative. TTF-1 and PanCK were positive in alveolar cells. No signs of malignancy were detected. Surgical margins were intact.

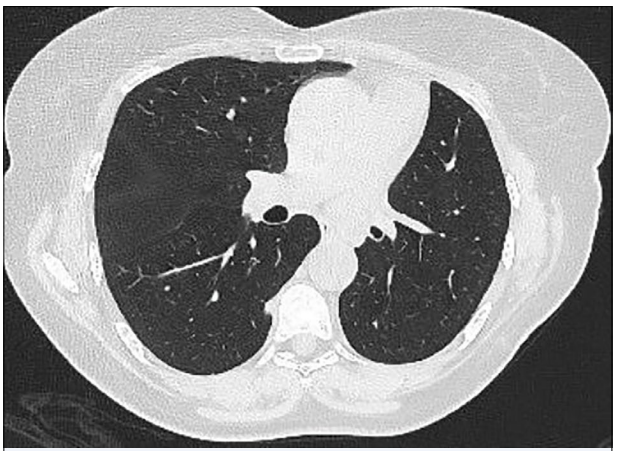

Fig. 4. The upper lobe of the left lung was not observed (operative). There were no signs of recurrence in the left lung. There was no difference in the size, nature and density of the right lung nodule.

followed the right lung nodule, according to Fleisher nodule follows up protocol. There was no change in the size and the character of the right lung nodule at the end of two years, and there was no recurrence of IMT on the left lung (Fig. 4).

Discussion

IMT is neoplasia with a low malignancy potential. IMT is a rare pathology that constitutes less than 1% of all lung neoplasms.[3,6,7] The exact etiology is not known; however, infection, trauma, surgical manipulation and radiation therapy are expected to be relevant risk factors.[8-10] There are some cases with IMT with other system malignancies and autoimmune diseases in the literature, which arises the suspicion that these somehow might be related to IMT. It is not certain that IMT has a female tendency, but, in general, IMT is more common in women than in men.[11-13] Approximately 70% of the pulmonary IMT patients are asymptomatic and detected incidentally on radiological examinations. Less frequently, patients present with symptoms, such as cough, chest pain, hemoptysis, dyspnea, fever, fatigue, and rarely weight loss.[3-5] Our patient was female, and she had comorbidities, allergic asthma, thymic hyperplasia, and graves' disease. She had no symptoms, and IMT was detected incidentally on thorax CT performed for another reason.

There is no characteristic radiological finding for pulmonary IMT. The most common thorax CT finding is the sharp, peripheral, solitary nodule. Calcification, cavity, necrosis, obstructive atelectasis are other possible radiological findings. PET CT has limited utility in distinguishing IMT from lung cancer because IMT is also hypermetabolic like carcinomas. PET CT is more beneficial just for follow-up rather than the diagnosis. Histological evaluation is required for the diagnosis. Bronchoscopic biopsy and percutaneous core biopsy are not recommended as tissue samples obtained by these ways are insufficient for histopathological evaluation, so the surgical biopsy is preferred.[14] Our patient had two peripheral solid nodules with sharp margins. PET CT showed that these nodules were hypermetabolic. The left lung nodule was composed of myofibroblasts. Microscopically, fibroblasts, myofibroblasts and inflammatory cells are present in varying concentrations in IMT.[15] The presence of any significant atypia suggests low-grade sarcoma.[3,6,7] Upon this knowledge, the left lung nodule was not a low-grade sarcoma as it had no atypia. It was an IMT with no suspicion with fibroblasts, myofibroblasts and inflammatory cells. Immunohistochemical methods are used to differentiate IMT from inflammatory sarcoma and spindle cell carcinoma. Anaplastic Lymphoma Kinase gene

(ALK) anomalies, which are associated with malignant lung tumors, may be present in IMT. ALK overexpression was detected in approximately half of IMT cases. The clinical significance of ALK abnormality is not fully understood but is thought to be associated with the risk of recurrence. The presence of chromosomal abnormalities suggests that IMT is a neoplastic proliferation of clonal origin and is associated with aggressive clinical behavior.[15,16] In our patient, there was no lesion at the surgical margins, pleural involvement was present, and spindle cells were stained positively by SMA immunohistochemically. ALK and Kaldesmon were negative. Necrotic granuloma was observed in one specimen without any malignancy.

The preferred treatment is complete surgical resection. Medical treatment may be considered in cases where surgery is contraindicated or in patients with locally invasive lesions or multifocal lesions or unresectable ones.[17,18] Five and ten-year survival rates after complete surgical resection are 91% and 77.7%, respectively.[19] The recurrence rate is 2% for complete resection and up to 60% for incomplete resection. Recurrence may develop after many years, so long-term follow-up is necessary. There are some publications on the efficacy of glucocorticoids, chemotherapy, or radiotherapy as a medical treatment option. According to the results of a recently published study, crizotinib, a tyrosine kinase inhibitor, has been suggested to be a new medical treatment option for patients with ALK mutations.[20] In our patient, there was no ALK mutation, and complete resection was performed for both diagnostic and therapeutic purposes. There was no recurrence after two years of follow-up.

Conclusion

Pulmonary IMT is a rare mesenchymal tumor. It is considered as neoplasia with low malignancy potential due to its ability to relapse and metastasis. Its etiology and biology are still unknown. Biological behavior is unpredictable in advance. The significance of genetic abnormalities in predicting biological behavior is unknown. Clinical and radiological findings are nonspecific, and diagnosis is based on the histopathological and immunohistochemical evaluation. Surgical complete resection is the treatment of choice. In cases where surgery cannot be performed, steroids, chemotherapy and radiotherapy may be used. As the pathogenesis of the disease is resolved at the genetic and molecular level, new drugs appear to be on the way.

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