Case Report

Common Presentation of Uncommon Disease: Inflammatory myofibroblastic tumor of the lung, case report

Safwat Eldaabossi\textsuperscript{a,b,*}, Lotfi Maghmoul\textsuperscript{a}, Elsaid Lotfy\textsuperscript{c}, Ahmad Antar\textsuperscript{d}, Yasser Elghoneimy\textsuperscript{e}, Hameed Aljawad\textsuperscript{f}, Ayman Ghoneim\textsuperscript{e}

\textsuperscript{a} Pulmonology Consultant, Almoosa Specialist Hospital, Al Ahsa, Saudi Arabia
\textsuperscript{b} Department of Chest Diseases, Al Azhar Faculty of Medicine, Egypt
\textsuperscript{c} Radiology Consultant, Almoosa Specialist Hospital, Al Ahsa, Saudi Arabia
\textsuperscript{d} Hematology-Oncology Consultant, Almoosa Specialist Hospital, Al Ahsa, Saudi Arabia
\textsuperscript{e} Cardiothoracic Consultant, Almoosa Specialist Hospital, Al Ahsa, Saudi Arabia
\textsuperscript{f} Pathology Consultant, Almoosa Specialist Hospital, Al Ahsa, Saudi Arabia

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ABSTRACT

Background: Inflammatory myofibroblastic tumor (IMT) of the lung is a rare lung tumor, accounting for 0.7% of all lung tumors. They are usually benign, but can invade surrounding structures, undergo malignant transformation, recur, or even metastasize.

Case report: We report a 44-year-old adult diabetic male from Saudi Arabia who had been suffering from cough with severe sputum and left shoulder pain for 2 weeks. Chest radiography (X-ray and computed tomography (CT)) revealed the presence of a mass lesion in the left lower upper lobe with central cavitation. The diagnosis of inflammatory myofibroblast lung tumor was confirmed by histological and immunohistochemical examination of the CT guided lung biopsy. The patient was successfully treated with surgical resection of the tumor by left limited thoracotomy with safety margin, and IMT was also documented.

Conclusion: A high degree of suspicion of a solitary pulmonary mass is required for diagnosis and management of an inflammatory myofibroblastic lung tumor. The clinical and radiologic presentation of an inflammatory myofibroblastic tumor is nonspecific and the diagnosis is rarely made before surgical biopsy. Histologic and immunohistochemical examination is usually required to confirm the diagnosis and prevent recurrence.

1. Introduction

Inflammatory myofibroblastic tumor (IMT) of the lung is a rare lung tumor, accounting for 0.7% of all pulmonary neoplasms \cite{1,2}. However, it’s more common in children and constitutes the most common pediatric primary lung tumor \cite{3}. They are usually benign and cured by complete resection, but it can invade surrounding structures, undergo malignant transformation, recur or even metastasize \cite{1,2}. It was first described by Brunn in 1939 \cite{4}. IMT has been described by various terms due to its variable cellular components, including plasma cell granuloma, inflammatory pseudotumour, and fibrous histiocytoma. The view that IMT is a reactive lesion or a neoplasm has been controversial \cite{5}.

\* Corresponding author. Pulmonology Consultant, Almoosa Specialist Hospital, Al Ahsa, Saudi Arabia.
E-mail address: safwatchest1@gmail.com (S. Eldaabossi).

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2. Case presentation

A 44 years old, Saudi male, married, ex-smoker for 4 years, with a smoking index of 10 packs/year. He has Type 2 diabetes for 10 years and takes basal insulin, metformin and sitagliptin. He has dyslipidemia on statins and vitamin D deficiency.

He had a productive cough for 2 weeks with large quantities of odorless yellow sputum, with intermittent pain in the left shoulder for 3 days. He denied fever, dyspnea, hemoptysis, or diarrhea. He denied any history of choking or dental caries.

The general examination was unremarkable. Local chest examination was positive for dullness in the left upper chest with fine inspiratory rales.

Chest x-ray showed opacity in the left hilum and para-hilar area (Fig. 1a). CT chest without contrast showed a soft tissue mass in the left upper lobe (lingual) with central necrosis (Fig. 2a, b).

Since he is diabetic and pulmonary tuberculosis is relatively common in our area, he was admitted to a negative pressure isolation room until tuberculosis can be confirmed or ruled out.

He was treated with intravenous amoxicillin and clavulanic acid as a case of pulmonary abscess and further investigations were performed. Sputum for acid fast bacilli 3 samples by stain and polymerase chain reaction were negative for tuberculosis as well as sputum Grams stain and culture were negative. The situation improved at first, as the chest pain subsided and the sputum became less and whiter. He was discharged after 3 days on oral co-amoxclav and ordered to follow up in the clinic after 10 days.

After a week he came for follow-up, complaining only of hemoptysis, a small amount of bloody sputum, with no other symptoms. Subsequent chest radiograph revealed persistent opacity of the left upper lobe and CT thorax showed 2.2 × 2.5 cm subpleural cavitary nodules in the left upper lobe/lingula associated with an adjacent area of patchy languor, minimal interlobular septal thickening, and tree-in-bud nodules (Fig. 2c, d).

He underwent a CT guided lung biopsy to diagnose the etiology, possibly lung cancer, and the procedure was performed as a day case under local anesthesia without complications (Fig. 2e).

A diagnosis of inflammatory myofibroblastic tumor of the lung was rendered based on histopathology evaluation and a panel of immunohistochemistry stains (Fig. 3a-e).

Sections of the tumor sampled in the core biopsy revealed spindle cells in a background of abundant mixed inflammatory infiltrate rich in plasma cells, lymphocytes, and few eosinophils. The spindle cells are arranged in fascicles. The immunohistochemistry stains revealed that the spindle cells were positive for Actin. They are negative for pankeratin, S100, CD68 and Alk1. The plasma cells in the background are proven to be polyclonal using Kappa and Lambda immunohistochemistry staining.

A follow-up CT thorax with contrast was performed to assess the size of the mass, other lung areas, and the mediastinum preoperatively (Fig. 2f).

A multidisciplinary team of pulmonology, oncology, thoracic surgery, and radiology was consulted and the decision was made to perform a surgical resection with a safety margin and rapid incision. A limited left thoracotomy was performed and a wedge resection with safety margin was performed. The resected lung lesion was confirmed by histopathological and immunohistochemical examinations as IMT of the lung.

Fig. 1. Chest x-ray posteroanterior view 23-August-2021: left paracardiac opacity with ill-defined margin and this appearance of the internal gas.

Fig. 1b: Chest x-ray posteroanterior view 13-Sep-2021: Slight Progressive size of the previously noted left paracardiac opacity with ill-defined margin and this appearance of the internal gas.
3. Discussion

The lung is one of the most common locations of IMTs, accounting for only about 0.7% of all lung tumors in the general population [1,2]. Sites outside the lung have also been reported, including the spleen, lymph nodes, stomach, esophagus, salivary glands, breast, central nervous system, epididymis, and soft tissues [4,5]. They may be associated with immunologic disorders [1] and chronic infections [6] and may occur in surgical lung scars.

The clinical and radiological manifestations are varied and non-specific. Therefore, it is difficult to establish the diagnosis unless surgical resection is performed [7]. They are usually asymptomatic and discovered incidentally as an abnormal chest radiograph, perhaps with nonspecific respiratory symptoms such as cough, chest pain, dyspnea, hemoptysis, and nonspecific inflammatory symptoms such as fever, malaise, and weight loss [1,2,6].

Radiological findings of pulmonary IMTs are nonspecific and may be solitary, well-circumscribed peripheral lung masses [8], with a predominance of lower lobes [9]. Aggressive behavior with invasion of adjacent structures has been described at CT [9]. The literature review seems to support a neoplastic origin for IMT with intermediate malignant potential in a small subset of cases described in the most recent World Health Organization classifications [10]. Cavitations - as in our patient - calcifications and lymphadenopathy are rare. In our case, neoplastic and infectious (bacterial, fungal and tuberculous) causes were initially excluded.

The diagnosis of IMT is difficult to make, and histological examination of the tissue is always required [11]. Transthoracic fine-needle aspiration biopsy and bronchoscopic specimens are usually too small and insufficient for a definite diagnosis [10]. Therefore, surgical excision of the lesion is the preferred method of diagnosis [12].

On macroscopic examination, the tumor is well circumscribed but not encapsulated, a yellowish gelatinous mass, firm and

Fig. 2. CT thorax lung window, 2b mediastinal window, 2c lung window coronal section 23-August-2021: A 2.2 × 2.5 cm subpleural cavitory nodule is seen in the anterior aspect of the left upper lobe/lingula, with an adjacent area of patchy ground glass opacity, minimal interlobular septal thickening, and tree-in-bud nodules.

Fig. 2b

Fig. 2c

Fig. 2d CT thorax mediastinal window, 2e lung window 23-August-2021: Interval growth of the subpleural mass in the left upper lobe of the anterior segment with interval growth of the associated left perivascular lymph nodes compared with the previous CT scan of August. Size is unchanged from the previous CT of September.

Fig. 2f CT thorax mediastinal window 23-Sep-2021: At CT, a true incisional needle biopsy was taken from a subpleural round lesion in the left upper lobe. The procedure was performed under local anesthesia without complications.
homogeneous, with sometimes some foci of necrosis, hemorrhage or calcification [13]. IMT is considered a neoplasm with intermediate biological potential that can recur and rarely metastasize. IMTs histologically consist of a variable mixture of fibroblasts, fibrous tissue, granulation tissue, and inflammatory cells. Immunohistochemistry is typically diffusely positive for actin, locally positive for FXIIIa, and negative for CD34 [14]. Histological examination revealed a variable proportion of myofibroblastic cells arranged in a myxoid, fibrous, or calcified stroma associated with a chronic inflammatory component including lymphocytes, plasma cells, and eosinophils distributed to varying degrees throughout the tumor [12,13].

Recent studies concluded that IMT is a true neoplasm rather than an inflammatory reactive lesion and reported that chromosomal abnormalities correlate with tumor incidence [13,15]. Approximately half of IMTs have a clonal abnormality overexpressing ALK, suggestive of a neoplastic process [16]. The main pathology differential diagnoses are organizing pneumonia, lymphoma, sarcoma, and fibrosis, but the pathologist is usually able to rule out malignancy [17].

A surgical lung biopsy is usually required to confirm the diagnosis, but a CT guided lung biopsy may also be diagnostic. Surgical resection (lobectomy and pneumonectomy) is the treatment of choice for IMTs to rule out malignancy and achieve cure. Spontaneous regression may occur, but local spread may be associated with significant morbidity and occasionally death. Complete surgical resection has an excellent prognosis with a low recurrence rate and a 5-year survival rate of more than 91%. When surgical resection is not possible, other treatments (radiotherapy, chemotherapy, or systemic corticosteroids) may be used as an alternative to surgery [9,10,12,15]. Chemotherapy is indicated in cases of multifocal, invasive lesions or in unresectable recurrent tumors [18]. The combined chemotherapy, carboplatin and paclitaxel, have shown response in some cases [19]. The use of corticosteroids for IMTs is controversial, with some studies showed good response and others have reported worsening of IMTs with steroids [20,21]. Tyrosine kinase inhibitors (Ex: ALK RT inhibitors and ROS-1 TK inhibitors) have been reported to be effective in advanced or unresectable ALK-rearranged IMTs or harboring ROS1kinase fusions [22,23].

Prognosis depends on the quality of surgical resection and tumor size [9,18]. After radical resection, the prognosis is excellent [11,18]. Even many years after the initial diagnosis, the disease may relapse. For this reason, patients should be monitored closely after
4. Conclusion

Inflammatory myofibroblastic tumor is a rare benign tumor. The clinical and radiological presentation is variable and nonspecific. The diagnosis is rarely made before surgical treatment. Only histological and immunohistochemical examination can confirm the tumor of the lung. Second, this would be the first case of inflammatory myofibroblastic tumor to be reported in Saudi Arabia.

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Declaration of competing interest

The authors have no conflict of interest to declare.

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