Inflammatory myofibroblastic tumor of the lung: A rare entity

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor usually seen within the first and second decade. They are extremely rare in adults, constituting less than 1% of adult lung tumors. It’s usually benign, but it had a tendency for local recurrence.

We report a case of asymptomatic inflammatory myofibroblastic tumor of lung in a 46-year-old non-smoker woman.

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor usually seen within the first and second decade [1,2]. They are extremely rare in adults, constituting less than 1% of adult lung tumors [1]. This tumor also known as plasma cell granuloma or inflammatory pseudotumor, used to be classified as a benign tumor [2,3]. However, it had a tendency for local recurrence.

Therefore, many recent study classified this tumor as an “intermediate malignancy” tumor [4]. IMT has been reported to occur in multiples anatomic sides which most commonly are lung, mesentery and omentum [5]. The treatment of choice is a complete surgical resection to exclude malignancy and to achieve a good prognosis [1]. We report a case of pulmonary inflammatory myofibroblastic tumor and review the literature related to demographical, clinical, biological characteristics as well as treatment efficacy of this tumor.

2. Case report

46-year-old non-smoker woman presented to chest department for incidental discovery of a suspected radiological opacity.

The patient had a history of systemic lupus erythematosus with joint and skin manifestations stabilized by plaquenil for 2 years. She had also a history of allergic asthma controlled by inhaled corticosteroid low doses.

The patient was asymptomatic. The clinical examination was normal.

Chest computed tomography (CT) showed a solitary nodule of right posterobasal segment, measuring 2 cm long axis with eccentric calcifications and without lymph node (Fig. 1). This nodule was highly suspicious of malignancy.

PET Scan was not performed for unavailability.

Bronchoscopy did not objectify a suspicious lesion and microscopy, culture and cytological examination of bronchial fluid were almost normal.

Surgery was carried out to obtain a diagnosis and achieve cure. Wedge resection was performed, and histological examination revealed the diagnosis of an inflammatory myofibroblastic tumor of the lung (Fig. 2).

At the follow-up (14 months after operation), the patient was symptoms free and there was no evidence of tumor recurrence on chest CT scan.

3. Discussion

IMT of the lung are rare. Its exact incidence is unknown. In the literature, the incidence reported is <1% of all lung tumors [4,6]. There were first described in 1939 [2]. IMT occur at any age, but have a predilection for children and young adults [7]. They are not limited to the lung, but any site can be involved, such as...
the brain, liver, spleen, lymph nodes, salivary glands, breast, soft tissues and skin [2]. The most common locations are lung, abdominopelvic region and retro peritoneum. This tumor usually affects a single organ, but multiple locations are possible [3].

Diverse nomenclature has been applied to describe these lesions, including plasma cell granuloma, xanthogranuloma, inflammatory myofibroblastic proliferation, inflammatory pseudotumor, fibrous histiocytoma, plasma cell histiocytoma complex and inflammatory fibrosarcoma [3,8]. All these entities reflect the uncertainty of the true origin of IMTs [2].

Although IMTs show typically benign clinical behavior, malignant evolution has been described in the literature, including recurrent (between 2 and 25% of cases) and metastatic disease (less than 5% of cases) [4,6].

The clinical symptomatology of pulmonary IMTs is various and nonspecific [8]. In approximately 70% of cases, the disease is discovered coincidentally on imaging exams requested for other reasons [1]. Patients may manifest some symptoms such as cough, chest pain, and shortness of breath, hemoptysis, fever and fatigue. These manifestations depend on the size and location of the tumor [4,9].

Radiographic findings showed in approximately 90% of cases a solitary peripheral lung nodule. There is a predilection for lower lobes, peripheral lung parenchyma and subpleural locations [4]. On computed tomography, the lesion appears as a heterogeneous mass with variable contrast enhancement. Calcifications -such as seen in our patient-, cavitations and lymphadenopathy are rare [3]. In 5% of cases, lung IMTs can extend towards mediastinum, diaphragm, pleura or chest wall [8]. PET can be useful to distinguish benign IMT from malignant lesions [3].

The diagnosis of IMT is difficult to establish, and histologic examination of tissue is always required [10]. Fine needle aspiration biopsy and bronchoscopic samples are typically too small and insufficient for confident diagnosis [3]. Therefore, surgical excision of the lesion is the preferred diagnostic method [2].

Histologic examination showed a varying proportions of myofibroblastastic cells, arranged in a myxoid, fibrous or calcified stroma associated to a chronic inflammation component including lymphocytes, plasma cells and eosinophils distributed to a variable degree throughout the tumor [11,12]. Three histological patterns have been described [4,13]. The first is myxoid and richly vascularized that has the appearance of nodular fasciitis or granulation tissue. The second pattern is a more densely spindle cell proliferation with focal nodular lymphoid hyperplastic resembling fibromatosis. The third type is that of a very sclera-hyalinized, slightly cellular stroma [4,13]. In this case, the histological pattern was hypocellular, densely collagenised and reminiscent of a fibrous scar.

Approximately, 50% of patients presents a cytogenetic translocation of the band chromosome 2p23 resulting in overexpression of ALK protein [4,14]. Recent studies demonstrated that these chromosomal abnormalities are correlated with tumor aggressiveness and local recurrence, and concluded that IMT is a true neoplasm rather than an inflammatory reactive lesion [12,15].

Complete surgical resection of the tumor is the treatment of choice [2,4]. Medical therapy such as chemotherapy, radiation and even corticosteroid therapy are recommended for patients with incomplete resection, multifocal disease, not resecable tumor or when surgery is contraindicated [3,16].

The prognosis depends on the quality of surgical resection and the tumor size [4,14]. After radical resection the prognosis is excellent [2,11]. Recurrent disease can occur even many years after the initial diagnosis. For this reason, patients should be followed closely after resection to detect local or distant recurrence [6].

4. Conclusion

Pulmonary inflammatory myofibroblastic tumor is a rare benign tumor. Clinical and radiological manifestations are non specific. Only
Histologic examination can confirm the diagnosis.

Despite the benign origin of IMTs, the risk of recurrence and distant metastasis require a complete surgical resection and prolonged follow-up.

Declaration of competing interest

The authors declare no conflict of interest.

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