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Case Report

Pulmonary Inflammatory Myofibroblastic Tumor with Massive Calcification and IVC Invasion in Adult: A Case Report and Literature Review

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Abstract

Inflammatory myofibroblastic tumor (IMT) is a mesenchymal neoplasm of intermediate biological potential and could occur at any anatomical site from the central nervous system to gastrointestinal tract. IMT of the lung is a relatively uncommon condition, with a reported incidence between 0.04 - 1.2% of all tumors of the lung. A 26-year woman was admitted to our hospital with complaint of irritating cough for the past six months. Chest computed tomography scan demonstrated a 6.8 cm x 6.9 cm, densely calcified mass in the right lower lobe neighboring the mediastinum. The tumor was radically resected via lobectomy and partial excision of right hemidiaphragm and pericardium, and the stump of tumor within inferior vena cava was electrocauterized. Histological examination combined with immunohistochemical study discovered an inflammatory myofibroblastic tumor with osseous component.

Keywords: *Inflammatory Myofibroblastic Tumors; Computed Tomography; Diagnosis; Treatment*

Introduction

According to the 2013 World Health Organization (WHO) Classification of soft tissue tumors, inflammatory myofibroblastic tumor (IMT) has been defined as an intermediate fibroblastic or myofibroblastic tumor, which is characterized by the presence of spindle-shaped myofibroblasts, and a chronic inflammatory infiltrate composed of eosinophils, lymphocytes, and plasma cells [1]. Pulmonary IMT was firstly described by Brunn in 1939. The incidence of pulmonary IMT is 0.04 - 1% of all tumors of the lung and the causes are always obscure [2]. Most of the patients are asymptomatic and detected incidentally on radiographic investigations. Pulmonary IMT may present with various appearances on CT, but most commonly they appear with heterogeneous attenuation and intensified enhancement after contrast injection. Calcified opacity within IMT occurs frequently in children, while these calcification patterns vary from amorphous, mixed or fine flecks to heavy mineralization [3]. Pulmonary IMT with densely calcifications and invasion to adjacent organs is presented in a 26-year woman. This unusual CT findings perplexed us before surgery and was finally confirmed to be IMT by histopathological and immunohistochemical analysis of the resected specimen. More interestingly, densely calcification was composed of mature osseous tissue on histopathological examination. Basing on the data of this case, a short overview of the literature is also given here.

Case Report

A 26-year young woman complained of irritating cough repeatedly for past six months. Her past medical history was unremarkable. Blood samples did not exhibit any abnormalities, including the hematological, biochemical profiles and the level of tumor markers. Chest CT scan showed a lobulated mass in the right lower lobe measuring approximately 68 mm × 69 mm, and multiple calcifications were also visible within the mass (Figure 1). After contrast injection, the solid component was significantly enhanced, and adhered to the pericar-

dium, diaphragm and inferior vena cava (Figure 2). There was no lymphadenopathy in the hilum and mediastinum. Further bronchoscopy revealed that the lumen of right lower lobe bronchus was externally compressing stenosis with almost occlusion, and biopsy was inconclusive. Exploratory thoracotomy was performed after the patient's consent. During surgery, there was a mass measuring approximately 10 cm × 8 cm × 8 cm in the lower lobe of right lung, which invaded the pericardium, diaphragm and IVC. The patient underwent right lower lobectomy and partial excision of right hemidiaphragm and pericardium, and the stump of tumor within IVC was electrocauterized. On gross examination, the mass was 7.5 cm in size, firm, and whitish-brown in color on cut surface (Figure 3). Histopathological examination revealed a proliferation of regular spindle cells in fascicles, admixed with lymphocytes, plasma cells and eosinophils (Figure 4). Dense calcification of the mass was identified to be mature osseous tissue (Figure 5). Histopathological examination combining with immunohistochemical analysis revealed pulmonary IMT with osseous tissue. The postoperative period was uneventful and she was discharged in a stable condition. A months later, the patient experienced three cycles of radiotherapy. There is no progress at 12 months of follow-up on chest CT (Figure 6).

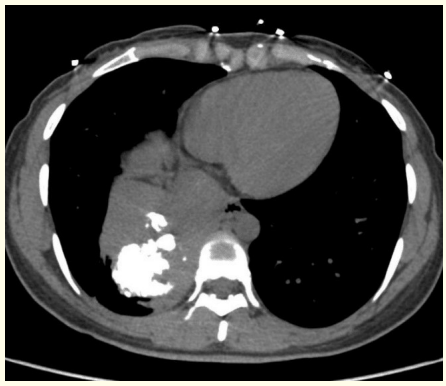

Figure 1: *Chest non-enhanced CT scan showed showed a lobulated mass together with multiple calcifications.*

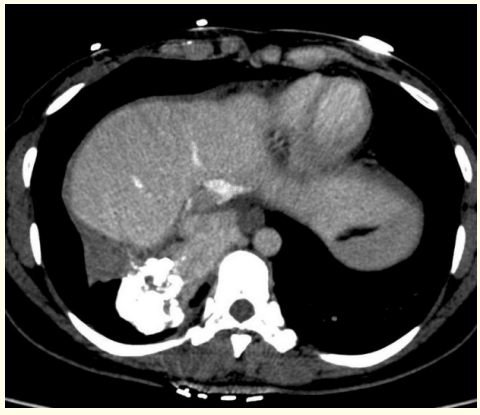

Figure 2: *Contrast-enhanced CT scan showed remarkable enhancement of solid proportion and filling defect in inferior vena cava.*

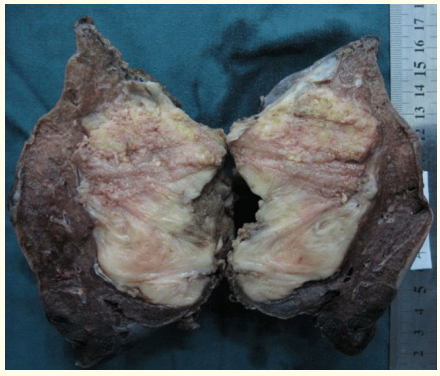

Figure 3: *Gross specimen showed the mass was firm, and whitish-brown on cut surface.*

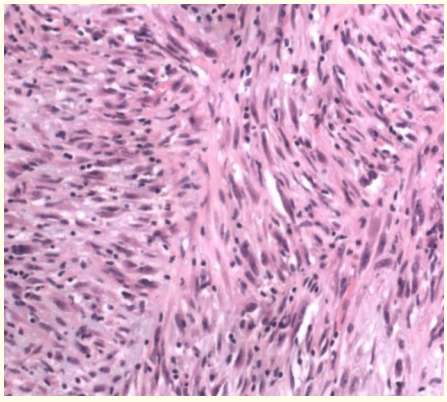

Figure 4: *Microscopic examination revealed a proliferation of regular spindle cells in fascicles, admixed with lymphocytes, plasma cells and eosinophils(HEx100).*

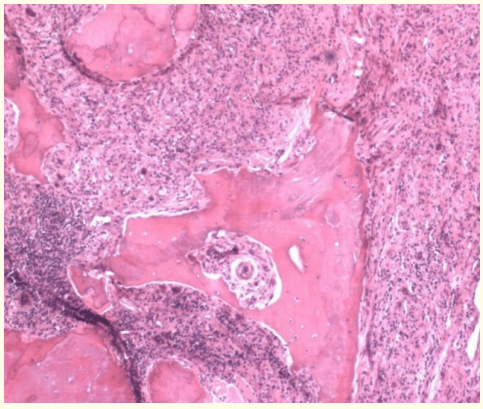

Figure 5: *Mature osseous tissue (dense calcification of the mass detected on CT) was located between spindle cells(HEx40).*

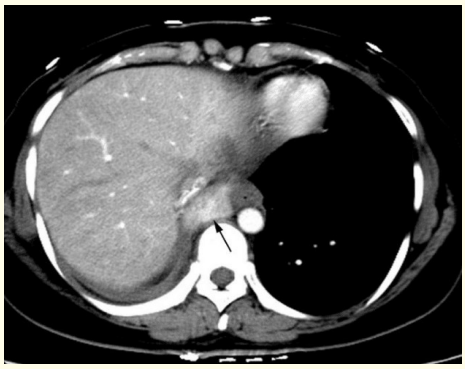

Figure 6: *Follow-up CT at postoperative 12 months showed residual mass and stable lesion in the inferior vena cava.*

Discussion

IMT was first observed in lung and described by Brunn in 1939 and was so named by Umiker., *et al*. in 1954 because of its propensity to clinically and radiologically mimic a malignant process [4]. IMT has ever been described by various terms because of its complexity and variable histological characteristics, which includes inflammatory pseudotumor, plasma cell granuloma, xanthogranuloma, fibrous histiocytoma, xanthoma and fibroxanthoma [5-8]. IMT most commonly involves the lung and the orbit, but it has been reported to occur in nearly every site in the body from the central nervous system to the gastrointestinal tract. IMT of the lung is rare and its incidence is reported to be 0.04 - 1% of all tumors of lung. IMT is the most common primary lung mass seen in pediatric patients, constituting approximately 50% of benign intrapulmonary tumor [9]. Being a rare lung tumor in adults, it only constitutes 0.7% of all lung tumors [5]. There are many uncertainties about the pathogenesis of IMT. Several hypotheses have been proposed such as an autoimmune mechanism or infectious origin. Indeed, 30% of cases are closely related to recurrent respiratory infections caused by several microorganisms. Malam M et al described a case of inflammatory pseudotumor of the lung associated with a paraneoplastic syndrome(Dermatomyositis) [10]. In some cases, IMT is thought to result from inflammation following minor trauma or surgery or to be associated with other malignancy or vasculitis. Other studies, however, suggests that it be a true neoplasm due to the presence of a fusion gene involving anaplastic lymphoma kinase (ALK) gene, which is initially found to be arranged in anaplastic large cell lymphomas. This fusion leads to constitutive overexpression of the ALK, causing cell proliferation. ALK gene could be a potential target molecule, and specific targeted therapy, such as tyrosine kinase inhibition, has been used in selected inflammatory myofibroblastic tumors with encouraging results, as noted by Butrynsky [11].

Primary IMT can simulate a malignant process both clinically and radiologically. From a clinical point of view, the percentage of asymptomatic IMT of the lung ranges between 30% and 70% of cases. Symptoms are strictly related to lesion location: parenchymal, mediastinal or endobronchial. Presenting symptoms include cough, weakness, chest pain, hemoptysis and fever. Occasionally, Pulmonary IMTs may cause atelectasis due to bronchial compression or endobronchial lesion. Laboratory tests are usually nonspecific, but increased levels of CRP and/or elevated erythrocyte sedimentation rate have been described in about 50% of IMT cases. In our patient, the patient only complained of irritating cough and all laboratory tests are nonspecific. Radiologically, IMT is commonly seen as a peripherally located solitary nodule or mass in the lower lobes. It may have lobulated contours and show varying degrees of heterogeneous enhancement on contrast-enhanced CT, and can contain focal calcification within the consolidative mass. Different forms of calcifications (e.g. peripheral, punctate or fine) are seen in 15% of IMTs, and are extremely rare in adult IMTs [12]. Densely calcified mass together with involving adjacent organs was detected in our patient, and the fact that huge calcification detected on CT was osseous tissue probably comes from osseous differentiation of precursor cell within IMT. To the best of our knowledge, no similar case has been reported previously. On magnetic resonance imaging, IMTs were reported to have heterogeneous signal characteristics with a little higher signal than the muscle on T1- and

T2-weighted images and show heterogeneous contrast enhancement as seen on CT [13]. There are long list of differential diagnosis for this tumor including, primary lung cancer, metastasis, hamartoma, chondroma or pulmonary granuloma. Endobronchial lesions may also be confused with carcinoid tumors if extensive contrast enhancement is observed.

Complete surgical resection, if possible, is the treatment of choice for most primary IMT. Tumor size of less than 3 cm or equal to 3 cm is the good prognostic factor after total resection. Several reports of spontaneous regression have been reported [14]. Radiotherapy, chemotherapy and steroid can be used for unrespectable or recurrent cases but the outcome has not been promising according to the literatures. Due to invasion of tumor to IVC and incomplete resection, postoperative radiotherapy was administered in our patient.

In conclusion, pulmonary IMT is a rare tumor in adults and may mimic malignancy both clinically and radiologically, it should be kept in mind in the differential diagnosis when a large tumor with lobulated contour or harboring coarse calcification is present. Elevated level of acute phase reactants and absence of hilar or mediastinal lymphadenopathy strongly suggest the diagnosis. Surgical resection is the recommended treatment in pulmonary IMTs.

Acknowledgments

No.

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