Central lung Inflammatory Myofibroblastic Tumor after aspergillosis and total occlusion of the left pulmonary artery

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Abstract

Inflammatory Myofibroblastic Tumor (IMT) is an exceptionally uncommon benign tumor. We present a 54-year-old man with IMT, frequent episodes of respiratory infections, aspergillosis, and hemoptysis, who has received 23 years of conservative treatment. A successful left intrapericardial pneumonectomy was performed. During chest closure, the left atrial auricle lesion caused a massive hemorrhage owing to contact with the staplers of the left pulmonary artery stapler line. The bleeding was immediately controlled. One year after surgery, the patient has fully recovered and no adverse events or relapses have occurred.

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Introduction

Inflammatory Myofibroblastic Tumor (IMT) is an uncommon mesenchymal tumor that manifests as a pulmonary or soft tissue mass with a propensity to recur.[1,2] It is typically associated with recurrent respiratory infections, but the precise cause is unknown. The predominant clinical manifestations are cough, dyspnea, hemoptysis, and fever.[3] IMT is a benign lesion for which the only treatment option is surgery. We present a 54-year-old man with IMT, frequent episodes of respiratory infections, aspergillosis, and hemoptysis, who has received 23 years of conservative treatment. A successful left intrapericardial pneumonectomy was performed. A signed authorization from the patient as assent to publish the case report's contents has been obtained.

Case Report

A 54-year-old male was admitted to the Department of Thoracic Surgery with complaints of cough, hemoptysis, shortness of breath, and chest tightness that had worsened over the past month.

In 1993, when he was treated for pulmonary thromboembolism, he experienced his first hemoptysis. Two years later, owing to repeated complaints of hemoptysis, shortness of breath, and fatigue, a Fiberoptic Bronchoscopy (FBS) with transbronchial lung biopsy was conducted. Computed Tomography (CT) scans revealed a lesion in the hilum of the left lung. The patient had been diagnosed with interstitial alveolitis and was treated with corticosteroids for six months.

Three years later, he was confined to a Pulmonology unit due to a hemoptysis of 200 mL in 24 hours, cough, shortness of breath, and fatigue. Again, FBS was performed, and segmental lower lobe bronchus bleeding was discovered. Etamsylate and terlipresin were used as a conservative treatment. A CT scan revealed a lesion involving the distal left pulmonary artery branches and left lower lobe. The broncho-alveolar lavage was positive for aspergillosis, as was the aspergillosis serum test (birataspergillus antigen). Six months of Itraconazole 2x200 milligrams were prescribed. The CT after that period revealed that the lesion had completely resolved.

A CT scan performed a year ago due to moderate hemoptysis revealed infiltration and stenosis of the left main and lobar bronchi, infiltration of the left main pulmonary artery and its lobar branches, most likely due to thrombo-mycotic infiltration, and minimal pericardial effusion (Figure 1). Positron Emission Tomography/Computed Tomography (PET/CT) revealed a central

lung lesion that was intensely metabolically active (Figure 2). FBS revealed that the proximal 1.5 cm of the left main bronchus was unobstructed, while the distal 1.5 cm was swollen and obstructed by a coagulum. In the distal portion of the left main and lower lobe bronchi, infiltrative alterations were detected and biopsied. The histology revealed myxoid (granulation) tissue that was proliferative. Negative fungal stains (grocot, mucicarmin, PAS) were observed.

A left-sided intrapericardial pneumonectomy was performed. Under general intubation anesthesia, a left lateral thoracotomy was performed using a Robert-Shaw tube. Adhesions between the visceral, costal, mediastinal, and diaphragmatic pleura partially occluded the pleural cavity, necessitating total debridement. A moderately dense tumor measuring approximately 45/45 mm was palpated in the hilum of the left lung, immediately adjacent to the left main pulmonary artery, left main bronchus, and superior branch of a solitary pulmonary vein. A circular pericardiotomy was performed behind the phrenic nerve and around the left pulmonary hilum. Multiple systemic arterial and venous vessels were identified surrounding the left pulmonary artery, common pulmonary vein, and left main bronchus upon dissection. Left pulmonary artery and the common trunk of the pulmonary vein were removed using vascular tourniquets, and the ligamentum arteriosum was sacrificed using a harmonic device. The left main bronchus, the common pulmonary vein, and the pulmonary artery were sewn together using staplers after a left intrapericardial pneumonectomy. A thorough lymph dissection was performed on the 5th, 7th, and 10th level lymph nodes that were enlarged and hyperplastic. When clear blood abruptly emerged from the catheter, a pleural catheter and pericostal sutures were positioned and tightened. About 400 ml of blood was aspirated from the pleural cavity after the sutures were removed. A 2 mm-diameter punctiform lesion was discovered on the left atrial auricle, most likely caused by contact with the last staple at the medial end of the pulmonary artery suture line. The lesion was sutured using Prolen 0000. For added security, an Atriclip 35 was installed below the suture line in the left atrium. Between the auricle and the remnant of the left pulmonary artery, hemostatic sponges were positioned.

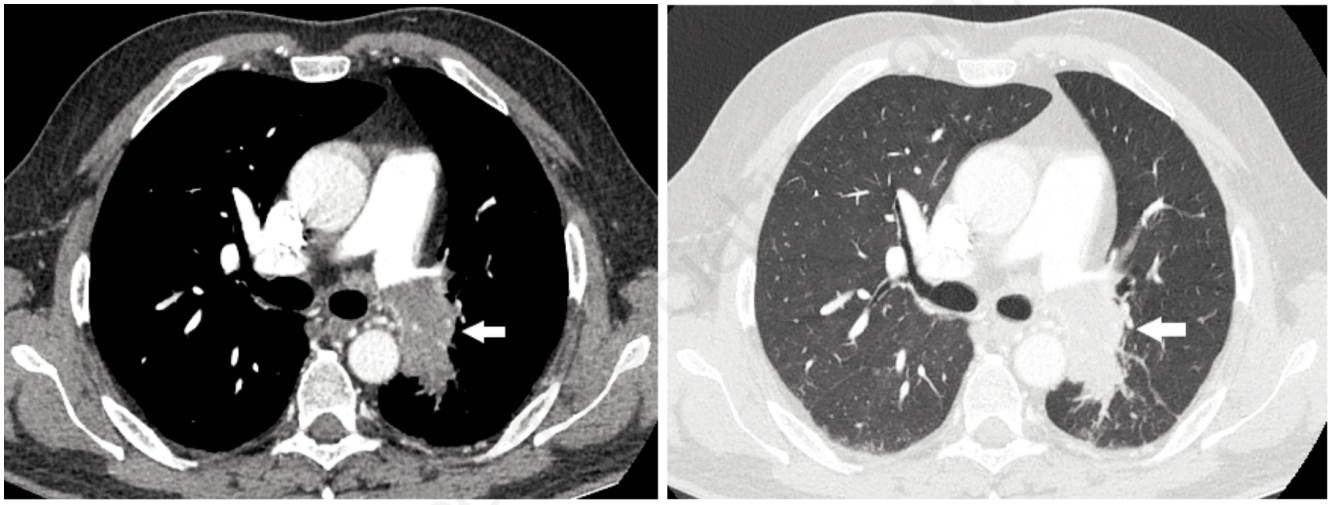

Figure 1. Computed Tomography (CT) axial images before surgery showed a central lung inflammatory myofibroblastic tumor, occluding the left main pulmonary artery.

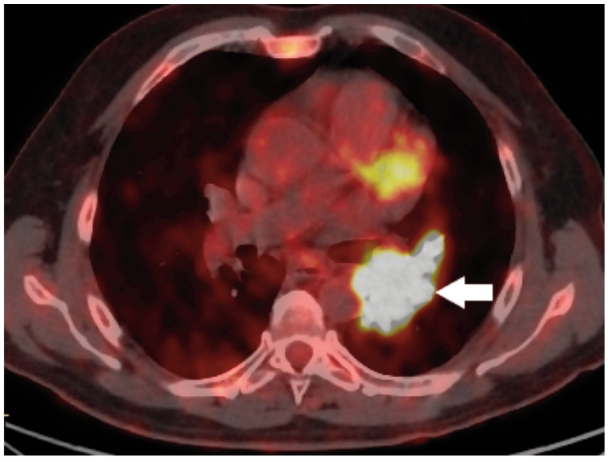

Figure 2. Positron Emission Tomography/Computed Tomography (PET/CT) showing an 18F-Fluorodeoxyglucose avid central lesion, diagnosed as lung inflammatory myofibroblastic tumor.

After the specimen was sectioned, an obstruction of the left pulmonary artery and a lesion in the left lung hilum, compressing the left main and lobar bronchi, were observed (Figure 3).

The pathohistological analysis revealed a solidly growing non-small cell tumor infiltrating the lung, polygonal to slightly elongated tumor cells with abundant cytoplasm and relatively monomorphic nuclei containing coarse chromatin. The described tumor directly infiltrated a peribronchial lymph node. Immunohistochemical study showed positive cytoplasmic reaction in tumor cells for vimentin (100%), ALK (85-90%), and CD31 (30%); weak and very weak cytoplasmic reaction in a small part of tumor cells for MSA and CKAE1/3 respectively; negative reaction in tumor cells for CD34, SMA, desmin, Bcl2, CD38, CD138, DOG1, CK7, TTF1, CK5/6, p40 and synaptophysin. The morphology and immune profile were characteristic of an inflammatory myofibroblastic tumor. It was determined that the lymphatic tissue surrounding the tumor as peribronchial lymph nodes was a component of the tumor and not an involved lymph node.

One year after surgery, the patient was discharged on the seventh postoperative day without any complications or relapse.

Discussion

IMT is a rare tumor that can affect numerous organs, including the larynx, orbita, bowels, breast, soft tissue, kidney, urinary bladder, and lungs, with the latter being the most prevalent site.[4] It accounts for approximately 0.7% of lung tumors,[5] affects patients 40 years of age with a reported mean age range of 27-50 years, and has no gender preference.[3] Immunohistochemistry, molecular biology, and cytogenetics are crucial diagnostic tools.[6] Histologically, IMT is constituted of myofibroblasts mixed with chronic inflammatory cells such as plasma cells, histiocytes, and lymphocytes.[7] Myofibroblastic spindle cells are arranged in fascicles and may express vimentin, desmin, smooth muscle actin, and muscle-specific actin, as well as epithelial markers such as cytokeratin and epithelial membrane antigen.[4] Fusions of ALK, ROS1, RET, and NTRK1 aid in differentiating IMT from reactive pseudotumors.[3]

Usually, small biopsied samples are insufficient due to the profusion of inflammatory cells;[7] therefore, the preoperative diagnosis of IMT is uncommon. In the presented case, preoperative histology revealed a myxoid tissue with blood vessels that was proliferating. It appears as a heterogeneous lesion with variable contrast enhancement on CT scan.[2] The majority of cases (87%) manifest with a solitary (multiple in 5%) lung mass measuring between 1 and 6 cm with sharp edges, pleural effusion (10%), and atelectasis (8%).[5] In IMT, surgical treatment is the preferred procedure.[7,8] Complete resection is the most significant prognostic factor, as the minimum volume of surgery without lymphatic dissection is recommended.[4] Endoscopic extirpation of central endobronchial lesions is also possible.[9] In one study, the following surgical procedures were reported: lobectomy (29.4%), wedge resection (29.4%), pneumonectomy (17.7%), segmentectomy (17.7%), and bronchial sleeve resection (5.8%).[4]

Bronchi and vessels may be infiltrated by myofibroblastic inflammatory tumors. We observed a central lesion in the hilum with left pulmonary artery trunk involvement. Because of this, a pneumonectomy was conducted. Multiple inflammatory changes of the lung parenchyma, parietal pleura, and adhesions that developed over time made the surgery exceedingly challenging. The intraoperative hemorrhage during thoracic closure was promptly and effectively controlled with sutures and Atriclip 35. During systole and diastole of the heart, it most likely arose due to contact between the left atrium and the staplers from the medial portion of the pulmonary artery stump. Between the pulmonary artery stump and the cardiac auricle, a flap of viable tissue, such as a pericardial

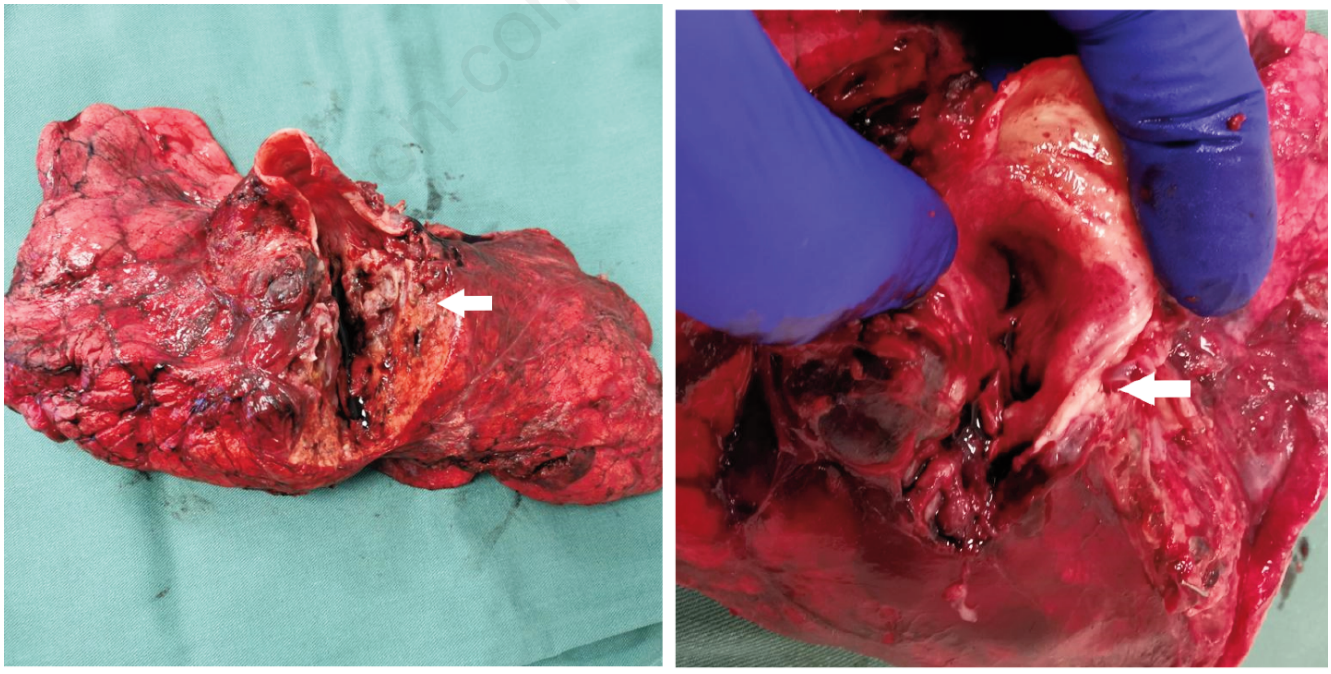

Figure 3. Postoperative specimen (left lung), showing a central lung inflammatory myofibroblastic tumor, obturating the left main pulmonary artery and bronchus.

fat pad, pediculated pericardium, or some hemostatic mesh, could be implanted to prevent this. A similar case with IMT causing left pulmonary artery stenosis was reported, which was inoperable and successfully treated with radiotherapy.[10]

Due to the potential malignancy of IMT and the reported recurrence rate of 14%, close monitoring is advised.[6]

Conclusions

Lung Inflammation Myofibroblastic Tumor is a rare disease for which preoperative diagnosis is frequently exceedingly difficult. Immunohistochemistry is vitally important. Particularly in the presence of hemoptysis, its primary treatment is surgical. Endoscopic extirpation is also possible in instances involving endobronchial centralization. We recommend the placement of a flap of viable tissue, such as a pericardial fat pad, a pediculated pericardium, or some hemostatic mesh, between the pulmonary artery stump and the cardiac atrium to prevent injury and bleeding of the latter from the pulmonary artery stump staplers during heart contraction.

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