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Case Report

Giant Inflammatory Pseudotumor of the Lung: a Case Report

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Abstract

The inflammatory myofibroblastic tumor, which was first described in 1939, is a rare neoplasm that accounts for 0.04%–0.7% of all lung neoplasms. These neoplasms occur most often in children, as they are the most common primary lung tumors in children. Preoperative diagnosis of such patients using bronchoscopy with endoluminal biopsy and transthoracic biopsy is not always informative and often the diagnosis can only be established during surgery. The presented case shows that on rare occasions, a giant myofibroblastic tumor of the lung may be encountered in adults, and radical intervention with subsequent rehabilitation can lead to full recovery.

Keywords

inflammatory myofibroblastic tumor, sternotomy, lung tumors

INTRODUCTION

Inflammatory pseudotumors were first described in 1905 in the orbital tissues of four patients with soft tissue infiltration by lymphocytes. The term 'pseudotumors' was coined because these tumors mimicked clinically and radiologically malignant neoplasms. The inflammatory myofibroblastic tumors are considered a subset of the inflammatory pseudotumors. These tumors, first described in 1939, account for 0.04% to 0.7% of all lung neoplasms.[1,2] These neoplasms occur most often in children as they are the most common primary lung tumors in children.[1,3] They are characterized by a lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils.[1] Inflammatory myofibroblastic tumors are usually benign and their removal leads to a complete cure. However, there are cases when they grow into surrounding structures, undergo malignant transformation, recur or metastasize.[4,5] Therefore, surgical treatment with complete removal of the tumor is the best way to treat this group of patients.

We present here a clinical case of successful removal of a large inflammatory myofibroblastic tumor in a 70-year-old female patient.

CASE REPORT

Patient Z, 70 years old, was consulted due to complaints of severe weakness and shortness of breath at minimal physical exertion (MRC III scale). Earlier in April 2021, she received a scheduled examination, which revealed a tumor in the lower lobe of the left lung, left-sided small hydrothorax (300 ml) and hydropericardium (500 ml). The patient was diagnosed with chronic autoimmune thyroiditis, subcompensated type 1 diabetes mellitus, bronchial asthma, hypertension grade

2, risk of cardiovascular events 3, and urolithiasis. A chest X-ray examination on April 29, 2021 revealed that the lung field was darkened from the 4th to the 12th rib of subcostal density, consolidated rib fractures on the left ***(Fig. 1A)****.*

The computed tomography (CT) scan of the chest, performed on March 23, 2021, found a mass in the lower lobe of the left lung (178×101 mm) with obturation of the lower lobe bronchus, and a layer of fluid (25 mm). According to the results of echocardiography on March 25, 2021, there was hydropericardium (500 ml) with an ejection fraction of 43%. Bone scintigraphy on April 6, 2021, revealed hyperfixation of the radiopharmaceutical agent in the left knee joint, a dim area in Th11, a pronounced uneven distribution in the thoracic spine. The MRI of the brain with intravenous contrast on March 26, 2021 revealed small foci of ischemic origin.

The patient received conservative treatment with diuretics (furasemide, 80 mg, prednisolone, 60 mg) and subjectively noted an improvement in her well-being. At the control echocardiogram on April 29, 2021, the systolic function of the left ventricle was satisfactory, no zones of impaired local contractility of the left ventricle were detected, slight mitral regurgitation, pulmonary regurgitation, moderate tricuspid regurgitation, moderate pulmonary hypertension, and no fluid in the pericardial cavity.

According to the results of bronchoscopy on April 30, 2021, no pathology was detected in the right lung. During bronchoscopy of the left lung, the lumen of the lower lobe bronchus was narrowed by 2/3 due to infiltration and the peribronchially located tumor tissue component: the mucosa in this area was pale, infiltrated, with a tortuous, deformed vascular pattern. On instrumental palpation, the tissues were dense. The lumen of B6 was narrowed by 1/2 due to mucosal infiltration. The lumen of the basal bronchi was narrowed due to infiltration and extraorganic compression. According to the results of the bronchoscopy, the patient had a peripheral formation of the lower lobe of the left lung with endoscopic signs of centralization, a peribronchial form of growth with formation of 3rd degree stenosis of the lumen of the lower lobe bronchus. According to the results of the histologic study, there was diffuse bilateral atrophic bronchitis with signs of inflammation.

Esophagogastroduodenoscopy and colonoscopy under general anesthesia were performed on April 30, 2021, during which no pathological changes were detected. Ultrasound of the pleural cavity was performed on May 4, 2021, with transthoracic biopsy. According to the biopsy results, no tumor growth was detected. Ultrasound of regional lymphatic vessels did not reveal pathological changes.

A CT scan of the abdominal cavity and retroperitoneal organs with intravenous contrast was performed in May, 2021, according to the results of which there was a mass located in the left hemithorax (184 mm in the longest part, 128 mm in the widest part), more typical for a fibrous tumor of the pleura with compression atelectasis of the lower lobe of the left

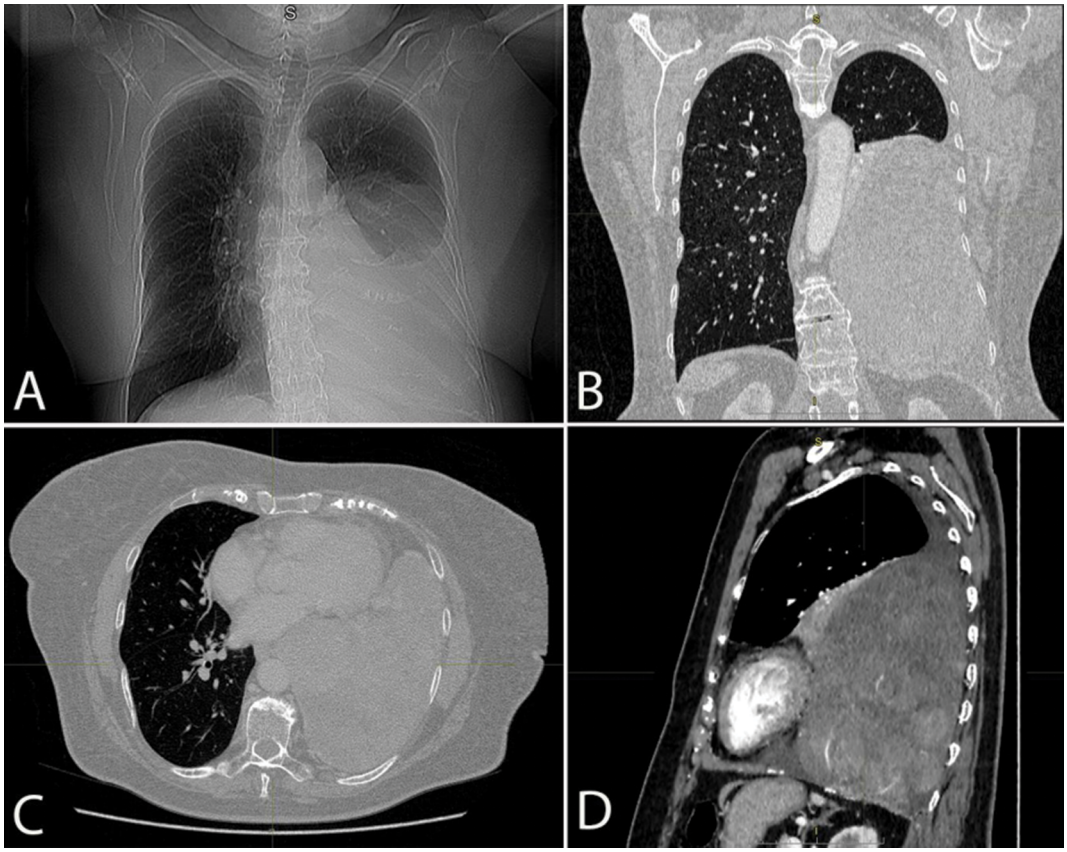

Figure 1. A. Chest X-ray; **B**. CT scan of the chest (frontal section); **C**. CT scan of the chest (axial section); **D**. CT scan of the chest (sagittal section).

lung, diastasis of the muscles of the anterior abdominal wall, dolichocolon, signs of osteoporosis, consolidated rib fractures on the left, and degenerative changes in the spine ***(Figs 1B, 1C, 1D)****.*

A multidisciplinary oncological consilium consisting of a surgeon-oncologist, chemotherapist, cardiologist, radiologist, and anesthesiologist was held. Taking into account the size of the formation and the impossibility of performing a minimally invasive surgical intervention, it was decided to administer surgical treatment under general anesthesia with separate bronchial intubation. Considering the size and topography of the mass, the operative approach was chosen through sternotomy.

On August 27, 2021, a lung segmentectomy was performed. After performing a sternotomy and revision of the pleural cavity, a volumetric formation measuring approximately 20 cm by 18 cm was visualized arising from the lower lobe of the left lung and fixed with dense fibrous adhesions to the diaphragm and surrounding tissues. We then performed separation of adhesions and allocation of the mass. The tumor was cut off with the Eshelon 60 mm (Blue Cassette) apparatus at the level of healthy lung tissues. The control of aerostasis and hemostasis was performed. Drainages were installed in the costophrenic sinus and the dome of the pleural cavity, brought out through counter-openings along the posterior and anterior axillary lines, followed by layer-by-layer stitching of the wound.

On macroscopic examination, the tumor was 18×16×14 cm, gray on section, lobulated with hemorrhage fields, necrosis, and cysts ***(Fig. 2)****.*

Microscopic examination showed that the tumor node is represented by intertwining fusiform cells with fibrous fields, weak lymphocytic infiltration, and resection margin without tumor growth (R0). In order to determine the histogenesis of the tumor, an immunohistochemical study was performed, according to the results of which the tumor cells did not express S100 and p53, weak focal reactions with SMA (in the spindle cell component), which allows us to conclude that the tumor was an inflammatory myofibroblastic tumor of the lung (ICD O - 8825/1) ***(Fig. 3)****.*

We continued the conservative therapy with diuretic and corticosteroids. The postoperative period was uneventful, the drains were removed 3 days after operation, and the patient was discharged 7 days after surgery. During control CT scan after 1 month, there was bilateral minimal hydrothorax, atelectasis of the lower lobes of both lungs, and degenerative changes in the spine ***(Fig. 4)****. At the follow-up examination at 6 months, there was no recurrence.*

DISCUSSION

There are many risk factors for developing inflammatory myofibroblastic tumors. Major among them are infectious diseases (pulmonary tuberculosis, pseudomonas infection of the lungs, *Moraxella catarrhalis*, actinomycetes, mycoplasmas, mycobacteria, Epstein-Barr virus, and human her-

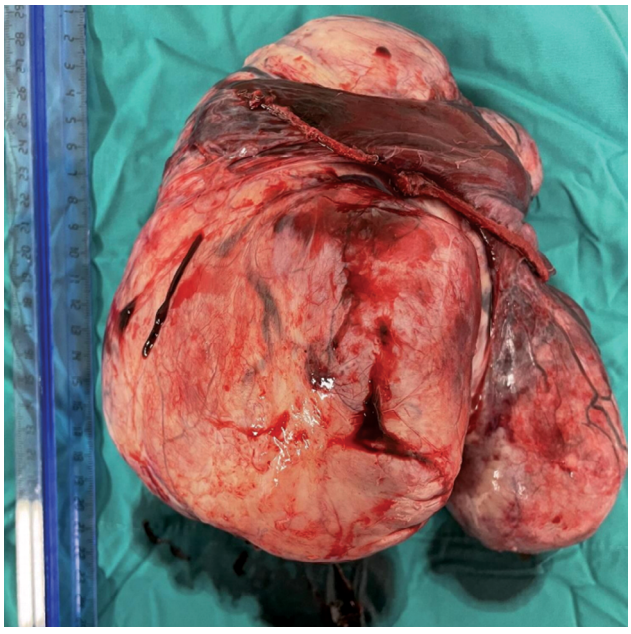

Figure 2. Macroscopic specimen of the tumor.

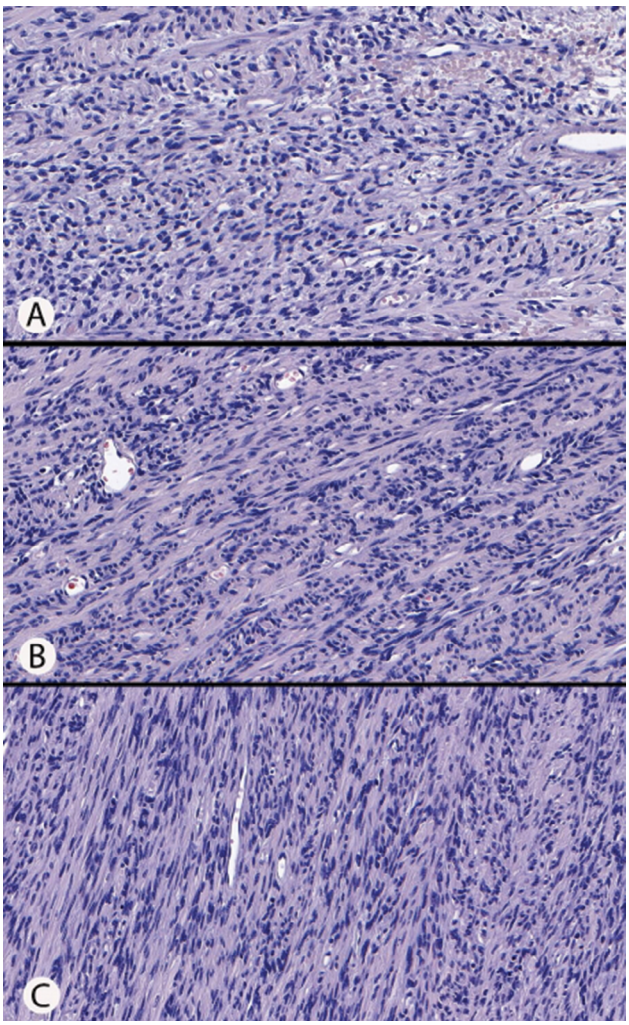

Figure 3. Histological picture of the tumor (**A**, **B**, and **C**).

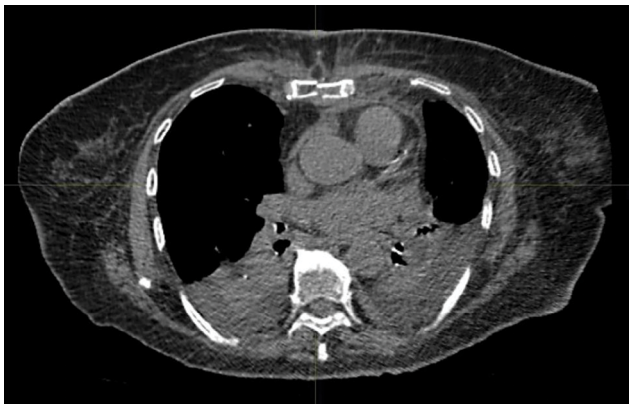

Figure 4. Control CT scan of the chest. Small bilateral hydrothorax with atelectasis of the lower lobes.

pes virus 8), autoimmune diseases (Schörgen's syndrome), oncological diseases (B-cell lymphoma), and patients after organ transplantation.[1]

Patients usually present with non-specific symptoms such as cough, dyspnea, hemoptysis, chest pain, fever, and weakness.[2,6]

Most often, inflammatory myofibroblastic tumors occur in the lower lobes with peripheral and subpleural localization.[3] On chest X-ray, pulmonary inflammatory myofibroblastic tumors appear as solitary, localized, lobulated lesions, predominantly localized in the lower lobes, sometimes with pleural effusions.[7] Differential diagnosis is carried out with primary or secondary neoplasms, hamartomas, hemangiomas or chondromas.[8] On CT, inflammatory pseudotumors have a variable and nonspecific appearance. On T1-weighted magnetic resonance images, these tumors have medium signal intensity and high signal intensity on T2-weighted images.[9] When the tumor becomes large, it can be difficult to locate the primary focus from which the tumor grew[10] Intralesional calcification is more common in children than in adults. The nature of the calcification can vary from amorphous, mixed, or fine-grained to severe mineralization. The size of the tumor varies in a wide range from 0.5 to 36 cm.[11] However, in most cases, tumors reach 1–6 cm in diameter.[2,6,12]

Surgery is considered the main method of treatment. Complete surgical resection, when possible, is the best method of treatment and histological verification.[7,8] A core-needle biopsy or endobronchial biopsy, although a standard of care, can be inconclusive. Depending on the volume of lung tissue involved in the pathological process, complete cure requires atypical lung resection, and in some cases lobectomy or pneumonectomy. At the same time, relapses occur in about 8% of cases.[4] In rare cases, when the tumor is localized within the trachea and respiratory tract, endoscopic resection may be an alternative. In these cases, bronchoscopy with endobronchial resection (surgical excision or laser) can become a possible alternative to surgery. If complete resection is not possible due to anatomical location or comorbidity, medical therapy (chemotherapy and hormonal therapy) combined with radiation therapy may be considered. This is especially true for multifocal, invasive processes or local recurrence.[6,13] It is also important to mention that surgery usually leaves behind large tissue defects of the thorax that often requires reconstruction.[14] The current case demonstrates that multidisciplinary centers where different specialist can consult and stabilize a patient with comorbid conditions are optimal for management of complicated cases.

CONCLUSION

Giant pseudotumors of the lungs are a rare group of diseases. Preoperative diagnosis of such patients using bronchoscopy with endoluminal biopsy and transthoracic biopsy is not always informative and the diagnosis can only be established during surgery. The presented case shows that a giant myofibroblastic tumor of the lung is rare, but can occur also in elderly patients. Radical intervention in these cases with subsequent rehabilitation can lead to full recovery.

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Гигантская воспалительная псевдоопухоль лёгкого: клинический случай

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Резюме

Воспалительная миофибробластическая опухоль, впервые описанная в 1939 г., является редким новообразованием, на долю которого приходится 0.04 – 0.7% всех новообразований лёгких. Эти новообразования чаще всего встречаются у детей, так как они являются наиболее распространёнными первичными опухолями лёгких у детей. Предоперационная диагностика таких больных с помощью бронхоскопии с эндолюминальной биопсией и трансторакальной биопсией не всегда информативна и часто диагноз удаётся установить только во время операции. Представленный случай показывает, что в редких случаях у взрослых может встречаться гигантская миофибробластическая опухоль лёгкого, а радикальное вмешательство с последующей реабилитацией может привести к полному выздоровлению.

Ключевые слова

воспалительная миофибробластная опухоль, стернотомия, опухоли лёгкого