Case Report
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Localized Fibrosing Mediastinitis Causing Pulmonary Infarction: A Case Report
폐경색을 유발한 섬유성 종격동염: 증례 보고

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A 44-year-old female patient visited our emergency room for hemoptysis and refractory chest wall pain of 2 months duration. She had no history of smoking or other medical conditions. Chest CT scan showed homogenously enhancing soft tissue mass without calcification at the left pulmonary hilum. Encasing and compression of the left lower pulmonary artery by the mass had resulted in pulmonary infarction in the left lower lobe. Laboratory tests for tuberculosis, fungus, and vasculitis were all negative. The patient underwent surgical biopsy and resection of infarcted left lower lobe that was histopathologically confirmed as fibrosing mediastinitis. Herein, we reported a rare case of surgically confirmed and treated localized fibrosing mediastinitis causing pulmonary infarction.

Index terms
Fibrosing Mediastinitis
Mediastinal Fibrosis
Sclerosing Mediastinitis

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INTRODUCTION

Fibrosing mediastinitis is a rare benign disorder that is characterized by deposition of dense fibrous tissue throughout the mediastinum and hilar regions (1). Resultant infiltrative or compressive effects on vital mediastinal organs such as central systemic vein, airway, and pulmonary vessels, could increase morbidity and in rare circumstances even mortality (2). We reported a rare case of fibrosing mediastinitis causing pulmonary infarction by encasing pulmonary artery that was detected on chest computed tomography (CT).

CASE REPORT

A 44-year-old female patient visited the emergency room (ER) of a tertiary referral university hospital due to acute hemoptysis and right chest wall pain of 2 months duration. Her chest pain was refractory to prior conservative treatment in a local clinic. The patient was a nonsmoker and had no particular medical history. On laboratory tests, serum erythrocyte sedimentation rate was mildly elevated but other parameters were within normal limits. Plain chest radiograph demonstrated patchy increased opacity in left lower lung zone and small amount of left pleural effusion (Fig. 1). Considering the refractory nature of her chest pain and the acute onset of hemoptysis, post-contrast chest CT was immediately conducted in the ER. Chest CT revealed soft tissue mass at the left hilar region, which showed homogenous enhancement and was encasing and provoking near total occlusion of left inferior pulmonary artery (Fig. 2A). There was no visible calcification within the mass. Wedge-shaped subpleural consolidation was seen in the left lower lobe, which was the corresponding vascular territory of the encased

left lower pulmonary artery (Fig. 2B). Therefore, this consolidative lesion could be regarded as pulmonary infarction. Laboratory screenings with antinuclear antibody, rheumatoid factor, and Anti-neutrophil cytoplasmic antibody were negative.

Without further investigation such as positron emission tomography-CT or bronchoscopy, open thoracotomy biopsy was performed for confirmative diagnosis owing to the patient's severe symptoms. Extensive left peribronchial fibrosis and soft tissue mass encasing left inferior pulmonary artery was seen intra-operatively. Except for superior segment, the remaining entire left lower lobe was rigid on palpation and there were multifocal scattered infarctions.

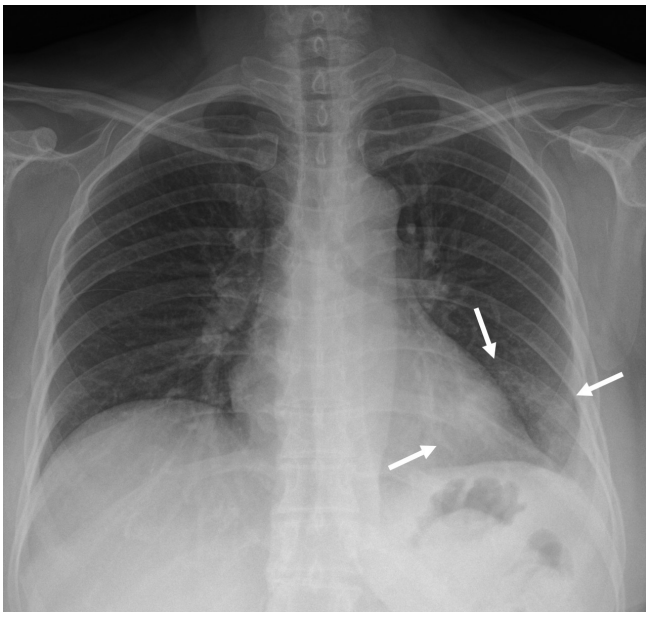
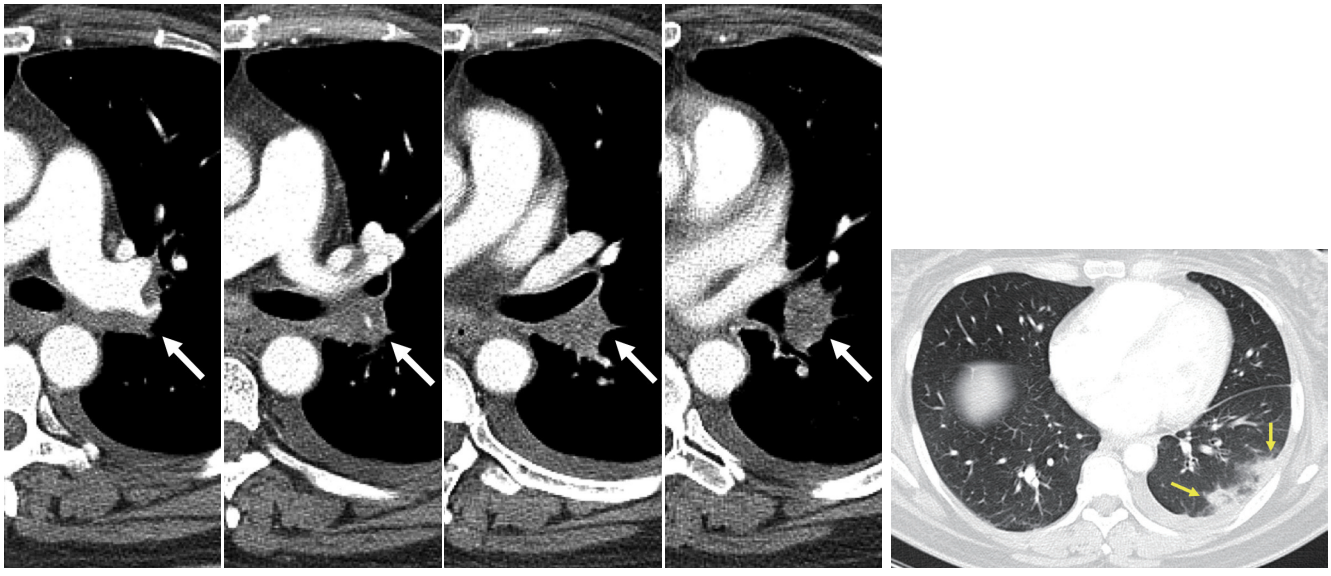


Fig. 1. Plain chest radiograph shows patchy increased opacity in left lower lung zone (arrows) and small amount of left pleural effusion.

Gross examination of resected specimen revealed firm white fibrous tissue surrounding the left hilum (Fig. 3A). Histopathologically, the mass consisted of dense fibrosis surrounding and infiltrating pulmonary vasculature with extension into the lung parenchyme (Fig. 3B). Occasional chronic necrotizing granulomas were seen within the dense fibrosis (Fig. 3C). Polymerase chain reaction (PCR) for tuberculosis was negative and Gomori methenamine silver (GMS) stain for fungal study was also negative. Consequently, the final diagnosis was fibrosing mediastinitis. The patient underwent follow-up for 3 years and was free from symptom and recurrence on imaging.

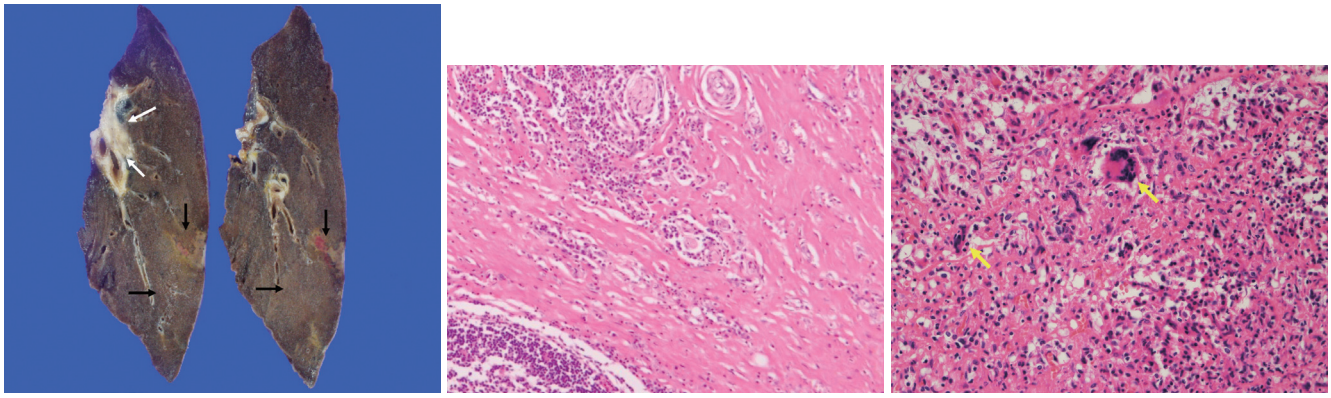
DISCUSSION

Fibrosing mediastinitis is a benign progressive non-neoplastic disease characterized by infiltration of excessive fibrous tissue within the mediastinum mediated by abnormal immune



A B

Fig. 2. Representative CT images of localized fibrosing mediastintis.
A. Preoperative post-contrast chest CT demonstrates mediastinal soft tissue mass extending into left hilar region and encasing left lower pulmonary artery, causing severe stenosis or occlusion (white arrows).
B. Multifocal wedge shaped consolidations with surrounding ground glass attenuation at subpleural area of left lower lobe suggesting pulmonary infarctions (yellow arrows).



A B C

Fig. 3. Gross and microscopic specimens of localized fibrosing mediastintis.
A. Photograph of cut surface of resected specimen demonstrates dense whitish fibrous tissue (white arrows) in left pulmonary hilum along the bronchovascular bundle with peripheral pulmonary infarctions (black arrows).
B. High-power photomicrograph (original magnification, × 100, hematoxylin-eosin stain) of peripheral lung demonstrates extensive dense fibrosis.
C. High-power photomicrograph (original magnification, × 100, hematoxylin-eosin stain) of left pulmonary hilum shows granulomatous inflammation (yellow arrows) with necrosis.

reaction to various antigens or underlying disease (3). Fibrosing mediastinitis is interchangeably known as mediastinal granuloma and mediastinal fibrosis, sclerosing mediastinitis, and granulomatous mediastinitis, based on dominant pathologic features (4). Although it is still controversial, Loyd et al. (5) postulated that granulomatous mediastinitis evolves into fibrosing mediastinitis; hence, granulomatous mediastinitis and fibrosing mediastinitis may represent 2 ends of a spectrum of abnormal inflammation and fibroproliferation in the mediastinum (6).

In our case, pathologic examination indicated extensive dense fibrosis surrounding and infiltrating pulmonary vasculature with extension into the lung parenchyme and occasional chronic necrotizing granulomas within the dense fibrosis. Thus, our case showed both dominant features of fibrosing mediastinitis and minor features of granulomatous mediastinitis.

Fibrosing mediastinitis is most commonly associated with histoplasma infection in North America (5). Other possible triggers of fibrosing mediastinitis are reportedly tuberculosis, fungal infections, and sarcoidosis (6). However, if certain identifiable cause is absent, the disease is categorized as idiopathic (6). In many cases, no specific microorganism is identified and thus categorized as idiopathic (7). In our case, laboratory examination and elastin stain for vasculitis were negative and investigation for immunoglobulin G4-related disease was also negative. In addition, PCR test for tuberculosis and non-tuberculosis *mycobacterium* were negative, as well as GMS tests for fungal study. Therefore, our case could be categorized as idiopathic fibrosing mediastinitis.

Fibrosing mediastinitis may present with various clinical manifestations according to the anatomical location involved. While some authors have reported that bronchial narrowing and superior vena cava (SVC) obstruction are the most common complication of fibrosing mediastinitis (8), others have reported pulmonary artery stenosis followed by SVC narrowing as the most common and significant manifestation (3). On CT scan, fibrosing mediastinitis is characterized by localized or diffuse soft-tissue mass in mediastinum obliterating fat planes and causing compression of adjacent structures. Additional abnormal findings associated with disease localization within the pleura and lungs could include pulmonary infarction from pulmonary artery obstruction, as in our case (3). CT can accurately identify tracheobronchial narrowing, major vessel obstruction, and degree of pulmonary arterial hypertension. In our patient, CT demonstrated the typical findings of fibrosing mediastinitis. Differential diagnosis included other mediastinal mass forming diseases such as Castleman's disease and lymphoma. The mass was non-calcified soft tissue mass and surgical tissue acquisition was necessary to exclude neoplastic diseases.

The treatment of fibrosing mediastinitis is controversial. The effect of systemic antifungal or antituberculous therapy is reported in cases with confirmed causative microorganism (9). However, in cases of granuloma or fibrosis causing compression of

mediastinal organ, surgical resection of affected tissue is necessary (10). In our case, the patient suffered from chest wall pain and blood tinged sputum as a result of pulmonary infarction. Surgical resection was necessary for symptom relief as well as histopathologic diagnosis. Consequently, the patient's symptoms were completely resolved for the following 3 years follow up period.

Our case demonstrated the rare occurrence of fibrosing mediastinitis with pulmonary artery occlusion and implication of surgical resection for local disease control.

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폐경색을 유발한 섬유성 종격동염: 증례 보고

서영균[1] · 김은영[1] · 강두경[1] · 박경주[1] · 고영화[2] · 선주성[1]*

흡연력이나 특이 과거력이 없는 44세 여성이 급성객혈과 두 달간 지속된 흉통을 주소로 응급실에 내원하였다. 내원하여 시행한 흉부 전산화단층촬영에서 좌측 폐문부에 균질한 조영증강을 보이면서 석회화가 없는 연조직 종괴가 관찰되었으며, 이 종괴는 좌하폐 동맥을 에워싸며 협착을 일으켜 좌하엽에 폐경색을 유발하고 있었다. 폐결핵, 진균 및 혈관염에 대한 임상검사는 모두 음성이었다. 개흉 수술을 통해 조직검사 및 경색된 좌하엽의 절제를 시행하였으며, 환자는 조직 병리학 검사 결과 섬유성 종격동염으로 확진되었다. 이는 수술적으로 치료 및 확진한 국소성 섬유성 종격동염의 드문 증례이다.

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