Case Report

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대한영상의학회지

Fibrosing Mediastinits Causing Obstruction of Left Lower Lobar Bronchus: A Pediatric Case Report

좌하엽 기관지 폐쇄를 유발한 섬유성 종격동염: 소아 증례 보고

Young Woo Sim, MD[1], Young Seon Kim, MD[1]*, Seung Eun Lee, MD[1], Min Hye Jang, MD[2]

Departments of [1]Radiology and [2]Pathology, Yeungnam University Hospital, Yeungnam University College of Medicine, Daegu, Korea

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***Corresponding author**
Young Seon Kim, MD
Department of Radiology, Yeungnam University Hospital, Yeungnam University College of Medicine, 170 Hyeonchung-ro, Nam-gu, Daegu 42415, Korea.

Tel 82-53-620-4129
Fax 82-53-653-5484
E-mail youngseon29@gmail.com

ORCID iDs
Young Woo Sim
https://orcid.org/0000-0002-3868-0238
Young Seon Kim
https://orcid.org/0000-0002-9168-8204
Seung Eun Lee
https://orcid.org/0000-0001-6693-4752
Min Hye Jang
https://orcid.org/0000-0002-4270-8897

Fibrosing mediastinitis is a rare benign disorder characterized by the proliferation of dense fibrous tissue within the mediastinum. It typically manifests as localized or infiltrative soft-tissue masses in the middle mediastinum or hilar area, which cause compression and encasement of adjacent mediastinal structures, such as the vessels or airway. Here, we report a rare case of fibrosing mediastinitis in a 13-year-old girl that presented as a middle mediastinal mass lesion on CT scan with obliterating left lower lobar bronchus. The patient's symptoms and follow-up chest CT showed significant improvement following systemic corticosteroid treatment. As fibrosing mediastinitis can improve with systemic steroid therapy, radiologists must be aware of its radiologic findings when discriminating between infiltrating soft tissue lesions in the mediastinum.

Index terms Computed Tomography, X-Ray; Mediastinal Fibrosis; Sclerosing Mediastinitis

INTRODUCTION

Fibrosing mediastinitis, also known as sclerosing mediastinitis and mediastinal fibrosis, is a rare benign disorder characterized by the proliferation of locally invasive fibrous tissue within the mediastinum (1). While several triggers have been associated with fibrosing mediastinitis, including fungal infections, tuberculosis, and sarcoidosis (granulomatous type), some fibrosing mediastinitis is categorized as idiopathic (non-granulomatous type) (1). According to the previous literature, there are two subtypes of fibrosing mediastinitis: the granulomatous type (usually focal form), and the non-granulomatous

type (usually diffuse form, also known as idiopathic type) (1, 2).

Fibrosing mediastinitis is characterized by the presence of extensive fibrous tissue throughout the middle mediastinum or hilar areas, leading to the compression, encasement, or invasion of anatomic structures within the mediastinum such as the superior vena cava, pulmonary veins, esophagus, or bronchi (2, 3). The imaging findings of fibrosing mediastinitis range from focal masses with or without stippled calcification to diffuse, infiltrative soft tissue density lesions encasing the adjacent anatomical structures (1-5). Long-standing obstruction of the vascular structure, esophagus, or bronchi causes various clinical symptoms. Chronic obstruction of the pulmonary vascular structure may lead to pulmonary infarction, hypertension, and cor pulmonale (3, 5, 6). while chronic obstruction of the bronchi may lead to dyspnea and recurrent pneumonia (7, 8).

Herein, we report a case of fibrosing mediastinitis in a 13-year-old girl admitted to our hospital with a chief complaint of dyspnea.

CASE REPORT

A 13-year-old girl presenting with acute-onset dyspnea was admitted to our institution. She also complained intermittent chest discomfort lasting for several months, without history of fever. Her oxygen saturation on pulse oximetry was 93% and her other vital signs were stable. The patient had no underlying disease or medication history and no critical family history.

The findings of routine laboratory examinations were unremarkable. The initial chest posteroanterior view demonstrated loss of retrocardiac lucency with a decreased left hemithorax volume, indicating left lower lobar atelectasis (Fig. 1A). A contrast-enhanced CT scan of the chest revealed an infiltrative lesion with soft tissue attenuation in the middle mediastinum that encased and narrowed the left lower lobar bronchus (Fig. 1B, C).

Considering the absence of clinical signs of infection, our first impression was infiltrative mediastinal masses such as Castleman's disease, lymphoma, or other malignancies. For pathological confirmative diagnosis, we performed a bronchoscopy-guided biopsy of the mediastinal lesion. During this examination, obstruction of the left main bronchus due to external compression was observed. However, the biopsy specimen showed mainly blood with some squeezed lymphocytes without evidence of malignancy. Therefore, the pathology department recommended a re-biopsy for accurate diagnosis.

Therefore, the patient underwent a surgical open mediastinal biopsy for pathological confirmation. The intraoperative findings revealed fibrotic tissue between the aorta and pulmonary artery, encasing the left main bronchus. The pathological findings revealed diffuse fibrosis with dense collagen deposition and squeezed lymphocytic infiltration, indicating a possible fibrosing mediastinitis diagnosis (Fig. 1D, hematoxylin and eosin stain, × 40). Gomori methenamine silver (GMS) staining results for fungal studies were negative. Granulomatous inflammation was not observed within the specimen.

The patient was treated with a systemic corticosteroid (Methysol®, Alvogen Korea, Seoul, Korea). After treatment, the patient's symptoms and serial follow-up chest radiographs showed significant improvement. Follow-up chest CT after 1 month also showed significant regression of the mediastinal lesion (Fig. 1E, F).

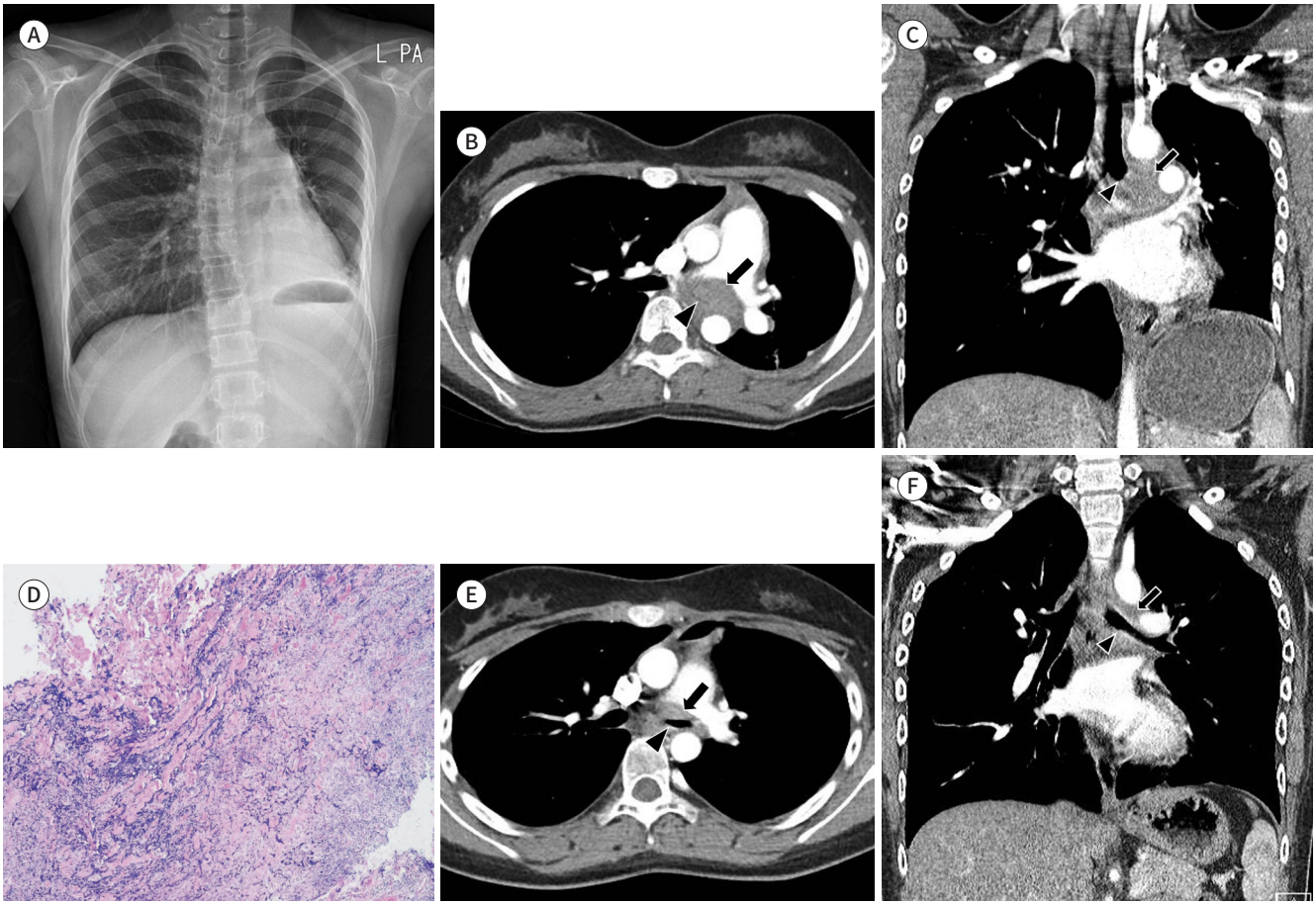
After discharge, laboratory studies performed for other infections such as tuberculosis and aspergillosis revealed negative results. The patients underwent follow-up for 2 months and remained free of symptoms, without recurrence on radiography.

This retrospective study was approved by the Institutional Review Board of Yeungnam University Hospital (IRB No. 2020-12-022), which waived the requirement for informed consent.

DISCUSSION

Fibrosing mediastinitis, also known as mediastinal or sclerosing mediastinitis, is an uncommon benign disorder characterized by the proliferation of dense fibrous tissue within

Fig. 1. A 13-year-old girl with fibrosing mediastinitis leading to obstruction of the left main bronchus.
A. Initial chest PA shows volume loss in the left hemithorax with elevated left hemidiaphragm and loss of retrocardiac lucency indicating left lower lobar atelectasis.
B, C. Axial (B) and coronal (C) views of the mediastinal setting of contrast-enhanced chest CT show an infiltrative lesion with soft-tissue attenuation in the middle mediastinum (arrows), with narrowing left lower lobar bronchus (arrowheads); the left upper and lingular bronchus are patent.
D. A representative photo of a mediastinal biopsy shows diffuse fibrosis with dense collagen deposition and squeezed lymphocytic infiltration (hematoxylin and eosin stain, × 40).
E, F. Following systemic corticosteroid therapy, follow-up chest CT after 1 month show significant regression of the mediastinal lesion (arrows) as well as improved luminal patency of the left lower lobar bronchus (arrowheads).
PA = posteroanterior



the mediastinum (1). While the pathogenesis remains unclear, the most generally accepted hypothesis is delayed hypersensitivity reactions to various antigens (1). In most cases, Fibrosing mediastinitis occurs following exposure to Histoplasma capsulatum or other infections such as tuberculosis, other fungi, and inflammatory conditions, such as sarcoidosis (2, 6).

Fibrosing mediastinitis has two major subtypes: granulomatous (focal) and non-granulomatous (diffuse or idiopathic). Granulomatous type fibrosing mediastinitis is commonly associated with prior histoplasmosis or tuberculosis infection, which commonly affects young patients (mean age, 35–46 years) (2). Non-granulomatous fibrosing mediastinitis is less common, representing approximately 10%–20% of fibrosing mediastinitis cases. Non-granulomatousfibrosing mediastinitis is considered an idiopathic reaction to medical treatment (e.g., methysergide), or autoimmune syndrome (2). Non-granulomatous fibrosing mediastinitis usually affects middle-aged or elderly individuals (2). However, in our case, GMS tests for fungal species were negative; moreover, laboratory examinations for tuberculosis and non-tuberculosis mycobacterium were also negative. Therefore, this case could be categorized as non-granulomatous (idiopathic) fibrosing mediastinitis.

On CT scans, granulomatous fibrosing mediastinitis typically manifests as a localized or infiltrative soft tissue density mass, usually with stippled or dense calcification (3). The mass usually obliterates adjacent mediastinal structures, such as pulmonary and systemic vessels or airways. Identifying the signs of previous histoplasmoses, such as calcified pulmonary granuloma, facilitates diagnosis (2).

The imaging findings of non-granulomatous fibrosing mediastinitis may overlap with those of granulomatous fibrosing mediastinitis, although calcification is typically absent and extrathoracic imaging findings such as retroperitoneal fibrosis may be observed in non-granulomatous fibrosing mediastinitis (2). Our case shows diffuse infiltrative lesion without calcification in the mediastinum, which is consistent with imaging findings of non-granulomatous (also known as idiopathic) fibrosing mediastinitis in previous literature.

The differential diagnosis of fibrosing mediastinitis on imaging includes primary lung cancer, mediastinal or hilar metastases, lymphoma, atypical sarcoidosis, and Castleman disease (2, 9, 10).

Many cases of fibrosing mediastinitis are reported, but pediatric cases are only a few. Our case shows pediatric patient manifestated as dyspnea, which is clinically important symptom. It was difficult to consider fibrosing mediastinitis as a first impression, but CT revealed typical finding of fibrosing mediastinitis; namely, a soft tissue mass with narrowing of the left main bronchus.

The prognosis of patients with fibrosing mediastinitis varies due to its unpredictable course (1). Most patients show slow disease progression over years; some patients remain stable and a few may experience spontaneous regression (1). Antifungal agents and corticosteroids have been used in the treatment of fibrosing mediastinitis; however, their efficacy has not been quantified. Surgical and nonsurgical procedures can be used to relieve the secondary symptoms of compression of mediastinal structures; however, the durability of these therapeutic interventions is frequently limited (1). In our case, the patient's symptoms and follow-up imaging studies showed improvement after the administration of systemic corticosteroid therapy.

In conclusion, we report a case of fibrosing mediastinitis that presented as a middle mediastinal mass lesion with obliterating left main bronchus on CT scan that responded to corticosteroid therapy. As fibrosing mediastinitis can improve with systemic steroid therapy, radiologists must be aware of the radiologic findings of fibrosing mediastinitis when discriminating against infiltrating soft tissue lesions in the mediastinum.

Author Contributions

Conceptualization, S.Y.W., K.Y.S.; data curation, all authors; formal analysis, all authors; investigation, all authors; supervision, K.Y.S.; writing—original draft, S.Y.W.; and writing—review & editing, K.Y.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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좌하엽 기관지 폐쇄를 유발한 섬유성 종격동염: 소아 증례 보고

심영우[1] · 김영선[1*] · 이승은[1] · 장민혜[2]

섬유성 종격동염은 종격동 내 고밀도 섬유 조직의 증식을 특징으로 하는 드문 양성 질환이다. 섬유성 종격동염은 일반적으로 중간 종격동 또는 폐문 부위의 국소적 또는 침윤성 연조직 종괴로 나타나며, 이는 혈관이나 기도와 같은 인접한 종격동 구조를 둘러싸거나 압박할 수 있다. 본 증례 보고에서 우리는 좌하엽 기관지의 폐색을 유발하는 종격동 종괴의 양상으로 나타난 13세 소녀의 섬유성 종격동염의 드문 증례를 보고하고자 한다. 환자는 전신 코르티코스테로이드 치료 후 증상과 추적관찰 흉부 CT 소견에서 호전되었다. 섬유성 종격동염은 전신 스테로이드 요법으로 호전될 수 있으므로 영상의학과 의사는 종격동의 침윤성 연조직 병변을 감별 진단할 때 섬유성 종격동염의 영상의학적 소견을 알고 있어야 한다.

영남대학교 의과대학 영남대학교병원 [1]영상의학과, [2]병리과