CASE REPORT

Fibrosing mediastinitis as an untypical complication of tuberculosis

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KEY WORDS

fibrosing mediastinitis, tuberculosis

ABSTRACT

Fibrosing mediastinitis (FM) is a rare benign disorder caused by proliferation of acellular collagen and fibrous tissue within the mediastinum. In the United States, most cases are thought to be caused by an abnormal immune response to *Histoplasma capsulatum* infection. Some cases of FM are related to tuberculosis. In most patients the cause of FM is unknown. The affected patients are typically young and present with signs and symptoms of obstruction or compression of the superior vena cava, pulmonary veins or arteries, central airways or esophagus. Computed tomography and magnetic resonance imaging are central to the diagnosis and management of this disorder. We present a rare case of FM as a sequela of tuberculosis.

INTRODUCTION Fibrosing mediastinitis (FM) is a rare benign disorder characterized by proliferation of dense fibrous tissue within the mediastinum.[1] The affected patients are typically young and present with signs and symptoms related to obstruction of vital mediastinal organs. Despite extensive investigation, the precise cause and pathogenesis of FM remains unknown. Most cases in the United States have been associated with *Histoplasma capsulatum* infection.[2] Sometimes other infections, especially tuberculosis, are implicated.[2]

We present a rare case of FM as an untypical manifestation of tuberculous infection.

CASE REPORT A 29-year-old woman was admitted to the National Research Institute of Tuberculosis and Lung Diseases with increasing shortness of breath and massive recurrent hemoptysis. Ten years earlier, in 1998, chest radiography performed due to persistent cough showed consolidations in the left upper lobe with the left hilar lymphadenopathy. Bronchoscopy revealed concentric narrowing of the lingular bronchus with a friable cicatricial lesion. Thoracoscopy was performed and microscopic examination of pulmonary tissue revealed tuberculous granulomas. Culture of the biopsy specimen was positive for *Mycobacterium tuberculosis*. Tuberculosis was diagnosed and the patient was treated with standard antituberculous drugs for 6 months. In 2004, she was again admitted to the local hospital because of massive hemoptysis with dyspnea. Contrast-enhanced computed tomography (CT) showed consolidations in the right upper lobe with mediastinal and hilar lymphadenopathy. During bronchoscopy fresh blood and concentric narrowing in the left main bronchus were visualized. The source of bleeding was not identified. Owing to suspicious nature of tuberculosis recurrence, the patient underwent antituberculosis therapy again. Clinical and radiological improvement was observed. In 2007, hemoptysis recurred. Contrast-enhanced CT scan demonstrated an infiltrating mediastinal mass with foci of calcification and ground-glass opacities in the right lower lobe. The mass encased the left pulmonary artery and the right pulmonary vein; the pulmonary trunk and the right pulmonary artery were considerably enlarged. We suspected hypoplasia of the pulmonary vessels and the inflammatory systemic disease, such as Wegener's granulomatosis. The mediastinal mass caused considerable concern.

The patient was admitted to our department. Magnetic resonance imaging (MRI) showed moderate enlargement of lymph nodes: mediastinal and left hilar nodes to 15 mm, lower left and right

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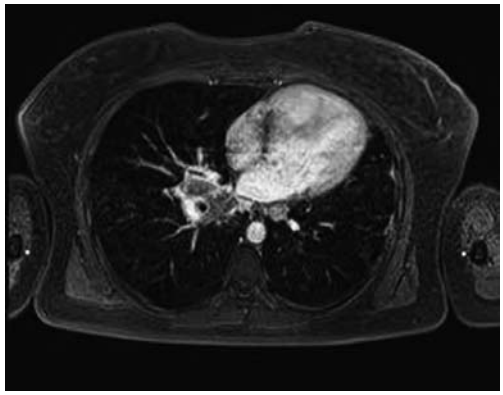

FIGURE 1 Transverse T1-weighted contrast-enhanced fat-suppressed image; abnormal mass in the right hilar region with narrowing of lower lobe bronchus and delicate contrast tissue enhancement; small abnormal tissue on the left side

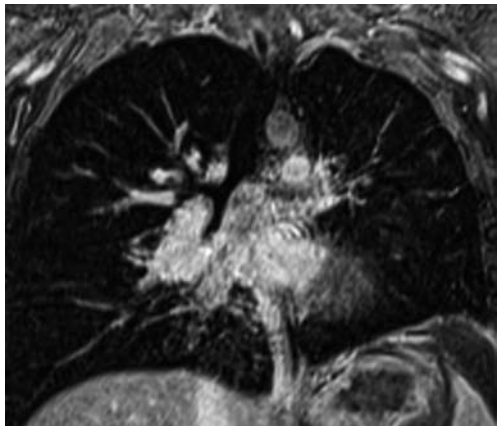

FIGURE 2 Coronal T1-weighted contrast-enhanced fat-suppressed image; abnormal hilar and mediastinal mass with contrast enhancement – narrowing of the intermedium bronchus

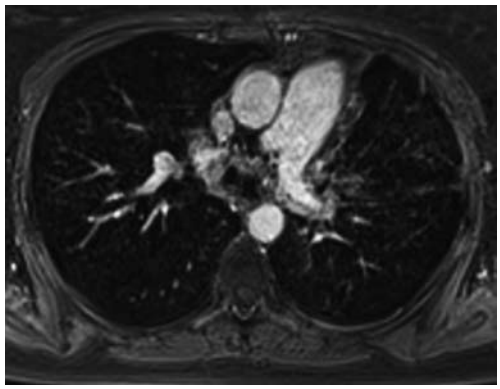

FIGURE 3 Transverse T1-weighted contrast-enhanced fat-suppressed image; abnormal left hilar mass narrowing the left pulmonary artery

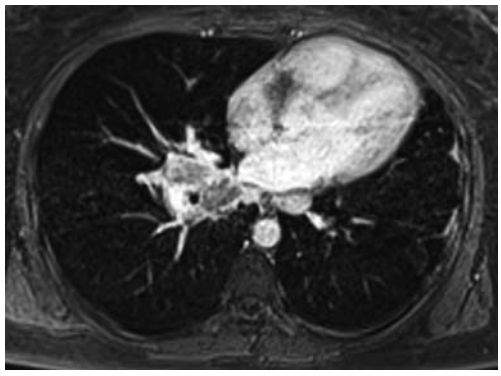

FIGURE 4 Transverse T1-weighted contrast-enhanced fat-suppressed image; abnormal right hilar mass narrowing the right pulmonary vein

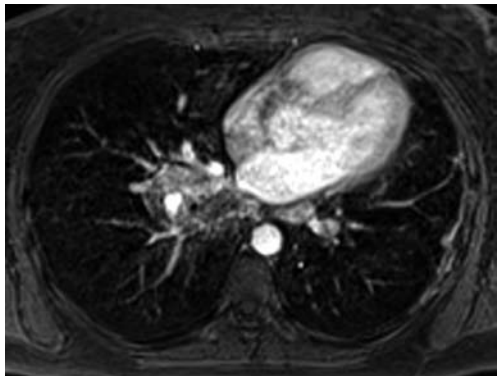

FIGURE 5 Chest magnetic resonance imaging performed 2 months after treatment showing enlarged right hilar and mediastinal mass and narrower right lower bronchus

paratracheal nodes to 12 and 13 mm, respectively, and subcarinal to 23 mm. Abnormal mass in both hilar regions caused narrowing of bronchial lumen (intermedium and both lower bronchi) and vessel structures. On the right side, the lower pulmonary vein was closed. On the left side, pulmonary artery was narrowed and both lobar arteries were completely closed, only the artery of 6 segment was normal. After contrast injection, lymph nodes and tissue masses showed a rim of peripheral enhancement (FIGURES 1, 2, 3, 4). Recently, FM was diagnosed and was associated with tuberculosis on the basis of the patient's history and calcifications found in subcarinal mass.

We attempted to treat the patient with prednisone 60 mg every other day, but MRI performed at 2 months showed that the mediastinal mass was bigger and the right lower bronchi was narrower (FIGURE 5). The treatment was terminated.

The patient is still under observation. The last time we examined her was in May 2009. She was in good general condition; there was no recurrence of hemoptysis, and the dyspnea subsided.

DISCUSSION FM is characterized by the proliferation of fibrous tissue and dense acellular collagen within the mediastinum, occasionally containing patchy infiltrates of mononuclear cells or less commonly granulomas. It is a rare, often progressive, and potentially lethal condition.[3]

Etiology is unknown in the majority of cases, but in endemic areas granulomatous infections have been implicated. The most common is *Histoplasma capsulatum* infection, particularly in North America,[2] while rare causes include tuberculosis, aspergillosis, mucormycosis, blastomycosis, acinomycosis, nocardiosis, coccidioidomycosis,and cryptococcosis.[2,4] FM has also been reported in cases of autoimmune disease, Behçet's disease, rheumatic fever, radiation therapy, trauma, Hodgkin's disease, silicosis, sarcoidosis, familial multifocal fibrosclerosis, and therapy with methysergide maleate.[2,4] Our patient had a history of pulmonary tuberculosis, which is a very likely cause of FM.

CT is considered the mainstay of diagnostic evaluation of patients with known or suspected FM.[5] It typically shows an infiltrative mass of soft-tissue attenuation that obliterates normal mediastinal fat planes and encases or invades adjacent structures, with calcification within the lesion, detection of which is an important diagnostic clue that may suggest tuberculosis as the cause.[5] MRI is comparable to CT in the detection and assessment of the extent to which the mediastinum and vascular structures have been affected. The differential diagnosis of FM includes sarcoidosis, malignant lymphoma, metastatic malignant tumors, thymoma, thymic carcinoma, malignant teratoid tumor, and vessel anomaly. Kozłowska et al. reported a patient frequently hospitalized to disclose the cause of a wide polycyclic pulmonary hilus. Digital subtraction

angiography showed the congenital anomaly of the pulmonary artery.[6]

Surgery is often performed to obtain specimens for histological diagnosis, but in some patients, like in our case, the typical CT findings may be sufficient to make tissue sampling unnecessary.[7] If the mediastinal mass does not contain calcification, the tissue should be obtained.

The clinical presentation of FM is a result of the anatomical location of the involved lymph nodes in relationship to mediastinal structures. Mechanical compression of the tracheobronchial tree, esophagus, superior vena cava (SVC), major pulmonary vessels and nerves causes symptoms. Initial symptoms typically include cough, dyspnea, hemoptysis (like in our patient), and chest pain. FM is the most common benign cause of SVC obstruction. The growing right paratracheal nodes or progressive fibrosis compress the SNC and cause the typical findings of SNC syndrome: venous distension of the neck and upper extremities, head and upper extremity edema, and enlarged collateral veins across the chest and back. Because the compression of the SVC develops at a slower pace than in malignant disease, the symptoms are less severe because it takes longer for collaterals to form. In such patients surgical treatment, such as SVC bypass grafting, is not needed.[4]

Treatment of FM is particularly challenging. Most of the available data are based either on case reports or small case series. There have been no prospective, randomized controlled trials conducted so far.[1] Yet, the limited data suggest that ketoconazole therapy and antituberculous agents administered in cases with evidence of active granulomas might stabilize the disease progression or, in some cases, lead to limited symptomatic improvement.[8] Most studies, including our report, have shown little or no beneficial effect of corticosteroid therapy.[7] Only few reports demonstrated significant reduction in the mediastinal lesion after corticosteroid therapy.[9] Clark et al. have been the first to show tamoxifen to be effective, which was later supported by several reports.[10]

In conclusion, FM could be an unexpected sequela of successfully treated tuberculosis. This is a benign, slowly progressing, inflammatory process causing sclerosis around any of the mediastinal structures. It very often has an unpredictable course; periods of both spontaneous remission and exacerbation of symptoms have been reported.[1] It can mimic a malignant process, which must be excluded. Systemic antifungal agents, antituberculous treatment, or corticosteroids have been recommended for treatment, if a specific cause is suspected, and in rare cases they may stabilize the process or relieve symptoms. In our patient steroid therapy was ineffective.

Reports of single cases of rare diseases may help extend our knowledge on the disease and its diagnosis, and perhaps to develop its effective therapy.

REFERENCES

1 Rossi SE, McAdams HP, Rosadso-de-Christenson ML, et al. From the archives of the AFIP: fibrosing mediastinitis. Radiographics. 2001; 21: 737-757.

2 Akman C, Kantarci F, Cetinkaya S. Imaging in mediastinitis: a systematic review based on aetiology. Clin Radiol. 2004; 59: 573-585.

3 Mole TM, Glover J, Sheppard MN. Sclerosing mediastinitis: a report on 18 cases. Thorax. 1995; 50: 280-283.

4 Gawdziński MP. [Mediastinal fibrosis]. Wiad Lek. 2001, LIV, 7-8, 429-436. Polish.

5 Devaraj A, Griffin N, Nicholson AG, et al. Computed tomography findings in fibrosing mediastinitis. Clin Radiol. 2007; 62: 781-786.

6 Kozłowska I, Kosiński K, Różański J, et al. [Anomalous pulmonary artery as a cause of wide, polycyclic pulmonary hilus – case report]. Pol Arch Med Wewn. 2005; 113: 359-363. Polish.

7 Dunn EJ, Ulicny KS Jr, Wright CB, Gottesman L. Surgical implications of sclerosing mediastinitis a report of six cases and review of the literature. Chest. 1990; 97: 338-346.

8 Urschel HC Jr, Razzuk MA, Netto GJ, et al. Sclerosing mediastinitis: improved management with histoplasmosis titer and ketoconazole. Ann Thorac Surg. 1990; 50: 215-221.

9 Ikeda K, Nomori H, Mori T, et al. Successful steroid treatment for fibrosing mediastinitis and sclerosing cervititis. Ann Thorac Surg. 2007; 83: 1199-1201.

10 Clark CP, Vanderpool D, Preskitt JT. The response of retroperitoneal fibrosis to tamoxifen. Surgery. 1991; 109: 502-506.

OPIS PRZYPADKU

Włókniejące zapalenie śródpiersia jako rzadkie powikłanie gruźlicy

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SŁOWA KLUCZOWE

gruźlica, włókniejące zapalenie śródpiersia

STRESZCZENIE

Włókniejące zapalenie śródpiersia jest rzadką chorobą o łagodnym przebiegu spowodowaną nadmierną proliferacją bezkomórkowego kolagenu i tkanki włóknistej w obrębie śródpiersia. W Stanach Zjednoczonych większość przypadków jest związana z nieprawidłową odpowiedzią immunologiczną organizmu na zakażenie *Histoplasma capsulatum*. Część zachorowań jest związana z gruźlicą. U wielu pacjentów przyczyna włókniejącego zapalenia śródpiersia nie jest znana. Pacjenci są przeważnie młodzi i zgłaszają objawy podmiotowe i przedmiotowe związane z uciskiem lub zwężeniem żyły głównej górnej, żył lub tętnic płucnych, dróg oddechowych lub przełyku. Tomografia komputerowa klatki piersiowej i rezonans magnetyczny odgrywają główną rolę w diagnostyce i leczeniu pacjentów z tą chorobą. Przedstawiamy rzadki przypadek włókniejącego zapalenia śródpiersia jako następstwa gruźlicy.

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