Endobronchial Findings of Fibrosing Mediastinitis

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Fibrosing mediastinitis is underdiagnosed because of the nonspecific character of the presenting symptoms. The endobronchial findings obtained via flexible bronchoscopy are not defined in the literature. We describe 3 cases of fibrosing mediastinitis, most likely caused by histoplasmosis. All 3 patients presented with hemoptysis and were found to have tracheobronchial concentric narrowing, severe hyperemia, and mucosal edema. The hyperemic blood vessels were treated with neodymium yttrium-aluminum-garnet (Nd:YAG) laser and argon plasma coagulation. We believe that recognition of specific endobronchial findings aids in prompt diagnosis of fibrosing mediastinitis. Key words: fibrosing mediastinitis, histoplasmosis, bronchoscopy, laser, embolization. [Respir Care 2003;48(11):1038–1042. © 2003 Daedalus Enterprises]

Introduction

Fibrosing mediastinitis is defined as “presence of excessive fibrotic tissue in the mediastinum that has the tendency to invade and obliterate normal structures.” Fibrosing mediastinitis is due to an autoimmune process or, more commonly, an idiosyncratic reaction to granulomatous mediastinal infection.1,2

The disease was first reported by Oulmont in 1856, but the role of Mycobacterium tuberculosis (TB) and Histoplasma capsulatum in its pathogenesis was not demonstrated until the late 1950s.3

Fibrosing mediastinitis has a benign course and produces nonspecific symptoms until it constricts major mediastinal structures, such as pulmonary arteries, veins, trachea, main bronchi, the superior vena cava, and the esophagus. Most patients present with symptoms such as cough, dyspnea, hemoptysis, pleuritic pain, recurrent pulmonary infections, wheezing, dysphagia, or superior vena cava syndrome. The interval between initial infection and late sequelae is several years and the mean interval between onset of symptoms and death is approximately 6 years.2

The physical examination, routine laboratory tests, and chest radiographs are usually noncontributory and, in association with the nonspecific character of the presenting symptoms, frequently lead to erroneous diagnoses such as asthma, pneumonia, and pulmonary embolism.

Thus the diagnosis is usually delayed and requires a high degree of clinical suspicion, especially in the areas endemic for TB and histoplasmosis. Diagnosis is based on computed tomography (CT) findings of excessive mediastinal soft tissue and calcification.1,2 Mediastinoscopy or thoracotomy may be necessary to exclude malignancy and to confirm the diagnosis.3

Our review of the literature found that the role of flexible bronchoscopy in fibrosing mediastinitis, especially with regard to the endobronchial findings, has not been defined.

In this report we describe 3 patients with fibrosing mediastinitis, most likely related to histoplasmosis, who presented with symptoms suggestive of airway involvement. Flexible bronchoscopy was performed and patients were found to have endobronchial abnormalities that could be explained by the pathophysiology of the disease.

Case 1

A 31-year-old woman was admitted for respiratory failure secondary to pulmonary hemorrhage following a dif-
difficult flexible bronchoscopy, which was performed for recurrent hemoptysis in the 6 months prior to admission.

A chest CT showed extensive calcified mediastinal lymphadenopathy, replacement of the normal fat of the right hilum by soft tissue, narrowing of the right pulmonary artery, and calcified granulomas involving the right lower lobe (Fig. 1). The initial echocardiographic and spirometric data were nonspecific. The ventilation-perfusion scan, on the contrary, revealed substantial compromise of the ventilation and perfusion of the right lung.

After extubation the patient was remarkable for mild anemia and flexible bronchoscopy findings consistent with concentric narrowing of the bronchial tree bilaterally from an extrinsic source and a highly hyperemic, oozing mucosa with a diffuse spider-web-like microvasculature (Fig. 2).

Because of the risk of recurrent hemorrhage, we performed arteriography of the thoracic vessels, which revealed 2 large hypertrophied bronchial arteries arising from the right side of the aorta and substantial intrapulmonary shunting into the right pulmonary artery. Selective bronchial artery embolization was successful.

The CT and bronchoscopy findings were both considered secondary to fibrosing mediastinitis, most likely from histoplasmosis. The mediastinoscopy reinforced the diagnosis by revealing dense fibrosis and lymphoid tissue negative for neoplasm or granuloma. TB etiology was further excluded by a negative purified protein derivative (PPD) of tuberculin test and bronchial cultures.

In the 3-year follow-up period the patient has been re-admitted to our hospital 6 times for recurrent hemoptysis. The rest of her evaluation is unchanged. She has had repeated bronchial artery embolizations and neodymium yttrium-aluminum-garnet (Nd:YAG) laser coagulation of the dilated endobronchial vessels, without complications.

Case 2

A 59-year-old male resident of the Ohio River Valley area underwent radio-frequency ablation for chronic atrial fibrillation. Several weeks after the procedure the patient developed hemoptysis and gradually worsening dyspnea. The initial evaluation included a spiral CT that showed thickening and narrowing of the pulmonary veins, especially on the left. The rest of the examination was remarkable only for multiple calcifications of the spleen and a negative PPD test.

The clinical evaluation, pulmonary function tests, and echocardiogram were unremarkable, but the quantitative lung perfusion scan revealed severely decreased flow to the left lung. The patient underwent balloon angioplasty of pulmonary veins and had some improvement.

Three months later the patient was reevaluated for recurrent episodes of hemoptysis and was found to have restenosis of the pulmonary veins. Balloon angioplasty was repeated.
Flexible bronchoscopy showed a severely hyperemic, "bleed on touch" mucosa with diffuse dilated telangiectatic vessels involving the left main bronchus (Fig. 4).

The bronchoscopy and CT findings were thought to be due to fibrosing mediastinitis from histoplasmosis. The evaluation was negative for TB, lymphoma, or neoplasm. He underwent argon plasma coagulation of the hyperemic blood vessels, without complications, and repeated balloon angioplasty of the occluded pulmonary veins.

Case 3

A 51-year-old woman was admitted for massive hemoptysis. Flexible bronchoscopy revealed marked extrinsic compression of the right main bronchus and the bronchus intermedius as well as active bleeding from the bronchus intermedius. The chest CT showed an infiltrative calcified process within the mediastinum, around the right main bronchus, and severe narrowing of the right main bronchus and bronchus intermedius.

Bleeding was not controlled with conservative measures, so a right pneumonectomy was attempted but was impossible because of a large broncholith in the right hilum, encasing the pulmonary artery and both the pulmonary veins. Biopsies were negative for neoplasm. Postoperative course was unremarkable, and the patient was discharged under close follow-up orders.

A few months later the patient was readmitted for another episode of hemoptysis. Flexible bronchoscopy revealed total obstruction of the right middle and lower lobes by an extrinsic process and an oozing mucosa (Fig. 5). The patient underwent embolization of right intercostal arteries, which were supplying collateral flow to the bronchial arteries of the right lung. In the last 3 years the patient has undergone 3 more bronchial artery embolizations for recurrent hemoptysis, without complications.

Discussion

Endoscopic findings of fibrosing mediastinitis have seldom been described in the literature. In our patients they
consisted of tracheobronchial concentric narrowing with severe hyperemia and mucosal edema. They mostly mimicked acute or chronic bronchitis and extrinsic compression due to mediastinal lymphadenopathy of infectious or neoplastic etiology, which can easily be ruled out by a proper history and physical examination. However, granulomatous infection and neoplasm need further evaluation. TB is reported to cause fibrosing mediastinitis, especially in areas with high prevalence of TB. Ruling out this condition is very important if the patient is to be treated with corticosteroids. All our patients had a negative PPD test, no history of exposure, and negative cultures for M. tuberculosis. On the other hand their epidemiologic characteristics strongly supported an etiology of histoplasmosis. They were all natives of the Ohio River Valley area, which is among the highly endemic areas of the United States for H. capsulatum. In that area skin test reaction to histoplasmin is present in over 80% of the population. Serology tests are helpful only with the active forms of the disease so they were not performed with our patients.

Biopsies of the mediastinal masses were performed in 2 of the 3 patients and were useful for excluding neoplasm, lymphoma, or active granulomatous disease. H. capsulatum was not found in our patients, as expected. Histologic stains and culture for fungi are negative in most lung and lymph node specimens from such patients, probably because of the inactivity of infection at the time of biopsy.

Mucosal biopsies were not obtained, because of the high risk of bleeding. Goodwin et al reported a single case in which biopsy showed chronic mucosal inflammation, submucosal invasion, and destruction by fibrous tissue.

Although radiographs, pulmonary function tests, and echocardiogram findings were nonspecific and therefore noncontributory, the chest CT was extremely valuable in the diagnosis of our patients. Fibrosing mediastinitis is strongly suggested by the obliteration of fat planes of the mediastinum and the presence of discrete masses or/and extensive calcified paratracheal, hilar, and subcarinal lymphadenopathy causing circumferential encasement of the mediastinal structures. Those characteristics could even obviate invasive diagnostic procedures in certain situations.

All of our patients presented with hemoptysis. Although cough was the initial symptom, hemoptysis was the main symptom that led to flexible bronchoscopy and eventually to diagnosis. The distal bronchial obstruction by the mediastinal fibrotic masses could cause partial obliteration of bronchial arteries and veins, leading to high intravascular pressures, rich collateral systems, and active bleeding.

Bronchial artery angiographic findings provide evidence about the pathogenesis of the endobronchial findings, by revealing bronchial artery hypertrophy and bronchial-pulmonary shunting. Ventilation-perfusion scanning is usually indicated to exclude the diagnosis of pulmonary embolism and to study any compromise in the blood flow to either lung. Cardiac catheterization is performed in selected cases, to look for pulmonary hypertension.

Management can be challenging, as the hemoptysis is usually severe and recurrent. Unfortunately, there is no evidence that complicated fibrosing mediastinitis due to histoplasmosis responds to any kind of antifungal or corticosteroid treatment. Corticosteroids and tamoxifen have been used to treat the idiopathic form of fibrosing mediastinitis, with various results. Surgical intervention is technically impossible and hazardous. Bronchial artery embolization is useful for managing severe hemoptysis; it achieves immediate control of bleeding in 75–90% of patients. In complicated fibrosing mediastinitis due to histoplasmosis, the recurrence of hemoptysis remains extremely high, because the primary fibrotic process does not respond to any treatment and nonbronchial collateral vessels also contribute to bleeding.

In our cases Nd:YAG laser coagulation of endobronchial vessels improved outcome. The Nd:YAG laser’s superior hemostatic properties and its ability to operate through a flexible bronchoscope were the major advantages of the system, but evidence of a sustained positive effect remains weak. In the future, argon plasma coagulation could be more useful.

Conclusion

Prompt evaluation of endobronchial and CT findings could be of value in diagnosing fibrosing mediastinitis and avoiding erroneous diagnoses and potentially hazardous interventions, especially with patients residing in areas highly endemic for histoplasmosis. We also believe that endobronchial modalities such as Nd:YAG laser and argon plasma coagulation can palliate symptoms of fibrosing mediastinitis.

REFERENCES


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