Chylothorax Associated with Idiopathic Mediastinal and Retroperitoneal Fibrosis

Quilotórax asociado a fibrosis retroperitoneal y mediastínica idiopáticas

To the Editor:

Mediastinal fibrosis is a disorder characterized by an excessive fibrotic reaction in the mediastinum which can result in compromise of airways, great vessels, and other mediastinal structures. It may develop following infection by *Histoplasma capsulatum*, mediastinal irradiation, or infection by *Aspergillus* or *Blastomyces* species. When no trigger exists, the condition is referred to as idiopathic mediastinal fibrosis, which is characterized by a pattern of diffuse involvement of the mediastinum without calcifications. In this latter case, fibrosing seems to be an autoimmune process associated with Riedel thyroiditis, orbital pseudotumor, and retroperitoneal fibrosis (also known as Ormond disease). We describe the case of a patient with chylothorax associated with idiopathic mediastinal and retroperitoneal fibrosis which resolved spontaneously.

Figure. Chylothorax in a patient with mediastinal fibrosis. Thoracoabdominal computed tomography scans (A and B): left pleural effusion (stars), and descending aorta coated by a 3-mm thick layer consistent with periaortic fibrosis (arrows).
The patient, a 68-year-old man and ex-smoker, was referred from general surgery with left pleural effusion. He had undergone surgery 5.5 years previously for acute urinary tract obstruction, paralytic ileus, and ascites, all secondary to retroperitoneal fibrosis. An abdominal computed tomography (CT) scan performed during a scheduled checkup revealed a left pleural effusion. Apart from experiencing nonspecific abdominal discomfort, the patient was asymptomatic. Biochemistry, blood and coagulation workups were all normal. The erythrocyte sedimentation rate was 21 mm. Diagnostic thoracentesis guided by pleural ultrasound resulted in the extraction of a milky liquid consistent with chylothorax: a triglyceride count above 110 mg/dL, associated with a pleural fluid to serum cholesterol ratio of less than 1 (109:251), and a pleural fluid to serum triglyceride ratio of more than 1 (1152:237). The thoracoabdominal CT scan showed left pleural effusion (severity level 2 out of a maximum classification of 4) partially occupying the upper part of the oblique fissure, with partial compressive atelectasia of the left lower lobe (Figure). The descending thoracic aorta at the subcarinal level had a 3-mm thick coating, with dense soft tissue planes and obliterated fat planes, all suggestive of periaortic fibrosis that had spread from the retroperitoneum through the retrocrural space. Of note in the upper abdomen—and associated with the retroperitoneal fibrosis—was a cuff of dense soft tissue planes around the abdominal aorta and the roots of the main branches. A follow-up x-ray 15 days after admission revealed a significant reduction in the pleural effusion. With minimum blunting of the left costophrenic angle evident on the x-ray, the patient was asymptomatic 30 days after discharge and remains stable at the time of writing.

Mediastinal fibrosis is a rare process that can appear as a late complication of infection by H capsulatum, which is the most frequent triggering factor. Histoplasmosis affects the mediastinal lymph nodes and may lead to the formation of a mediastinal granuloma, or, less frequently, the development of mediastinal fibrosis. The microorganism cannot be cultured from biopsied material, but it is thought that the mediastinal fibrosis may originate as a response to a hypersensitivity reaction secondary to the passage of the fungal antigens from the lymph nodes to the mediastinal space. Serologic tests for H capsulatum were negative in our patient, and, given the CT scan indication of an absence of enlarged mediastinal lymph nodes or calcifications and the absence of other known triggering factors, and bearing in mind the patient’s history of retroperitoneal fibrosis demonstrated by surgical biopsy, the preliminary diagnosis was idiopathic mediastinal fibrosis.

The invasiveness of mediastinal fibrosis helps explain the left chylothorax in the patient. Lymph flow was possibly interrupted as a consequence of compression and/or traction exercised on the thoracic duct at the point where it passes through the mediastinum. Chylothorax, however, is a rare complication of mediastinal fibrosis; most of the very few cases described in the literature are secondary to mediastinal fibrosis associated with sarcoidosis and mediastinal radiation. Only 1 case described chylothorax associated with mediastinal fibrosis with no triggering factor: that of a 14-year-old girl who developed sequential bilateral chylothorax as a complication of idiopathic mediastinal fibrosis.3

Another noteworthy aspect of our case is the association between mediastinal and retroperitoneal fibrosis. Both are associated with positive antinuclear antibodies and hyperglobulinemia, both have been described following the administration of metysergide,4 both sometimes respond to corticosteroids, and both have similar histologic features. They are generally considered to be manifestations of the same process,5 sharing the same pathophysiological patterns in which autoimmune mechanisms in genetically predisposed individuals may be implicated. The overall incidence of all forms of mediastinal fibrosis is extremely low, even in areas where histoplasmosis is endemic. This has led to a search for an association between mediastinal fibrosis and the human leukocyte antigen (HLA), with a relative risk of 3.3 determined for this condition in individuals with HLA-A2.6 HLA-A2 typing for our patient was negative.

In our case, the left chylothorax associated with mediastinal fibrosis developed in a patient with a history of idiopathic retroperitoneal fibrosis. The association between both diseases may be due to fibrotic progression from the retroperitoneum to the mediastinum. Alternatively, and as occurs with other autoimmune diseases, there may simply be an association based on similar pathophysiological processes.

References


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