A Rare Cause of Mediastinal Enlargement: Sinus Histiocytosis with Massive Lymphadenopathy

Una causa poco común de agrandamiento mediastínico: histiocitosis sinusal con linfadenopatías masivas

To the Editor:

Sinus histiocytosis with massive lymphadenopathy (SHML), which was described for the first time by Rosai and Dorfman in 1969, is a rare disorder characterized by a non-neoplastic proliferation of histiocytes in the lymph nodes and in the lymphatic vessels of extranodal locations. The etiology of the disease is unknown (although it has been speculated that some viruses, such as human herpes 6 and Epstein Barr, may intervene in its pathogenesis) and more than 90% of patients present massive, painless bilateral cervical lymphadenopathies.[1,2] The disease mainly affects the lymph nodes, but there may be a simultaneous affectation in extranodal locations (eyelids, eye sockets, skin and subcutaneous tissue, gastrointestinal tract, upper airways and central nervous system) in 40% of the cases.[1] Mediastinal affectation is extremely rare, and there are few cases reported in the literature.[3-5]

A 34-year-old woman who presented brownish, itchy lesions was remitted to our department due to a prominent mediastinal enlargement seen on PA chest radiograph (fig. 1A). The patient had presented generalized hyperpigmented brownish skin lesions in the lower limbs over the last 15 years and had been diagnosed with sinus histiocytosis by means of biopsy of these lesions. The patient had no lung symptoms and the thoracic physical exploration was normal. Chest CT showed an invasive infiltrating mass around the mediastinal and vascular structures (fig. 1B). Laboratory analyses showed normal serum biochemistry and complete hemogram, except for a high globular sedimentation rate (69 mmHg) and an increase in the levels of globulin (albumin: 3.4 g/dL and globulin: 5.3 g/dL). Given the suspicion of an invasive thymoma, lymphoma or pulmonary carcinoma, a diagnostic left anterior mediastinotomy was performed. The anatomopathologic examination of the mediastinal biopsies revealed the presence of a chronic mononuclear inflammation (mainly histiocytes and plasma cells) that intensified around the vascular structures. The histiocytes were positive for the S-100 protein. After the confirmation of mediastinal affectation by sinus histiocytosis, the patient was controlled with no treatment. Three years after the diagnosis, the patient remains stable and thoracic CT has shown no progression.

This disease usually appears between the ages of 20 and 40, and the patients generally present bilateral cervical swelling that is massive and painless. Our patient presented no lung symptoms and only had skin lesions. In the previously-published cases of mediastinal affectation, one patient presented dyspnea and thoracic pain, and another had fever, cough, night sweats and weight loss.[4,6] As in our case, two patients with mediastinal affectation were reported to have an absence of symptoms at the time of the initial presentation.[3,5]

The diagnosis of the disease is fundamentally based on histopathological characteristics and specifically on the proliferation of histiocytes with unique characteristics.[5] These cells are large and contain vesicular nuclei and a pale, voluminous cytoplasm, and are often accompanied by phagocytosis of viable lymphocytes.[5] The histiocytes intensely express the S-100 protein in practically all cases, as well as macrophage antigens such as CD68, HAM-56 and Mac-387.[5]

Given it is frequently a benign and self-limited disease, in most patients treatment is not necessary.[1,2] Treatment may be needed in only a minority of patients that present massive lymph node or extranodal growth that affects the function of the proximal organs.[1,2] Otherwise, the ideal treatment for SHML has not yet been defined.[2]

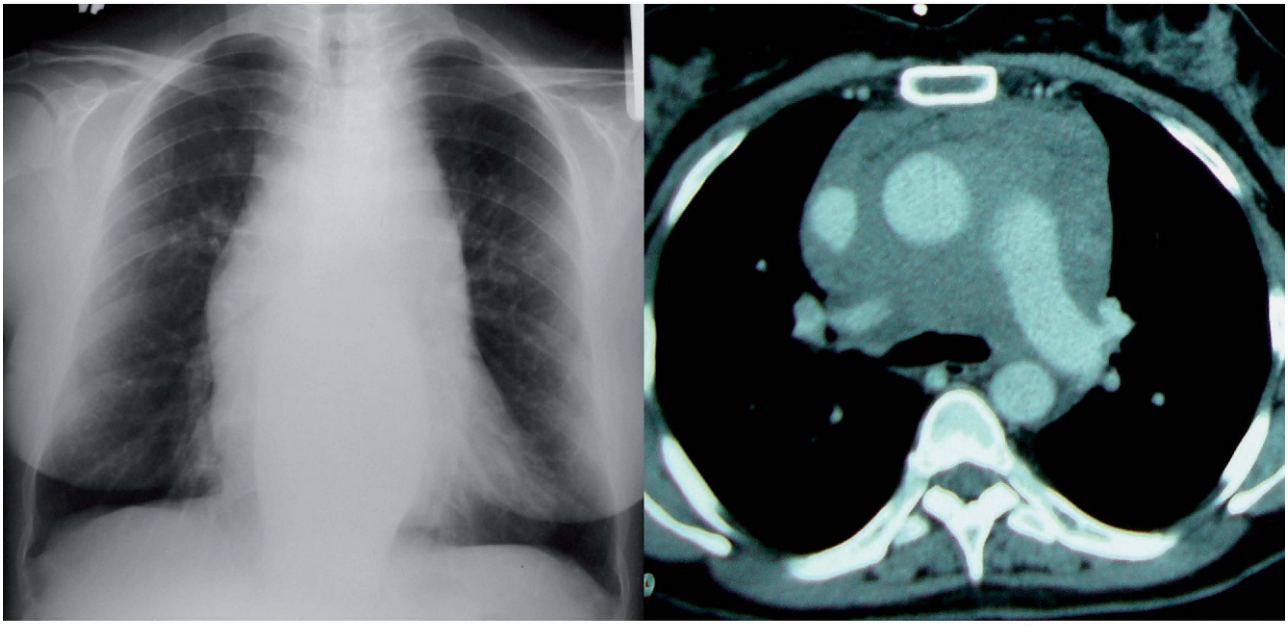

Figure 1. Chest radiograph, where a mediastinal enlargement can be observed (A), and thoracic computed tomography showing an infiltrating mass around the vascular and mediastinal structures (B).

Conflict of Interest

The authors declare having no conflict of interest.

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Tevfik Ozlu,[a] Yilmaz Bulbul,[a,*] Mehmet Muharrem Erol[b]

[a]*Department of Chest Diseases, Karadeniz Technical University, School of Medicine, Trabzon, Turkey*
[b]*Department of Thoracic Surgery, Karadeniz Technical University, School of Medicine, Trabzon, Turkey*

*Corresponding author.
E-mail address: bulbulyilmaz@yahoo.com (Y. Bulbul).

Haemotopsis as a First Sign of Pulmonary Venous Stenosis Secondary to Radiofrequency Ablation of Atrial Fibrillation

Hemoptisis como forma de presentación de estenosis de las venas pulmonares secundaria a ablación por radiofrecuencia de la fibrilación auricular

To the Editor:

We present two clinical cases, both middle-aged men who came to our consultation due to persistent hemoptoic sputum and dyspnea on great exertion. Regarding their personal history, bother were ex-smokers and had been diagnosed with paroxysmal atrial fibrillation. Both had undergone circumferential ablation of the pulmonary veins of the left auricle after the failure of pharmacological treatment.

The patients presented unremarkable physical examinations. In the first of the two cases, the complementary tests, including PA and L chest radiograph as well as on thoracic computed tomography (CT) with contrast, revealed several alveolar infiltrates in different areas of the pulmonary parenchyma, confluent in some areas, and changing over time. As the symptoms were persistent, and given the lack of etiologic diagnosis, we ordered an angiotomography of the pulmonary veins. This revealed complete amputation of the opening of the left upper pulmonary vein, while the right upper venous confluent maintained a normal diameter. The right and left lower venous confluents presented reduced diameters at the height of the ostium (fig. 1). Given the diagnosis of stenosis of the pulmonary veins, the patient was sent to the Hemodynamics unit, where balloon angioplasty was performed at the level of the occlusion of the left upper pulmonary vein.

The second patient debuted with massive hemoptysis and underwent emergency surgical intervention. Left upper lobectomy was performed, with good post-op evolution. One month later, the patient was followed-up in our consultation, at which time we ordered a follow-up angio-CT. This revealed severe stenosis short of the ostium of the lower left pulmonary vein, with faint and delayed enhancement of the previous venous pathway. Given the diagnosis of stenosis of the pulmonary veins, the patient was sent to the Hemodynamics unit, where, after being evaluated, a stent was successfully inserted in the left upper pulmonary vein. To date, the patient enjoys a good clinical evolution.

Discussion

Stenosis of the pulmonary veins is an underdeveloped pathology with regards to its diagnosis and therapeutic actions.[1] The lack of symptoms is very characteristic. In symptomatic patients, what is most prevalent is debut with effort dyspnea, or with persistent catarrhal symptoms, while hemoptysis is quite infrequent.[2] As for the radiological findings, there is no specific pattern in this patology.[3] For all these reasons, it is a pathology that is usually under diagnosed.

The imaging test for diagnosis is pulmonary angiography,[4] but it is a very specific test, and for this reason many cases are never diagnosed.

The definitive treatment consists in achieving the recanalization of the stenosed segment.[5] In order to do so, balloon dilatation can be done, as in the first case. This is a more conservative method, with less surgical risk but with greater risk of relapse. Another therapeutic alternative is the implantation of a *stent* in the area of the occlusion of the vessel, as done in the second case. It is an effective method, with less risk of recurrence but with a higher rate of complications. Nowadays, transcatheter ablation is being used more and more as a therapeutic procedure in patients with chronic symptomatic atrial fibrillation. This pathology is currently being taken into consideration, and from a cardiology viewpoint prevention by means of technique perfection is the most important point. From the pulmonology standpoint, it is primordial to keep this pathology in mind in order to determine early, correct diagnosis.[6]