Behçet’s Disease and Hemoptysis

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Behçet’s disease (BD) is a very uncommon inflammatory and autoimmune condition that was first described in 1937 by the Turkish physician Hulusi Behçet. It is characterized by recurrent oral ulcers; genital, ocular, cutaneous, and vascular changes; and a positive pathergy test result [1-4]. Venous vessels are more often affected than arteries (aorta, femoral, lower extremity, mesenteric, coronary, renal, subclavian, and pulmonary). Importantly, the rupture of the pulmonary artery or aneurysms are factors of a poor outcome and the main cause of death in BD [1]. Pleural and parenchymal involvement, including infarcts, atelectasis, alveolar infiltrates, and ground-glass areas, and a tree-in-bud pattern may be found in less than 10% of the cases; pleural involvement in BD may cause nodules and effusions and reactive lymphadenopathies. Chest computed tomography is the best tool to diagnose and follow the course of these manifestations of BD [1]. Pericarditis, endomyocardial fibrosis, and coronary artery involvement have rarely been reported [2].

In a very interesting editorial recently published in this journal, Çağrıncı and Kılınç [5] briefly commented on key learning points associated with massive hemoptysis. They highlighted the lack of consensus on the definition of this severe condition, with published values ranging from 200 mL to 1000 mL of blood expectoration during a 24-h period. Moreover, they emphasized the life-threatening cardiopulmonary hemodynamic instability due to hemoptysis, which must be considered “massive,” regardless of the blood loss amount. Hemoptysis may be caused by conditions such as arteriovenous malformation, pseudoaneurysm, bronchiectasis, tuberculosis, mycoses, abscess, polyangiitis, and pulmonary malignancy [5]. Additional comments were on the best timing of a surgical procedure in cases of hemoptysis from arterial (after embolization and clinical stabilization) or venous (immediate) vessels [5].

Abarca et al. [1] described the case of two male patients with BD and massive hemoptysis due to aneurysms in the pulmonary segmental and interlobar arteries, which were successfully controlled. The authors commented that in up to 29% of the cases, aneurysms may be an initial manifestation. The patients were 14- and 42-year-old males and presented with pulmonary thrombosis. The younger patient underwent total resection of the right lower lobe and received prednisone 20 mg daily, whereas the older patient had a good clinical outcome after receiving corticosteroids and cyclophosphamide.

The purpose of the comments included is to broaden the clinical awareness of non-specialists about uncommon entities that may also be the etiology of massive hemoptysis. In Brazil, although ethnic groups are not well characterized in relation to the risk of BD, this issue would be of primary interest for primary healthcare workers in the Mediterranean area, considering the high prevalence (71 per 100,000 people per year) in Turkish descendants [3]. Didactic editorials and case reports may enhance the suspicion index about rare conditions.

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