Giant pulmonary arterial saccular aneurysms in Behçet syndrome

Francisca Godinho Oliveira*; Inês Barreto; Joana Barbosa; Elsa Fragoso; Carlos Lopes; Pilar Azevedo; João Rodrigues Inacio; Cristina Barbosa

1Department of Pulmonology, North Lisbon University Hospital Centre, Portugal.
2Department of Radiology, North Lisbon University Hospital Centre, Portugal.

*Corresponding Author: Francisca Godinho Oliveira
Department of Pulmonology, North Lisbon University Hospital Centre, Portugal.
Email: franciscammgo@gmail.com

Clinical image description

A 28-year-old non-smoker male with Behçet syndrome presented with recurrent oral/genital ulcers and erythema nodosum, giant pulmonary arterial saccular aneurysms (Figure 1 A-D) with recurrent hemoptysis and a pulmonary nodule with biopsy suggestive of necrotizing vasculitis.

The patient was initially treated with methylprednisolone and cyclophosphamide with no clinical or imaging response (Figure 1 A-D). Immunosuppression was changed to infliximab and azathioprine with corticosteroid therapy at higher doses. Due to persistent hemoptysis, the patient underwent endovascular arterial embolization with coiling and cyanoacrylate-lipiodol mixture of three large pulmonary aneurysms, resulting in decrease in size of aneurysms and temporary control of the symptoms (Figure 1 E-H). Surgical lung resection was not considered appropriate due to the extension of the disease and the patient was referred for lung transplantation. The patient died shortly afterwards as a result of fulminant hemoptysis in the intensive care unit.

Behçet syndrome is a multisystemic vasculitis affecting arteries and veins from all sizes. Pulmonary arterial aneurysms are the most common pulmonary vascular lesions in Behçet syndrome [1] and are more common among males [2]. Hemoptysis is the most frequent presenting symptom [1]. Pulmonary arterial aneurysms suggest a poor prognosis being a life-threatening complication of Behçet syndrome. Medical, endovascular and surgical treatments have been used, usually with poor outcomes [2]. The choice of appropriate treatment should take into account the severity of the hemoptysis and also the number and localization of pulmonary arterial aneurysms. Recently, anti-TNF alpha treatment has shown promising results in severe refractory Behçet’s disease [3].

Figure 1: (A-D) Computed tomography scan of the chest demonstrating pulmonary aneurysms without endovascular treatment; (E-H) Pulmonary aneurysms after endovascular embolization.

References

