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Clinical Image

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Giant pulmonary arterial saccular aneurysms in Behçet syndrome

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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 promissive results in severe refractory Behçet´s disease [3].

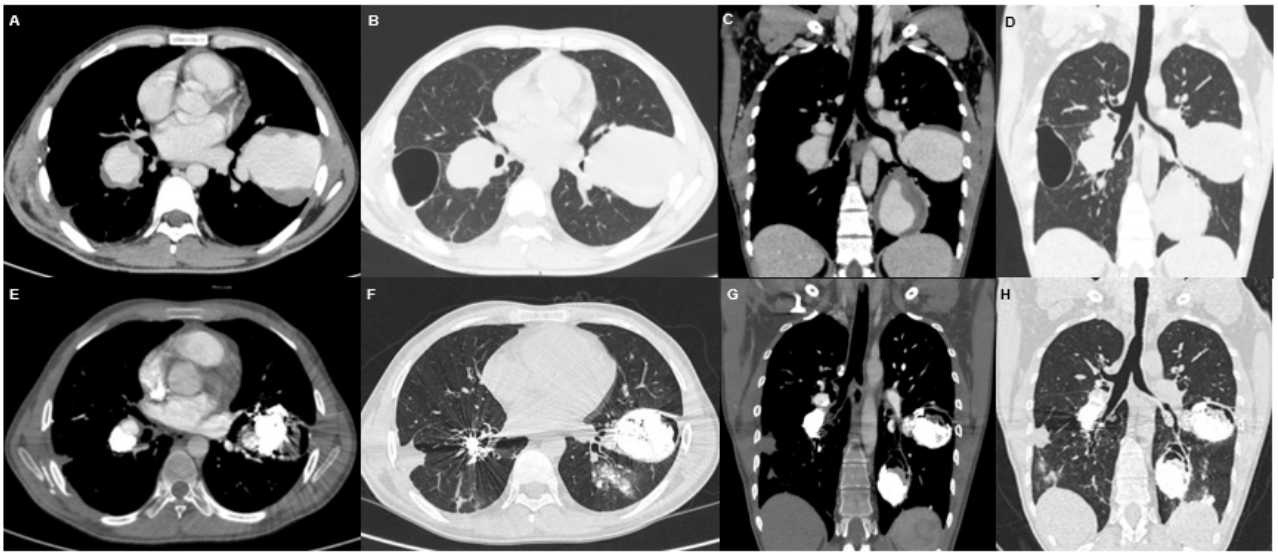

Figure 1: (A-D) Computed tomography scan of the chest demonstrating pulmonary aneurysms without endovascular treatment; **(E-H)** Pulmonary aneurysms after endovascular embolization.

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