

# Reversible pulmonary hypertension with operation of large intramediastinal pseudoaneurysm and anti-inflammatory treatment in patients with Behcet disease

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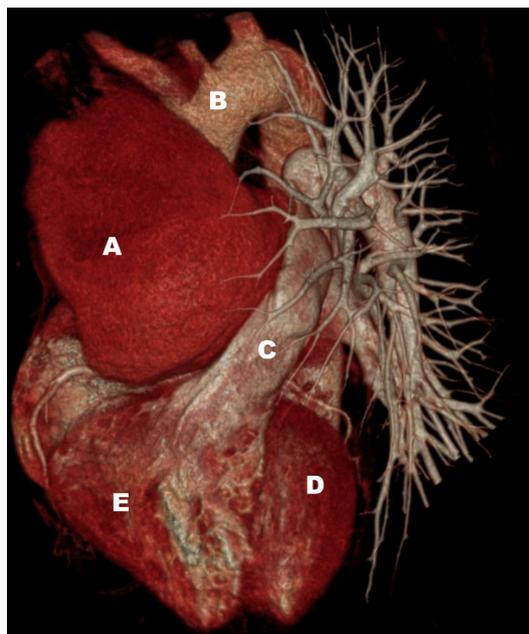
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## DESCRIPTION

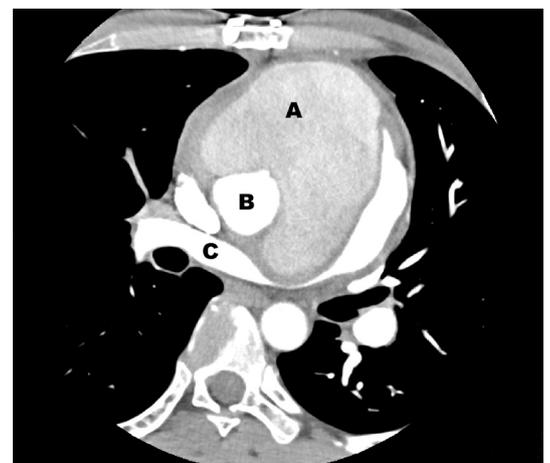
We present the case of a 35-year-old Asiatic man, with no medical relevant history, who debuted in October 2018 with nocturnal cough and orthopnoea. He was diagnosed by transthoracic echocardiogram of severe acute aortic regurgitation due to aortic root dilatation, with normal left and right ventricular functions and no signs of pulmonary hypertension. The patient decides to move to his country, where he undergoes cardiovascular surgery. Aortic root and aortic valve replacement were performed with a mechanical valved tube according to the Bono-Bentall technique. The anamnesis revealed frequent oral and genital aphthosis and folliculitis on his back. Based on those symptoms and a remarkable pathergy phenomenon, Behcet's disease (BD) was diagnosed. After surgery, he started treatment with glucocorticoids and monthly cyclophosphamide intravenous pulses (a total of 6, cumulative dose 9.8 gr.). After finishing induction treatment, maintenance treatment with azathioprine was prescribed.<sup>1,2</sup>

The patient remains asymptomatic for 10 months until he begins again with a progressive dyspnoea and dry cough, finally readmitting in our hospital in August 2020. The echocardiography documented a large intramediastinal pseudoaneurysm, severe pulmonary hypertension (PH) with systolic pulmonary artery pressure (sPAP) of 65 mm Hg, preserved left ventricle function, severe tricuspid regurgitation, and an impaired and dilated right ventricle: basal diameter of 49 mm, tricuspid annular plane systolic excursion (TAPSE) of 14 mm. CT scan documented partial dehiscence of the distal anastomosis of the dacron graft to the ascending aorta, and confirmed the presence of a giant aortic pseudoaneurysm (10×7.3×8.4 cm) compressing the pulmonary trunk and both pulmonary arteries (figures 1 and 2).

Given the dimensions of the pseudoaneurysm, it is agreed that the patient would be a better candidate for surgical intervention rather than endovascular procedure. Optimisation of medical treatment was carried out replacing azathioprine with infliximab 5 mg/kg (300 mg per dose),<sup>2</sup> receiving two doses 15 days apart, achieving normal C reactive protein values prior to the cardiac surgery. Surgery was carried out under extracorporeal circulation and in moderate hypothermia (25°C). The voluminous aortic pseudoaneurysm was resected and



**Figure 1** CT scan, 3D reconstruction. (A) Aortic pseudoaneurysm. (B) Aortic arch. (C) Pulmonary trunk. (D) Left ventricle. (E) Right ventricle.



**Figure 2** CT scan: (A) aortic pseudoaneurysm; (B) ascending aorta; (C) pulmonary trunk.



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## Images in...

replaced with a new 28 mm Dacron vascular prosthesis. The immediate postoperative period was uneventful.

At 2 months, the echocardiography showed preserved left ventricle function, dilated and normocontractile right ventricle (TAPSE 19 mm, basal diameter 50 mm), mild tricuspid regurgitation and normalisation of pulmonary pressures with sPAP of 27 mmHg.

BD is currently classified as ‘variable vessel vasculitis’ (Chapel Hill, 2012). It is a chronic, multisystemic, inflammatory rheumatic disease. Its main manifestations are mucocutaneous, neurological, cardiovascular, pulmonary and musculoskeletal.<sup>3</sup> Vascular involvement of BD, the so-called ‘angio-Behcet’, appears in 7%–29% of patients.<sup>4</sup> Characteristically, both vascular

territories can be affected. Venous pathology and thrombotic complications are the most common.<sup>1</sup> Arterial complications are less frequent and usually occur in a 1%–14%.<sup>3</sup> While arterial stenoses, pseudoaneurysms and ruptured large vessels are less frequent, they represent a major cause of morbidity and mortality. In most studies, patients usually receive immunosuppressive treatment before and after the intervention, including anti-TNF agents such as infliximab, thus reducing the recurrence rate.<sup>2</sup>

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### Learning points

- ▶ Behcet’s disease (BD) is a chronic, multisystemic, inflammatory rheumatic disease. Among its manifestations the cardiovascular ones stand out.
- ▶ Aortic pseudoaneurysms are a common complication of vascular surgery in BD. The recurrence rate of aneurysms is high, being estimated at around 50%, developing during the first year in 24% and in 92% in the same location.
- ▶ Perioperative control of disease activity is essential to reduce this risk.

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