An unusual case of chronic and reversable pulmonary hypertension

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Abstract

Behcet's Disease (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. Vascular involvement of BD, so-called "angio-Behcet", appears in 7%-29% of patients. Arterial complications, such as arterial stenoses, pseudoaneurysms, and ruptures occur in a 1-14% range and represent a major cause of morbidity and mortality in BD patients. We, herein, report the case of a large aortic pseudoaneurysm producing severe pulmonary hypertension in a patient with angio-Behcet who had previously undergone aortic root replacement.

INTRODUCTION

We report the case of a large aortic pseudoaneurysm producing severe pulmonary hypertension (PH) in a patient with angio-Behcet.

PATIENT AND METHODS

We present the case of a 35-year-old Asian man who in 2018 had an echocardiographic diagnosis of severe aortic regurgitation due to aortic root dilatation, with normal left and right ventricular functions and no signs of PH. In the same year the patient underwent aortic root and aortic valve replacement with a mechanical composite valve graft at his local hospital. During the following months he suffered frequent oral and genital aphthosis and folliculitis on his back. Based on these symptoms and a remarkable pathergy phenomenon, Behcet's disease (BD) was diagnosed. Thus, treatment with glucocorticoids and monthly cyclophosphamide intravenous pulses (up to 6) was started. After finishing induction treatment, maintenance treatment with azathioprine was prescribed[1-2].

In 2020 the patient was admitted to our hospital because of a 3-month history of progressive dyspnea and dry cough. The echocardiography documented a large intra-mediastinal pseudoaneurysm, severe PH with systolic pulmonary artery pressure (sPAP) of 65 mmHg, 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 14mm. 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 (10x7.3x8.4 cm) compressing the pulmonary trunk and both pulmonary arteries (Figures 1 and 2).

Optimization of medical treatment was carried out replacing azathioprine with infliximab 5mg/kg (300mg per dose)[2-3]. Surgery was carried out under extracorporeal circulation and in moderate hypothermia (25°C). The voluminous aortic pseudoaneurysm was resected and repaired with a new 28mm dacron graft. The immediate postoperative period was uneventful.

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

COMMENTS

Vascular involvement of BD, so-called "angio-Behcet", appears in 7%-29% of patients[3]. Arterial complications occur in a 1-14% range[4]. For this reason, patients with angio-Behcet who undergo cardiovascular surgery require a close therapeutic and echocardiographic follow-up.

CONCLUSION

We presented a case of a patient with angio-Behecet who developed a large aortic pseudoaneurysm producing severe and reversible PH.

LEGEND

Fig.1 CT scan, 3D reconstruction. A: aortic pseudoaneurysm; B aortic arch; C: pulmonary trunk; D: left ventricle; E: right ventricle.

Fig.2 CT scan: A: aortic pseudoaneurysm; B ascending aorta; C: pulmonary trunk

Author contributions

Paola del Coromoto León Suárez: Conceptualization. Writing the original draft. Iñigo Rúa-Figueroa Fernández de Larrino: Conceptualization. Writing the original draft. Stefano Urso: Writing the original draft. José Domingo Marín Esmenota: Supervision. Eliú Nogales: Visualization. Francisco Portela: Supervision

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