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AORTITIS IN BEHÇET'S DISEASE: AN UNUSUAL CLINICAL MANIFESTATION

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BACKGROUND

Behçet's syndrome (BS) is an inflammatory, multisystemic disease, of unknown etiology, with significant genetic susceptibility, having HLA B*51 as the main predictor of genetic risk. Classified as a vasculitis of variable vessels, its classic manifestations being recurrent oral and genital ulcers, uveitis and skin lesions. However, it has less common manifestations, but with important lethality, such as the involvement of large vessels and the central nervous system. In relation to the involvement of large vessels, arterial involvement has a prevalence between 4–17%, occurring in a greater proportion in young men. The main presentations are aneurysms (45–70%), occlusions (36–80%) and arterial strictures (13%), and the main locations are aortic (abdominal, thoracic, femoral, pulmonary and iliac).

CASE REPORT

A 34-year-old female patient hospitalized for etiological investigation of aortic insufficiency associated with aneurysm of the ascending aorta root demonstrated on transthoracic echocardiography and thickening of the descending aorta with thickening of the intimal layer, without atheroma plaques demonstrated on transesophageal echocardiography, after routine evaluation to authorize physical exercises. After a targeted evaluation for autoimmune diseases, it was found that the patient had a personal history of recurrent vaginal lesions and episcleritis, as well as a first-degree personal history of BS with cardiac involvement (aortic insufficiency and aortic aneurysm with multiple surgical corrections), and positive HLA B*51 research. As a follow-up to the investigation, a pathergy test, with a negative result, and cardiac angioresonance were requested, which demonstrated significant aortic insufficiency, with moderate global left ventricular dysfunction, thoracic aorta with thickening of the vessel walls, with signs compatible with diffuse

Figure 1. Cardiac angioresonance: 1. Left ventricle with significant increase in volumes and moderate global dysfunction at the expense of diffuse hypokinesia. 2. Major aortic insufficiency (57% regurgitant fraction). 4. Thoracic aorta with thickening of the vessel walls, with signs compatible with diffuse inflammation/edema. 5. Abdominal aorta of normal caliber, with no signs of involvement of the walls, with aortic branches without stenosis or aneurysms.

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inflammation/edema (Fig. 1). After diagnosis of BS, pulse therapy with methylprednisolone 1 g/day, for 3 days, was chosen, and maintenance with only 1 mg/kg/day of corticosteroid therapy, until a surgical procedure was performed to correct aortic insufficiency, which occurred after 4 months, without complications. Therefore, maintenance treatment with azathioprine 150 mg/day and tapering corticosteroid dosage were started. After 2 years from the start of treatment, the patient remains asymptomatic, corticosteroid free, being followed on an outpatient basis by both rheumatology and cardiology.

CONCLUSION

The BS has typical manifestations such as oral and genital ulcers. However, the involvement of large vessels is part of this syndrome, and should always be remembered in affected patients.

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