



Aortic Complications Of Behcet's Disease: A Case Report

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Abstract :

Behçet's disease is a rare systemic vasculitis that can involve arteries and veins of all calibres, often presenting in young adults with a male predominance. Cardiovascular complications are infrequent, but when they do occur, they are severe and include arterial aneurysms, heart failure and valvular disease. The present case report concerns a 30-year-old female patient with Behçet's disease who was diagnosed at the age of 18 and presented with progressive exertional dyspnea (NYHA class III). A clinical examination revealed a diastolic murmur, and imaging studies confirmed an ascending aortic aneurysm measuring 7.6 cm in diameter, accompanied by massive aortic insufficiency.

The patient underwent an urgent Bentall procedure involving replacement of the ascending aorta and aortic valve, with coronary artery reimplantation under cardiopulmonary bypass. Histopathological analysis of the surgical specimen revealed extensive inflammatory infiltration of the vascular wall, with neutrophils, lymphocytes, plasma cells, and multinucleated giant cells, consistent with Behçet's disease. Postoperative recovery was favourable, with no major complications, and the patient was maintained on immunosuppressive therapy to prevent recurrence of inflammation. This case underscores the potential severity of vascular complications in Behçet's disease, particularly when involving the aorta. Early detection and intervention are critical to preventing catastrophic outcomes. Surgical management, such as the Bentall procedure, remains the cornerstone for treating symptomatic or critically sized aneurysms. Long-term management necessitates meticulous clinical and radiological follow-up, in addition to the sustained administration of immunosuppressive therapy, with the objective of controlling systemic inflammation and mitigating the risk of recurrence. The optimal management of such complex cases is contingent on multidisciplinary collaboration.

Introduction

Behçet's disease is a rare systemic vasculitis characterized by inflammation affecting arteries and veins of all calibers. This disease predominantly affects young adults, with a male predominance. Cardiovascular complications, although rare, are severe and include arterial aneurysms, heart failure, and valvular disease. We present the case of a 30-year-old female patient with Behçet's disease, presenting with an ascending aortic aneurysm associated with massive aortic insufficiency.

Case Report

A 30-year-old female patient had been followed for Behçet's disease diagnosed at the age of 18. Episodes of inflammatory flares had been regularly treated with colchicine, resulting in good clinical response. She presented with progressively worsening exertional dyspnea, classified as New York Heart Association (NYHA) functional class III.

On clinical examination, the patient was eupneic at rest, with normochromic conjunctivae. Her blood pressure was measured at 135/60 mmHg, and peripheral pulses were well palpable. Cardiac auscultation revealed a diastolic murmur of aortic regurgitation, graded 3/6, at the aortic area.

Transthoracic echocardiography (TTE) revealed a dilated left ventricle with preserved ejection fraction (56%), massive aortic insufficiency, and aneurysmal dilation of the proximal ascending aorta. These findings were confirmed by thoracic CT angiography, which showed critical dilation of the ascending aorta, reaching a maximum diameter of 7.6 cm.

Given the critical size of the aneurysm and the massive aortic insufficiency, the patient underwent ascending aortic replacement with a Bentall procedure under cardiopulmonary bypass. Histological analysis of the surgical specimen revealed inflammatory infiltration involving the media, adventitia, and perivascular vasa vasorum. The infiltrate consisted of neutrophils, lymphocytes, and plasma cells, with occasional eosinophils and multinucleated giant cells, confirming the inflammatory etiology linked to Behçet's disease.

Postoperative evolution was favorable without major complications. Immunosuppressive therapy was continued to prevent inflammatory relapses. Medium-term clinical and radiological follow-up has been satisfactory.

Discussion :

Behçet's disease is a systemic vasculitis that can affect vessels of all calibers, although venous involvement is more common. Arterial involvement is less common but often severe, manifesting mainly as aneurysms or arterial occlusions. Aneurysms, present in around 5-10% of Behçet patients, mainly affect large arterial trunks such as the aorta. This arterial involvement is often asymptomatic until it reaches an advanced stage, underscoring the importance of regular clinical follow-up.

Chronic inflammation of the vessels is the cornerstone of aneurysm formation in Behçet's disease. Destruction of the media and damage to the vasa vasorum by an inflammatory infiltrate rich in neutrophils, lymphocytes and plasma cells progressively weaken the vascular wall. This fragility favours aneurysm formation and increases the risk of rupture, which constitutes a vital emergency.

In the case reported, massive aortic insufficiency associated with ascending aortic aneurysm represents a rare but serious complication of Behçet's disease. Aortic insufficiency is generally associated with chronic inflammation and dilatation of the aortic annulus, leading to valvular regurgitation. This inflammatory process also contributes to the progression of the aneurysm, creating a vicious circle.

Management of these complications relies primarily on ascending aortic replacement surgery, such as the Bentall procedure. This technique simultaneously replaces the dilated ascending aorta and the aortic valve, while reimplanting the coronary arteries. However, tissue fragility due to inflammation increases the risk of post-operative complications, such as prosthetic disinsertion and false aneurysm formation. To minimize these risks, a period of clinical and biological remission is recommended before any major surgery.

Post-operative follow-up is just as crucial. Regular clinical and radiological monitoring enables early detection of complications such as inflammatory recurrence or residual aneurysmal dilatation. Prolonged immunosuppressive therapy is often required to prevent inflammatory recurrence. The most commonly used agents include corticosteroids and immunosuppressants such as azathioprine or cyclophosphamide, although biotherapies such as TNF-alpha inhibitors are increasingly used in refractory forms.

Finally, the prognosis of Behçet patients with aortic involvement depends on a number of factors, including the earliness of diagnosis, the extent of vascular involvement, the efficacy of medical treatment and the success of surgical management. Multidisciplinary management involving rheumatologists, cardiovascular surgeons and interventional radiologists is essential to optimize outcomes. Prevention of long-term complications relies on rigorous follow-up and strict control of systemic inflammation.

Conclusion

This case illustrates the severity of vascular complications in Behçet's disease, particularly when they affect the ascending aorta. Careful monitoring of patients with this disease is essential for early diagnosis and management of complications. Surgical treatment remains the only curative option for symptomatic or critically sized aortic aneurysms. However, preoperative management of inflammation and postoperative follow-up are essential to improve the long-term prognosis of these patients.

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