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What the Cardiologist Should Know About Cardiac Involvement in Behçet Disease

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ABSTRACT

Behçet disease (BD) is a chronic multisystem inflammatory vasculitis affecting mainly young adults and is characterized by a remitting-relapsing course. In North America, the prevalence is 5.2 per 100,000 population. It is believed that cardiac involvement is one of the most severe complications in patients with BD despite its sporadic occurrence, being greatly correlated with mortality.

RÉSUMÉ

La maladie de Behçet (MB) est une vasculite multisystémique chronique touchant principalement les jeunes adultes; elle évolue par poussées entrecoupées de rémissions. En Amérique du Nord, la prévalence est de 5,2 par 100 000 habitants. L'atteinte cardiaque est l'une des complications les plus graves chez les patients souffrant de la MB en dépit de sa survenue sporadique; elle est fortement corrélée à la mortalité.

We present the case of a 64-year-old woman of Greek descent who presented with acute heart failure and pulmonary edema. The echocardiogram showed severe aortic and mitral regurgitation. The patient underwent surgical intervention with double mechanical valve replacement. Because of spontaneous dissection of the ascending aorta during the operation, it was decided to proceed with a Bentall procedure. The pathology report for both valves and aorta were consistent with chronic inflammation with a lymphoplasmocytic infiltrate. In the follow-up period, the patient experienced multiple arterial aneurysms and underwent a vascular operation for a femoral artery aneurysm.

Cardiovascular involvement secondary to Behçet disease (BD) is rare but is frequently associated with a poor prognosis. Valvular disease can be severe, as illustrated by the present case. Its preoperative recognition is paramount because the appropriate use of immunosuppressive agents and modification of surgical techniques regarding possible aortic involvement may greatly improve overall survival. The diagnosis of BD with cardiac involvement should be suspected by the cardiologist in the presence of significant valvular regurgitation and the full spectrum of the disease or in the presence of significant valvular regurgitation with the pathology report showing acute/chronic lymphoplasmocytic infiltrates in the valves, a finding strongly suggestive of BD.

Case Description

A 64-year-old woman presented with acute-onset heart failure and pulmonary edema. An echocardiogram was obtained, which showed severe mitral insufficiency. The patient was then transferred to our institution for mitral valve replacement. However, a second echocardiogram was obtained preoperatively, and this time it displayed severe mitral and aortic regurgitation with a preserved ejection fraction (Video 1, view video online). The valvular involvement was unusual, with thickened mitral leaflets and aortic cusps in the absence of calcifications (Video 2, view video online). Prolapse of aortic cusps with aneurysmal changes were also described. It was decided to proceed with a double mechanical mitral and aortic valve replacement. Because of spontaneous dissection of the ascending aorta during the operation, it was decided to proceed with a Bentall procedure in addition to aortic and mitral valve replacement. The postoperative course was uncomplicated. The pathology report for the mitral and aortic valves was consistent with chronic inflammation with a lymphoplasmocytic infiltrate,

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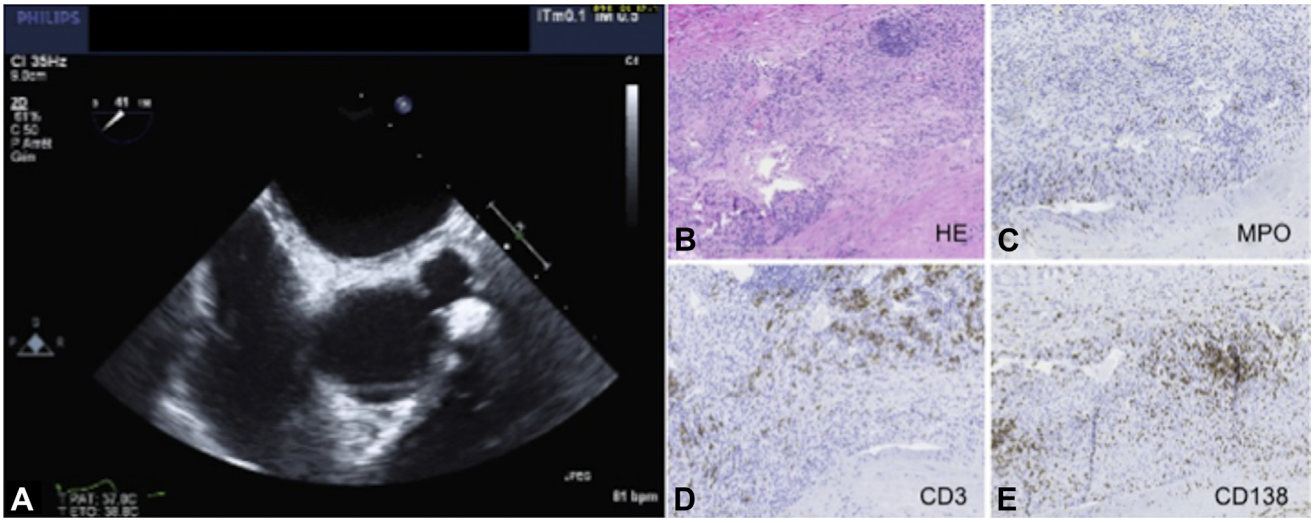

Figure 1. (**A**) Transesophageal echocardiographic image of a large (3 × 2 cm) aneurysm of the proximal left main coronary artery (esophageal position, 2-chamber plane, 30° rotation). (**B-E**) Histologic appearance of left femoral artery showing mixed acute and chronic vasculitis. (**B**) Inflammatory cell infiltrates observed on routine hematoxylin-eosin (HE) stain are confirmed by immunohistochemistry to be acute, with (**C**) myeloperoxidase (MPO) in neutrophils. They are shown to be chronic with (**D**) CD3 in T lymphocytes and (**E**) CD138 in plasma cells.

and lymphoplasmocytic aortitis was also documented. A control echocardiogram was obtained 2 months later and was normal. Thereafter, the patient was hospitalized for fever and *Streptococcus salivarius* bacteremia. Antibiotics were given for possible endocarditis, but transesophageal echocardiography was normal. The patient was treated with a prolonged course of intravenous antibiotics. However, transthoracic echocardiography revealed an important aneurysm (3 × 2 cm) of the left main coronary artery (Fig. 1A). Three months later, she was hospitalized for left groin pain. A pseudoaneurysm of the left femoral artery measuring 5 × 4 cm was demonstrated by Doppler ultrasonography. A contrast computed tomographic scan confirmed the pseudoaneurysm but also demonstrated the presence of multiple arterial aneurysms (celiac tree, gastric artery, left renal artery, and splenic artery). The patient underwent a pseudoaneurysm cure with an ileofemoral bypass. The diagnosis of vasculitis was confirmed by the pathologic characteristics of the femoral artery, showing a lymphoplasmocytic vasculitis similar to that in the pathology report for the mitral and aortic valves and the aorta (Fig. 1, B-E). This histologic pattern was strongly suggestive of BD. A further questionnaire revealed that the patient had had oral aphthous ulcers in the past. A pathergy skin test was also done and produced positive results. The clinical presentation and pathologic examination were highly suggestive of BD with secondary valvular disease, involvement of the ascending aorta, and multiple arterial aneurysms. Prednisone treatment was started and the patient did well thereafter without any adverse events.

Discussion

BD is a chronic multisystem inflammatory vasculitis characterized by a remitting-relapsing course and affecting mainly young adults.[1] Ethnicity is important in the incidence of the disease, with Asian and Mediterranean individuals being affected more often. In North America, the overall incidence is 0.38 per 100,000 population, and the prevalence is 5.2 per 100,000 population. Carrying the human leukocyte antigen B51 increases the risk of BD developing by 1.5 to 16 times, but it is not correlated with the severity of the disease. The etiologic factors remain to be clarified.[2] International study group criteria for the disease were revised in 1990 and include recurrent oral aphthous ulceration and 2 of the following: genital ulceration, skin manifestations, eye lesions, and a positive skin test result (pathergy test).[2] However, there is no specific diagnostic test for BD. It is believed that cardiac involvement is one of the most severe complications in patients with BD, despite its sporadic occurrence, being greatly correlated with mortality.[3] The prevalence of cardiac involvement is variable between studies (1%-29%), and men seem to be predisposed (male-to-female ratio 14:1).[2] The cardiac involvement is varied and includes pericarditis, myocarditis, endocarditis with valvular regurgitation, intracardiac thrombus, endomyocardial fibrosis, coronary arteritis with or without myocardial infarction, and aneurysms of the coronary arteries or sinus of Valsalva.[3] Vascular involvement, arterial and venous, is also more frequent in patients with heart disease.[3] Echocardiographic criteria for valvular involvement have been proposed and include cusp aneurysmal change, echocardiographic free space, and vegetation-like lesions.[2] Some authors have reported that redundant motion of elongated aortic cusps or prolapse of aortic cusps with aneurysmal changes may be highly suggestive of valvular involvement in BD. However, these findings can also be found in other diseases such as infectious endocarditis, Libman-Sacks endocarditis, and marantic endocarditis. In the case presented here, the patient mainly had vegetation-like lesions on the mitral valve and aneurysmal changes of the aortic cusp (Table 1). Our case is unusual because only a few reports of bivalvular involvement secondary to BD can be found in the literature. The aortic valve is usually involved more often than the mitral valve and is accepted as a predictor of mortality. Approximately 25% of patients with BD and cardiac