CASE REPORT
DOI 10.4070/kcj.2011.41.4.217

Print ISSN 1738-5520 / On-line ISSN 1738-5555

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An Atypical Mitral Valve Prolapse in a Patient With Behçet's Disease

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ABSTRACT

We report the case of a 42-year-old male who was admitted to the hospital with progressive dyspnea. Cardiomegaly and diffuse pulmonary edema were visible on chest X-ray and multiple oral and genital ulcers on physical examination. On admission, echocardiography revealed mitral valve prolapse (MVP) predominantly involving a basal portion of the posterior leaflet, with severe mitral regurgitation. A successful mitral valve replacement with St. Jude #29 was performed, after pre-treatment with prednisolone for 2 weeks. Fifteen months following the operation, the patient expired from severe pulmonary edema and secondary pneumonia. This case demonstrates, for the first time in the literature, an unusual feature of mitral prolapse in the basal portion with severe mitral regurgitation in a patient with Behçet's disease. As suggested by this case, we should consider an atypical type of MVP as a possible inflammatory involvement of the heart in patients with Behçet's disease. **(Korean Circ J 2011; 41:217-219)**

KEY WORDS: Behçet's syndrome; Heart valve prolapse; Mitral valve; Inflammation.

Introduction

Mitral valve prolapse (MVP) is a relatively common and highly variable clinical syndrome of the mitral valve apparatus, resulting from diverse pathogenic mechanisms. It is commonly associated with myxomatous degeneration and is a frequent finding in patients with heritable disorders of connective tissue, including Marfan syndrome, osteogenesis imperfecta, and Ehler-Danlos syndrome. Most of the time, prolapse of the mitral valve occurs in the tip or middle portion of the valve, resulting from the redundancy or rupture of chordae with degenerative changes of the chordae and valve. In this case, however, the atypical prolapse of the basal portion of the mitral valve with a relatively preserved valve tip portion, arose from the ruptured basal chordate, and was due to inflammation. We hereby present an unusual feature of mitral prolapse in the basal portion with severe mitral regurgitation in a patient with Behçet's disease.

Received: July 2, 2010
Revision Received: August 9, 2010
Accepted: August 17, 2010
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• The authors have no financial conflicts of interest.

Case

A 42-year-old male was admitted to hospital with progressive dyspnea. He had no other specific past history except smoking. Cardiomegaly and diffuse pulmonary edema was evident on chest X-ray and multiple recurrent oral and genital ulcers on physical examination. Initial laboratory examination revealed mild leukocytosis (white blood cell 10,300/mm^2) and elevated N-terminal pro-B-type natriuretic peptide (522 pg/mL), erythrocyte sedimentation rate (ESR) (111 mm/hr), and C-reactive protein (CRP) (8.99 mg/dL). The ANA and c-ANCA/p-ANCA were negative.

On admission, transthoracic echocardiography (TTE) and transesophageal echocardiography revealed MVP, predominantly involving a basal portion of the posterior mitral leaflet (mainly P2 region), with severe mitral regurgitation and aneurysmal changes of the aortic valve without aortic regurgitation (Fig. 1A, B and C). Because of suspicion of an inflammatory involvement of Behçet's disease, considering the history of recurrent oral and genital ulceration, uveits on ophthalmolo-

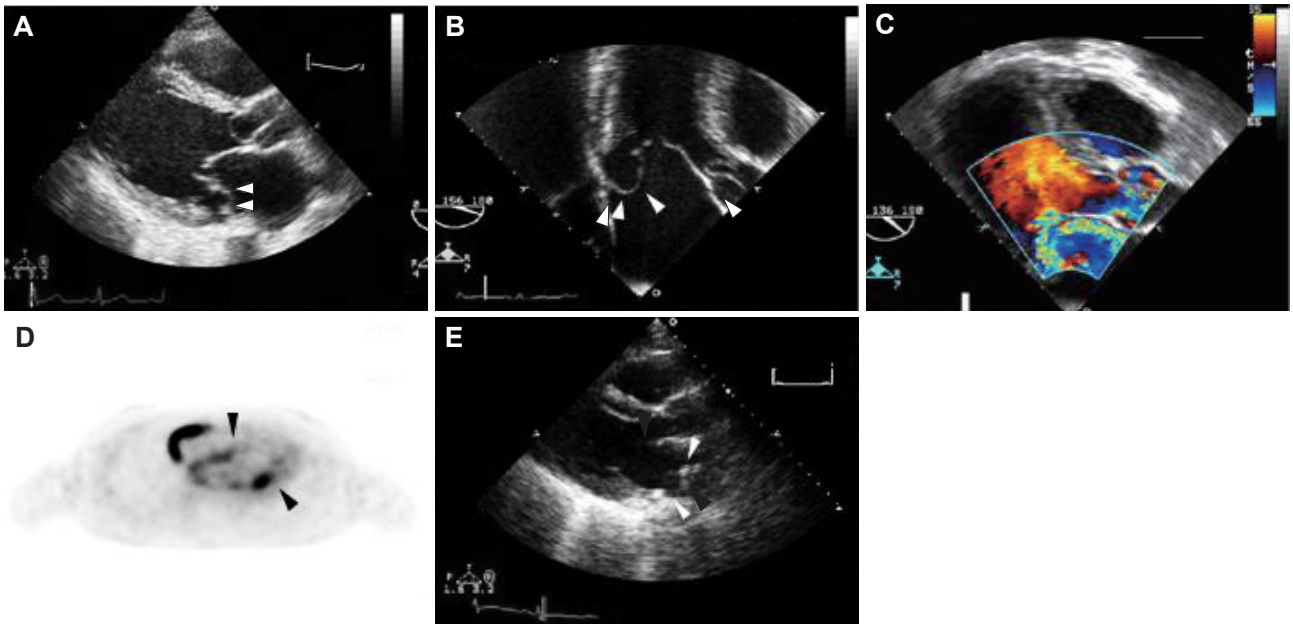

Fig. 1. A: parasternal long view showing mitral valve prolapse, predominantly involving a basal portion of the posterior mitral leaflet (white arrowheads) and aneurysmal changes of the aortic valve in TTE. B: TEE (156 degree) showing mitral valve prolapse in basal portion of posterior mitral leaflet (white arrowheads). C: TEE (136 degree) showing severe mitral regurgitation. D: PET (18-FDG) showing increased uptake along the right atrial wall and mitral annulus (black arrowheads). E: parasternal long axis view showing almost totally detached prosthetic mitral valve and displacement into the left atrium (white arrowheads).

gic examination and elevated ESR and CRP, positron emission tomography (18F-fluorodeoxyglucose) was performed, revealing strongly increased uptake along the mitral annulus (Fig. 1D).

A mitral valve replacement with St. Jude #29 was performed after pretreatment with prednisolone (60 mg/day) for 2 weeks. Pathologic finding revealed inflammatory cell infiltration composed of lymphocytes on mitral valve. The laboratory examination was performed every three months and ESR and CRP after 3 months were 43 mm/hr and 1.17 mg/dL (initial: ESR 111 mm/hr, CRP 8.99 mg/dL), respectively. Following the successful operation, the patient continued to take prednisolone (10 mg/day) and azathioprine (150 mg/day) with anticoagulation.

Fifteen months following the operation the patient was admitted to the hospital again with fever and severe dyspnea. TTE revealed almost total detachment of the prosthetic mitral valve from the annulus and displacement into the left atrium (Fig. 1E). The levels of ESR and CRP were greatly elevated (ESR 120 mm/hr, CRP 39.6 mg/dL), suggesting the possibility of reactivation of mitral annular inflammation. He suffered with severe dyspnea, cough, sputum and fever from severe pulmonary edema and secondary pneumonia and expired seven days later.

Discussion

MVP is a relatively common and highly variable clinical syndrome resulting from diverse pathogenic mechanisms of the mitral valve apparatus. It is commonly associated with myxomatous degeneration and is a frequent finding in patients with heritable disorders of connective tissue, including Marfan syndrome, osteogenesis imperfecta, and Ehler-Danlos syndrome.[1)]

Behçet's disease is a chronic relapsing, inflammatory process manifesting as recurrent oral and genital aphthous ulcerations. Higher incidences of interatrial septal aneurysm (31% vs. 6%), MVP (25% vs. 3%), mitral regurgitation (40% vs. 6%), and aneurysmal dilatation of the sinus valsalva and ascending aorta are observed in the Behçet's disease patients compared with normal subjects.[2)3)] Despite this high incidence of MVP in patients with Behçet's disease, the mechanism of MVP is not clear. The association of MVP and Behçet's disease was first reported in China.[4)] Shen et al.[5)] from Shanghai, postulated that MVP may be the result of structural damage and functional derangement caused by the underlying vasculitis of Behçet's disease. The prognosis of cardiac involvement of Behçet's disease is poor, including relapse of valvular disease after operation.[6)]

The length and tension of the chordae between the papillary muscle and the mitral valve is larger in the tip portion of the mitral valve than in the middle or basal portions. Consequently, most prolapse of the mitral valve occurs in the tip or middle portion of the valve; this comes from the redundancy or rupture of chordae with degenerative changes of the chordae and valve. However, in this case, the atypical prolapse of the basal portion of the mitral valve, with a relatively preserved valve tip portion, arose from the ruptured basal chordae due to inflammation; additionally, aneurysmal changes of the

basal portion of mitral valve were induced.

As suggested by this case, we should consider the possibility of inflammatory involvement of the mitral apparatus in patients with the unusual feature of mitral prolapse in the basal portion of mitral valve. This case demonstrates the atypical feature of mitral prolapse in the basal portion with severe mitral regurgitation, for the first time in a patient with Behçet's disease.

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