中文題目：一個心臟內的兩個故事：貝西氏症相關的嚴重主動脈瓣逆流和感染性心內膜炎相關的嚴重二尖瓣逆流

英文題目：A story of two tales: Behcet's disease related severe aortic regurgitation and infective endocarditis-associated severe mitral regurgitation

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Introduction: Behcet's disease (BD) is a chronic inflammatory disorder with pathophysiology of vasculitis that may affect multiple systems. Cardiac involvement has a diversity of manifestation, among which, valvular disease is the most prevalent one. However, different mechanism other than autoimmune-related inflammation is still a possible cause for valvulopathy. Here, we presented a rare case of BD related severe aortic regurgitation (AR) and a simultaneous infective endocarditis-associated severe mitral regurgitation (MR).

Case presentation: A 68-year-old female with BD presented to the clinic with progressive exertional dyspnea for six months. Physical examination revealed a grade III/VI pansystolic murmur at apex and a grade III/VI diastolic murmur at right upper sternal border. Her N-terminal pro-brain natriuretic peptide level was 22065 pg/mL. The transthoracic echocardiogram (TTE) showed left ventricular end-diastolic dimension (LVEDD) 62 mm with an ejection fraction (EF) of 51%, severe MR with suspicious mitral valve (MV) aneurysmal formation, and significant AR. Transesophageal echocardiogram (TEE), both 2D and 3D, confirmed severe MR caused by MV-P2 perforation and severe AR due to aortic valve (AV) retraction with central malcoaptation. Cardiac catheterization showed wide pulse pressure (131 mmHg) with a low diastolic blood pressure (DBP) (36 mmHg) and elevated LV end-diastolic pressure (LVEDP) (29 mmHg) - a compounding effect from both severe MR and AR. She underwent surgical double valve replacement. Intra-operative findings revealed AV edge thickening and retraction, particularly non-coronary cusp, causing malcoaptation, and thickened MV with P2 perforation and pseudoaneurysm formation. Histopathology showed mixed acute and chronic inflammatory cell infiltration for MV, and focal myxoid degeneration for AV. Post-operatively, her blood pressure normalized (118/74 mmHg) and TTE showed reduced LVEDD (47mm) with an improved LVEF (60%). Her symptoms resolved significantly.

Discussion: BD is a systemic inflammatory disease. Cardiac involvement had been reported in 7 – 46% of patients. Of these cardiac lesions, valvular disease is the most prevalent, especially for AR (65%), followed by MR (5%). Although BD may cause non-bacterial thrombotic endocarditis (NBTE), the histopathology of the vegetation usually shows deposition of fibrin and platelets and is devoid of inflammation. Valve destruction was also rare in NBTE. This unusual case showcased chronic BD-related post-inflammatory changes on both AV and MV with MV superimposed by bacterial endocarditic changes. The patient's symptoms were caused by two different mechanisms of severe regurgitation presented simultaneously.

Conclusion: BD is a chronic inflammatory disorder capable of multi-organ involvement. This unusual case highlighted the importance of regular cardiac surveillance in patients with autoimmune disease for early detection of and timely intervention for cardiac involvement before progression to irreversible LV intrinsic dysfunction. This case also demonstrated the essentiality of 2D and 3D TEE for valvular mechanism-elucidation.