

Pulmonary valve involvement and left ventricular thrombosis in Behçet's disease: a case report and literature review

Sirs,

Behçet's disease (BD) is an autoimmune systemic vasculitis. When the heart is involved, it is called cardiac BD, and it can affect all parts of the heart (1). Cardiac involvement is rare but fatal.

A 22-year-old Chinese man was admitted to our hospital for a recurrent rash and oral ulcer lasting 2 years, recurring fever and arthralgia for 1 year, and progressive chest pain for 4 months. The rash was mainly located on the lower limbs, oral cavity and forehead (Fig. 1). The patient had initially been diagnosed with hip synovitis, but antibiotic treatment was unsuccessful. He presented with chest pain initiating in the substernal area and radiating to the left shoulder. The electrocardiogram (ECG) showed ischaemic changes (Supplementary Fig. S1). Laboratory investigations showed an elevated erythrocyte sedimentation rate and C-reactive protein. Skin biopsies showed chronic inflammation. Transthoracic echocardiography revealed a low-echogenic mass attached to the pulmonary valve, which was confirmed via computed tomography (CT) scanning (Supplementary Fig. S2), but pulmonary CT angiography (CTA) showed no pulmonary embolism. The jet velocity of the pulmonary valve was

1.8 m/s, and the right ventricular size and thickness were normal. No bacteria were found, and antibiotic treatment was unsuccessful. Based on these results, BD was the most likely diagnosis. According to the International Criteria for BD, oral aphthosis, skin lesions and positive pathergy tests, the patient scored a minimum of four points and was diagnosed with BD. Because the patient presented with chest pain and ECG changes, coronary CTA was performed. The coronary artery contained no stenosis; the left ventricular endocardial thickness was increased, and the apex showed several irregular filling defects (Supplementary Fig. S3). Methylprednisolone, low-dose cyclophosphamide, and low-molecular-weight heparin completely relieved the patient's symptoms; the left ventricular lesions were attenuated, and the ECG gradually improved until normal (Supplementary Fig. S5, S6).

Mitral valve prolapse and aortic valve prolapse are the most common manifestations of valve involvement in BD, especially in East Asia (2). This is the first report of pulmonary valve involvement. Intracardiac thrombus (ICT) is a rare complication of BD. Most ICT cases occur on the right side of the heart and often involve the ventricle (3, 4). This may be partly attributed to the extended thrombi in the vena cava and lower pressure on the right side of the heart. ICT is uncommon on the left side of the heart. Most patients reside in the Mediterranean basin regions or the Middle East; however, some cases have been reported in China (5, 6).

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Competing interests: none declared.

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Fig. 1. Rash on the lower limbs, oral cavity and forehead.