

### Duration of anti-TNF treatment in vascular Behçet's disease: better to prolong treatment?

Sirs,

Behçet's disease (BD) is a chronic, autoimmune, multisystem disorder of unknown origin, with vasculitis as its main underlying pathological process. In cases of vascular and neurological involvement, BD is associated with poor prognosis. Intracardiac thrombus (ICT) is a rare but severe complication of BD. In a large study consisting of 626 BD patients, ICT was reported in 12 (1.9%) subjects (1). Anti-TNF agents, infliximab (IFX) and adalimumab, are widely used in the treatment of severe and/or refractory BD due to their high and rapid efficacy but there is ongoing debate on duration of treatment (2). Herein, we report an interesting case of BD with ICT who responded well to IFX but immediately relapsed after cessation of therapy.

A 37-year-old male patient admitted to our clinic with chest pain, fever and dyspnea. He had had diagnosis of BD in 2010 with oral aphthous ulcers, uveitis and erythema nodosum and prescribed to colchicine. In 2013 he presented with fever, palpitation and chest pain. His pertinent investigation revealed thrombi in bilateral pulmonary arteries and left single pulmonary artery aneurysm. Human leucocyte antigen (HLA)-B51 was positive and detailed thrombophilia screening was unrevealing. Patient was treated with intravenous pulse methyl prednisolone 1 gr for three consecutive days and cyclophosphamide 1 g/month along with daily 1 mg/kg prednisone. Patient had lower gastrointestinal bleeding on admission therefore anticoagulants were never used. His symptoms were remarkably improved with immune-suppressive treatment, and he turned to his normal life after 2 months. Glucocorticoid dose was gradually tapered. Ten months after treatment with monthly 1 g cyclophosphamide he complained of fever and dyspnea again. His erythrocyte sedimentation rate (ESR) was 40 (N:0-20) mm/h and C-reactive protein (CRP) was 55 (N: 0-5) mg/L. Thoracic spiral computed tomography showed thrombus in inferior branches of left pulmonary artery and right ventricle. Transthoracic echocardiography revealed 10x9 mm mass-like thrombus in the apex of the right ventricle and 9x8 mm thrombus adherent to interatrial septum in the right atrium. His treatment was switched to IFX 5 mg/kg per infusion, plus 1 mg/kg prednisone and dramatic response was observed. In periodic echocardiographic screening cardiac thrombi were completely disappeared after one-year treatment with IFX. This was con-

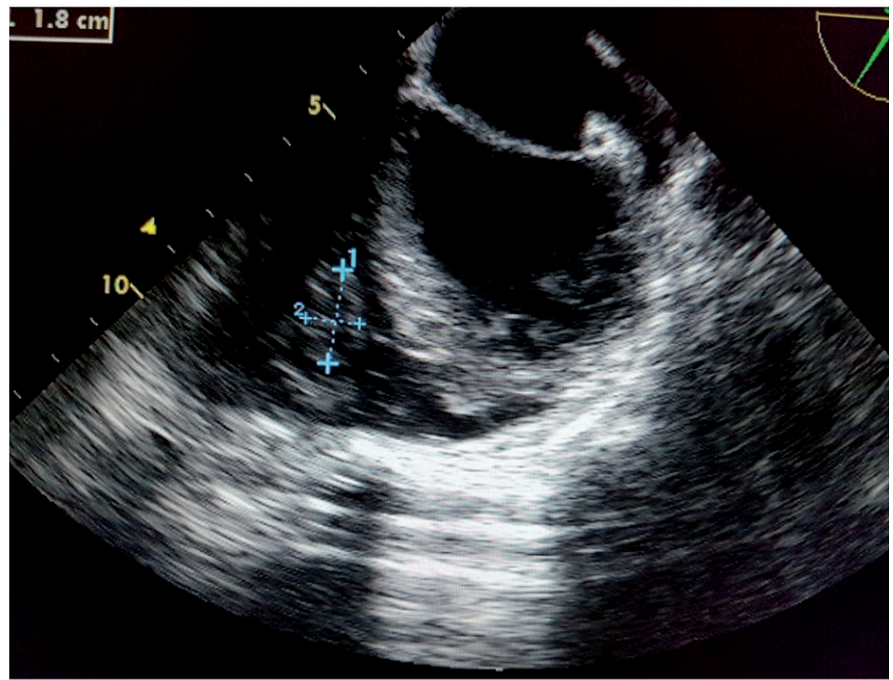


Fig. 1. Transesophageal echocardiography with thrombus in the right ventricle.

firmed with annual echocardiographic surveillance. Since he was in complete remission for three years, IFX was ceased and 3 mg/kg azathioprine was prescribed. Three months after the withdrawal of IFX, patient developed fever, dyspnea, weakness and anorexia. On physical examination, there were many oral ulcers, fever (38.6°C) and erythema nodosum lesions. Blood analysis, echocardiography and thoracoabdominal computed tomography was performed. ESR and CRP levels were elevated to 75 mm/h and 129 mg/L, respectively. The transesophageal echocardiography showed a 18x10 mm thrombus in the right ventricle, adherent to interventricular septum (figure) and IFX was re-instituted with a dramatic improvement in clinical symptoms and acute phase response.

There is no consensus on duration of anti-TNF treatment after achieving clinical remission in BD. In a recent retrospective small study, after median 2 years of treatment with anti-TNFs, 41% of patients remained in complete remission for at least 3 years after withdrawal of anti-TNF treatment (3). Our case experienced a relapse within 3 months after IFX discontinuation which suggests anti-TNFs does not restore key pathways in BD. Although relapses may occur in any time in BD, which warrants close follow up (4), it is prudent to prolong anti-TNF treatment, particularly in those BD patients with life-threatening manifestations such as vascular and neurological involvements.

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