Cardiology in the Young (2014), **24**, 382–383
doi:10.1017/S1047951113000498

Brief Report

A giant left ventricular pseudoaneurysm in Behçet's disease: a case report

Najat Mouine, Rajae Bennani, Rachida Amri

Department of Cardiology B, Mohammed V University Souissi Rabat Morocco, Rabat, Morocco

Abstract Behçet's disease is a chronic autoimmune disease with vascular complications that are most frequently manifested as thromboembolism in veins and pseudoaneurysm in arteries. We report the case of a 13-year-old boy admitted for clinical and biological signs of rheumatic fever associated with chest pain. The clinical examination found heart sounds with a discrete systolic murmur of mitral regurgitation. The electrocardiogram showed a microvoltage with diffuse repolarisation disorder. Biologically, he had inflammatory syndrome. Transthoracic echocardiography showed circumferential pericardial effusion with anterosepto-apical hypokinesia of the left ventricle with systolic dysfunction, and a minimal mitral regurgitation. The patient was treated by corticotherapy and antibiotherapy. The outcome was marked by orogenital aphthous ulceration and decreased visual acuity related to intermediate uveitis. The retinal angiography showed a vasculitis. The late appearance of this symptom led to the right diagnosis of Behçet's syndrome. Transthoracic echocardiography showed a hypokinetic dilated cardiomyopathy left ventricular with septo apical and anterior akinesia and severe systolic dysfunction, with a defect of the inferior septal with a collar communicating the left ventricle with a giant pseudo aneurysm. Magnetic resonance imaging showed a giant pseudoaneurysm communicating with the left ventricle. The coronary computed tomography was normal. The patient had undergone surgical treatment for the pseudoaneurysm with good outcomes.

Keywords: Pseudoaneurysm; left ventricle; Behçet's disease

Received: 3 December 2012; Accepted: 31 March 2013; First published online: 10 May 2013

Case report

A 13-year-old boy, who had a history of angina since 10 years of age, was admitted for clinical and biological signs of rheumatic fever associated with chest pain. The clinical examination found heart sound with a discrete systolic murmur of mitral regurgitation. The electrocardiogram showed a microvoltage with diffuse repolarisation disorder. The chest radiography showed cardiomegaly with a cardiothoracic ratio of 0.57.

Biologically, he had inflammatory syndrome with a very high sedimentation rate, c protein-reactive level, and antistreptolysin O titre.

Transthoracic echocardiography showed circumferential pericardial effusion (15 mm) with anterosepto-apical hypokinesia of the left ventricle with systolic dysfunction, and a minimal mitral regurgitation.

The patient was treated by corticotherapy and antibiotherapy for 12 weeks with good clinical and biological outcomes.

After 1 year, the outcome was marked by orogenital aphthous ulceration and decreased visual acuity related to intermediate uveitis. The retinal angiography showed vasculitis and the pathergy test was positive. The late appearance of this symptom led to the right diagnosis of Behçet's syndrome.

Transthoracic echocardiography showed a hypokinetic dilated cardiomyopathy left ventricular with septo apical and anterior akinesia and severe systolic dysfunction (EF = 38%), with a defect of the inferior septal with a collar measuring 11 mm communicating the left ventricle with a pseudo aneurysm measuring (75 × 80 mm) a seat of spontaneous contrast and small thrombi (Figs 1 and 2).

Correspondence to: N. Mouine, Department of Cardiology, Mohammed V Military Hospital, Rabat, 10012, Morocco. Tel: +00212661202733; Fax: 00212537710721; E-mail: mouine2@yahoo.fr

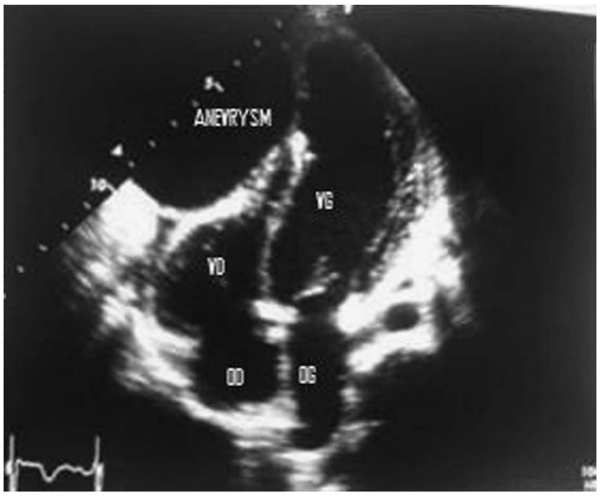

Figure 1.
Transthoracic echocardiography showing a giant left ventricular pseudoaneurysm in the apical four-chamber plane.

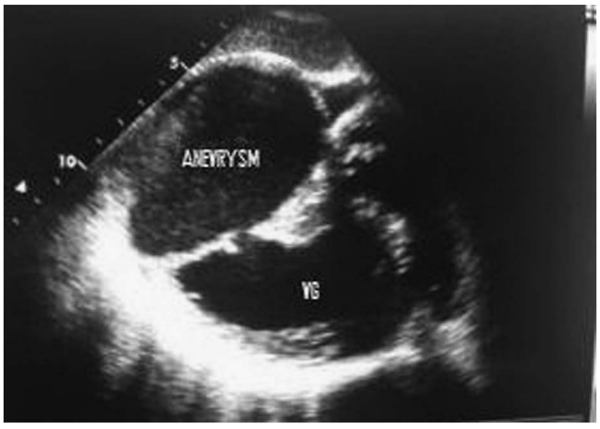

Figure 2.
Transthoracic echocardiography showing a giant pseudoaneurysm communicating with the left ventricle.

Magnetic resonance imaging showed a giant pseudoaneurysm communicating with the left ventricle (Fig 3). The coronary computed tomography was normal.

The patient had undergone surgical treatment for the pseudoaneurysm with good outcomes.

In 1937, Hulusi Behçet, a Turkish dermatologist, first described a chronic autoimmune disease characterised by orogenital aphthous ulceration and uveitis.[1] The primary pathology is a vasculitis affecting the skin and joints, as well as pulmonary, gastrointestinal, urinary, and nervous systems.[2] Its vascular complications are most frequently manifested as thromboembolism in veins and pseudoaneurysm in arteries.[3]

Although pseudoaneurysms are the most common form of arterial involvement in Behçet's disease, we could only find one case reported by

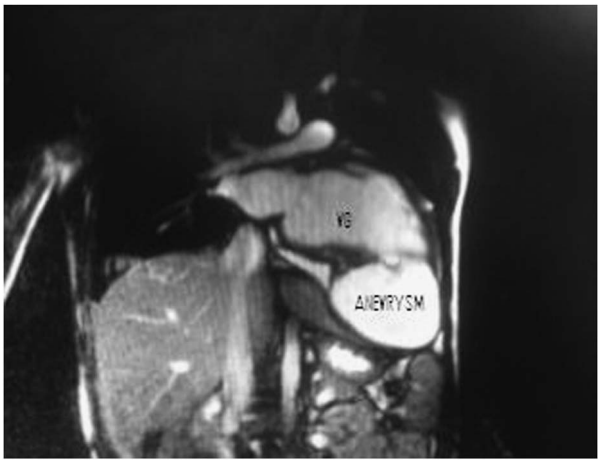

Figure 3.
Magnetic resonance imaging showing a giant pseudoaneurysm of the left ventricle.

Rolland et al.[4] with Behçet's disease and cardiac pseudoaneurysm. Occasional cases of cardiac pseudoaneurysms have been reported in association with rheumatoid arthritis,[5] like in our case; he was treated for rheumatoid arthritis and then he presented with symptoms of Behçet's disease complicated by a giant left ventricular pseudoaneurysm. There are no data on the long-term prognosis of patients with cardiac pseudoaneurysms in Behçet's disease; however, long-term survival could not be expected given the diffuse involvement of cardiac structure and vascular elements.[6] Considering its fatality and non-specific manifestations, one should consider cardiac pseudoaneurysms as a potential risk in any patient with Behçet's disease. Thanks to early diagnosis and surgery, our patient was treated successfully.

References

1. Cakir O, Eren N, Ulka R, Nazaroghi H. Bilateral subclavian arterial aneurysm and ruptured abdominal aorta pseudo aneurysm in Behçet's disease. Ann Vasc Surg 2002; 16: 516–520.
2. Alkim H, Gurkaynak G, Sezgin O, Oguz D, Saritas U, Sahin B. Chronic pancreatitis and aortic pseudoaneurysm in Behçet's disease. Am J Gastroenterol 2001; 96: 591–593.
3. Silistreli E, Karabay O, Erdel C, et al. Behçet's disease: treatment of popliteal pseudoaneurysm by an endovascular stent graft implantation. Ann Vasc Surg 2004; 18: 118–120.
4. Rolland JM, Bical O, Laradi A, et al. False aneurysm of the left ventricle and coronary aneurysms in Behçet's disease. Arch Mal Coeur Vaiss 1993; 86: 1383–1385.
5. Rittoo D, Sutherland GR. Posterior left ventricular pseudoaneurysm after aortic valve replacement in a patient with rheumatoid arthritis: diagnosis by transesophageal echocardiography. J Am Soc Echocardiogr 1994; 7: 429–433.
6. Gurgun C, Ercan E, Ceyhan C, et al. Cardiovascular involvement in Behçet's disease. Jpn Heart J 2002; 43: 389–398.