Left ventricular pseudoaneurysm revealing Behçet's disease: a case report

Sirs,

Behçet's disease (BD) is a multisystemic vasculitis characterised by recurrent oral and/or genital aphthous ulcers accompanied by cutaneous, ocular, articular, gastrointestinal, and/or central nervous system involvement (1, 2). Heart involvement has been reported in 1 to 6% of BD patients and include pericarditis, myocarditis, endocarditis, intracardiac thrombosis, endomyocardial fibrosis, coronary arteritis and aneurysms of the coronary arteries or sinus of Vasalva (3). Left ventricular pseudoaneurysm (LVPA) is exceptional. Here, we report a case of a 30-year-old woman from Mayotte who was admitted in university hospital for the management of a LVPA. The history started fifteen days before with a symptomatic unprovoked deep vein thrombosis (DVT) of the right superficial femoral vein. A treatment by tinzaparin had been started. The day after the DVT diagnosis she complained of chest pain and a pulmonary computerised tomography (CT) excluded a pulmonary embolism. Seven days later, because of persisting and increasing chest pain with the occurrence of a fever at 38.3°C, a second CT chest scan was performed showing a 73 mm diameter pseudoaneurysm of the left ventricular apex. A second interpretation of the first CT scan showed that the pseudoaneurysm was already present, 10 mm smaller. After deep history taking she reported asthenia for 8 months and a weight loss of 7 kg in 3 months, recurrent oral ulcers with three flares in 8 months, genital ulcers and an erythema nodosum 6 months before (Fig. 1). Physical examination showed genital scarring. Biologically she had an inflammatory syndrome persisting for 8 months. The C-reactive protein concentration at admission was 122 mg/L (normal, <5 mg/L). The transthoracic echocardiography confirmed the diagnosis, with an akinesia of the apex. The coronary angiography was normal. The cardiac magnetic resonance imaging revealed a necrosis of one myocardial wall's segment of the apex with a pseudoaneurysm of 82 x 48 mm and an orifice of 7.5 mm, with gadolinium enhancement of a left pleural effusion (Fig. 2). Three days after the hospital admission, she underwent surgical left ventricular reconstruction. She had a smooth postoperative course. BD was diagnosed after the surgery according to the International Study Group for Behçet's Disease Criteria. Abdominal CT scan, fluorine-18-fluorodeoxy-glucose positron emission tomography, brain magnetic resonance imaging and ophthalmologic examination were unremarkable. Medical therapy, started after surgery, consisted of prednisone 1 mg/kg per day (60 mg), with a goal of 0.5 mg/kg per day at 3 months and 10 mg per day at 6 months, azathioprine at 2 mg/kg per day (125 mg), warfarine (target international normalised ratio 2.0–3.0) and colchicine at 1 mg per day. The fever and inflammatory syndrome decreased dramatically and she was discharged 28 days after her admission. Prednisone was discontinued after 16 months, and warfarin after 17 months and replaced with aspirin. At her 23-month follow-up, she was still in remission. Seven other cases of LVPA related to BD are reported in literature (Table I) (4-10). As for five of them, BD was diagnosed in our patient after heart complication onset (6-10). Between the cases already described, the arterial involvement is frequent, with five patients presenting coronary implication (mainly aneurysm), including one patient with pulmonary artery aneurysm and one with femoral artery pseudoaneurysm following coronary revascularisation (4, 6-9). Cardiac pseudoaneurysms are mainly caused by the postinfarction rupture of the myocardium. Even if coronary occlusion was not found in our patient and histological examination was not performed, small-vessel vasculitis could not be ruled out. Surgery is mandatory because a high risk of rupture and embolic events from the noncontractile cavity. Steroids and immunotherapy are necessary to avoid suture leakage or relapse, but a full description of the medical treatment was reported only in one case with an association of prednisone, azathioprine colchicine and anticoagulants (8). We also chose this treatment and prednisone was tapered as proposed for BD with arterial involvement (11). Even if in literature the initial outcome is often good, long-term follow-up is rarely reported, with a maximum of 2 years (5, 8).

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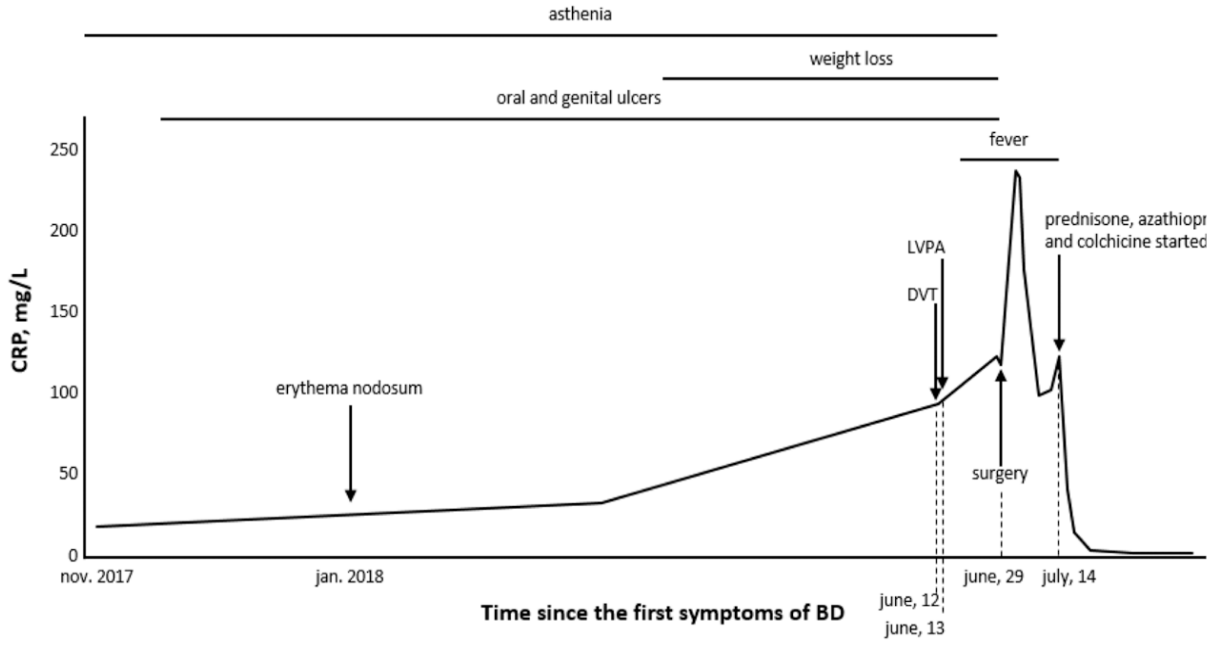

Fig. 1. Timeline of symptoms and treatment.
DVT: deep vein thrombosis; LVPA: left ventricular pseudoaneurysm; CRP: C-reactive protein; BD: Behçet's disease.

Table I. Characteristics of Behçet's disease patients with left ventricular pseudoaneurysm.

Reference	Age	Sex	Origin	Clinical signs at presentation	BD diagnosis criteria	Delayed BD diagnosis	Cardiac involvement	Other vascular involvement	Laboratory exams	Histologic findings
Rolland	29	M	NA	NA	NA	No	LVPA CAA	NA	NA	NA
Marashi	13	M	Iran	Cough Chest pain Fever	Oral ulcers Orchitis Uveitis Pseudofolliculitis Arthritis	No	LVPA	DVT	NA	Fibrous pseudoaneurysm, old haemorrhage, thrombosis, chronic inflammation
Delgado	23	M	North Africa	Chest pain Fever	Oral and genital ulcers	No	LVPA CAO	PVT	↗CRP ↗ESR	Fibrous tissue, acute and chronic inflammation with neutrophils, thrombus
Harrison	40	M	Spain	Chest pain Fever	Oral ulcers	Yes	LVPA CAO CAPA	FAPA	↗CRP ↗ESR	Chronic inflammation of the arterial media of coronary artery, thrombus
Sacré	39	F	Haiti	Cough Dyspnea Fever	Oral and genital ulcers Erythema nodosum Polyarthritis	Yes	LVPA CAA	None	↗CRP	NA
Lai	28	M	China	Haemoptysis	Oral ulcers Erythema nodosum Pathergy test	Yes	LVPA CAA	PAA PE	↗CRP ↗ESR	Destruction of the elastic structures of the pulmonary artery, neutrophils and lymphocytes into the wall
Mouine	13	M	Morocco	Chest pain Fever	Oral and genital ulcers Uveitis Pathergy test	Yes	LVPA	None	↗CRP ↗ESR	NA
Lapébie	30	F	Mayotte	Chest pain Fever	Oral and genital ulcers Erythema nodosum	Yes	LVPA	DVT	↗CRP	NA

M: male; F: female; BD: Behçet's disease; LVPA: left ventricular pseudoaneurysm; CAO: coronary artery occlusion; CAA: coronary artery aneurysm; CAPA: coronary artery pseudoaneurysm; DVT: deep vein thrombosis; PVT: portal vein thrombosis; FAPA: femoral artery pseudoaneurysm; PAA: pulmonary artery aneurysm; PE: pulmonary embolism; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate.

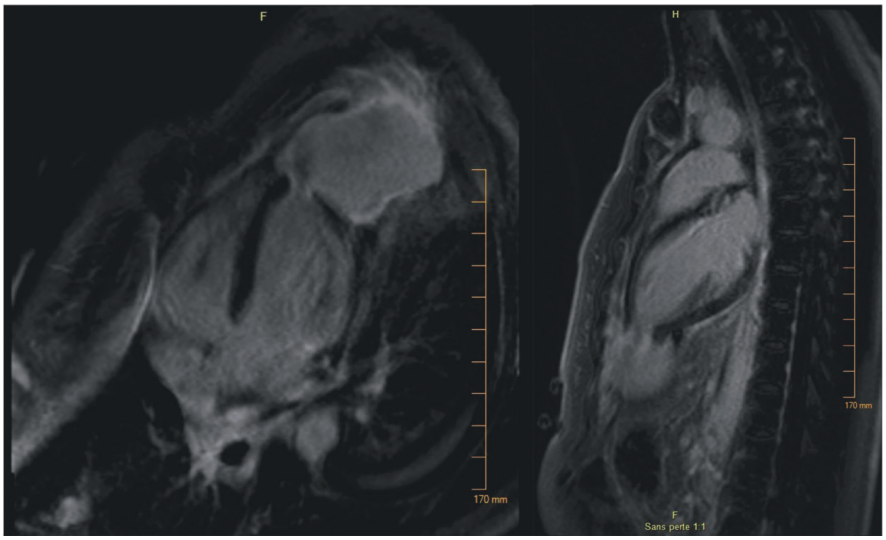

Fig. 2. Magnetic resonance imaging of the heart showing the left ventricular pseudoaneurysm and a limited transmural necrosis of the apex (T1-weighted sequences with late gadolinium enhancement).

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Competing interests: none declared.

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