CASE REPORT

COMPLETE RECOVERY OF RIGHT INTRAVENTRICULAR THROMBUS AND PULMONARY ARTERITIS IN BEHÇET’S DISEASE

D. LE THI HUONG, C. DOLMAZON, D. DE ZUTTERE,* B. WECHSLER, P. GODEAU and J.-C. PIETTE

Department of Internal Medicine, Groupe Hospitalier Pitié-Salpêtrière, 83 bd de l'Hôpital, 75013 Paris, *INSERM U 251 Hôpital Bichat, 46 rue Henri Huchard, 75018 Paris and Department of Physiology, Hôpital du Perpétuel Secours, 4 rue Kléber, 92300 Levallois-Perret, France

SUMMARY

We report on a patient who presented with sustained fever, weight loss, haemoptysis and elevated erythrocyte sedimentation rate. The diagnosis of Behçet’s disease was based on recurrent oral and genital aphthae, pseudofolliculitis and a history of thrombophlebitis. A right intraventricular thrombus and bilateral pulmonary aneurysms were discovered. Their complete recovery was observed within 6 months after a combination of prednisone, azathioprine, colchicine and aspirin therapy was started.

KEY WORDS: Behçet’s disease, Intraventricular thrombus, Arterial aneurysm.

In 1937, Hulusi Behçet reported on three patients with recurrent oral and genital aphthae and relapsing uveitis. This triad defined the disease that bears his name. Behçet’s disease (BD) is now recognized as a systemic disorder mainly affecting young adults in Mediterranean, Middle Eastern and Far Eastern countries. Its aetiology remains unknown. Various cardiovascular manifestations have been reported, but are rare. We report a new case of right intraventricular thrombus associated with bilateral pulmonary aneurysms that recovered completely under corticosteroids, azathioprine, colchicine and aspirin therapy.

CASE REPORT

A 48-yr-old Moroccan man, living in Morocco, was admitted to our department in November 1994 for a longstanding 39–40°C fever and 15 kg weight loss insensitive to antibiotics and antituberculous therapy. He had a history of left tibial thrombophlebitis in 1985. He complained of recurrent oral and genital aphthae, and recent haemoptysis. Physical examination revealed pseudofolliculitis and no ophthalmological abnormalities. Pathergy test was negative. ESR was 80 mm at the first hour. HLA B5 antigen was present; anticardiolipin antibodies and lupus anticoagulant were absent. Chest radiograph disclosed bilateral hilar opacities. EKG showed evidence of right atrial enlargement and incomplete right bundle branch block. Arterial and venous ultrasonography Doppler examination found left femoro-popliteo-tibial thrombophlebitis. Pulmonary scintigraphy displayed multiple bilateral pulmonary defects. Echocardiography demonstrated a 33 × 15 mm right intraventricular apical mass (Fig. 1) and an elevated estimated systolic pulmonary arterial pressure (30 mmHg). Thoracic CT scan showed that the right intraventricular mass was associated with bilateral pulmonary aneurysms involving the right superior lobar and the left inferior lobar arteries (Fig. 2), and distal pulmonary defects. A combination of prednisone 30 mg/day, azathioprine 100 mg/day, colchicine 1 mg/day and 100 mg/day aspirin therapy was started with rapid relief of fever and normalization of ESR within 2 weeks. Bilateral external compression stockings were prescribed. He was seen 6 months later. Haemoptysis and fever did not recur. He had 15 kg weight gain. ESR, chest radiograph, EKG, echocardiogram and thoracic CT scan were normal. Lung perfusion scintigraphy showed mild persistent defects. Ultrasonography Doppler examination showed persistent occlusion of left iliac, superficial femoral veins, but patent left popliteal and tibial veins. The prednisone dosage was then progressively reduced to 10 mg/day; azathioprine, colchicine and low-dose aspirin therapy were maintained with no relapse on >12 months of follow-up.

COMMENTS

Our patient had recurrent oral and genital ulcers, and pseudofolliculitis that fulfilled the International Study Group criteria for the diagnosis of BD [1]. To our knowledge, right intraventricular thrombus has been previously reported in 12 BD patients [2–10], of whom two were not detailed [10]. Haemoptysis was the most prominent revealing symptom. It was related to pulmonary embolism and/or pulmonary arteritis that were associated in eight cases, including ours [3–7, 10]. A tricuspid regurgitation murmur was present in two cases [4, 9]. Intraventricular thrombus was generally suspected by echocardiography [3–6, 8, 9] which was systematically carried out in four cases [3, 5, 6] or by angiography [3]. Intraventricular thrombus was only discovered at autopsy in two cases [2, 7]. Disappearance of intraventricular thrombus was observed after heparin [3, 8, 9] or warfarin therapy associated with corticosteroids [3, 6, 8, 9] and immunosuppressive therapy (azathioprine or oral cyclophosphamide) in three cases [3, 6, 8]. Ineffective...
combination of heparin, prednisone and azathioprine after 5 days therapy duration led to successful replacement of heparin by streptokinase in one case [3]. It should be underlined that one case of intraventricular thrombus recurred after surgical excision despite warfarin therapy with no corticosteroids [6]. In our case, pulmonary arterial aneurysms presented a problem with fibrinolytic and/or anticoagulant therapy. We demonstrated that prolonged combination of corticosteroids, azathioprine, low-dose aspirin and colchicine therapy may also be efficient. A persistent tumour-like mass under heparin alone [3] or no anticoagulant therapy [3, 6] led to endomyocardial biopsy [3] or ventriculotomy [3, 6].

Besides coincidental cardiac tumour, the major differential diagnosis of intracardiac thrombus in a patient with BD is endomyocardial fibrosis. This complication seems rare since only seven cases have been previously reported in the literature [2, 3, 10, 11]. Persistent intraventricular mass despite corticosteroids [11] or with no therapy [3] led to diagnostic endomyocardial biopsy [11] or surgery [3].

The striking feature of nearly all patients with intraventricular thrombus is prominent vascular involvement that defined the vasculo-Behçet’s disease, observed in 3–46% of the patients with BD [12].

Thrombophlebitis is its major feature. Arterial lesions are rare, but have a poor prognosis, particularly when they are located on pulmonary arteries. Because aneurysms expose patients to the risk of rupture, surgery or embolization should be considered in addition to medical therapy consisting of corticosteroids generally associated with cyclophosphamide or azathioprine. However, most pulmonary aneurysms are only stabilized. Between 38 and 75% of these patients died within 2 yr after diagnosis [11–14]. To the best of our knowledge, complete radiologic resolution was demonstrated in only three cases [13–15]. We previously described complete recovery of bilateral pulmonary aneurysms under prednisone, immunosuppressive and heparin therapy [14]. This patient remains in remission on colchicine and low-dose aspirin therapy. Hamuryudan et al. [14] observed that one out of 24 patients recovered under prednisolone and cyclophosphamide therapy, and Stricker et al. [15] reported one remission under prednisone alone.

REFERENCES
