Fever, haemoptysis and a mass in the heart

M J Steward¹ D P D'Cruz²
L Lang-Lazdunski³ J Chambers¹

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Behçet's syndrome is a complex multisystem disease and at presentation the classical triad of diagnostic criteria are not always evident.

CASE HISTORY

A 36-year-old man was admitted with fever, weight loss and malaise. Investigation at the referring hospital had revealed a right atrial mass (Figure 1) presumed to be a myxoma. However, at surgery there was a cyst arising from the superior margin of the coronary sinus filled with an acute inflammatory exudate and fibrin, but no organisms. Blood cultures were normal and bone marrow aspiration excluded malignancy. On subsequent questioning he had had oral and scrotal ulceration with several pustular lesions on his back and forehead within the previous year. He discharged himself but was readmitted with breathlessness and haemoptysis. There were two ulcers on the lateral border of the tongue. Computerized tomography (CT) pulmonary angiography showed bilateral pseudoaneurysms in the branches of the pulmonary artery. The right-sided aneurysm was embolized to prevent rupture. (Figure 2a and b). He remained febrile (38.2°C) with a normochromic, normocytic anaemia (Hb 8.5 g/dL), leucocytosis (white cell count $15.9 \times 10^9/\text{ml}$) and raised inflammatory markers (C-reactive protein 188 mmol/l and ESR 120 in the first minute). Urinalysis was normal. Antinuclear antibodies, antineutrophil cytoplasmic antibodies and rheumatoid factor were negative and complement levels were normal. Echocardiography showed no residual intracardiac abnormality. He was treated initially with intravenous Ceftriaxone because of the evidence of sepsis; however, the fever persisted. There were new lesions on the CT suggesting emboli and an abdominal CT revealed thrombus in the inferior vena cava, extending from L5 to the hepatic portion of the vessel. Lupus anticoagulant was strongly positive by Dilute Russell Viper Venom Time on two separate occasions, and he was anticoagulated with warfarin.

Figure 1 Transthoracic echocardiogram showing a right atrial mass. This is a magnified four-chamber view. There is a cystic lesion arising from the edge of the coronary sinus as it enters the right atrium.

Although there was no evidence of pathergy or eye disease, a diagnosis of Behçet's syndrome was made based on the presence of oral and genital ulceration, recurrent pustular lesions and pulmonary pseudoaneurysms. A diagnosis of antiphospholipid syndrome was based on the persistent lupus anticoagulant and thrombosis in the inferior vena cava with pulmonary emboli. After intravenous methylprednisolone and cyclophosphamide with high intensity warfarin (INR 3–4) there was a dramatic clinical response. He was subsequently readmitted with severe haemoptysis and required ligation of the left pulmonary artery to control bleeding from a bronchial erosion, from which he has made a good recovery.

DISCUSSION

Behçet's disease is a vasculitis classically manifesting as oral and genital ulceration with uveitis. The presentation is variable, however, and the only criterion regarded as essential by the international study group¹ is oral ulceration occurring at least three times in a 12-month period. This can be accompanied with a history of two of the following: recurrent genital ulcers, eye lesions, skin lesions or a positive pathergy test. Pulmonary pseudoaneurysms have been reported before with sarcoma in the differential diagnosis.² A number of treatment options for pulmonary pseudoaneurysms have involved surgery, aneurysm embolization and immunosuppression. There have not been controlled therapeutic trials carried out and there is therefore debate concerning the best modality. Antiphospholipid syndrome is documented in Behçet's disease,^{3,4} but thrombosis may occur even in its absence.⁵ Despite the multisystem involvement of the disease, cardiac pathology is

¹Department of Cardiology and ³Cardiothoracic Surgery, Cardiothoracic Centre, St. Thomas' Hospital, London, UK

²The Louise Coote Lupus Unit, St. Thomas' Hospital, London, UK

Correspondence to: Dr M J Steward

E-mail: MichaelSteward@doctors.net.uk

(a)

(b)

Figure 2 Bilateral pseudoaneurysms. (a) Chest radiograph showing prominent hilum with the right sided embolization coil. Patchy left sided opacification is visible, which could have been caused by an earlier bleed. (b) Contrast computerized tomogram: the left sided pseudoaneurysm arising from the left pulmonary artery branch is shown.

rare, although the non-specific features may be mistaken for endocarditis.⁶ This is the first report of a right atrial mass as a cyst arising from the superior margin of the coronary sinus, showing that Behçet's can also mimic a myxoma.

Competing interests None declared.

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