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CASE REPORT

Behçet's Disease with Resolving Pulmonary Artery Aneurysm and Intracardiac Thrombus

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ABSTRACT

The combination of pulmonary artery aneurysm and intracardiac thrombus is rare in Behçet's disease. We report on a 39-year-old man from Iraq with a clinical diagnosis of Behçet's disease based on a non-healing mouth ulcer, recurrent venous thromboses in both lower limbs, and pulmonary embolism. Computed tomographic pulmonary angiography demonstrated a pulmonary artery aneurysm and a transoesophageal echocardiogram revealed a right ventricular thrombus. After receipt of medical treatment, the pulmonary artery aneurysm and intracardiac thrombus showed partial and complete radiological resolution, respectively.

Key Words: *Aneurysm; Behcet syndrome; Pulmonary artery; Thrombosis*

中文摘要

貝賽特氏症併發消退中的肺動脈瘤及心腔內血栓

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貝賽特氏症（Behçet's disease）患者很少同時出現肺動脈瘤及心腔內血栓。本文報告一名伊拉克籍39歲男性因不能癒合的口腔潰瘍，雙腿出現復發性靜脈血栓形成及肺栓塞，而被診斷為患有貝賽特氏症。CT肺血管造影顯示有肺動脈瘤，經食管超聲心動圖發現有右心室血栓。治療後肺動脈瘤有部分減退的跡象，而心腔內血栓則完全消退。

INTRODUCTION

Behçet's disease (BD) is a chronic, relapsing, inflammatory disease of unknown aetiology with the clinical triad of recurrent oral ulceration, genital ulceration, and uveitis.[1] It is endemic in South East Asia and along the Silk Route throughout the Middle East, but is uncommon in other ethnic origins.[1] Recurrent oral ulceration is the hallmark of the disease.[2] It can also affect the gastrointestinal, central nervous, and vascular systems.[1] Pulmonary artery aneurysm (PAA) and intracardiac thrombus are known but rare complications of BD with potentially lethal consequences if not detected early and treated appropriately.[3]

CASE REPORT

A 39-year-old man from Iraq migrated to Australia eight years ago in year 2000 with a background history of unexplained recurrent deep vein thromboses and pulmonary embolism between 2004 and 2006 and a long-standing mouth ulcer (lasting more than 16 years). In 2003, he had had an episode of sagittal sinus thrombosis that was regarded as related to right

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mastoiditis. He was recently admitted to the infectious diseases unit with a 6-month history of rigors, night sweats, tiredness, and 10 kg loss of weight.

On physical examination, he was febrile (temperature of 38ºC). Several oral ulcers and bilateral folliculitis of the thighs were noted. He had no lymphadenopathy, genital ulceration, or ocular lesion, however. Full blood examination was normal. Inflammatory markers were elevated; his erythrocyte sedimentation rate was 53 mm/h (normal range, 0-20 mm/h) and his C-reactive protein level was 103 mg/l (normal level, <10 mg/l). A computed tomography (CT) of the chest and transoesophageal echocardiogram (TOE) were performed to investigate his pyrexia. The CT demonstrated a fusiform aneurysm measuring 18 mm in diameter at a fourth-generation right lower lobe pulmonary artery branch (Figure 1). It contained a mild-to-moderate eccentric mural thrombus. No haemorrhage was demonstrated. The TOE revealed a large mass within the right ventricular apex. A biopsy via a transfemoral venous approach confirmed it to be a thrombus (Figure 2).

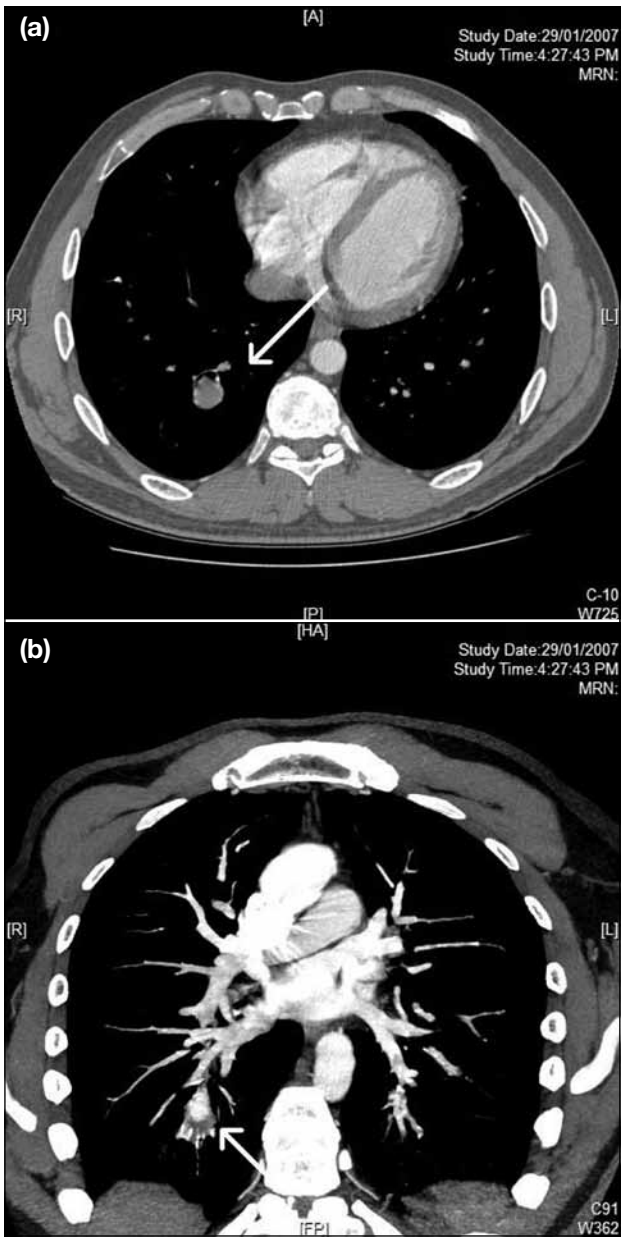

Figure 1. Computed tomographic pulmonary angiography in the (a) axial plane and (b) oblique coronal plane, demonstrating an 18-mm fusiform aneurysm that contains an intralumenal non-occlusive thrombus at the right lower lobe pulmonary artery branch (arrows).

A clinical diagnosis of BD was made based on the above clinical and imaging findings. Prednisolone and azathioprine were commenced. For the next 12 months, warfarin was prescribed because of the intracardiac thrombus. Follow-up CT of the chest three months after discharge showed partial resolution of the right lower lobe PAA, which had reduced to 5 mm in diameter (Figure 3). Four months after discharge, a transthoracic echocardiogram showed complete resolution of the right ventricular thrombus (Figure 4).

DISCUSSION

Vascular involvement in BD occurs in 14 to 62% of patients[4] and is more common in males.[5] It most commonly manifests as superficial vein thrombophlebitis and lower limb deep venous thrombosis.[6] Arterial involvement is less common, occurring in 1 to 7% of patients with BD,[6] manifesting as thromboses, stenoses, occlusions, aneurysms, and haemorrhage. About 1% of patients with BD exhibit PAAs.[3] It is important to recognise this condition, as earlier diagnosis and treatment confer a better prognosis, and notably about 33% of the thrombi in such PAAs increase in

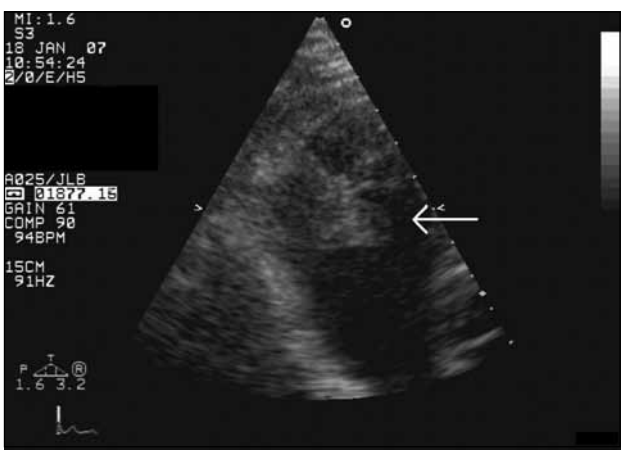

Figure 2. A transoesophageal echocardiograph showing a mildly echogenic 1.2 x 2.4 cm mass lesion (arrow) adhering to the wall of the right ventricle.

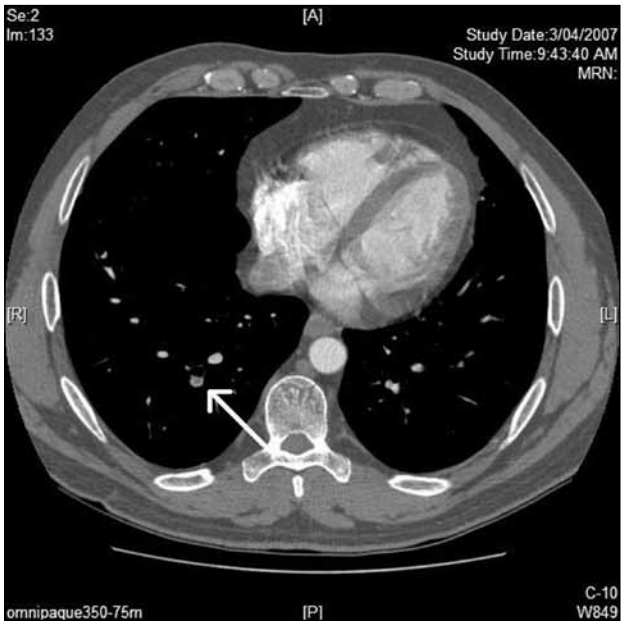

Figure 3. Computed tomography of the chest showing a partial resolution of the right lower lobe pulmonary artery aneurysm (arrow) after three months of corticosteroid and azathioprine treatment. A small thrombus is still visualised within the lumen of the artery.

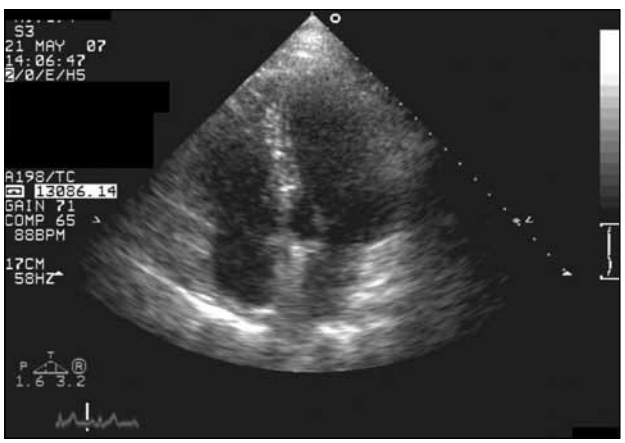

Figure 4. A transthoracic echocardiograph confirms the resolution of the thrombus in the right ventricle.

size during treatment before regressing.[6] The main presenting symptom of PAA is haemoptysis, though not in our patient. PAA is a life-threatening condition with a short-term mortality of 50%.[7] Over a three-month period, our patient showed almost complete resolution following corticosteroid and azathioprine treatment. There have been only eight case reports and one study in the literature regarding the reversibility of PAA with medical treatment of BD.[8] The histopathological process in PAA seems to be inflammation of the vasa vasorum of the tunica adventitia.[7] This inflammation causes necrosis and fragility of the vessel wall resulting in pseudoaneurysm formation.[9] After some time, adventitial fibrosis and thrombosis occur. We postulate that early immunosuppressive treatment prior to fibrosis may result in reversibility of the aneurysm or that the fibrosis of the aneurysmal wall may contract the lumen leading to a decrease in aneurysmal size.

Intracardiac thrombus is also a rare finding in BD. Mogulkoc et al[10] undertook a systematic review of the literature, which cited 21 case reports covering 25 patients. The combination of intracardiac thrombus with PAA is even rarer. Thus far, there have only been seven previous case reports of this combination in the literature.[10]

CONCLUSION

BD is a chronic relapsing inflammatory disease with no specific serological markers. A diagnosis of BD is completely dependent on clinical features. Although this condition is relatively uncommon, this diagnostic possibility should be considered in any patient with an unknown cause of mouth ulcers. Whilst vascular manifestations are not obligatory for the diagnosis of BD, PAA and intracardiac thrombus are definitely consistent with its known complications. It is important to recognise that these complications can be reversed following appropriate medical treatment, as demonstrated in our patient, and potentially life-threatening consequences can be avoided.

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