

CASE REPORT

Fatal Haemoptysis due to Pulmonary Artery Aneurysm in Behçet's Disease

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Introduction

Behçet's disease (BD) was described by the Turkish dermatologist, Hulusi Behçet, in 1937.¹ The main symptoms of the disease vary according to the affected organs such as skin, mucosa, eyes, joints, gastrointestinal tract, vascular system and nervous system.² For many years, the known vascular complications of BD have been attributed to thrombophlebitis. However, several reports have been published in recent years relating to cases with aneurysm and arterial occlusion.³⁻¹¹ The aetiology is still unknown; however, vasculitic changes are common in all involved organs.^{2,8}

Pulmonary artery aneurysm (PAA) is uncommon in patients with BD. PAA may be bilateral⁴ and may also rupture causing sudden death due to massive haemorrhage or haemoptysis from an arteriobronchial fistula.⁵ The incidence of vascular involvement in our 183 cases with BD was 28.4% (52 cases) in the last 15 years, PAA was observed in four (2.2%) patients who are reported in this paper.

Case Reports

Case 1

A 26-year-old male patient was admitted with a 2 day history of haemoptysis. BD had been diagnosed 3 years prior to admission. Chest X-ray revealed bilat-

eral perihilar well-circumscribed round opacities (Fig. 1). Computerised tomography (CT) of the thorax showed bilateral PAA (Fig. 2), verified at pulmonary angiography. Bronchoscopy revealed significant bleeding from the left lower lobe bronchus. Because of increased bleeding, the patient was urgently taken to theatre. Although a left lower lobectomy was carried out, the patient died due to massive haemoptysis on the third postoperative day. A post mortem study was

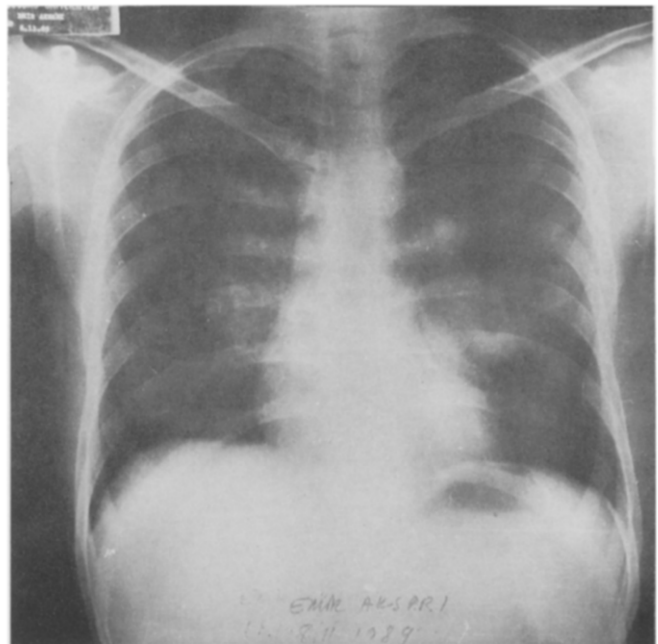


Fig. 1 Chest X-ray in patient 1 showing bilateral perihilar round opacities.

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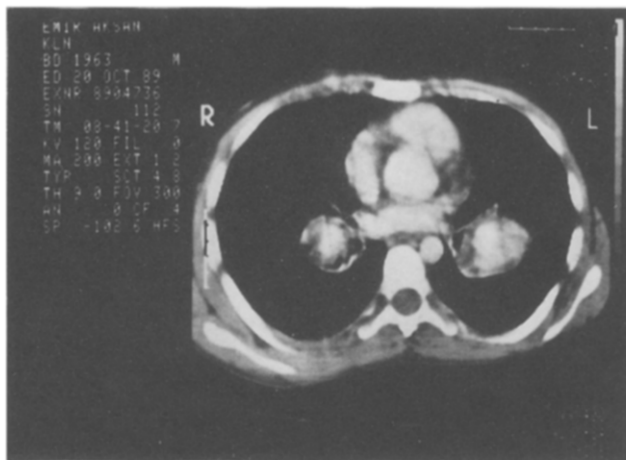


Fig. 2 The computerised tomography of the thorax in patient 1 showing bilateral PAA.

not performed, but the massive bleeding was attributed to rupture of the aneurysm in the right lung.

Case 2

A 20-year-old male patient was admitted with profuse haemoptysis. He had been known to have BD for 3 years. Chest X-ray and CT of the thorax revealed a giant rounded opacity on the left pulmonary artery and multiple rounded opacities in the right lung fields. Digital subtraction angiography (DSA) showed multiple small aneurysms of the branches of the right pulmonary artery and one giant aneurysm of the left pulmonary artery (Fig. 3). Bronchoscopy showed bleeding from the right main bronchus. The haemoptysis did not stop and on the second day the patient was operated on. Four saccular aneurysms on the right side were excised. A pneumonectomy was not performed because there was another giant aneurysm on the left side. On the second postoperative day, a massive haemoptysis occurred and bronchoscopy revealed bleeding from the left bronchial tree. High dose corticosteroid therapy was begun and mechanical ventilation was instituted, but the patient died on the 10th postoperative day due to continued bleeding and septicaemia.

Case 3

A 50-year-old male patient complained of dull pain on the left side of the chest and mild haemoptysis for 3

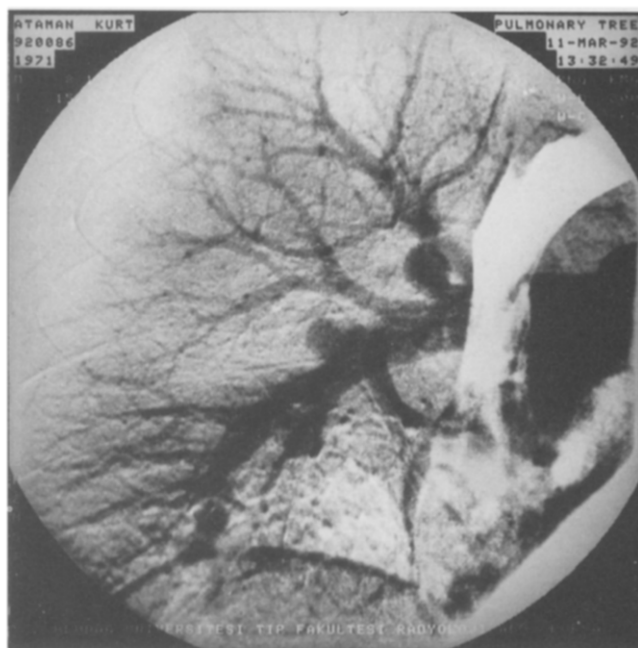


Fig. 3 Digital subtraction angiography in patient 2 demonstrating multiple small aneurysms of the branches of the right pulmonary artery and one giant aneurysm of the left pulmonary artery.

days. He had been followed up for BD for the last 10 years. Chest X-ray and CT of the thorax (Fig. 4) revealed a giant rounded opacity on the left pulmonary artery. It was thought that this could be a PAA and the patient was advised to have a DSA but he refused further investigation and treatment. He was therefore discharged and died due to massive haemoptysis 3 months later.

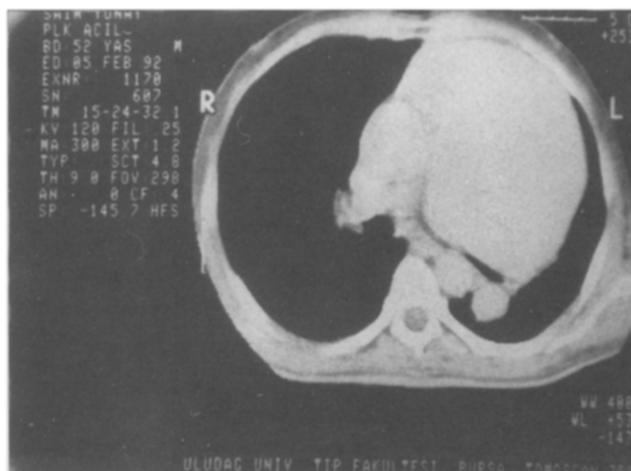


Fig. 4. The computerised tomography of the thorax in patient 3 showing a giant rounded opacity of the left pulmonary artery.

Case 4

A 35-year-old man was admitted with haemoptysis for 10 days. He had been diagnosed as having BD 3 years before admission and was blind owing to iridocyclitis. Chest X-ray and CT revealed bilateral round opacities in the lung fields which were confirmed as PAA at angiography. He was treated with prednisone azathioprine and colchicine and the haemoptysis ceased. He was not considered suitable for surgical treatment because of bilateral multiple aneurysms. The patient was subsequently admitted to the hospital several times due to haemoptysis and the same medical treatment given with success. He died due to a massive haemoptysis at home 11 months after the first episode of haemoptysis.

Discussion

Behçet's Disease is not a rare condition in Middle and Far East countries. The current prevalence is 1 in 10 000 in Japan and 8 in 10 000 in Turkey.^{2,9,12} This disease is mostly seen among young adults between the age of 20–40 and affects both sexes equally.^{12,13} However, all of our patients with PAA were male as has been indicated in most reports.^{4–7} Young male patients (15–25 years old) have a more severe course compared to female and older patients.¹⁴ Our patients had been diagnosed clinically by the International Study Group criteria¹⁵ at least 3 years before the first episode of haemoptysis. The pathergy test represents hyperactivity of the skin to simple trauma, such as a needle prick. A characteristic papule or a pustule forms in 24–48 h following the test. The prevalence of the pathergy test among Behçet's patients shows a geographic variation. The test has been found to be positive in 60–70% of patients in Turkey and Japan.² The test was positive in all four of our cases.

The incidence of vascular involvement is 25% and venous system involvement is seen more frequently than the arterial system.^{16–18} The incidence of vascular involvement in our 183 cases with BD was 28.4% (52 cases). Because only patients presenting with haemoptysis underwent further investigation, the true incidence of PAA may be higher than the 2.2% found by us. There are four types of vascular lesions in BD; (1) arterial occlusion, (2) arterial aneurysm, (3) venous occlusion and (4) varicose veins. Although isolated PAA was diagnosed in our patients, arterial aneurysms are most commonly seen in the abdominal aorta. Furthermore, femoral, subclavian and popliteal arterial aneurysms are also seen with decreasing

frequencies.^{16,18,19} The pathogenesis of arterial aneurysm is thought to be due to obliterative endarteritis of the vasa vasorum, destroyed elastic fibres in the medial layer resulting in dilatation, aneurysm and pseudoaneurysm formation or perforation.²⁰

Aneurysms of the pulmonary artery are rare and may also be caused by syphilis, mycotic aneurysms, trauma, and chronic pulmonary hypertension, particularly when associated with congenital anomalies of the heart and great vessels.¹⁰ Some authors believe that BD should be considered in the differential diagnosis of haemoptysis in areas of high prevalence such as Japan and Turkey.^{2,10,22} The reported prevalence of pulmonary involvement in BD ranges from 1.1% to 19%.^{3,21} Although PAA are uncommon, they can be bilateral and may rupture causing death due to either massive internal haemorrhage or haemoptysis from an arteriobronchial fistula.⁸ Haemoptysis is usually the first and the worst prognostic sign in BD leading to death within a few years.^{9,10}

Perihilar round shadows on chest X-ray are seen in the majority of the cases. The most accurate differential diagnosis is made by CT of the thorax, pulmonary angiography or pulmonary DSA. Although, CT was performed in all cases, the definite diagnosis was confirmed by angiography in three of our cases. In addition, bronchoscopy may show an aneurysm eroding into the bronchial tree or diffuse pulmonary haemorrhage in some patients.²² In three of our patients, the eroded area was not seen during the bronchoscopy due to massive bleeding. Various treatment modalities with different results have been described including surgery, embolisation of the aneurysm, immune suppressive drugs such as cyclophosphamide, azathioprine and recently cyclosporin alone or in combination with steroids.⁷ Recurrent or massive haemoptysis may require urgent pneumonectomy or lobectomy if the haemorrhage can be located during bronchoscopy. Lung resection may be considered if there is one single or multiple aneurysms in the same lung. If the surgery is performed for massive bleeding in patients with multiple aneurysms, rupture of the contralateral aneurysm may occur as observed in two of our patients. Primary repair or excision of the aneurysm can be performed in suitable cases. Transcatheter embolisation has also been suggested in cases with multiple or bilateral aneurysms.^{9,22}

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