

CASE REPORT

Bilateral pulmonary artery aneurysms in Behçet's disease

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Bilateral pulmonary artery aneurysms in Behçet's disease. Y. Almog, G. Polliack, M. Dranitzki Elhalel, M. Shalit, E. Rosenmann. ©ERS Journals Ltd 1993.

ABSTRACT: A patient presented with fever, haemoptysis and large bilateral perihilar masses. Histology proved that these were pulmonary artery aneurysms due to pulmonary vasculitis, as a manifestation of Behçet's disease. It is suggested that a routine dynamic computed tomographic (CT) scan should be performed in the evaluation of such cases.

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Behçet's disease is a chronic multisystem vasculitis, affecting many organs. Pulmonary involvement is rare in Behçet's disease, approaching 1-5%. We report a case of a young female with large bilateral pulmonary artery aneurysms due to Behçet's disease. The clinical, radiological and pathological aspects of this rare complication are discussed. This unique and potentially fatal condition should be well-recognized, since early steroid treatment can be of therapeutic value.

Case report

A 16 year old white Arab girl presented with low grade fever and haemoptysis of 6 month duration. The patient recalled having non-painful buccal aphthae several times in the last year, and episodes of migratory arthralgia in both knees and in the right hip.

Physical examination revealed an afebrile, pale and tachycardic (120 beats·min⁻¹) patient, with decreased breath sounds in the left lung base. Clubbing, white nails and Dupuytren's contractures were noticed. During the course of hospitalization, a non-painful oral ulcer appeared, and healed completely within three days. Relevant laboratory studies revealed erythrocyte sedimentation rate (ESR) of 60 mm·h⁻¹ (Westergren method) and profound microcytic hypochromic anaemia, with haemoglobin of 58 g·l⁻¹.

The postero-anterior and lateral chest radiograph (fig. 1 a and b) and the computed tomographic (CT) scan, which were obtained on admission, revealed bilateral, large rounded pulmonary masses. The maximal diameters measured 7.5 cm on the left side and 5 cm on the right. The mass on the left-side reached the diaphragm and the pleura of the lateral and posterior chest wall.

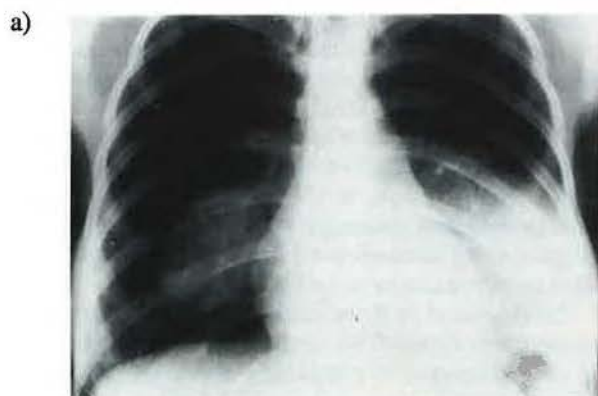


Fig 1a. - Postero-anterior chest radiograph showing bilateral pulmonary masses cast upon perihilar regions. Silhouetting of the left diaphragm, and extension to left lateral chest pleura are noted. b) Lateral view of the same lesions, demonstrating perihilar location, posterior extension, and silhouetting of the left mid-diaphragm.



Fig. 2. - Dynamic *i.v.* contrast computed tomograph (CT) of the chest, demonstrating the two large pulmonary artery aneurysms with mural thrombi, and on the right, surrounding air bubbles.



Fig. 3. - Left lung removed at surgery showing the huge pulmonary artery aneurysm and bronchoarterial fistula (probe).

CT with dynamic intravenous contrast injection, (fig. 2) revealed the enhanced lumen of a huge pulmonary artery aneurysm - bilaterally, surrounded by an unenhanced mural thrombus. On the right side, air bubbles were perceived in the thrombus, indicating a possible communication with the respiratory tract. Bronchoscopy and biopsies were non-contributory. At this stage an open lung biopsy was performed during which massive bleeding occurred. The left lung was resected in an unsuccessful attempt to save the patient's life. Gross examination of the resected lung showed a huge perihilar aneurysm, measuring 7.5 cm in diameter. Its wall proved to be a large pulmonary artery. Bronchoarterial fistulae were observed (fig. 3). Further microscopic evidence of healed vasculitis was found in the main and smaller pulmonary arteries. Recent and old organized and recanalized thrombi were conspicuous both in arteries and veins.

Discussion

The patient suffered from subfebrile illness associated with arthralgia, recurrent oral ulcers, haemoptysis, pro-

found iron deficiency anaemia, and bilateral perihilar pulmonary masses. Pathologically, these two masses proved to be large pulmonary artery aneurysms, resulting from a diffuse vasculitic process.

The air bubbles that were noted on CT proved to be, on pathological examination, a consequence of a fistula between the left pulmonary artery and the left main bronchus.

The occurrence of pulmonary vasculitis and aneurysms of the main pulmonary arteries in a patient with buccal aphthae and arthralgia was highly suggestive of Behçet's disease.

Behçet's disease is a chronic multisystem vasculitis of unknown aetiology, affecting many organs including skin, joints, eyes, central nervous system, gastrointestinal tract, kidneys, epididymis, heart, lung, large arteries and veins [1]. The vascular system (*e.g.* veins and arteries) is affected by an inflammatory process in 25% of the cases.

Pulmonary artery aneurysms are very rare. Recognized causes include syphilis, mycotic aneurysms derived from right-sided endocarditis, trauma and long-standing pulmonary hypertension [2]. Behçet's disease in its incomplete form, the Hughes-Stovin syndrome should be added to those diseases which cause aneurysms of large pulmonary arteries [2]. In fact, it is the only known primary vasculitic disease which causes such aneurysms.

Pulmonary involvement is rare in Behçet's disease, approaching 1-5% in different series [3]. Dyspnoea, chest pain and haemoptysis are the major clinical manifestations. The radiological findings include pulmonary infiltrates, pleural effusion, prominent pulmonary arteries, and, rarely, pulmonary artery aneurysms [4]. STRICKER and MALINVERNI [5] have recently reported a similar case with multiple large aneurysms of pulmonary arteries, and with similar CT findings that were confirmed angiographically. Complete and persistent resolution of signs and symptoms was achieved with prednisone treatment.

We conclude that in the clinical context of Behçet's disease, in the presence of hilar pulmonary masses, the possibility of pulmonary artery aneurysm should be considered. A CT with dynamic *i.v.* contrast injection is the examination of choice in these cases. It is superior to angiography since it lacks the complications, is noninvasive and brief [4]. In addition, it demonstrates the outer wall and thrombus. Furthermore, gas bubbles in these masses, which can only be seen on CT, suggest the presence of a bronchoarterial fistula, as was also found in this patient. Nuclear magnetic resonance (NMR) is another potential diagnostic tool although its advantage over dynamic CT in these cases remains to be proved.

The prognosis in these cases is very poor, because of severe haemoptysis. This case emphasizes the importance of early diagnosis of pulmonary artery involvement in Behçet's disease, since early steroid therapy, cyclosporin and cytotoxic drugs can be of therapeutic value and provide a potential cure [4, 5].

A routine CT with dynamic *i.v.* contrast injection is, thus, suggested in all patients suspected of having the disease.

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