

EDITORIAL

Pulmonary involvement in Behçet's disease

J.D. O'Duffy*

The diagnosis of Behçet's disease is made when recurring oral aphthous ulcerations are seen in patients having any two of the following features: recurring genital ulcers, uveitis or retinal vasculitis, cutaneous lesions, *i.e.* pustules or nodules, and a pathergic skin reaction to needle trauma [1]. To be valid, the lesions should be observed by a physician. Ordinarily, this poses no problem since the oral ulcerations, which are the best clue to the disease, are present most of the time. Important additional clinical features omitted from the diagnostic criteria include: meningo-encephalitis, synovitis, phlebitis and arteritis. Since modern treatment with immunosuppressive drugs can preserve both sight and life in Behçet's disease, we must sharpen our diagnostic wits so as not to miss the diagnosis.

ALMOG *et al.* [2], in this edition of the Journal, present the case of a young patient with recurring oral ulcers, arthralgias, and recurring haemoptysis. She did not fulfil the diagnostic criteria for Behçet's disease. However, the pulmonary arteritis and aneurysms against a background of recurrent oral ulcerations, arthralgia, and recurring haemoptyses make a compelling case for one diagnosis, *i.e.* Behçet's disease.

Haemoptysis is the dominant clinical feature of Behçet's pulmonary arteritis. It was the direct cause of death in 11 of 28 Behçet's patients having lung involvement in the review by EFTHIMIOU *et al.* [3]. A thrombo-angiitis affects the pulmonary vessels in this unique type of vasculitis [4-6]. The clinical syndrome is easily understood when one comprehends the pathology. The internal elastic lamella and elastic fibres of the media of the arteries are disrupted, in a piecemeal vasculitic process that results in infarcts, fibrosis, and, in 75% of histologically studied cases, aneurysms that communicate with bronchi [3-10]. All sizes of vessels can be involved, from the main pulmonary arteries to the capillaries and veins. Repeated bouts of haemoptysis interspersed with periods of few symptoms are explained by the varying age of vasculitis lesions, showing phases of vessel healing, scarring, and destruction. Haemoptysis is due to an artery-to-bronchus fistula. Most patients are young men, and evidence of veno-occlusive disease outside the lung is common [7, 11, 12].

Despite the unique presentation, the diagnosis is frequently missed. Indeed, the Hughes-Stovin syndrome, described in 1959 as comprising pulmonary "thromboses", veno-occlusive disease and intracranial venous hyper-

tension, in 1993 reads like a missed diagnosis of Behçet's disease, with pulmonary arteritis [12].

A check-list to help us recognize and manage Behçet's pulmonary arteritis presenting as haemoptysis is as follows:

1. Single or recurrent haemoptysis: ask about oral ulcers.
2. Chest X-ray: abnormal in 90%. Alveolar infiltrates are due to haemorrhage [10]. Perihilar opacities may be aneurysms in pulmonary or segmental arteries [3, 7, 10].
3. Ventilation-perfusion (V-Q) scan: expect it to be positive, but the infarction may be due to pulmonary arteritis. Do not anticoagulate: Behçet's disease patients seldom die of pulmonary infarction [11] - even those having recurrent deep vein occlusion. Haemoptysis is promoted by anticoagulants.
4. Bronchoscopy: may fail to localize the source of bleeding [3, 5].
5. Pulmonary arteriography: will show the extent of aneurysms, unless thrombosis occludes them [7]. Angiography can provoke bleeding in some patients.
6. Computed tomography (CT) with contrast: can delineate the aneurysms, but can also miss them [3, 5, 7]. If the situation is not an emergency, consider options 5 and 6.
7. If the situation is an emergency, urgent transfusion and surgical consultation are mandatory. Dramatic haemoptysis can force the issue. Local resection of the involved lobe, or even lung, can be life-saving. Local resources and the degree of urgency will determine whether CT can be obtained. If not, chest X-ray and bronchoscopy will be the surgical guides.
8. In less urgent cases, as well as in any patient who has survived aneurysm resection, begin prednisone 1 mg·kg⁻¹ *q.d.* plus chlorambucil 0.1 mg·kg⁻¹ *q.d.* [13]. Corticosteroids alone do not suffice to prevent a fatal outcome [3, 8]. Immunosuppressive treatment can be tapered over one year. This regimen is based on its successful use in Behçet's patients having ocular or cerebral disease.

With the above measures it should be possible to lower the mortality rate in pulmonary Behçet's disease from its estimated current unacceptable level of nearly 50% [4].

References

1. International Study Group for Behçet's disease. - Criteria for diagnosis of Behçet's disease. *Lancet* 1990; 335: 1078-1080.
2. Almog Y, Polliack G, Elhalel MD, *et al.* - Bilateral pulmonary artery aneurysms in Behçet's disease. *Eur Respir J* 1993; 6: 1067-1069.
3. Efthimiou J, Johnston C, Spiro SG, Turner-Warwick M. -

* Dept of Internal Medicine, Division of Rheumatology, Mayo Clinic, Rochester, Minnesota 55905, USA.

Pulmonary disease in Behçet's syndrome. *Q J Med* 1986; 227: 259-280.

4. Kohno S, Fujikawa M, Kanda T, *et al.* - A case of Behçet's syndrome with rupture of a pulmonary aneurysm. Autopsy findings and a literature review. *Jap J Med* 1986; 25: 293-300.
5. Lie JT. - Cardiac and pulmonary manifestations of Behçet's syndrome. *Path Res Pract* 1988; 183: 347-352.
6. Slavin RE, deGroot WJ. - Pathology of the lung in Behçet's disease: case report and review of the literature. *Am J Surg Pathol* 1981; 5: 779-788.
7. Raz I, Okon E, Chajek-Shaul T. - Pulmonary manifestations in Behçet's syndrome. *Chest* 1989; 95: 585-589.
8. Stricker H, Salvetti M. - Seltene vaskuläre Komplikationen des Morbus Behçet's (sogenannter AngloBehçet's). *Schweiz Med Wschrn* 1989; 119: 1290-1295.
9. Bouacha H, Azzabi S, Hamzaoui A, Kanzari A, Daoues A, Maalet M. - Aneurysme de l'artere pulmonaire et infarctions pulmonaires multiple inaugurant la maladie de Behçet. *Ann Med Interne (Paris)* 1991; 142: 544-546.
10. Gibson RN, Morgan SH, Krausz T, Hughes FRV. - Pulmonary artery aneurysm in Behçet's disease. *Br J Radiol* 1985; 58: 79-82.
11. Wechsler B, Picte JC, Conard J, Du LTH, Bletry O, Godeau P. - Les thromboses veineuses profondes dans la maladie de Behçet: 106 localisations sur une serie de 177 malades. *Presse Méd* 1987; 18: 661-664.
12. Hughes JP, Stovin PFI. - Segmental pulmonary artery aneurysms with peripheral venous thrombosis. *Br J Dis Chest* 1959; 53: 18-27.
13. O'Duffy JD. - Vasculitis in Behçet's disease. *Rheum Dis Clin North Am* 1990; 16: 423-431.