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Vascular Thrombotic Problems in Behçet's Disease

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We have read with great interest the case report by Sanchez-Burson et al. [1] on Behçet's disease (BD) with deep vein thrombosis (DVT) and subsequent pulmonary embolism. They describe a 46-year-old female patient with a 6-month history of BD. She had uveitis and was being treated with corticosteroids and thalidomide at the time of vascular complications. Although anticoagulant therapy was begun, she had recurrent DVT and pulmonary embolism within a week. That time she was treated successfully with the thrombolytic agent urokinase and at the end of the 2 years of therapy, she was free of thrombotic attacks.

In a recent study, we have evaluated 224 patients with BD of which 36 had DVT of various systems [2]. The first 2 years of the disease were the most critical period for DVT. Other risk factors were an age ≤30 years and the presence of ocular involvement [3]. Pulmonary arterial thrombotic complications generally occur in the presence of DVT of the calf veins and isolated involvement of the pulmonary artery is rare. In a previous retrospective study during 1968–1988, 3 patients with pulmonary arterial occlusion were reported from our institute [4]. These patients were symptomatic with chest pain and dyspnoea. Diagnosis was confirmed by angiographic examinations. Actually, thrombotic disease of the pulmonary arteries is much higher than expected as evidenced by abnormalities on perfusion scintigraphy, computed tomography scan and angiographic investigations in asymptomatic BD patients [5].

The main problem in BD is to prevent recurrent serious complications – such as vascular thrombotic disease and ocular involvement and their recurrences. It is hard to say that, with thrombolytic therapy, recurrences of DVT might be prevented. The course of BD is unpredictable and the disease generally leads a course with exacerbations and remissions. Not all patients with DVT are as lucky as the patient presented by Sanchez-Burson et al. [1]. In a large proportion of patients, BD runs a mild to moderate course, where conventional forms of therapy such as colchicine and thalidomide may control mucocutaneous and articular symptoms [6, 7]. However, these conventional drugs and corticosteroids have very little if any effect on the course of vascular thrombotic complications. In recent years, alpha-interferon (α-IFN) has been gaining popularity in the treatment of BD especially in severe forms of the disease with vascular and ocular complications. We have tried α-IFN in patients with vascular and ocular complications and obtained favourable results in decreasing the attack rate [8, 9]. Since the first 2 years of the disease seem the most critical period for major complications [2, 3], we feel that it is reasonable to treat patients preferably with α-IFN during this period.

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