Behçet’s disease (BD) is a multisystem inflammatory disease of unknown aetiology, characterized by oral and genital ulcers and cutaneous, ocular, arthritis, vascular, central nervous system and gastrointestinal involvement (1, 2).

Vasculo-Behçet’s disease affects arteries, veins, and blood vessels of all sizes; it occurs in about 7.7-43% (3-5).

Venous thrombosis (VT) is the most common manifestation, notably superficial thrombophlebitis, in as many as 1/3 of patients. Involvement of large veins, such as thrombosis of the superior (SVC) or inferior vena cava (IVC), is rare.

We propose to study the clinical features, the treatment, and the outcome of BD patients with vena cava thrombosis (VCT).

We have performed a retrospective review of the records of 430 cases diagnosed as BD in the department of Internal Medicine, La Rabta University Hospital in Tunis, Tunisia (a tertiary referral centre), over a 20-year period (1989-2009). Diagnosis of BD was made according to the criteria of the International Study Group for BD (6). Our data were analysed using the SPSS (version 11).

The diagnosis of VT was made using venous ultrasonography in all patients, with computed tomography in cases with VCT.

The patients were 295 males and 135 females (sex-ratio=2.2), with a mean age of 33 years. The clinical and genetic features of the patients are summarized in Table 1.

Twenty nine patients had VCT (6.74%). They were all male, with a mean age of the disease of 24.78 years (18-38 years) at the beginning and 32.34 years (18-48 years) at the time of VCT diagnosis. The average time to VCT diagnosis was 7 years. The VCT revealed the disease in one case.

Twelve patients had isolated superior VCT (46.4%) and 11 (39.2%) had isolated inferior VCT. The 2 localizations occurred simultaneously in 6 cases (20.68%).

The comparison of demographic data and frequency of clinical manifestations between patients with and without VCT is presented in Table 2.

All patients were treated by anticoagulation. Corticosteroids (3 pulses of methylprednisolone 1gr/day and then Prednisolone: 1mg/kg/day; mean duration: 24 months) and immunosuppressive therapy was indicated in 22 cases (Intravenous cyclophosphamide: 20 cases, azathioprine: 2 cases). Eight patients were lost to follow up. The outcome was good in 12 cases; extension occurred in 4 cases (Sus hepatic and jugular vena).

In our study, we found a significant association of VCT with younger age (p=0.001), male gender (p<0.018), pseudofolliculitis (p=0.05) and a strong association with positive pathergy test (p=0.07), while retinal vasculitis was less frequent (p=0.07). It was significantly associated to lower limb, sus hepatic and jugular thrombosis (p=0.01).

VCT is a rare but well-recognized manifestation of BD, observed in 0.2 to 10% of cases, more frequently in West Mediterranean and European patients (6, 7). Its prevalence was 6.74% in our study; it was 2.1% in the Lebanon study of Tahmé et al. (4), 33.8% in the study of Düzgün et al. (3), and 1.6% in the largest cohort of Gürler et al. (8).

In the study of Koç et al. (9), patients with subcutaneous thrombophlebitis were more likely to develop major venous occlusions (22.2%) in the lower extremities and inferior vena cava; no other study compared the clinical manifestations in BD patients with or without VCT.

The recommendations of the European League Against Rheumatism of BD indicate corticosteroid and immunosuppressive therapy for the treatment of VCT (10). Anticoagulation is still discussed. In our series, 20 patients were treated by cyclophosphamide and all received anticoagulation.

In conclusion, the frequency of VCT in our study is comparable to those of other studies. It is usually associated with other thrombosis. And to our knowledge, no study has specifically systematically examined VCT in BD. We must think of the diagnosis of BD in patient with VCT.
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References
