Vena cava thrombosis in Behçet's disease

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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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Accepted Date: 21.11.2013 **Available Online Date:** 05.03.2014

DOI:10.5152/akd.2014.5042

Table 1. Clinical features of the patients

1- Clinical features	Patients (n=430) n (%)
Oral ulcers	430 (100)
Genital ulcers	341 (85)
Pseudofolliculitis	320 (74.4)
Positive pathergy test*	176/305 (57.7)
Articular involvement	195 (45.7)
Ocular involvement	200 (46.5)
Neurological involvement	140 (32.6)
Vascular involvement	150 (34.9)
Deep vein thrombosis	136 (31.6)
Arterial aneurysms	23 (5.3)
Arterial thrombosis	6 (1.4)
Intestinal involvement	7 (1.6)
HLA B51 +	84/177 (47.5)
*done in 305 patients	

Table 2. Comparison of patients with and without Vena cava thrombosis

	without TVC n=401 n (%)	with TVC n=29 n (%)	*P*
Mean age at onset	29.37	25.65	0.018
Males	266/135	29/0	<0.001
Genital aphthosis	316 (78.8)	25 (86.2)	NS
Cutaneous aphhtosis	339 (84.53)	27 (93.1)	NS
Pseudofolliculitis	294 (73.31)	26 (89.65)	0.05
Erythema nodosum	70 (17.45)	3 (10.34)	NS
Positive pathergy test*	165 (56.7)	12 (80)	0.07
Ocular involvement	190 (47.38)	10 (34.48)	NS
Uveitis	178 (44.38)	9 (31)	NS
Retinal vasculitis	118 (29.42)	4 (13.79)	0.071
Articular involvement	184 (45.88)	11 (37.93)	NS
Arterial lesions	25 (6.2)	3 (10.34)	NS
Neurologic involvement	111 (27.68)	10 (34.48)	NS
HLA B51*	81 (47.9)	3 (33.3)	NS
*done in 178 patients			

Conflict of Interest: None declared.

Peer-review: Externally peer-reviewed.

Authorship contributions: Concept - A.H.; Design - A.H.; Supervision - A.H., H.M.H.; Resource - A.H.; Materials - A.H.; Data collection&/or processing - A.H., S.K.M.; Analysis &/or interpretation - J.F; Literature search - A.H., J.F; Writing - A.H.; Critical review - A.H., B.S.T.; Other - B.G.I., L.M.

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