J Neurol (2011) 258:908–909
DOI 10.1007/s00415-011-5967-3

SHORT COMMENTARY

Response to comment on "Cerebral venous thrombosis in Behcet's disease: a systematic review" by Afshin Borhani-Haghighi and Anahid Safari

Diana Aguiar de Sousa · Tiago Mestre · José Ferro

Published online: 15 March 2011

We read with interest the comments by Borhani-Haghighi and Safari, which corroborate the particular character of cerebral venous thrombosis (CVT) in Behçet's disease (BD).

The physiopathology of CVT in BD was beyond the scope of this article. However, we took your suggestion and reviewed in all the included studies the presence of venous infarction or intracranial hemorrhages, including hemorrhagic transformation. This information is presented in Table 1. As Borhani-Haghighi and Safari suspected these intracerebral lesions are absent in most of the cases of CVT in BD.

The high frequency of extra-neurologic vascular events in patients with BD and CVT has been repeatedly highlighted, favoring the "vasculo-Behçet" hypothesis. Tunc et al. [1], and Wechsler et al. [2] found this association in 64% of patients with CVT, but only in 36% without CVT. Likewise, Al-Fahad and Al-Araji [3] reported that 55% patients who presented with intracranial hypertension (IH) had other venous thromboses, but this occurred only in 14% of patients with other neurological manifestations. This association was also referred to by Saadoun et al. [4]. However, this author also found that prothrombotic risk factors were present in one-third of patients with CVT and in most (56%) of those relapsing. The odds of thrombosis relapse was higher in those patients with a prothrombotic

Table 1 Presence of venous infarction or hemorrhagic transformation in patients with CVT and BD (included studies)

Study	Reference to venous infarction or intracranial hemorrhages	%
Farah et al.	–	
Ramos et al.	–	
Wechsler et al. [2]	In four patients, focal hypodensities were observed (lenticular, temporal, diencephalic, mesencephalic, and pontine).	4/25
Akman-Demir et al.	–	
Borhani-Haghighi et al.	–	
Tohme et al. 1997	–	
Mnif et al.	–	
Houman et al. 2002	–	
Al-Fahad et al.	–	
Daif et al.	–	
Mossadeq et al.	Typical pattern of Rosenthal vein thrombosis	
Rougemont et al.	–	
Wechsler et al. 1993	One case of hemorrhagic infarct (of venous origin) (rim of hypodensity surrounding the hyperintense lesion, both on T1 and T2	1/10
Pamir et al.	–	
Houman et al.	–	
Al-Araji et al.	–	
Saadoun et al.	Focal T2-weighted hypersignals (lenticular, temporal, diencephalic, mesencephalic, and pontine) were observed in 6 patients	6/64
Kidd et al.	One case of cortical vein thrombosis presented with a hemisphere lesion	1/4

This comment refers to the article by Sousa et al. doi:10.1007/s00415-010-5885-9

D. A. de Sousa · T. Mestre · J. Ferro (✉)
Department of Neurosciences (Neurology), Hospital de Santa Maria, University of Lisboa, Lisbon, Portugal
e-mail: jmferro@fm.ul.pt

Table 1 continued

Study	Reference to venous infarction or intracranial hemorrhages	%
Tohmé et al. 2009		
Siva et al.		
Yesilot et al.	One case presenting with isolated intracranial hypertension had a small (10 9 15 mm) hypodensity in the right frontal premotor cortex on the CT scan. On T2 weighted MRI, the lesion was evaluated as a hemorrhagic infarction. The other BD patient had a right frontal venous infarction.	2/36
Kural-Seyahi et al.	–	
Benamour et al.	–	

risk factor (4.9) than in patients with peripheral venous thrombosis (2.8).

The available information on therapeutic options and outcome, including sequels, is presented in our paper in Table 5. Definite conclusions on what treatment could provide the best outcomes, in respect to sequels and prevention of relapses, can only be provided by a specifically designed randomized controlled study.

Conflict of interest None.

References

1. Tunc R, Saip S, Siva A, Yazici H (2004) Cerebral venous thrombosis is associated with major vessel disease in Behçet's syndrome. Ann Rheum Dis 63:1693–1694
2. Wechsler B, Vidailhet M, Piette JC, Bousser MG, Dell Isola B, Blétry O, Godeau P (1992) Cerebral venous thrombosis in Behçet's disease: clinical study and long-term follow-up of 25 cases. Neurology 42:614–618
3. Al-Fahad SA, Al-Araji AH (1999) Neuro-Behcet's disease in Iraq: a study of 40 patients. J Neurol Sci 170:105–111
4. Saadoun D, Wechsler B, Resche-Rigon M, Trad S, Le Thi Huong D, Sbai A, Dormont D, Amoura Z, Cacoub P, Piette JC (2009) Cerebral venous thrombosis in Behçet's disease. Arthritis Rheum 61:518–526