



Intracerebral Hemorrhage in Behcet's Disease

Ali M. Mursi*

Department of Rheumatology Benha Teaching hospital, General Organization of Hospitals and Institutes, Benha, Egypt

*Corresponding author: Ali M. Mursi Department of Rheumatology Benha Teaching hospital, General Organization of Hospitals and Institutes, Benha, Egypt. Email: alimursi2015@gmail.com

Citation: Mursi AM (2018) Intracerebral Hemorrhage in Behcet's Disease. Int J Musculoskelet Disord: IJMD-112 DOI: 10.29011/IJMD-112.0000012

Received Date: 29 September, 2018; **Accepted Date:** 6 November, 2018; **Published Date:** 14 November, 2018

Abstract

Background: Behcet's disease is a heterogeneous, multisystem, inflammatory disorder of unknown etiology. The classic triad of oral and genital ulcerations in conjunction with uveitis was originally described by the Turkish dermatologist Hulusi Behcet in 1937, but associated symptoms of the cardiovascular, central nervous, pulmonary, and gastrointestinal systems were later identified. In fact, BD with neurological involvement (neuro-Behcet's disease) is not uncommon. Patients with Neuro-BD typically exhibit a diverse array of symptoms, most commonly in the brainstem and diencephalic regions. Here in, we report two unusual cases of Neuro-BD in a patient who presented with a Lt basal ganglia hemorrhage and one with subarachnoid hemorrhage.

Cases presentation: Case-1: A 28-year-old Egyptian male was admitted to our hospital with complaints of a sudden RT side weakness, arthralgia, urine incontinence and depressive syndrome. CT angiogram and MRI revealed a Lt basal ganglia hemorrhage.

Case-2: A 36-year-old Egyptian male known case of BD was admitted to our hospital complaining of severe headache and red eye CT was done and revealed subarachnoid hemorrhage.

Conclusions: These findings suggest that the patient's cerebellar hemorrhage could have been due to intracranial vasculitis in a rare, if not unique, complication of neuro-Behcet's disease.

Keywords: Behcet's disease; Case report; Intracerebellar hemorrhage; Neuro-Behcet's disease

Abbreviations

BD	:	Behcet's Disease
MR	:	Magnetic Resonance Imaging
Neuro-BD	:	Neuro-Behcet's Disease,
Rt	:	Right
Lt	:	Left

Background

BD is a heterogeneous, multisystem, inflammatory disorder of unknown etiology. The classic triad of oral and genital ulcerations in conjunction with uveitis was originally described by the Turkish dermatologist Hulusi Behcet in 1937, but associated symptoms of the cardiovascular, central nervous, pulmonary, and gastrointestinal systems were later identified. In fact, BD with neu-

rological involvement (neuro-BD) is not uncommon.

Patients with neuro-BD typically exhibit a diverse array of symptoms, most commonly in the brainstem and diencephalic regions [1]. However, these Central Nervous System (CNS) abnormalities tend to resolve over time. Cerebral venous thrombosis is commonly evident on neuroimaging analyses [2]. In this report, I describe an unusual presentation of two cases of neuro-BD in a patient who presented with a Lt basal ganglia and subarachnoid hemorrhage.

Cases presentation

Case-1

A 28-year-old Egyptian male was admitted to our hospital with complain of a sudden RT side weakness, arthralgia, urine incontinence and depressive syndrome. He had been initially diagnosed with BD in 2010, with oral and genital ulcerations and uveitis. There had been no recent head trauma. CT angiogram and MRI revealed a Lt basal ganglia hemorrhage.

On admission, vital signs were normal, physical examination showed Rt side muscle weakness and hyper reflexia (Figure 1). ophthalmological examination showed anterior uveitis, all laboratories were average or negative for autoimmune profile. The patient received pulse methylprednisolone/day for 5 days with no response so we added cyclophosphamide pulse with minimal improvement of uveitis and cerebral hemorrhage, so we decided to give infliximab 5mg/kg/dose doses 0,2,6 and every 8 weeks uveitis completely improved and CT showed complete disappearance of hemorrhage after 5 months of therapy.

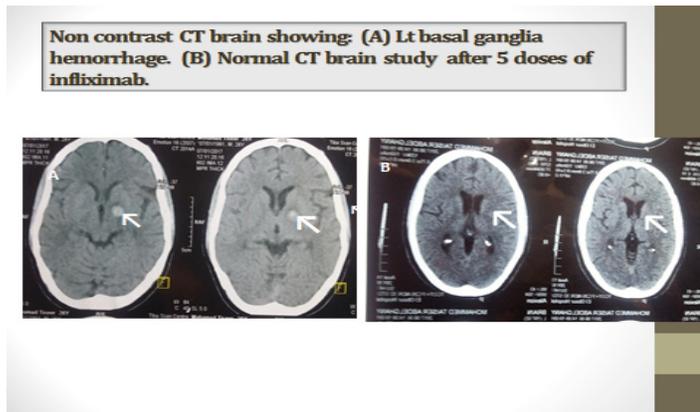


Figure 1: Non contrast CT showing: (A) Lt basal ganglia hemorrhage. (B) Normal CT brain study after 5 doses of infliximab.

Case-2

A 36-year-old Egyptian male known case of BS was admitted to our hospital (ICU) complaining of severe headache and red eye. CT was done and revealed subarachnoid hemorrhage (2010). He was diagnosed as BS since 2008 as he had recurrent painful genital, mouth ulcers and uveitis.

CT angiogram of the brain was done no vascular malformations or aneurysms. From 2011 till now he had many attacks of mouth and genital ulcers, IHD treated by nitroglycerine when needed (2013) (Figure 2). Attack of brain infarction and thrombosis of all brain sinuses plus Rt testicular gangrene with urgent surgical removal (2016). He had attack of hematemesis which diagnosed as thrombosed Rt pulmonary artery aneurysm which was treated and improved by infliximab after failure of methylprednisolone pulse and cyclophosphamide pulses (2017).

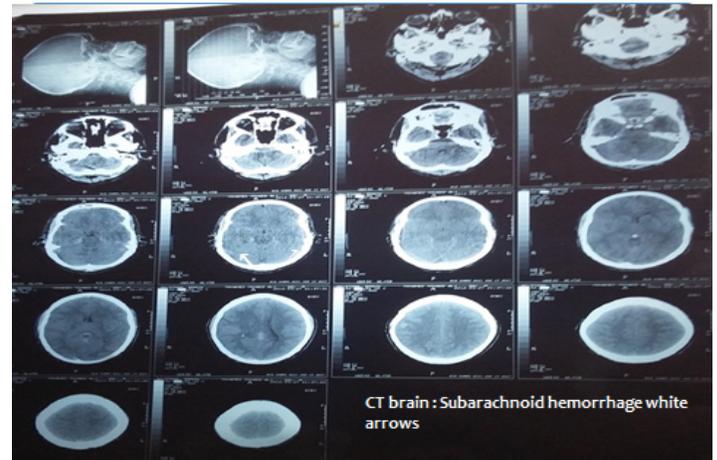


Figure 2: CT brain: Subarachnoid hemorrhage white arrows.

Discussion

The vasculitis is considered to be a key feature of neuro-BD [3]. The veins and arteries of any size can be affected. Venous manifestations appear to be more prevalent than those of arteries [4]. Neuroradiologic studies suggested that neuro-BD resulted in the development of inflammatory lesions in the brain stem as well as in a breakdown in blood-brain barrier [5].

Neuropathologic studies; the syndrome has been described as representing a vasculitis with cellular infiltrates mainly affecting post capillary venules with reports of intracranial hemorrhage being rare [6]. Vascular lesions are common in many of the complications of BD. Most of these lesions are thought to contribute to the occlusive process or to aneurysm formation in large vessels. Most cerebrovascular disturbances in BS are occlusive in nature such as in Dural Sinus thrombosis, while hemorrhage is rare. Reported one case of neuro-BD presenting with intracerebral hemorrhage. (A 39-year-old women with Rt cerebellar hemorrhage). The vascular complications include cerebral venous thrombosis, and subarachnoid hemorrhages associated with intracranial aneurysms [7].

Few reports have described cerebral hemorrhages that develop in the absence of aneurysms and vascular abnormalities. described 5 cerebellar hemorrhagic BD cases at which the hemor-

rhagic lesions, three were subacute, and two were acute. Hemorrhagic lesions were identified in the mesodiencephalic junctions of three patients, the tectum of one patient, and the posterior perforate substance of another patient. suggested that the cerebellar hemorrhage could have been due to intracranial vasculitis, which is a rare, if not unique, complication of neuro-BS and decided that hemorrhagic complications of cerebral arteritis. developing subsequent to arterial inflammation and vessel wall weakening. The cerebellar hemorrhage was attributable to similar venous changes, although other possible causes cannot be excluded such as venous thrombosis-associated hemorrhage, an infarct that subsequently underwent hemorrhagic transformation, or neuro-BS of uncertain etiology.

As like our patients- there is no hypertension, a vascular anomaly, no venous thrombosis or coagulation abnormalities. So he stated that BG hemorrhage particularly; caused by the vasculitic changes associated with BS as they appeared at the same site of the inflammatory lesions in the acute phase of the disease. Also, they stated that the hemorrhagic lesions resulted from disruption of the integrity of small vessels. Unlike our patients, there were one case caused by prolonged use of cyclosporine > 5 years. (as cyclosporine induces endothelial cell damage and also induces hypertension which not in our cases).

Conclusion

Despite cerebral hemorrhage is a rare manifestation of BS, review of literatures showed only 14 cases reported all over the world till now. Neuro-BD should be considered as a diagnosis on cases presented with cerebral hemorrhage and in particularly pontine hemorrhage especially in absence of causes of hemorrhage.

Can anti-TNF monoclonal antibodies treat hemorrhagic presentation of BD?

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

1. Kocer N, Islak C, Siva A, Saip A, Akman C, et al. (1999) CNS involvement in neuro-Behcet syndrome: an MR study. *AJNR Am J Neuroradiol* 20: 1015-1024.
2. Wechsler B, Vidailhet M, Piette JC, Bousser MG, Dell Isola B, et al. (1992) Cerebral venous thrombosis in Behcet's disease: clinical study and longterm follow-up of 25 cases. *Neurology* 42: 614-618.
3. Shahien R, Bowirrat A (2010) Neuro-Behcet's disease: a report of sixteen patients. *Neuropsychiatr Dis Treat* 6: 219-225.
4. Owlia MB, Mehrpoor G (2012) Behcet's disease: new concepts in cardiovascular involvements and future direction for treatment. *ISRN Pharmacol* 2012: 760484.
5. Kidd D, Steuer A, Denman AM, Rudge P (1999) Neurological complications in Behcet's syndrome. *Brain* 122: 2183-2194.
6. Siva A, Kantarci OH, Saip S, Altintas A, Hamuryudan V, et al. (2001) Behcet's disease: diagnostic and prognostic aspects of neurological involvement. *J Neurol* 248: 95-103.
7. Zsigmond P, Bobinski L, Bostrom S (2005) Behcet's disease, associated with subarachnoidal heamorrhage due to intracranial aneurysm. *Acta Neurochir (Wien)* 147: 569-571.