International Journal of
Neurology and Neurotherapy

Cação et al. Int J Neurol Neurother 2016, 3:044
DOI: 10.23937/2378-3001/3/2/1044
Volume 3 | Issue 2

ISSN: 2378-3001

Case Report: Open Access

# Trigeminal Neuropathy as a Relapse in Behçet Disease

***Gonçalo Cação*, Isabel Moreira, José Eduardo Alves, Fátima Farinha and Ernestina Santos***

*Neurology Department, Centro Hospitalar do Porto, Porto, Portugal*

***Corresponding author:** *Gonçalo Cação, Neurology Department, Centro Hospitalar do Porto, Largo do Professor Abel Salazar, 4099-001 Porto, Portugal, Tel: +351 912860834, Fax: +351 222 053 218, E-mail: goncalo.cacao@gmail.com*

**Abstract**

A 49 years old woman, diagnosed with Neuro-Behçet, was admitted with complains of numbness, aching and periods of electric shock-like pain in right side of the face. Associated with painful oral ulcers, anorexia, nausea and gait instability. Neurological examination revealed dysesthesia on right trigeminal nerve territory, horizontal-rotatory nistagmus on horizontal gaze bilateral with fast phase to the right and tandem instability. Brain MRI revealed new lesions. It was assumed a rhombencephalitis in the context of Neuro-Behçet relapse. Started on cyclophosphamide and methylprednisolone with symptoms resolution. Although brainstem involvement in Behçet disease is common, relapses with trigeminal neuropathy are very rare.

**Keywords**

Behçet disease, Neuro-Behçet syndrome, Trigeminal neuropathy, MRI.

## Introduction

Behçet disease (BD) is a chronic multisystem relapsing inflammatory disorder of unknown cause [1]. Neuro-Behçet syndrome (NBS) is defined as the occurrence of neurologic symptoms in a patient with BD that is not better explained by any other well-known systemic or neurologic disease [2,3]. The prevalence of NBS in BS is between 3% and 9% in non-selected large series [4]. The most common neurological presentation is a subacute meningoencephalitis, accounting for 75% of cases in NBS with parenchymal involvement [4]. Typical MRI acute lesions are hyperintense in T2,show enhancement in T1 and are often associated with oedema [5].

## Case

A 49 years old woman with Behçet disease HLA-B51 positive with previous neurological involvement without sequelae and under treatment with azathioprine, was admitted with complains of numbness, aching and periods of electric shock-like pain in the right side of the face. Associated with painful oral ulcers, anorexia, nausea and gait instability. Neurological examination revealed dysesthesia on right trigeminal nerve territory, horizontal-rotatory nistagmus on horizontal gaze bilateral with fast phase to the right and tandem instability. Brain MRI revealed new brainstem lesions (Figure 1). It was assumed a rhombencephalitis in the context of NBS relapse. Treated with intravenous methylprednisolone pulses for 3 days and intravenous cyclophosphamide, with symptoms resolution. Discharged under treatment with oral prednisolone, presenting no further attacks.

## Discussion

According to the International Classification of Headache Disorder [6], our case meets the criteria of painful trigeminal neuropathy attributed to other disorders. In NBS, brainstem relapses are relatively common and although reported in a few cases [7,8], relapses as a trigeminal neuropathy are very rare. Recommended treatment options for acute/sub-acute parenchymal attack is a course

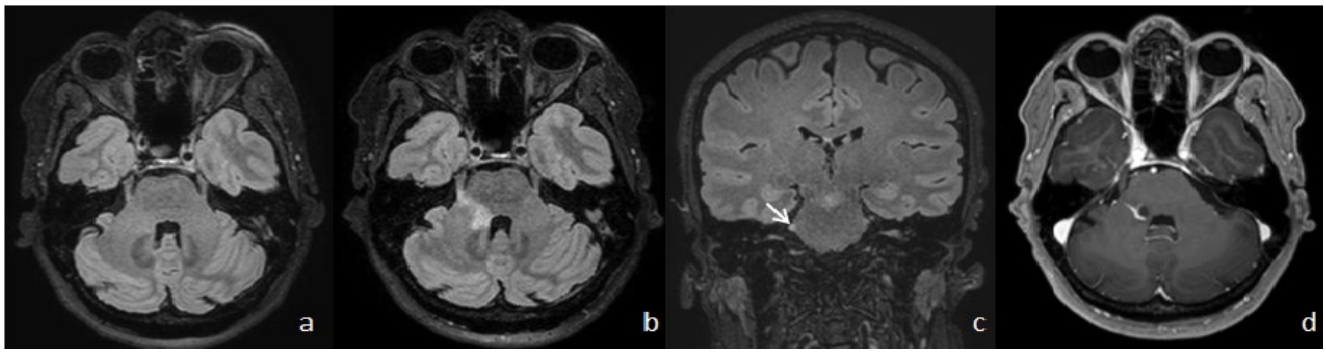


**Figure 1:** Axial 3D T2-weighted fluid-attenuated inversion recovery (FLAIR) images from April (**a**) and November (**b**) show a "de novo" hyperintense lesion in the right posterior pons and right trigeminal root entry zone. 3D T2-weighted FLAIR image (**c**) reveals thickening and hyperintensity in the cisternal segment of the right trigeminal nerve (white arrow). Post-gadolinium 3D T1-weighted turbo field echo (TFE) image (**d**) shows partial peripheral enhancement of the right pontine lesion.

ClinMed International Library

**Citation:** Cação G, Moreira I, Alves JE, Farinha F, Santos E (2016) Trigeminal Neuropathy as A Relapse in Behçet Disease. Int J Neurol Neurother 3:044. doi.org/10.23937/2378-3001/3/2/1044

**Received:** February 25, 2016: **Accepted:** March 26, 2016: **Published:** March 29, 2016

of corticosteroids, preferably intravenous methylprednisolone for 3-10 days followed by a maintenance oral corticosteroid for a few months (up to 6 months) [3]. Regarding disease modifying therapy, azathioprine is recommended as first line, with mycophenolate mofetil, methotrexate and cyclophosphamide being considered as alternatives [3,9]. As far as we know, no reported case of NBS with trigeminal neuropathy presented this perfect clinical and image findings correlation.

## Authors' Conflicts of Interest

The authors declare no conflict of interest.

## References

1. Krause I, Weinberger A (2008) Behçet's disease. Curr Opin Rheumatol 20: 82-87.

2. Peño IC, De las Heras Revilla V, Carbonell BP, Di Capua Sacoto D, Ferrer ME, et al. (2012) Neurobehçet disease: clinical and demographic characteristics. Eur J Neurol 19: 1224-1227.

3. Kalra S, Silman A, Akman-Demir G, Bohlega S, Borhani-Haghighi A, et al. (2014) Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. J Neurol 261: 1662-1676.

4. Saip S, Akman-Demir G, Siva A (2014) Neuro-Behçet syndrome. Handb Clin Neurol 121: 1703-1723.

5. Al-Araji A, Kidd DP (2009) Neuro-Behçet's disease: epidemiology, clinical characteristics, and management. Lancet Neurol 8: 192-204.

6. Headache Classification Committee of the International Headache Society (IHS) (2013) The International Classification of Headache Disorders, 3rd edition (beta version). Cephalalgia 33: 629-808.

7. Farah S, Al-Shubaili A, Montaser A, Hussein JM, Malaviya AN, et al. (1998) Behçet's syndrome: a report of 41 patients with emphasis on neurological manifestations. J Neurol Neurosurg Psychiatry 64: 382-384.

8. Studer U, Ruehe B, Waldegg G, Vajtai I, Escher R, et al. (2012) Atypical presentation of Behcet's disease with central nervous system involvement successfully treated with infliximab. Rheumatol Int 32: 1431-1435.

9. Diri E, Espinoza LR (2006) Neuro-Behçet's syndrome: differential diagnosis and management. Curr Rheumatol Rep 8: 317-322.