



Trigeminal Neuropathy as a Relapse in Behçet Disease

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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.

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

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

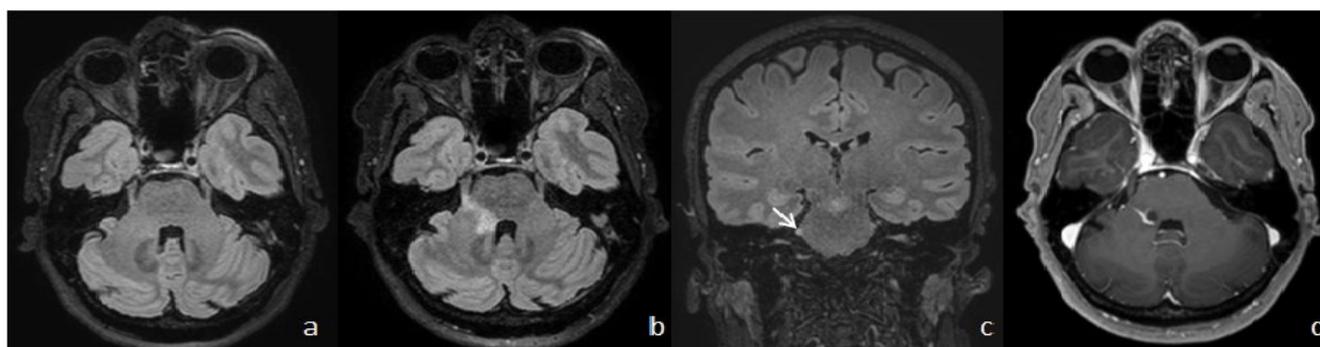


Figure 1: Axial 3D T2-weighted fluid-attenuated inversion recovery (FLAIR) images from April (a) and November (b) show a "de novo" hyperintense lesion in the right posterior pons and right trigeminal root entry zone. 3D T2-weighted FLAIR image (c) reveals thickening and hyperintensity in the cisternal 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.

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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.

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