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Neuro-Behçet as a cause of secondary headache: a case report

Lucas de Oliveira Vieira, Cyro Luis Feitoza Cavalcante, Yann Gusmão Alves Dias Tavares, Renata Nunes Oliveira, Anna Paula Matos de Jesus, Gabriel Ribeiro Leão Barroso, Taimisson Melo Barbosa, Jamyllo Sales Brito, Camilla Souza Fonseca de Lacerda, Wemelly da Costa Silva Barros, João Mário Aguiar Abrantes Dourado, Cárita Victoria Carvalho de Santana, Laysa Raiane de Oliveira Carneiro

Clériston Andrade General Hospital, Feira de Santana, Bahia, Brazil

Lucas de Oliveira Vieira
lucasvieiranet@hotmail.com

Introduction
Secondary headaches arise from diverse underlying pathologies including vascular, traumatic, infectious, and metabolic etiologies. Despite accounting for only 2% to 7% of emergency visits, early diagnosis is imperative due to high morbidity and mortality. The SNNOOP10 mnemonic is essential for identifying red flags such as sudden onset, focal neurological deficits, or late-onset headache. Behçet's syndrome is an inflammatory vasculitis of unknown etiology with multisystemic manifestations, and neurological involvement represents its most severe form, requiring early diagnosis to prevent irreversible sequelae.

Case presentation
A 23-year-old male was admitted with holocranial headache, left lower limb paresis, peripheral facial palsy, and axial ataxia. He had developed analgesic-refractory headache and oral ulceration in the preceding weeks, with a history of recurrent orogenital ulcerations. During hospitalization, his condition worsened with pyramidal signs and urinary incontinence. The pathergy test was positive. Cranial computed tomography revealed a hypodense area in the right cerebral peduncle not conforming to a vascular territory. Magnetic resonance imaging showed T2/FLAIR hyperintense foci in the internal capsules, brainstem, and cerebellar hemispheres, displaying the classic "waterfall sign". Cerebrospinal fluid showed isolated elevated protein. Neuro-Behçet was diagnosed and methylprednisolone pulse therapy was initiated, with gradual motor improvement.

Discussion
Behçet's disease is a chronic autoinflammatory vasculitis mediated by Th17 pathways. Neuro-Behçet occurs in 5% to 10% of cases, preferentially affecting the brainstem in up to 50% of patients. Headache affects over 80% of patients with Behçet's disease, and secondary headache attributed to Neuro-Behçet falls under ICHD-3 criterion 6.4.3 — headache attributed to secondary angiitis of the central nervous system. Differential diagnosis with ischemic stroke is frequent in young patients, and the "waterfall sign" on MRI is highly suggestive of Neuro-Behçet. First-line acute treatment is intravenous methylprednisolone pulse therapy (1 g/day for 3-5 days), followed by immunosuppressive agents according to severity.

Conclusion
Early recognition of secondary headache as a sentinel manifestation is essential for identifying severe conditions such as Neuro-Behçet. The correlation between clinical, laboratory, and neuroimaging findings — particularly the "waterfall sign" — enables confident diagnosis. Immediate pulse therapy promotes significant improvement, whereas therapeutic delay may cause irreversible sequelae. In young patients with headache and red flags, Neuro-Behçet should be included in the differential diagnosis.

Keywords:
Secondary headache
Neuro-Behçet
Red flags
Neuroinflammation
ICHD-3

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