Acute presentation of pseudotumoral lesion in Behçet's disease: a challenging diagnose

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INTRODUCTION

Behçet disease (BD) is a multisystem vasculitis of unclear aetiology charachterized by recurrent oral and genital ulcers, skin lesions and uveitis. Other manifestations include arthritis, trombophlebitis and gastrointestinal ulcerations. Neurological involvement (neuro-Behçet disease NBD) is a very rare condition (1.3- 59 % of cases) with a poor prognosis[1]. Central nervous system involvement includes, in 81% of cases, a parenchymal form manifesting with multifocal infiammatory and necrotic lesions in multiple areas of the basal ganglion region, brainstem or internal capsule and non-parenchymal form caracheterized by cerebral venous thrombosis or arterial aneurysm lesions[2].

The presence of pseudotumoral lesions on brain MRI is very rare in Neuro Behçet's disease (NBD) especially at the onset of the disease[3].

CLINICAL CASE

P. D. ♂ 31 years-old

History of Present Illness:
Acute severe headache and right hemiparesis

Personal and Family history
- Nothing relevant

Past health:
- Nothing relevant

Neurological examination
- horizontal nystagmus
- right facio-brachio-crural hemiparesis
- brisk reflexes throughout all extremities with right prevalence
- positive Hoffman responses and Babinski signs bilaterally

Diagnostic work-up

- **Blood examinations:** E.S.R 49 (n.v. 0 - 15), C.R.P. 1.70 (< 0.5), fibrinogen 537 mg/dl (n.v. 200 - 400)
- **Brain MRI:** on T2 weighted images, an hyper-intense lesion in the bulb and pons extending to involve midbrain, the left cerebral peduncle and the internal capsule (Fig. 1)
- Later the patient reported that he had suffered from oral ulcers and visual impairment
- **Eye examination** revealed uveitis and papillitis on the left eye (Fig. 2)
- **Blood examination:** ANA, ENA, ANCA, nDNA, Ab antiphospolipid and Ab anticardiolipin were normal
- **HLA- typing studies:** presence of HLA- B5 (51)

Neuro-Behcet's disease

- **Treatment: a)** 1 g IV methylprednisolone for five days
  b) Oral steroid (Prednisone 50 mg/die) and Cyclosporine (100 mg/die)
- **Follow-up:** after three mounts the patient showed a marked improvement of his clinical condition

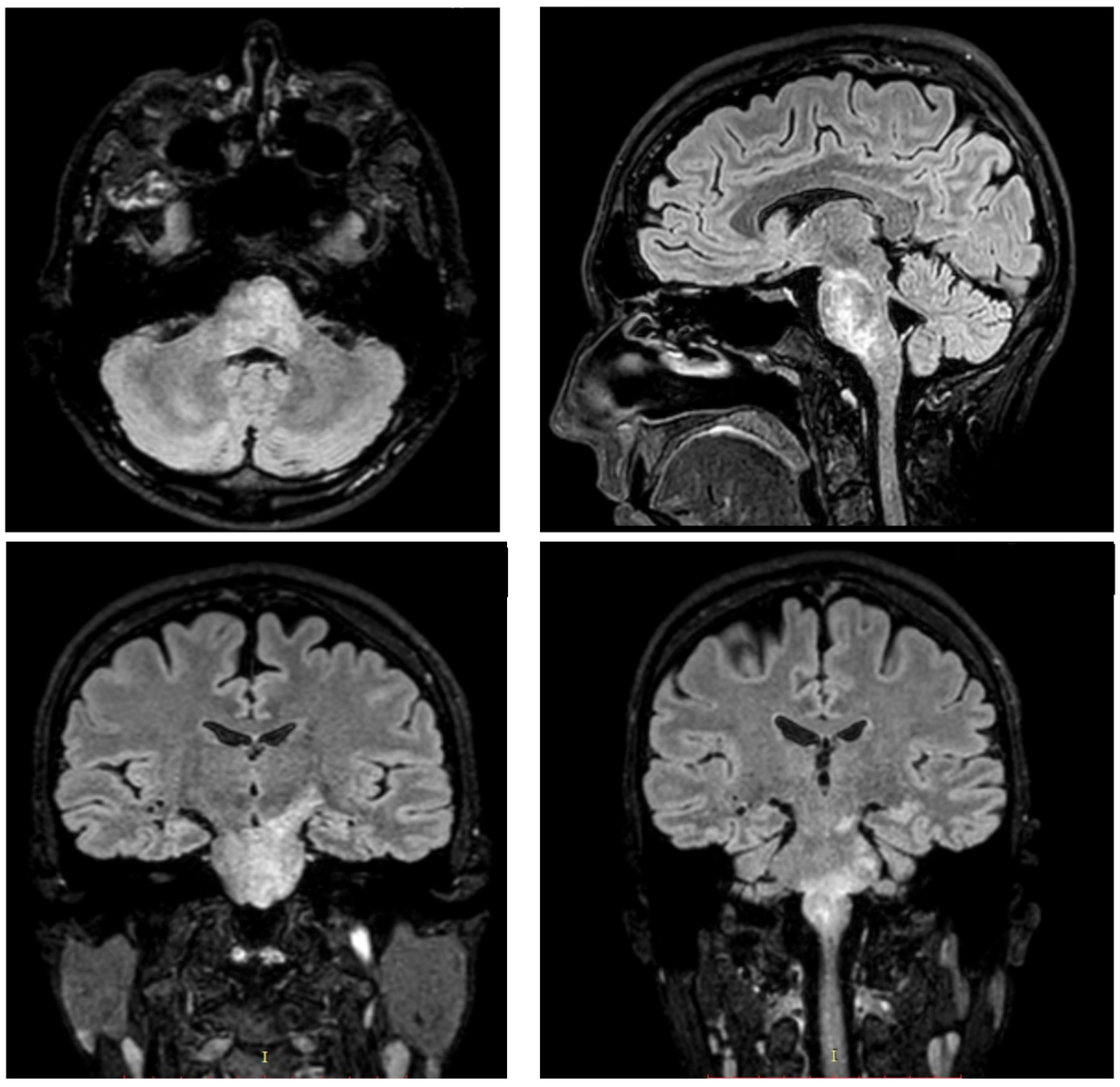

Fig. 1 Brain MRI: FLAIR sequences hyper-intense lesion in the bulb and pons extending to involve midbrain, the left cerebral peduncle and the internal capsule

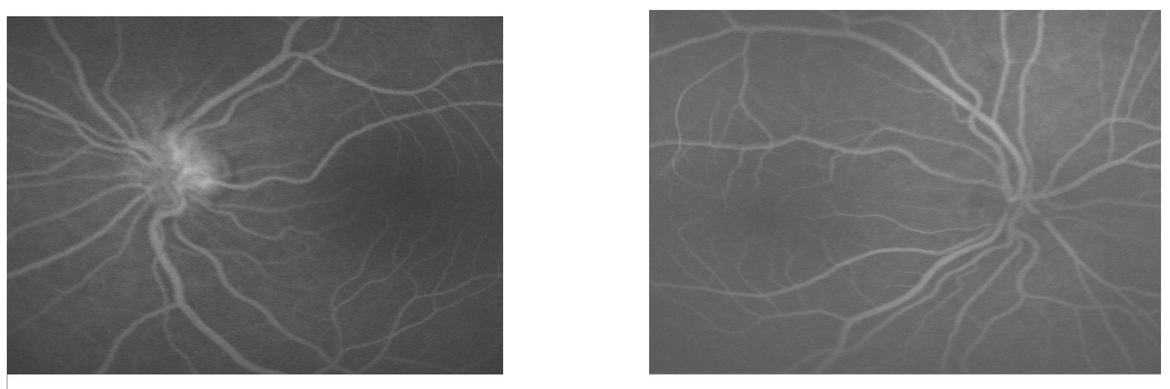

Fig. 2 FAG: papillary and perivascular hyperfluorescence that increases in the late stages

DISCUSSION

This case of neuro-Behcet's disease, manifesting with a pseudotumoral lesions, reflects the need to consider this diagnosis in a young patient who presents with an acute neurological syndrome. Pseudotumoral NBD is a relatively uncommon disorder of the central nervous system. These lesions can mimic stroke, demyelinating and inflammatory diseases of CNS, infections and tumor; for this reason neurologists must consider this diagnosis because appropriate treatment may ameliorate clinical status and prognosis.

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