NEURO-BEHÇET: A CASE OF INTRACRANIAL HYPERTENSION SECONDARY AN INFLAMMATORY DISEASE OF THE BRAIN WITHOUT TROMBOTIC MANIFESTATIONS

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BACKGROUND

Behçet's disease (BD) is an inflammatory multisystem disease. Neuro-Behçet's disease (NBD) has variable prevalence depending on the series but can represent around 5–10% of the affected patients in a large series. The diagnostic criteria for NBD are the current diagnosis of BD plus the presence of neurological symptoms not otherwise explained by known systemic or neurological diseases. In this report, we would like to present a case of NBD with an unusual initial presentation of papilledema and intracranial hypertension and emphasize on NBD as rare but important cause of bilateral papilledema due to intracranial hypertension.

CASE REPORT

FB, 42 years old, male, with a history of oral, nasal ulcers and recurrent genital ulcers for > 2 years associated with inflammatory joint manifestations and pseudofolliculitis. He was diagnosed with BD only in 2020. Prednisone 20 mg/day, colchicine 0.5 mg/day and azathioprine 100 mg/day for 6 months (irregular treatment) with partial response. In 2020, 1 month after a viral infection (COVID-19 with exclusively respiratory manifestations) evolved with mucocutaneous worsening. Followed by severe headache, decreased bilateral progressive visual acuity, visual blurring, nausea and vomiting. Magnetic resonance showed hypersignal on T2 and flair in the supratentorial white matter and trunk. Hydrocephalus with signs of incipient cerebrospinal fluid (CSF) transudation. In the cranial venous and arterial phase computed tomography angiography thrombosis was not evidenced. Cerebrospinal fluid puncture showed high opening pressure, without signs of infection. With other infectious serologies (HIV, syphilis, hepatitis B, C, cytomegalovirus and Epstein–Barr virus) negative. Autoantibodies (anti-dsDNA, ANA, anti-SM, C3 and C4) was also negative. The diagnosis of NBD with intracranial hypertension was made, with pulse therapy with methylprednisolone 1 g/day for 5 days, with induction with cyclophosphamide 0.75 mg and measures for intracranial hypertension. The patient evolved with improved light perception of vision and resolution of cranial hypertension, with normalization of CSF opening pressure. He was discharged with prednisone 1 mg/kg/day and scheduled for another 6 pulses of cyclophosphamide. Currently, the patient maintains visual sequelae, but controlled and stabilized.

CONCLUSION

Neurological involvement occurs in 9% of cases, vascular complications occur in 14%. Intracranial hypertension secondary to inflammatory disease of the brain without thrombotic manifestations is most uncommon. In conclusion, we emphasize the importance of considering NBD in the differential diagnosis when patients present with bilateral papilledema, complaining of visual changes and intracranial hypertension.

KEYWORDS

Neuro-Behçet, Intracranial hypertension, Inflammatory disease.