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Atypical Presentation of Idiopathic Intracranial Hypertension Simulating Demyelinating Disease of the Central Nervous System

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Introduction
Idiopathic intracranial hypertension (iih) is a syndrome of increased intracranial pressure of unclear etiology that most often occurs in obese women of childbearing age, the prevalence ranges between 0.5 and 2 per 100,000 habitants and is expected to increase further given the worldwide increase in obesity. Headache and papilledema are common signs, but asymmetric papilledema in iih is an uncommon finding that can raise concern for alternate diagnoses, such as unilateral optic neuropathy (on), present in demyelinating diseases of the central nervous system (cns) manifesting unilateral or bilateral visual loss, mainly in young women such as iih.

Objective
To highlight the importance of revisiting the diagnosis of atypical cases of iih and especially in cases of asymmetric involvement of optic nerves, we must differentiate optic neuropathy from iih, considering that these conditions have different treatments and early diagnosis can prevent permanent visual loss.

Case Report
Woman, 46-year-old, obese and with previous and unnoticed total visual loss in her right eye for a year, presented to the emergency room with an atypical headache that worsened with exercise. Fundoscopy showed pallor of the right optic disc and edema of the left optic disc, the acuity of which was 20/20. In addition to a lumbar puncture (lp) opening pressure of 36cmh2o and 10 cells on cerebrospinal fluid analysis, she had a visual field test with left peripheral temporal loss. Orbital mri showed faint contrast enhancement in the bilateral optic nerve and nerve sheath, suggesting optic neuritis (on). The anti-aquaporin 4 antibody (aqp-4) was negative. After 2 days, lp was repeated and the opening pressure was 5cmh2o and due to the possibility of inflammatory disease of the optic nerve, 1g methylprednisolone was administered for 5 days. After that, the patient's symptoms improved. During outpatient follow-up, low-dose azathioprine and prednisone were started and, although the patient used the medications irregularly, her disease stabilized until three years later, when she began to complain of transient episodes of visual loss in her left eye. Ophthalmological evaluation demonstrated concentric visual loss and edema of the left optic disc. New cranial and orbital mri showed signs of intracranial hypertension, previously absent. A new lp revealed an opening pressure of 41cmh2o. Another aqp-4 test using the cba method was again negative. Then, four years after the first symptom, the patient was diagnosed with optic neuropathy caused by iih.

Conclusion
In our case, the patient was admitted with previous unilateral visual loss and progression to involvement of the other optic nerve. The first mri showed signs of inflammation in the optic nerve without signs of intracranial hypertension. Both lp and immunosuppressive treatment were instituted simultaneously. It was not possible to differentiate which of the two the patient developed a response to and it was subsequently decided to continue continuous treatment of the inflammatory condition. Until the recurrence of symptoms and imaging findings, visual field with concentric visual loss and persistence of intracranial hypertension years later revealed the diagnosis of iih. The clinical similarities between iih and inflammatory on, such as vision loss and the occasional presence of oligoclonal bands in csf, have brought to light the notion that antiglial antibodies associated with neuromyelitis optica may also participate in the pathogenesis of iih. Thus, some studies have focused on looking for the presence of antibodies aqp-4 or against myelin oligodendrocyte glycoprotein (anti-mog) in patients with iih. however, none of the studies showed these antibodies in patients with iih. medical reports in which these conditions have occurred concomitantly are rare and the phenotypic polymorphism of both conditions makes differentiating between these conditions a diagnostic challenge.

Keywords: Idiopathic Intracranial Hypertension; Demyelinating Disease; Differential Diagnosis.