Short communication

Optic neuropathy in a case of recurrent idiopathic hypertrophic pachymeningitis unresponsive to steroids and immunosuppressants

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\textbf{A B S T R A C T}

Case report: A 38-year-old female patient with bilateral papilledema who presented with loss of vision in her left eye. The Magnetic Resonance Imaging (MRI) showed thickening of the dura mater, and the intracranial pressure was elevated. A cancer, infectious origin, and autoimmune origin were ruled out.

Discussion: The initial response to high doses of corticoids was satisfactory, with disappearance of the optic disk edema, with visual acuity and an improvement in the MRI. However, after one year without treatment she had a new outbreak of the disease. Despite renewed treatment with corticoids and azathioprine, the patient developed a left optic neuropathy and irreversible visual loss.

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Neuropatía óptica en un caso de paquimeningitis hipertrófica idiopática recurrente sin respuesta a esteroides e inmunosupresores

\textbf{R E S U M E N}

Caso clínico: Mujer de 38 años con pérdida visual en ojo izquierdo y papiledema bilateral. La resonancia magnética nuclear (RMN) mostraba engrosamiento de la duramadre y la presión intracraneal estaba elevada. Se descartó enfermedad infecciosa, tumoral y autoinmune.

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**Introduction**

Hypertrophic pachymeningitis is an uncommon condition characterized by inflammatory thickening of the dura mater or less frequently the spinal cord.\(^2\)\(^3\) It has been described in association with infections, trauma, tumors, self-immunity and rheumatological diseases. Idiopathic cases are diagnosed by exclusion.

**Clinic case**

Female, 38, with diminished visual acuity for years with diminished visual acuity (VA) in the left eye (LE) down to hand movements together with retro-ocular pain with one week evolution. Relative afferent pupil defect was observed in LE and bilateral papillary diffusion, with VA=1 in the right eye (RE).

Brain nuclear magnetic resonance (NMR) revealed diffuse thickening of dura, more acute around the left optic nerve (ON) (Fig. 1A). Suspecting neurosyphilis due to neonatal congenital syphilis, empirical treatment was established with high dose of corticoids (methylprednisolone 1 g per day during 3 days, followed by oral prednisone 1 mg/kg/day) associated to intravenous penicillin. Cerebrospinal fluid (CSF) exhibited high opening pressure (27 cm H\(_2\)O), while the rest of the parameters were normal. All the following tests VDRL in CSF, TPHA, serology (Brucella, rubella, herpes, cytomegalovirus, smallpox, syphilis) and testing of autoantibodies (ANCA, ANA, FR) were negative. In the absence of concomitant disease the diagnostic was idiopathic hypertrophic pachymeningitis. Accordingly, antibiotic treatment was suspended.

After 10 days of treatment a significant reduction of the thickness was observed (Fig. 1B) with a recovery of VA in the LE up to 0.6. The patient remains stable for 18 months with low dose of oral prednisone (10 mg/day), which was gradually withdrawn.

One year after termination of said corticoids treatment the patient exhibited relapse with progressive loss of vision in LE to finger counting with associated headache. In the RE, VA remained at 1. Inferior diffusion of right papilla and temporal paleness of the left papilla was observed, with right retinal edema in optic coherence tomography (Stratus OCT, Carl Zeiss Meditec) and diminished thickness of the peripapillary retinal nerve fiber layer in LE (Fig. 2). NMR revealed a new nodular increase in the meninges which compressed the left ON (Fig. 1C). Prednisone and oral azathioprine treatment were established (both at 50 mg/day) with resolution of the right edema (Fig. 2) and improvement of VA in LE up to 0.5. However, the condition evolved to left optic atrophy (Fig. 3). Two

**Fig. 1** Brain nuclear magnetic resonance (NMR): changes during follow-up: (A) NMR during debut, showing diffuse dura thickening with increased linear pattern at the temporal level and left lower tentorial as well as around the left optic nerve. No space-occupying lesions were observed and tumor disease was discarded. Thickened dura was isointense in T1 sequences and hypointense in T2, with intense contrast after administering gadolinium. Pia mater and arachnoid were respected. (B) Significant reduction of meningeal thickening 10 days after corticoid treatment (methylprednisolone pulses 1 g per day during 3 days, followed by oral prednisone at a dose of 1 mg/kg/day), with minimum dural contrast at the frontotemporal and tentorial level and adjacent to the left optic nerve. (C) Recurrence after one year without corticoid treatment. New nodular meningeal thickening in anterior region of sellar diaphragm which compresses the left optic nerve causing optical atrophy. Culture and VDRL in CSF were again negative.
years after recurrence, the patient exhibits central defect in automated perimetry (Octopus, Haag-Streit) in LE (Fig. 4), with hands movement VA and preserved VA in RE.

**Discussion**

At the clinical level, the most frequent expressions of this disease are headache, cranial pair paresis (including oculomotor nerves with diplopia and extraocular motility alterations), ataxia and optic nerve involvement.\(^1^\)\(^2\) Even though the patient of this case exhibited intracranial hypertension, this expression is less frequent.\(^1\)\(^2\)

Papilla edema can appear due to compression by inflamed meninges, to intracranial hypertension or both. The patient of this case exhibited unilateral VA loss with bilateral edema, probably due to the asymmetric involvement with greater increase of meninges around the left ON. Evoked visual potentials revealed significant dysfunction in the LE, correlated to visual loss.

Brain NMR is the most utilized neuroimaging for diagnostic purposes. It is characterized by diffuse or zonal thickening of dura with linear or nodular pattern.\(^1\)\(^2\) It can appear simultaneously or sequentially, as in the present case. In addition, it generally exhibits clinical–radiological correlation which is useful for assessing response to treatment.\(^1\)

In idiopathic cases, CSF analysis is negative for tumoral cells and infection. Lymphocytosis and increased proteins have been described\(^1\)\(^4\) although in most cases the values are

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**Fig. 2** – Optic coherence tomography (OCT) in recurrence after one year without corticoid treatment (temporal domain OCT Stratus, signal quality 6/10). (A) Papilla edema in inferior sector and diminished retina nervous fiber layer (RNFL) in left eye. Diminished RNFL in temporal sector of left eye. (B) Tomograph after combined treatment with oral corticoids and azathioprine. Resolution of right eye papilla edema and discrete RNFL thickness reduction in right eye with preserved VA of 1. Left optical atrophy despite treatment.
within normality. The opening pressure is high with associated intracranial hypertension.

Meninges biopsy provides certainty diagnosis and is indicated in the presence of NMR worsening or clinical deterioration despite the treatment. It shows a chronic inflammatory infiltrate with lymphocytes, plasmatic cells and histiocytes with occasional fibrosis. The patient declined biopsy after recurrence.

Even though some cases of spontaneous resolution have been described, corticoid therapy is the core treatment as it produces clinical and radiological improvement. Response varies depending on the thickening pattern, being faster and sustained in linear involvement (initial episode) and generally insufficient in nodular pattern (recurrence), suggesting associated fibrotic reaction. Recurrences may occasionally appear when reducing corticoids dosage, making it necessary to add immunosuppressants (methotrexate, azathioprine), generally with good response.

In conclusion, despite its low incidence, ophthalmologists should be familiar with the condition discussed above as its debut is not infrequent with neuro-ophthalmological symptoms. NMR is the imaging test featuring the highest sensitivity, making it useful to assess clinical and radiological response to corticoid treatment. Immunosuppressants are applied to spare the use of corticoid or in the presence of recurrences. The peculiarity of the case discussed herein is the lack of response to combined therapy with prednisone and azathioprine, producing left optical atrophy with irreversible visual loss.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES