HYPERTROPHIC PACHYMENINGITIS AND
OPHTHALMOLOGICAL DISTURBANCES: DESCRIPTION
OF TWO CASE REPORTS

PAQUIMENINGITIS HIPERTRÓFICA Y ALTERACIONES
OFTALMOLÓGICAS: DESCRIPCIÓN DE DOS CASOS CLÍNICOS

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ABSTRACT

Case reports: We report two cases of hypertrophic pachymeningitis with ophthalmological disturbances. One patient suffered from hypertrophic pachymeningitis supposedly secondary to a focal neurosarcoidosis and had a sixth nerve paresis. The second patient suffered from a diffuse idiopathic hypertrophic chronic pachymeningitis (a rare form) and displayed disturbances in vision and ocular motility.

Discussion: Hypertrophic pachymeningitis is a condition with a wide spectrum of etiologies and clinical manifestations, and needs to be considered as the cause in patients with alterations in ocular motility (Arch Soc Esp Oftalmol 2008; 83: 497-500).

Key words: Hypertrophic pachymeningitis, neurosarcoidosis, retrobulbar optic neuritis, sixth nerve paralysis, idiopathic hypertrophic chronic pachymeningitis.

RESUMEN

Caso clínico: Presentamos dos casos de paquimeningitis hipertrófica con alteraciones oftalmológicas asociadas. Uno de los pacientes presentó paquimeningitis hipertrófica presuntamente secundaria a neurosarcoidosis focal y paresia del sexto par. La segunda paciente presentó paquimeningitis hipertrófica crónica idiopática (tipo poco frecuente) difusa con alteraciones de la motilidad ocular y en la visión.

Discusión: La paquimeningitis hipertrófica es una patología con un amplio espectro de etiologías y manifestaciones clínicas, que debe ser tenida en cuenta en el diagnóstico de alteraciones de la motilidad ocular.

Palabras clave: Paquimeningitis hipertrófica, neurosarcoidosis, neuritis óptica retrobulbar, parálisis del sexto par craneal, paquimeningitis hipertrófica crónica.

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INTRODUCTION

Hypertrophic pachymeningitis is a condition derived from the thickening of the dura mater due to acute or chronic inflammation (1), infection, neoplasia or self-immune disorder, which causes neurological alterations due to compression of adjacent structures. It may appear as a neurological deficit in the ophthalmological domain.

CASE REPORTS

Case report 1

A 59-year old male who attended the practice due to horizontal binocular diplopia combined with headache and gait instability. His personal history comprised non-insulin dependent diabetes mellitus with several years of evolution, high blood pressure and hypercholesterolemia. Upon exploration a right eye abduction limitation was observed. The red-maddox exploration revealed an external base of 8 horizontal dioptres in near vision and an external base of 12 prismatic dioptres for far vision. A nuclear magnetic resonance (NMR) with contrast was made, which identified a meningeal thickening of the right side tentorium and petrous and tribal regions, consistent with tentorium hypertrophic pachymeningitis (fig. 1). The high numbers for angiotensin converting enzyme (ACE) both in serum and in cerebrospinal fluid gave rise to a suspected neurosarcoidosis. A systemic corticoid treatment was established. The chest X-ray and CAT (Computerized Axial Tomography) as well as the lung function test, the nasal endoscopic exploration, body scan with gallium, muscular NMR and SPECT (single photon computerized tomography) yielded normal results.

From the ophthalmological viewpoint, after a press-on test prisms with an external base of 5 prismatic dioptres to the usual optic correction were prescribed. With time, the patient exhibited recurring bilateral paresia episodes of paresis of the Sixth pair (7 and 10 months after the clinical symptoms began) as well as isolated episodes of paralysis of the Seventh and Eighth homolateral cranial pairs (14 months after the beginning of clinical symptoms). At present, the patient is in treatment with immunosuppressants (azathioprine 1.5 mg/kg/day) with satisfactory control of symptoms.

Case 2

A 37-year old female who visited the emergency ward due to headache and horizontal diplopia. Her personal history did not include relevant antecedents. Upon exploration a limitation in the right eye abduction was identified, with a horizontal external base of 14 dioptres both in near and far vision in red-maddox. A contrast MNR was made which revealed the presence of a diffuse thickening of the dura mater (fig. 2). All the analyses were within normal limits, discarding systemic pathologies which involve dura mater thickening such as neoplasia (particularly of hematologic origin), acute infectious or chronic diseases (such as tuberculosis) and granulomatous diseases. The case was defined as idiopathic hypertrophic pachymeningitis. In evolution, the patient exhibited paresia of the Sixth
contralateral pair. The condition improved with oral systemic corticosteroid treatment. Three months later, together with the corticoid reduction pattern, the patient exhibited visual acuity reduction (0.2) with afferent right eye relative pupil defect. The ophthalmoscopic observation of the optic disc was normal. A Humphrey SITA 24-2 visual field assessment was made which revealed a cecal-central defect in the right eye involving fixation. A new imaging study did not show changes. The diagnosis was retrobulbar optic neuritis, establishing treatment with large dosages of intravenous steroids (1 mg of methylprednisolone for 5 days) followed by an oral descending pattern plus azathioprine as immunosuppressant therapy. The patient remains free of symptoms to this date.

**DISCUSSION**

Hypertrophic pachymeningitis causes a thickening of the dura mater which may have different etiologies (neoplasia, infection or self-immune conditions).

The case 1 patient exhibited a localized form of hypertrophic pachymeningitis. Considering the high rates of ECA found, together with a compatible clinical case, the possible diagnostic of neurosarcoidosis was discussed (2). However, the presence of granulomas could not be demonstrated or systemic sarcoidosis in this patient (meningeal biopsy was not made due to the patient’s non-consent).

Neurological alterations occur in 5% of sarcoidosis patients. In half of all cases, neurological alterations are the first expression of sarcoidosis. However, it is not frequent that neurosarcoidosis is the only expression of said condition (2). Clinical paralysis of cranial pairs occurs in up to 75% of patients.

In some cases there is no explanation for said condition, and the category of idiopathic hypertrophic pachymeningitis is adopted, an infrequent exclusion condition (3). The highest prevalence of this condition is in the seventh decade of life (1).

The clinical expressions are headache, cranial pairs paralysis and ataxia (1). Our patient exhibited a retrobulbar optic neuropathy.

According to some authors, the diagnostic can be made by magnetic resonance in the absence of concomitant pathologies explaining said pachymeningitis and on the bases of a favorable response to corticoids, even though the test of choice continues to be meningeal tissue biopsy (4). However, this was not done because the patient did not consent.

In the case of idiopathic forms, the treatment of choice is corticosteroids, with pharmacological immune suppression as alternative treatment so as to reduce corticoid use and in relapses of the condition.

The literature includes references to subcutaneous injections of methotrexate as a therapeutic tool for treating hypertrophic meningitis (12.5 mg per week for 6 months, followed by 6.25 mg subcutaneous for 4 months and 2.5 mg orally a further 2 months) in a single patient with good results (5).

In summary, hypertrophic meningitis is a neurological condition which may appear with symptoms of neurological deficit in the ophthalmological area. Accordingly, the ophthalmologist must be knowledgeable about said condition.
REFERENCES


