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CASE REPORT

Idiopathic Hypertrophic Cranial Pachymeningitis Associated With Intermediate Uveitis

Sevil Ari Yaylali,[1] Aylin Ardagil Akcakaya,[1] Nihal Işik,[2] Hasan Hasbi Erbil,[1] Ali Olgun,[1] Zeki Aslan,[1] and Tulay Kansu[3]

Departments of [1]Ophthalmology and [2]Neurology, Istanbul Goztepe Educational Hospital, Istanbul, Turkey, and [3]Department of Neurology, Hacettepe University Medical Faculty, Ankara, Turkey

ABSTRACT

The authors report a case with idiopathic hypertrophic cranial pachymeningitis associated with intermediate uveitis. The patient complained of decreased vision in both eyes, especially the right. Ophthalmic examination revealed right optic disc pallor, bilateral vitritis, and cystoid macular edema. Magnetic resonance imaging revealed marked enhancement of a dural lesion. The macular edema responded well to medical treatment. Intermediate uveitis has not yet been reported in the context of idiopathic hypertrophic cranial pachymeningitis.

KEYWORDS cystoid macular oedema, idiopathic hypertrophic cranial pachymeningitis, intermediate uveitis

INTRODUCTION

Hypertrophic cranial pachymeningitis is a clinical disorder due to localised or diffuse thickening of the dura mater, with or without an associated inflammation.[1] Malignancy and infectious or autoimmune disorders such as tuberculosis, syphilis, and human T-lymphotropic virus type I (HTLV-I), Lyme, rheumatoid arthritis, Wegener granulomatosis, and Behçet disease are recognised causes.[2–4] When the evaluation fails to reveal a cause, a diagnosis of idiopathic hypertrophic cranial pachymeningitis (IHCP) is made.

The term intermediate uveitis is used for patients with intraocular inflammation predominantly involving the vitreous and peripheral retina as suggested by International Uveitis Study Group.[5] Intermediate uveitis may exist as an isolated idiopathic disorder, but it has also been associated with inflammatory disorders such as sarcoidosis and multiple sclerosis, and infectious diseases such as Lyme disease, HTLV-I, and hepatitis C infection.[6,7]

In this report we describe a patient with IHCP who developed intermediate uveitis.

CASE REPORT

A 24-year-old male was referred to our clinic in April 2008 with bilaterally decreased vision. He first presented in 2001 with headache and diplopia due to a 6th cranial nerve palsy. A gadolinium-enhanced magnetic resonance imaging (MRI) scan at that time revealed thickening of the left posteromedial temporal dura and tentorium cerebelli. The following investigations were performed to rule out infectious, granulomatous inflammatory causes and neoplastic causes (i.e., tuberculosis, sarcoidosis, lymphoma, metastasis) and were found to be normal or negative: chest X-ray; computerised tomographic (CT) chest scan; serum angiotensin I-converting enzyme; cerebrospinal fluid composition (protein, glucose, angiotensin I–converting enzyme, cultures and stains for bacteria, tuberculosis, and fungi); transbronchial biopsy; and bronchoalveolar lavage. A meningeal biopsy was attempted but the tissue sample was inadequate and could not be evaluated. The patient declined a second biopsy. A diagnosis of idiopathic hypertrophic pachymeningitis was made. Despite clinical improvement after pulse steroid treatment followed by oral steroid

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Correspondence: Sevil Ari Yaylali, MD, Department of Ophthalmology, Istanbul Goztepe Educational Hospital, Istanbul, Turkey. E-mail: sevil.yaylali@gmail.com

and azathioprine, recurrences occurred resulting in a left 3rd cranial nerve palsy in February 2002 and a right optic neuropathy in January 2004.

The best corrected visual acuity evaluated at admission with early treatment diabetic retinopathy study (ETDRS) chart at 4 m was 15 letters in the right eye and 45 letters in the left eye. Anterior segment examination was unremarkable. Funduscopic examination showed right optic disc pallor, bilateral macular oedema, and 1+ vitritis. Fluorescein angiography (FA) revealed cystoid pattern of macular oedema and peripheral vascular leakage (Figure 1a–c). Humphrey perimetry revealed a central scotoma, enlargement of the blind spot and a peripheral scotoma in the right eye (Figure 2a). The left visual field was normal.

There was no restriction of the ocular movements. Contrast-enhanced MRI demonstrated a lesion extending from the right cavernous sinus to the prechiasmatic part of right optic nerve (Figure 3a) and residual enhancement of dura previously involved in 2001. The total white blood cell count was 7400 mm^3, erythrocyte sedimentation rate was 49 mm in the first hour. Extensive testing for autoimmune antibodies associated with immunological disorders (i.e., Wegener disease, Sjögren syndrome: perinuclear antinuclear cytoplasmic antibody [p-ANCA], cytoplasmic ANCA [c-ANCA], antinuclear antibody [ANA], anti-Ro [SS-A], anti-La [SS-B]) did not reveal any abnormality. Serologic tests for syphilis (venereal disease research laboratory [VDRL] test, fluorescent treponemal antibody absorbed [FTA-ABS] test), rheumatoid factor, hepatitis C markers, and pathergy test were negative. The patient had been taking 125 mg of oral azathioprine per day and 15 mg of deflazacort every other day.

The MRI findings were interpreted as indicating a recurrence of the pachymeningitis and the dose of deflazacort was increased to 75 mg daily. One week later the visual acuity increased to 50 letters in the right eye and 53 letters in the left eye. Humphrey perimetry performed 2 weeks later revealed partial regression of the scotomas (Figure 2b). Examination at 1 month revealed resolution in the macular oedema and decrease of the peripheral vascular leakage on FA (Figure 1d–f). The deflazacort daily dose was tapered gradually. There was no relapse of the macular oedema over a 2-year follow-up. The last MRI performed in July 2010 showed residual enhancement of the left posteromedial temporal dura, tentorium cerebelli (Figure 3b), and the right cavernous sinus lesion.

DISCUSSION

The most common symptoms of ICHP are headache and cranial nerve palsies. The first presentation of the disease in our patient was a headache and left eye limitation of abduction due to 6th nerve palsy in 2001. Despite the clinical improvement after pulse steroid treatment followed by oral steroid and azathioprine recurrences occurred. Riku and Kato described recurrences or progression of the disease in their patients in spite of the treatment and also a long-term improvement in one of the patients receiving combined therapy with steroid and azathioprine.[8]

Visual loss in patients with pachymeningitis due to optic nerve involvement has been reported.[8–10] The MRI of our patient performed in 2008 revealed a lesion involving the prechiasmatic part of the right optic nerve and this was probably the cause of the visual field abnormalities in the right eye. Nerve encasement and ischaemic changes caused by thickening of the dura are thought to be the probable pathogenesis for

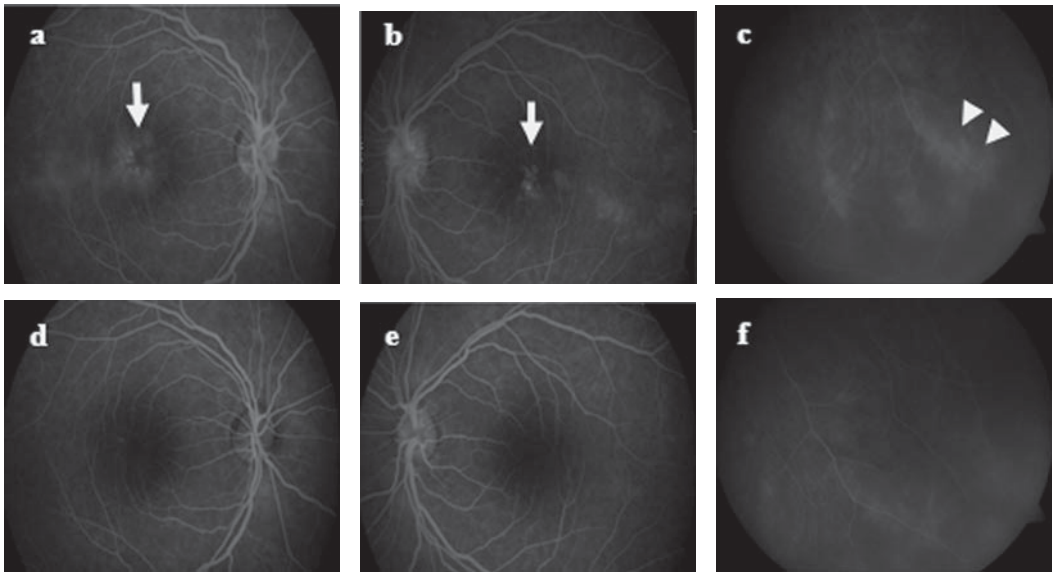

FIGURE 1 Fluorescein angiography images show cystoid macular oedema (arrows) and peripheral vascular leakage (arrowheads) (a, b, c). One month later images show resolution of macular oedema and a decrease in the peripheral vascular leakage (d, e, f).

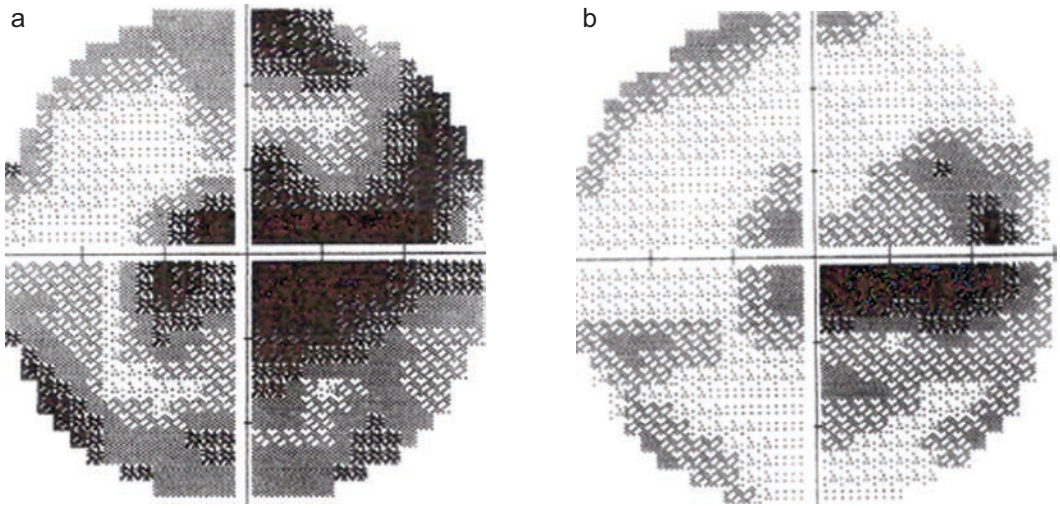

FIGURE 2 Humphrey visual field test shows central scotoma, enlargement of the blind spot, and peripheral scotoma in the right eye (a). The test performed 2 weeks later showed partial regression of scotomas (b).

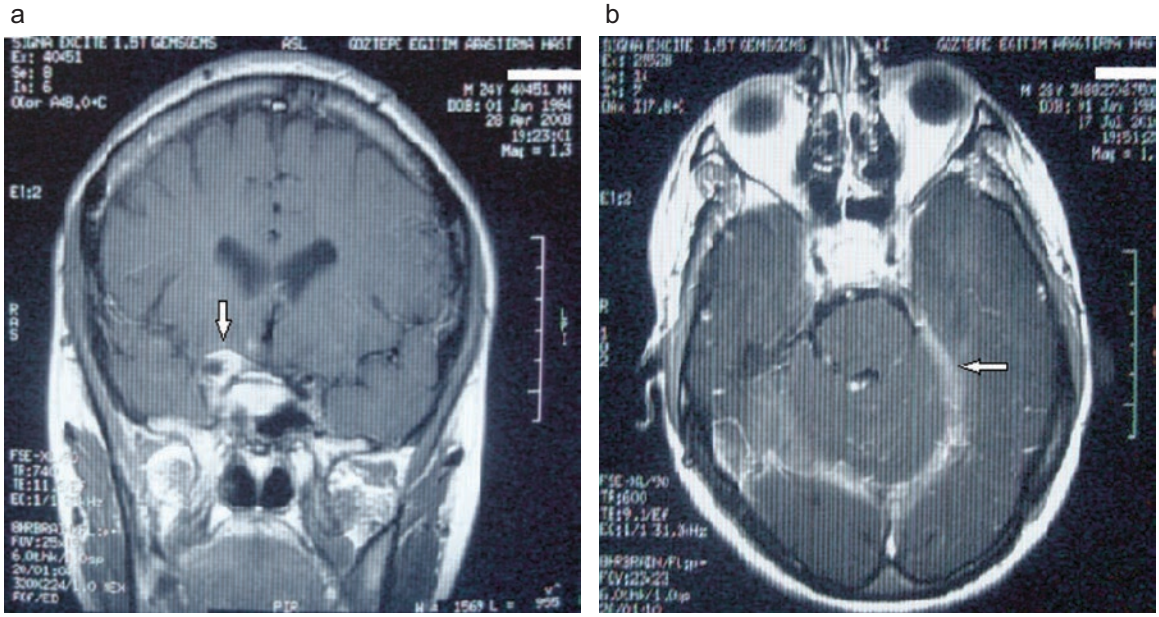

FIGURE 3 Coronal post-gadolinium image performed in April 2008 shows the lesion extending from right cavernous sinus to the prechiasmatic part of right optic nerve (a, arrow). Axial post-gadolinium image of the brain 9 years after diagnosis shows residual thickening of the tentorium cerebelli (on prednisone and azathioprine therapy) (b, arrow).

this damage and other cranial neuropathies caused by IHCP. In addition to optic nerve involvement, cystoid macular oedema associated with intermediate uveitis was responsible for the visual loss in our patient. Focal vascular endothelial decompensation caused by inflammation may be responsible for cystoid oedema and also peripheral vascular leakage.[11]

Sarcoidosis, as one of the diseases associated with intermediate uveitis, may be a possible diagnosis in our patient. Despite repeatedly performed tests such as serum angiotensin-converting enzyme (ACE) levels and chest CT scans, it is extremely difficult to exclude neurosarcoidosis without meningeal biopsy. However, during the 9-year follow-up period, no systemic symptoms or findings related to sarcoidosis were observed. Autoimmune mechanisms have been postulated in the pathogenesis of hypertrophic pachymeningitis as well as intermediate uveitis.[12] Episcleritis[9] and orbital pseudotumor,[13] which have both been reported in the patients with idiopathic hypertrophic pachymeningitis, are also often associated with autoimmune diseases. In our case, no cause was found for intermediate uveitis or pachymeningitis. We hypothesise that common autoimmune or other mechanisms were responsible for their presence in our patient. The presence of two reports in literature that describe presentation of uveitis with ICHP support our hypothesis. However, there was no information about the type of uveitis in one of the cases and the second described panuveitis.[14,15] In conclusion, intermediate uveitis

associated cystoid macular oedema may be the reason for the visual loss in patients with idiopathic hypertrophic pachymeningitis: such oedema responds well to steroid treatment.

***Declaration of interest*:** The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper. The authors have no proprietary or commercial interest in any materials discussed in this article.

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