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Case report

Optic perineuritis: A further cause of visual loss and disc edema in children

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Abstract

Background: Optic perineuritis is a rare form of orbital inflammatory pseudotumor in which the specific target tissue is the optic nerve sheath. Patients are mainly represented by adult women. Differential diagnosis with demyelinating optic neuritis is essential in terms of prognosis and treatment. *Case presentation:* An 8-year-old Caucasian girl presented with bilateral loss of vision, disc edema, eye movement impairment, and diplopia. Brain MRI findings were suggestive of optic perineuritis. The patient received steroid pulse therapy followed by prolonged course of oral steroid therapy. The visual acuity recovered dramatically within 2 days. Two months later, a new MRI investigation was normal. No clinical relapse was observed at the follow-up. *Discussion:* We first report on a child affected by optic perineuritis. Our observation suggests that optic perineurits should be considered in the differential diagnosis of children presenting with visual loss and disc edema. An early and correct diagnosis may lead to an appropriate therapeutic approach with very good outcome.

Keywords: Optic neuritis; Demyelinating disease; Diplopia; Pediatrics; Steroids; Optic nerve

1. Introduction

Optic perineuritis (OPN) is an uncommon disorder first recognized by Edmunds and Lawford [1] in 1883. It is now considered a type of orbital inflammatory pseudotumor [2–4], in which the specific target tissue is the optic nerve sheath. The inflammatory infiltrate is loosely organized around the optic nerve which explains why in many cases of OPN the nerve function has been reported to be normal [4]. OPN is occasionally preceded by viral upper respiratory tract infections. Specific etiologies include syphilis, Herpes zoster, sarcoidosis, tuberculosis, giant cell arteritis, and Wegener's granulomatosis [5]. OPN has also been reported to be associated with disorders like acute leukemia [6] and Crohn disease [7]. However, the majority of cases are idiopathic as no specific inflammatory disorders are identified. OPN shows variable clinical profile and uncertain visual prognosis [4,5,8]. It should be adequately differentiated from a distinct disorder such as demyelinating optic neuritis (ON) [2]. The aim of this report is to increase awareness of such entity describing a young girl presenting with OPN.

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2. Case report

An 8-year-old girl, born to non-consanguineous, healthy parents, presented with no family history of ophthalmologic, neurological, or autoimmune disorders. Her previous medical history was uneventful and she did not take any medication. She first came under our care at the age of 7 years because of difficulties in visual task. Two weeks earlier she had an episode with fever, headache and vomiting. On admission the child appeared wakeful and oriented. A physical examination showed normal cardiac, abdominal and lung objectivity. Ophthalmological examination revealed severe visual loss (visus 1/10 bilaterally), relative afferent pupillary defect, and eye movement impairment, bilaterally. The child complained mild dyschromatopsia, bilateral ocular discomfort, and diplopia. Fundoscopy revealed bilateral disc edema. The remaining of the neurological examination was normal. Brain MRI showed a high signal intensity area around the optic nerves with mild flattening of the posterior pole of ocular globes. Optic nerve sheaths were abnormally increased in size with tortuosity of their course. There was no enhancement of optic nerve and meningeal sheaths after gadolinium administration. All these MRI features were congruent with inflammation around the optic nerve sheaths (Fig. 1). Polymerase chain reactions on the cerebrospinal fluid for Enterovirus, Herpes simplex virus type 1 and 2, Herpes virus hominis 6 e 7, Adenovirus, Cytomegalovirus, Epstain Barr virus, Varicella Zoster virus, and Influenza virus types A and B, were negative. Serology was negative for CMV, EBV, Mycoplasma pneumoniae, Borrelia Burgdorferi, and Toxoplasma Gondi. No infectious agents were detected in blood, urine and throat swab. Electrolytes, liver and kidney functions were within normal limits. Visual evoked response testing showed a mild increase of P_{100} latency and mild reduction of P_{100} amplitude. The patient was placed under steroid pulse therapy consisting of methylprednisolone intravenously (20 mg/Kg/day for 5 days) followed by oral administration of prednisone 1.5 mg/Kg/day for 1 month. Clinical findings dramatically improved. Diplopia and dyschromatopsia disappeared soon after 2 days of therapy. A fully recovery of visual acuity was noted after 5 days. Two months later, a new brain MRI investigation was normal (Fig. 2). Fundoscopic and ophthalmological examinations remained normal during follow-up.

3. Discussion

OPN is a disorder clinically characterized by typical visual field signs, pain with eye movements and normal or swollen optic disc [5]. Although clinical overlapping with ON there exist, some clinical features may help to distinguish between these entities [4,5]. Age at onset of OPN is broader than ON. When globally considered patients with OPN are mainly represented by women older than 50 years of age [8]. Visual loss is often paracentral or arcuate in OPN, while it is usually central in ON. Clinical progression in OPN might be slower (over weeks) than in ON.

To the best of our knowledge, we describe the first pediatric patient presenting with OPN reported so far. The child showed visual loss associated with disc edema and mild dyschromatopsia mimicking ON. As in previous reports [9,10], visual loss was bilateral. The patient

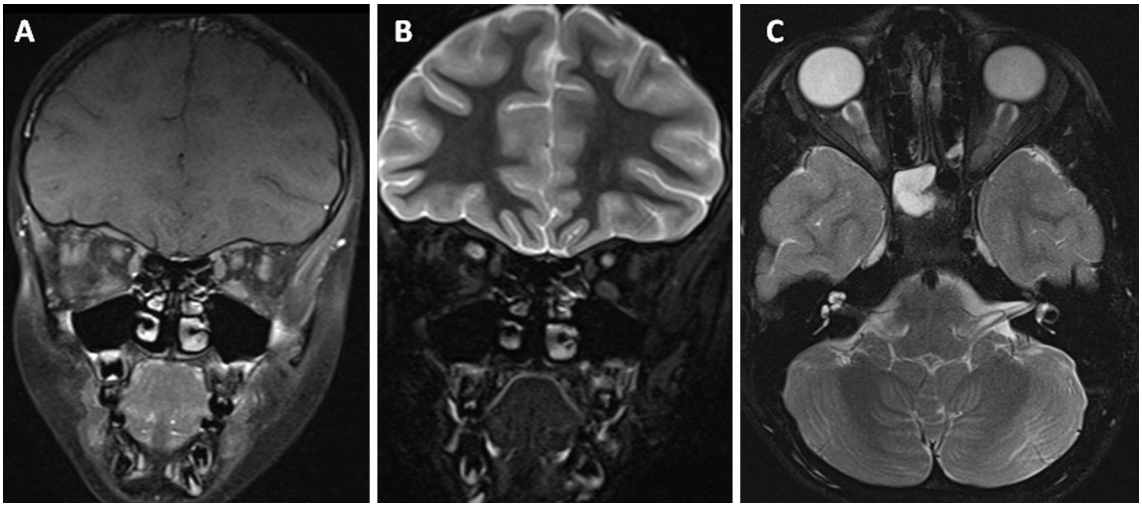

Fig. 1. (A) Post-contrast fat-suppressed coronal T1-weighted image. There is no enhancement of optic nerve and meningeal sheaths after gadolinium administration. Optic nerve sheath appears increased in size, bilaterally. (B) Fat-suppressed coronal T2-weighted image demonstrates a high signal intensity area around the optic nerve and confirms that optic nerve sheath is abnormally increased in size, bilaterally. (C) Fat-suppressed axial T2-weighted image shows that the high signal intensity area around the optic nerves is associated with tortuosity of the course of optic nerve sheath and mild flattening of the posterior pole of ocular globe, bilaterally.