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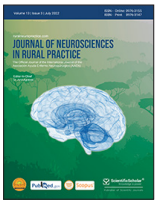

Case Report

Bilateral idiopathic optic nerve sheath meningocele: A case report and literature review

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ABSTRACT

Optic nerve (ON) sheath meningocele is an enlargement of the ON sheath, consisting in a cerebrospinal fluid collection along the perineural space of the optic nerve. It should be considered primary when it is not associated with orbital-cerebral neoplasm or with cranio-orbital junction malformations. Here, we report a case of bilateral primary idiopathic ON meningocele with gradual vision loss, treated with acetazolamide, which showed a maintained visual recovery and partial improvement during a 6-month follow-up period. The literature review retrieved eight cases of primary idiopathic ON sheath meningocele: ON sheath fenestration is considered in patients with progressive and severe vision loss, otherwise, acetazolamide treatment is indicated with good results on symptoms control, as confirmed in our case report.

Keywords: Optic nerve, Meningocele, Vision loss

INTRODUCTION

Optic nerve (ON) sheath dilatation or meningocele is a rare condition described as an enlargement and dilation of primarily the ON sheath.[1] It generally consists of a collection of cerebrospinal fluid (CSF) along the perineural space of the ON and should be considered primary when it is not associated with orbital-cerebral neoplasm or with cranio-orbital junction malformations. We describe a case of bilateral primary idiopathic ON meningocele with gradual vision loss and treated with acetazolamide, which showed a maintained visual recovery and partial improvement during a 6-month follow-up period.

CASE REPORT

A 46-year-old female presented with a 7-year history of blurring of vision in the right eye, chronic headache, and periorbital pain episodes. Her medical history included Hashimoto's thyroiditis, gastric bypass surgery, and cholecystectomy. On examination, her pupil reactions were normal, and her ocular motility was unrestricted. Her best-corrected visual acuity was 9/10 in both eyes. A first visual field test reported a partial temporal hemianopsia in the right eye. Visual evoked responses revealed a conduction delay in the right eye. A first color vision was tested with Ishihara color plates and it was normal on both sides. Anterior segment examination was normal in both eyes. Examination of the fundus reveled bilateral optic atrophy; there was no disk swelling. Her intraocular pressure was measured at 16 mmHg in both eyes.

After 6 months, a second visual evoked responses confirmed evidence of pathway delay in the right eye, equal to the first examination. Otherwise, color vision was partially reduced in the right eye with a score of 16 correct responses out of the first 21 Ishihara plates. There was no evidence of a hyperopic shift in her refraction, any evidence of choroidal folds and currently macular pathology has not been noted.

Investigations including thyroid function tests, full blood count, serum B12, folate, urea and electrolytes, erythrocyte sedimentation rate, and a treponemal screen were normal. Anti-nuclear antibodies were negative. A lumbar puncture was performed with an opening pressure of 29 cm of water.

The patient underwent a brain magnetic resonance imaging (MRI) scan that ruled out conditions that can lead to chiasmal compression and excluded a case of idiopathic intracranial hypertension (IIH). MRI showed bilateral ON sheath dilatation and enlargement, with normal sized optic nerves and with no associated inflammation, orbital, or cerebral neoplasm at the apex of the orbit [Figures 1 and 2]. Except for empty sella, MRI did not reveal imaging features that support the diagnosis of IIH, and diagnostic

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criteria for IIH were not satisfied. Subsequently, the patient underwent a phase-contrast cine MRI which demonstrated the absence of hyperdynamic flow of CSF through the aqueduct.

The patient is being followed conservatively with serial visual examinations and with a course of acetazolamide 250 mg twice daily (Tablets) for 3 months. At present, with this therapy, she did not report a further vision loss.

DISCUSSION

The term "ON head meningocele" (ONHM) was first coined in 1990 in Garrity *et al.*'s landmark paper in which 13 patients were described with this condition:[1] They all had dilated ON sheaths with an expanded CSF space and with no associated inflammation, orbital or cerebral neoplasm at the apex of the orbit. The pathophysiology of ONHM is not well understood. One theory relies on the difference in osmotic gradient between the cerebral subarachnoid space and perioptic subarachnoid space; in fact, a raised protein level was found in the CSF drained from an ON cyst in a 1997 case report.[2] Another theory is that congenital narrowing of the optic or cranial-orbital junction could cause the CSF accumulation in the perioptic subarachnoid space.[3] Cases of ONHM have spanned both sexes and all age groups.[1] The presentation symptoms are visual disturbance (either a drop in visual acuity or a field disturbance), headache, proptosis, and periorbital pain.[3,4] On ocular examination, many patients have a visual field defect with optic disk pallor or edema.[2] Some authors have noticed a subgroup of ONHM that consists of middle-aged men with progressive hyperopia, choroidal folds, and macular pathology.[2] Other authors have noticed a subgroup of cases with elevated CSF pressure.[1,4,5] In a case series by Dailey *et al.*, four out of the seven patients reported with hyperopia and choroidal folds had ON meningoceles.[6] ON sheath enlargement can be seen in cases of neurofibromatosis Type I, Marfan syndrome, or IIH.[7-10] The diagnosis is usually made by obtaining MRI of the orbits. The dilated ON sheath which is filled with CSF can easily be visualized. Coronal T2-weighed images can show the so-called bull's eye appearance, which represents expanded CSF spaces around the ON.[1] MRI is useful to rule out conditions that can lead to secondary enlargement of ON sheath such as orbital masses and ON gliomas. The prognosis for this condition varies. Progressive visual loss can occasionally be seen. Treatment is usually tailored to the patient's vision. Carbonic anhydrase inhibitors can be beneficial in some cases: Three patients on acetazolamide therapy reported no new loss of vision.[3,11,12] Surgical interventions in the form of ON sheath fenestration may be of benefit in cases of progressive vision loss. In a 1997 case review, 13 of 33 patients with ON sheath meningocele had some type of surgical decompression – four frontal craniotomies, one pterional craniotomy, and eight lateral orbitotomies. Five of the 13 patients who had the procedure experienced improvement of their symptoms while six had no change in their symptoms.[2] In a 2017 case report, an ON sheath fenestration was performed in a 10-year-old boy, and he experienced an improvement and stabilization of vision during a 1-year follow-up period.[13]

The current evidences are summarized in [Table 1].

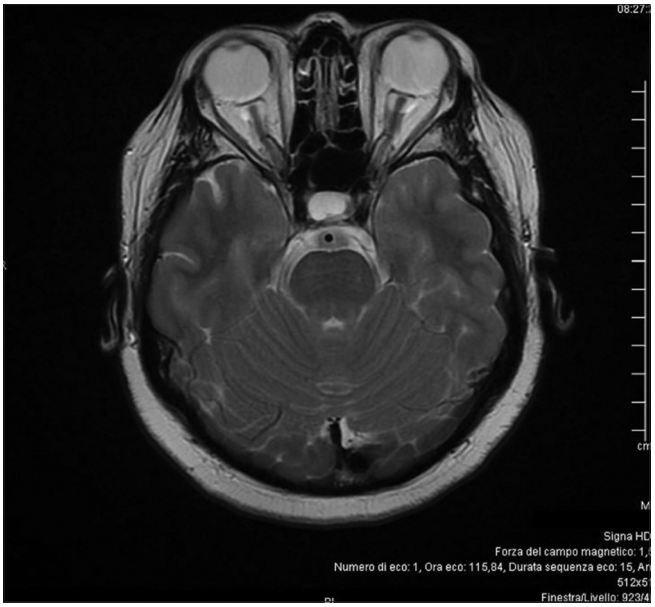

Figure 1: T2-weighted axial view showing the optic nerve meningocele.

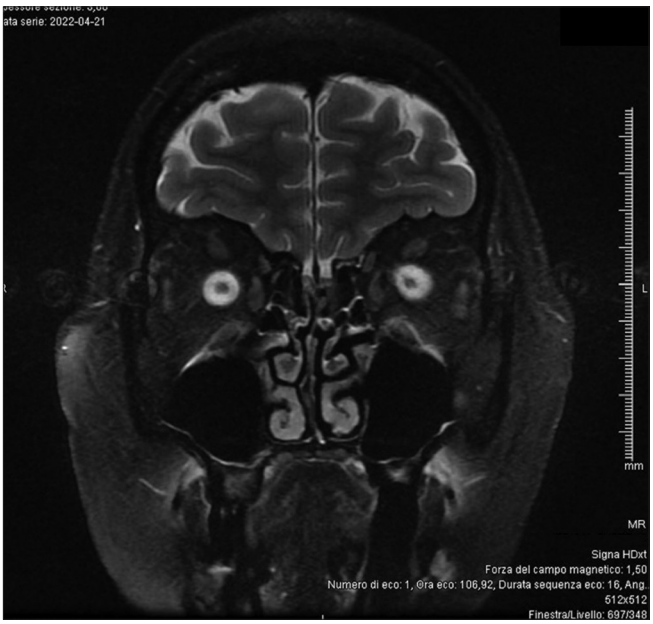

Figure 2: Coronal T2-weighted image showing the "bull's eye" appearance.

Table 1: Summary table of cases reported in the literature.

Author, year	Type of article	Cases (sex)	Presentation	Cerebrospinal fluid pressure	Patients underwent a surgical procedure	Associated findings	Associated medical conditions
Garrity *et al*., 1990	Review	13 patients (7 M-6F)	6 Visual deficit 4 headaches 3 both	Elevated in 2 patients	3	Postoperatively, one patient improved	3 patients had type I neurofibromatosis; 5 patients had optic nerve type defects
Lunardi *et al*., 1997	Review	33 patients	Visual deficit and headache	-	13	Postoperatively, 5 patients improved	9 patients presented hyperopia
Shanmuganathan *et al*., 2002	Case report	1 M	Visual deficit, orbital pain and proptosis	Elevated	-	-	Unilateral cystoid macular edema
Mesa-Gutiérrez *et al*., 2008	Case report	1 M	Visual deficit	Normal	-	With acetazolamide, not reported new loss of vision	Choroidal folds
Halimi *et al*., 2013	Case report	1 F	Visual deficit and periorbital pain	Normal	-	With acetazolamide, not reported new loss of vision	Optic disk drusen and pseudopapilledema
Mahatma *et al*., 2017	Case report	1 M	Visual deficit	-	1	Postoperatively, patient improved	Bilateral colobomatous cavitary disk anomalies
Algarni *et al*., 2018	Case report	2 (1 M – 1 F)	Visual deficit	Elevated	-	-	Swelling of the optic disk and choroidal folds
Jain *et al*., 2019	Case report	1 F	Visual deficit and proptosis	-	-	With acetazolamide, not reported new loss of vision	Hyperopia and choroidal folds
Shaikh *et al*., 2019	Case report	1 F	Soft-tissue mass covering eye	-	-	-	Patient had type I neurofibromatosis

CONCLUSION

ON sheath meningocele refers to dilatation of the ON sheath, which can have variable presentation. ON dysfunction can be seen, and it can range in severity from subtle involvement to a progressive vision loss. The diagnosis is made with orbit MRI. ON sheath fenestration may be an option in patients with progressive and severe vision loss; otherwise, acetazolamide treatment is indicated with good results on symptoms control, as confirmed in our case report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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