Delayed visual loss from optochiasmatic arachnoiditis after resection of craniopharyngioma

Case report

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Visual loss following surgery for craniopharyngioma is usually the result of operative injury or tumor recurrence. The authors present the case of a 12-year-old boy who developed progressive visual field constriction 11 years after gross-total resection of a solid and cystic craniopharyngioma. No tumor recurrence was evident on multiple MRI studies, and it was only at surgical exploration that the diagnosis of optochiasmatic arachnoiditis was established. Lysis of the adhesions around the optic nerves and chiasm resulted in substantial and sustained visual improvement.

Key Words • visual loss • optochiasmatic arachnoiditis • optic chiasm • gross-total resection • optic nerve • craniopharyngioma • oncology

We present the case of a 12-year-old boy who developed progressive visual field (VF) constriction 11 years after undergoing gross-total resection of a solid and cystic craniopharyngioma. It was only at surgical exploration that the diagnosis of the arachnoiditis was established. Lysis of the arachnoidal adhesions around the optic nerves and chiasm resulted in substantial visual improvement.

Case Report

A previously healthy 6-month-old boy was suddenly noted by his mother to have gone blind with a new onset of roving eye movements. Examination revealed loss of light perception with absent startle response to the room lights being switched on and off. The patient’s pupils were widely dilated and responded very sluggishly to light. Computed tomography scanning revealed a 3-cm cystic craniopharyngioma compressing the optic chiasm (Fig. 1). Preoperative endocrinological assessment revealed only mild hypothyroidism. Gross-total excision of a solid and cystic craniopharyngioma was performed. Postoperative imaging confirmed gross-total excision and revealed a small asymptomatic infarction in the anterior limb of the internal capsule (Fig. 2).

Within days of the tumor removal, the patient’s vision had improved. By 6 months postoperatively, his visual function had returned to normal for his age. His vision remained stable, and by the age of 8 years, when he was able to cooperate with testing, visual acuity at 20/20 was recorded in each eye, normal scores on the Ishihara chart were noted for each eye, mild optic disc pallor was exhibited, and a stable incongruous left homonymous hemianopia was found on Goldmann perimetry (Fig. 3). No radiation or other adjuvant therapy was given. The patient was confirmed to have normal endocrine function, including thyroid function, at assessment 4 months postoperatively.

Abbreviation used in this paper: VF = visual field.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
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At the age of 11 years, while the patient remained endocrinologically normal, the right VF was noted to be reduced. Over the next month, the patient’s VF continued to deteriorate with further reduction in the superior temporal quadrants bilaterally (Fig. 4). At this time, MRI showed no recurrent tumor, and no papilledema was seen. Steroids were not administered.

In the absence of visualized tumor recurrence, reexploration was not considered until the VF changes were confirmed to be progressive. With this information and with no other option for treatment, it was decided to explore the optic nerves and chiasm to ensure that no tumor was present and to treat any lesion resulting in optic nerve and/or chiasmal distortion. Optochiasmatic arachnoiditis and empty sella syndrome were considered.

The patient’s frontal pterional craniotomy was reopened, and the frontal lobe was elevated to provide visualization of the optic nerves and chiasm. The diaphragma sellae was intact and there was no tumor recurrence, but both optic nerves were enveloped with thick and scarred arachnoid (Fig. 5 left). The arachnoidal adhesions extended to involve the A1 segments of the anterior cerebral arteries and their branches. The adhesions were lysed, and the suprasellar cistern was widely opened. Optochiasmatic arachnoiditis was diagnosed based on the findings and the absence of other evident etiologies (Fig. 5 right).

In the immediate postoperative period, further VF deterioration was noted, but the visual acuity remained stable at 20/20 in each eye. By 5 months postexploration, the patient’s visual function had improved significantly and his Goldmann VFs had returned to those recorded at the age of 10 years (Fig. 6). He exhibited optic disc pallor in the right eye. The patient’s visual function has remained stable for the last 5 years.

Discussion

Craniopharyngioma is a benign tumor arising from remnants of Rathke’s pouch. Its estimated incidence is 13 cases per 1 million individuals/year. The ideal method of treatment remains highly controversial. However, a gross-total resection is thought to be associated with the best outcomes regarding postsurgical recurrence-free survival. Postoperative visual loss can occur, particularly in patients with significant preoperative visual dysfunction. Visual failure occurring several
years after resection usually results from tumor recurrence causing optic nerve or chiasmatic compression. Optochiasmatic arachnoiditis as a cause of visual failure has been reported in cases of tuberculous leptomeningitis and traumatic brain injuries.\textsuperscript{2,3,6,8,9,12} When the optic nerve is involved, this entity presents with gradual and progressive monocular concentric VF reduction. The gradual VF reduction may subsequently occur in the other eye.\textsuperscript{2,5,9,12} The process can also be bilateral and simultaneous. Additionally, loss of visual acuity can present itself in the forms described for VF loss.

This patient demonstrated recovery of excellent vi-

Fig. 3. Visual fields obtained in 2005 after initial resection and recovery. The field remained stable for 11 years.

Fig. 4. Upper: Visual fields obtained in 2006 showing field constriction in the left eye 1 year prior to reexploration. Lower: Visual fields obtained in 2007 prior to optic nerve and chiasmal reexploration.
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Fig. 5. Left: Left lateral view of the chiasmatic cistern showing thickened arachnoid enveloping both optic nerves. Right: Left lateral view of optic nerves showing left nerve constriction after partial arachnoidolysis.

Fig. 6. Visual fields on postoperative Day 115 (in 2008), preserved to the present.

be considered in patients exhibiting progressive VF loss after surgical treatment of tumors in the suprasellar region if MRI does not identify a specific etiology. Surgical exploration is recommended to exclude this entity.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References

9. Jacquemin PJ, Frrippiat M: [Opto-chiasmatic arachnoiditis: clinical study of various cases; attempt at classification of symp-

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