Intrachiasmatic craniopharyngioma: a rare cause of chiasmal thickening

Case report

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The authors describe the case of a young man who presented with a central scotoma in one eye and a temporal hemianopsia in the other. Magnetic resonance imaging showed distinctive bilobed thickening of the chiasm and a “potbelly” expansion of the contiguous optic nerves. The distal portions of the intracranial optic nerves appeared normal. At craniotomy, incision of the lamina terminalis exposed intraventricular craniopharyngioma with anterior extension into the chiasm and proximal optic nerves. Partial resection of the tumor restored normal vision.

KEY WORDS • craniopharyngioma • brain neoplasm • optic chiasm • visual loss

When progressive loss of vision occurs in a patient with a thickened chiasm, the probable cause is chiasmal glioma. When the patient is an adult the tumor is usually malignant, progressing to blindness within several months and to death within a year.2 Rarely, craniopharyngioma occupies the chiasm and produces symptoms and surgical findings mimicking those of chiasmal glioma, as exemplified in a case described by Duff and Levine in 1983.1 The present report describes a case of intrachiasmatic craniopharyngioma, its diagnosis by magnetic resonance (MR) imaging, and its management at craniotomy.

Case Report

This 33-year-old tow-truck operator on the San Francisco Bay Bridge noted difficulty with depth perception and trouble in focusing his eyes for 5 months. During work breaks he would fall asleep and then have trouble resuming his tasks. He began having severe headaches and his libido diminished.

Examination. On examination, his vision was 20/20 in the right eye and 20/200 in the left. There was a relative afferent papillary defect in the left eye. He had a partial temporal hemianopsia in the right eye and a central scotoma in the left. The optic discs appeared normal.

The patient underwent MR imaging. The images showed a bilobed enlargement of the chiasm extending forward into both optic nerves (Fig. 1). There was a clear demarcation between the normal-appearing distal optic nerves and the thickened proximal portions. Multiple small dark spots indicated mixed signal intensity within the substance of the chiasm. A round, smoothly margined mass expanded the anterior third ventricle. There was no hydrocephalus. The inferior portion of the septum pellucidum was thickened. A sagittal T1-weighted image showed a mass of mixed signal intensity in the chiasmal area. A parasagittal view showed a fat chiasm with a “potbelly” expansion of the contiguous optic nerves (Fig. 2).

Operation. A right frontal craniotomy was performed, with exposure of the optic nerves, anterior chiasm, right anterior cerebral artery, and lamina terminalis. The chiasm and contiguous optic nerves were enlarged to about two and one-half times their normal dimensions (Fig. 3). The enlarged optic nerves extended forward for a distance of 5 to 6 mm before they abruptly returned to normal size. An incision was made in the lamina terminalis. Lying within the third ventricle was a yellow tumor that had a frond-like surface speckled with fine dots of purple and purple-green. It had the gross appearance of a craniopharyngioma (Fig. 3).
Intrachiasmatic craniopharyngioma

FIG. 2. Sagittal and parasagittal T1-weighted (TR 600 msec, TE 20 msec) magnetic resonance images. The sagittal image (left) shows a mass of mixed signal intensity in the chiasmal area. The parasagittal image (right) shows a "potbelly" expansion of the proximal optic nerve.

FIG. 3. Operative photographs of the optic chiasm. Upper Left: View showing the enlarged chiasm and optic nerves. Arrow indicates the point at which the normal optic nerve diameter begins. Upper Right: Incision in the lamina terminalis. Lower Left: The tumor can be seen prolapsing from the third ventricle into the operative field. Lower Right: The appearance of the chiasm after tumor removal.

In that case an enhancing suprasellar mass was revealed on computerized tomography scans, and an enlarged chiasm was found at craniotomy. Incision into its dorsal surface revealed craniopharyngioma, which was biopsied but not resected.

In our case there were several clues in the MR images that suggested intrachiasmatic craniopharyngioma. Bilateral optic nerve expansion extended only a few millimeters forward from the chiasm. Optic nerve expansion from glioma would be expected to extend all the way to the optic canals. This MR finding correlates with the "potbelly" optic nerve expansion in the necropsy specimen described by Lindenberg, et al.4 The bilobed appearance of the chiasm on the MR coronal sections in our case also correlates with the bilobed chiasm in their specimen. We have not seen this sign
in MR images of chiasmal gliomas. The mixed signal intensity of the tumor may correspond to the polycystic nature of the tumor as shown by Lindenberg, et al. Chiasmal gliomas usually have a uniform signal intensity. Lanzieri, et al., 3 described thickening of the septum pellucidum in four patients who had third ventricular craniopharyngiomas. In all cases, biopsy of the area showed only reactive gliosis. In one case the septum pellucidum was buckled upward by the tumor and its leaves were separated by the tumor. Again there was no evidence of tumor invasion. They emphasized that this sign is nonspecific and occurs with other third ventricular tumors.

Recovery of vision in our case was complete in spite of the fact that the intrachiasmatic portion of the tumor was not resected. Since debulking the tumor restored normal vision, the patient’s visual loss was probably caused by compression of the visual pathways by the third ventricular component of the tumor.

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References


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