Reversible visual deficit following debulking of a Rathke’s cleft cyst: a tethered chiasm?

Case report

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Delayed chiasmal syndromes after emptying of a Rathke’s cleft cyst have not been reported previously. When these deficits occur following the treatment of parasellar lesions they are usually associated with the descent of a scarred optic system into an empty sella, and vision often improves promptly when the optic system is elevated. Two months after transsphenoidal surgery with emptying of a large intrasellar cyst, a 22-year-old man developed recurrent bitemporal visual field deficits over a 3-day period. Sagittal magnetic resonance imaging demonstrated an enhancing band of tissue extending anteriorly from the normally placed chiasm down to the anterior portion of the sella turcica. At craniotomy the enhancing tissue was found to be scar extending from the anterior border of the chiasm to the diaphragma sellae. The anterior portion of the diaphragm was resected as widely as possible without dissecting the scar itself from the chiasm. A membrane consistent with the wall of a Rathke’s cleft cyst was found attached to the resected tissue. The patient’s vision was improved 2 days after surgery. This case illustrates that traction by scar extending from the chiasm to the diaphragm, even when the chiasm is in its normal anatomical location, may cause progressive visual loss; and that untethering of the chiasm by resecting the diaphragm while leaving the scar intact can result in improved vision.

Key Words • Rathke’s cleft cyst • postoperative visual deficit • optic chiasm

Delayed chiasmal syndromes have been reported after radiation therapy or excision of pituitary macroadenomas, transsphenoidal hypophysectomy for breast cancer, and antibiotic treatment of tuberculous meningitis.2-7 We report this phenomenon after surgical emptying of a Rathke’s cleft cyst. Scarring extending from the chiasm to the diaphragma sellae was demonstrated by magnetic resonance imaging; neither the chiasm nor the diaphragm was displaced into the sella. Vision improved promptly after resection of the diaphragm adjacent to the scar.

Case Report

This 22-year-old male electrician had for 1 year found it difficult to see a poster on his bedroom wall when he awoke in the morning. He sought medical help after he had driven his car into the curb on numerous occasions.

Examination. The patient weighed 112 lbs and was 5 ft 8 in. tall. He was youthful in appearance and of shorter stature than other members of his family. Facial hair was sparse but he had normal axillary and pubic hair and genitalia. Neurological examination revealed normal optic fundi, a bitemporal hemianopsia on confrontation testing, and visual acuity of 20/200 in the right eye and 20/20 in the left using a near card. Magnetic resonance imaging demonstrated a nonenhancing mass enlarging the sella turcica, elevating the chiasm and extending to the foramen of Monro (Fig. 1 left). The serum testosterone level was mildly depressed (334 ng/dl; normal range 360 to 990 ng/dl), and an isolated serum growth hormone content was 1.5 ng/ml. Thyroid function, levels of follicle-stimulating hormone, luteinizing hormone, and serum prolactin, and a Cortrosyn (corticotropin) stimulation test were normal.

First Operation. On September 1, 1992, the patient underwent craniotomy to explore the tumor via a transsphenoidal approach. Upon opening the dura of the sella turcica, a soft yellow material with mucous texture and the histological appearance of acellular, necrotic debris exuded into the sphenoid sinuses in cohesive glob. This material came away cleanly from the walls of the sella. The diaphragm appeared to hang down at the sides but was suspended at its apex.

On examination 3 weeks later the visual fields were normal on confrontation testing, and visual acuity was
FIG. 1. Midline magnetic resonance T1-weighted images with gadopentate dimeglumine enhancement, sagittal projection. 

Left: Image obtained prior to the first operation. A homogeneous mass is seen expanding the sella (curved arrow) and extending into the suprasellar cistern (black arrows). The enhancement along the lesion's posterior aspect (open arrow) could represent scarring, displaced pituitary tissue, or tumor. 

Right: Image obtained before the second operation showing an enhancing band extending from the optic chiasm to the pituitary fossa (curved arrow) just anterior to the normally enhancing pituitary stalk (straight arrow).

20/30 in the right eye and 20/20 in the left using a near card. Pituitary function remained unchanged. However, on October 21, 1992, the patient noted return of his visual deficit without headache or other symptoms. Examination in the clinic on October 26, 1992, revealed a bitemporal hemianopsia with visual acuity of 20/40 in the right eye and 20/20 in the left, as confirmed by formal testing (Fig. 2 upper). Magnetic resonance imaging showed no residual tumor; sagittal views revealed an enhancing band of tissue anterior to the normal pituitary stalk that extended up to the anterior portion of the chiasm (Fig. 1 right). The patient was given dexamethasone therapy.

Second Operation. At a second operation, on October 31, 1992, the suprasellar region was explored via a transcranial approach. The anterior border of the chiasm was bound to the diaphragma sellae by dense scar tissue (Fig. 3 left) and as the diaphragm was incised it gaped as if it were under tension. The diaphragm was resected widely, leaving the scar attached to the anterior border of the chiasm (Fig. 3 right). By the 2nd postoperative day, the patient's vision had improved both subjectively and on confrontation testing. Two weeks later, his visual fields were improved, although a right superior temporal defect persisted (Fig. 2 lower). Ten days after craniotomy, a cerebrospinal fluid rhinorrhea was repaired via a transsphenoidal approach. Subsequent recovery was uneventful.
Pathological Examination. Examination of the resected specimen revealed a dense fibrous tissue with an attached epithelial lining consistent with a Rathke's cleft cyst (Fig. 4). The epithelium varied considerably in thickness and was composed of stratified or pseudostratified rarely ciliated columnar cells, with occasional cells positive for periodic acid-Schiff and mucicarmine staining. There were also regions in which superficial columnar cells rested on several layers of flattened epithelial cells with microcysts and focal calcification. Immunoperoxidase reactions (using keratine and epithelial membrane antigen preparation) confirmed the character of the lesion.

Discussion

Ten patients have previously been reported; ours is the eleventh, in whom loss of vision occurred following radiation, surgical, or medical treatment of sellar or suprasellar pathology, and in whom the chiasm was subsequently explored surgically. Eight patients had pituitary macroadenomas, treated with radiation therapy, surgical excision, or a combination of the two (Table 1). Treatment for the remaining three patients consisted of transsphenoidal hypophysectomy for breast cancer, antibiotic drugs for tuberculous meningitis, and transsphenoidal emptying of a Rathke's cleft cyst in our patient. In these 11 cases, the most common visual field deficits were chiasmal syndromes (Table 2). Three patients in addition to ours had a bitemporal hemianopsia; three patients were virtually blind in one eye and hemianoptic in the other, two patients had unilateral hemianopsia, and in two patients the visual field defects were not stated.

Scarring of the chiasm was the most common operative finding in these cases and was often accompanied by displacement of the chiasm and optic nerves into the sella. The condition of the chiasm was not described in two cases. Descent of the optic system into the sella was noted in six patients. In two patients,
TABLE 2

Visual deficits in 11 patients with tethered chiasm

<table>
<thead>
<tr>
<th>Visual Field Deficits</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>bitemporal hemianopsia</td>
<td>4</td>
</tr>
<tr>
<td>unilateral temporal field cut</td>
<td>2</td>
</tr>
<tr>
<td>blind or near blind in one eye, temporal cut in other</td>
<td>3</td>
</tr>
<tr>
<td>not stated</td>
<td>2</td>
</tr>
</tbody>
</table>

* Case presented in this report.

TABLE 3

Improvement after treatment of tethered chiasm

<table>
<thead>
<tr>
<th>Surgical Treatment</th>
<th>No. of Cases</th>
<th>Improved Vision</th>
<th>Days to Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>diaphragm elevated by packing sella</td>
<td>3</td>
<td>3</td>
<td>6, 7, &lt;42</td>
</tr>
<tr>
<td>hole placed in lamina terminalis</td>
<td>1</td>
<td>1</td>
<td>?</td>
</tr>
<tr>
<td>lysis of adhesions or scar or incision of diaphragm</td>
<td>3</td>
<td>3</td>
<td>1, 2, ?</td>
</tr>
<tr>
<td>chiasm explored only</td>
<td>2</td>
<td>0</td>
<td>?</td>
</tr>
<tr>
<td>details of surgery not given</td>
<td>2</td>
<td>1</td>
<td>?</td>
</tr>
<tr>
<td>total cases</td>
<td>11</td>
<td>8</td>
<td></td>
</tr>
</tbody>
</table>

* Case presented in this report.

ours and the one reported by Scott, et al., the chiasm was above the sella. In three, the relationship between the chiasm and the sella was not stated. Magnetic resonance imaging was helpful in the preoperative evaluation of our patient because it showed both the enhancing scar and the position of the chiasm.

Surgery resulted in improved vision in eight of the 11 patients (Table 3). In the two patients reported by Lee and Adams, the chiasm was simply explored and vision was not improved. The surgical details of the two patients described by Poppen were not given; only one patient improved. Three patients had improved vision after elevation of a depressed diaphragm to which the chiasm was adherent. Welch and Stears commented that elevating the diaphragm released tension on the optic nerve. Two patients were improved by simply freeing the chiasm of adhesions. The chiasm had descended into the sella in one of these cases but not in the other. In our patient, improvement occurred after the diaphragm had been widely excised without dissecting the scar between the diaphragm and the normally positioned chiasm. Finally, one patient improved following puncture of the lamina terminalis, a maneuver that did not result in improvement in another patient with a primary empty sella. The precise timing of recovery is uncertain in the eight patients whose vision improved (Table 3). In three patients besides ours, vision improved within 1 week after surgery, and in two of these cases, improvement was noted within 2 days. Another patient was noted to have improved when examined 6 weeks after surgery. However, in three patients there was no reference to the time of improvement.

If there is a common mechanism for this reversible disturbance of chiasmal function, it has not been found; some authors believe it is multifactorial. Traction, kinking, vascular strangulation, and radiation injury, either alone or in combination, have all been implicated. Scarring or tethering of the chiasm to the diaphragm or to the wall of the sella appears to be the common element, while displacement of the optic system into the sella occurs in many but not all cases (for example our patient and the one reported by Scott, et al.).

Bergland and Ray argued that rapid recovery following decompression indicates an ischemic etiology of the loss of vision associated with pituitary macroadenomas. They demonstrated that the vessels to the chiasm extended upward around the infundibulum and became particularly numerous at the angle formed by the anterior aspect of the infundibulum and the inferior surface of the chiasm. The authors postulated that compression of these vessels caused rapidly reversible ischemia of the chiasm. Based on this argument, the rapid recovery seen after untethering of the chiasm in our patient or after "chiasmapexy" reported by others would suggest an ischemic etiology for the loss of vision. Contraction of the scar could cause distortion of the chiasmal vessels at the top of the pituitary stalk. A local vascular compromise may occur, causing neurological dysfunction, as has been postulated in cases of spinal cord tethering. Mortara and Norrell found that puncture of the lamina terminalis resulted in improved vision in one patient. This was thought to be accomplished by diverting the pulsations of the cerebrospinal fluid in the third ventricle away from the chiasm. However, it is difficult to imagine how this procedure could improve blood flow to the optic system unless the operation included freeing the chiasm from adjacent scar.

References


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