

CASE REPORT

What Comes Down Must Go Up—Chiasmal Prolapse: A Field Report

Akshay Gopinathan Nair¹, Shikha R. Bassi¹, Veena Olma Noronha¹,
Krishnaprasad Moodambikana², and K. R. Suresh Babu²

¹Sankara Nethralaya, A Unit of Medical Research Foundation, Nungambakkam, Chennai, India and ²St. Isabel's Hospital, Mylapore, Chennai, India

ABSTRACT

We report a case of a 45-year-old man who had experienced considerable improvement in vision following surgery to excise a non-functioning pituitary adenoma. However, 1 year after surgery, during a regular follow-up, secondary visual field changes were detected that prompted neuroimaging. Magnetic resonance imaging revealed an empty sella with chiasmal prolapse, with no recurrence of the tumour. The patient underwent an endoscopic endonasal transsphenoidal chiasmectomy. Post-operatively the visual fields and the visual acuity showed improvement. This case emphasises the need for monitoring of visual fields, even post-operatively in cases of pituitary tumour. Chiasmal herniation can cause secondary visual field changes, early recognition of which can help in prompt surgical intervention to reverse the field loss.

Keywords: chiasmal prolapse, chiasmectomy, empty sella syndrome

CASE REPORT

A 45-year-old male presented to us with complaints of progressive diminution of vision in his left eye. His best corrected visual acuity (BCVA) in the right eye was 6/6, N 6 and counting fingers at 1 m in the left eye. According to the patient, this vision loss had been gradual over the past 3 months; however, he could appreciate a very rapid deterioration in vision in the preceding 2 weeks, which prompted this hospital visit. Pupillary evaluation revealed a relative afferent pupillary defect in the left eye. Colour vision was tested and found to be normal in the right eye, but in the left eye the patient could not identify the test plate on Ishihara's tests. Intraocular pressures were normal in both eyes. The fundus was normal in the right eye. However, optic disc pallor was noted in the left eye. Humphrey's visual field (HVF) charting in the right eye showed an incomplete temporal hemianopic defect and a full-field 120-point screening test for the left eye showed a temporal hemianopia with spill-over defects crossing the vertical midline (Figure 1a). Magnetic resonance (MR) imaging showed a large, circumscribed, lobulated mass in the sella with suprasellar extension, compressing the optic chiasma

(Figure 2a). Exhaustive hormonal assays revealed no abnormal levels. A neurosurgical opinion was sought, based on which a transsphenoidal excision of the mass was performed and histopathological examination confirmed the diagnosis of a non-functioning pituitary adenoma.

Examination of visual function 6 weeks post surgery revealed a visual acuity of 6/6, N6 in the right eye and 6/24, N36 in the left eye. He was re-examined 6 months post surgery when BCVA in the right eye was 6/6, N6 in the right eye while it was 6/24, N36 in the left eye. HVF charting in the right eye showed enlargement of the blind spot with defects in the superotemporal quadrant; the left eye showed a temporal hemianopia (Figure 1b). The patient was re-assessed after 6 months (a year from the pituitary surgery) when the BCVA was 6/6 N6 in the right eye but the visual acuity had dropped to 6/36 N 36 in the left eye. The visual fields in the right eye showed enlargement of the blind spot with an increase in the defect size. The left eye showed a temporal hemianopia with defects crossing the vertical midline (Figure 1c). The visual fields had acceptable reliability indices. Suspecting a recurrence of the tumour, MRI of the brain was performed, which showed an empty

Received 11 October 2010; revised 15 April 2011; accepted 16 April 2011

Correspondence: Dr. Akshay Gopinathan Nair, MBBS, Sankara Nethralaya, A Unit of Medical Research Foundation, 18, College Road, Nungambakkam, Chennai 600 006, India. E-mail: akshaygn@gmail.com

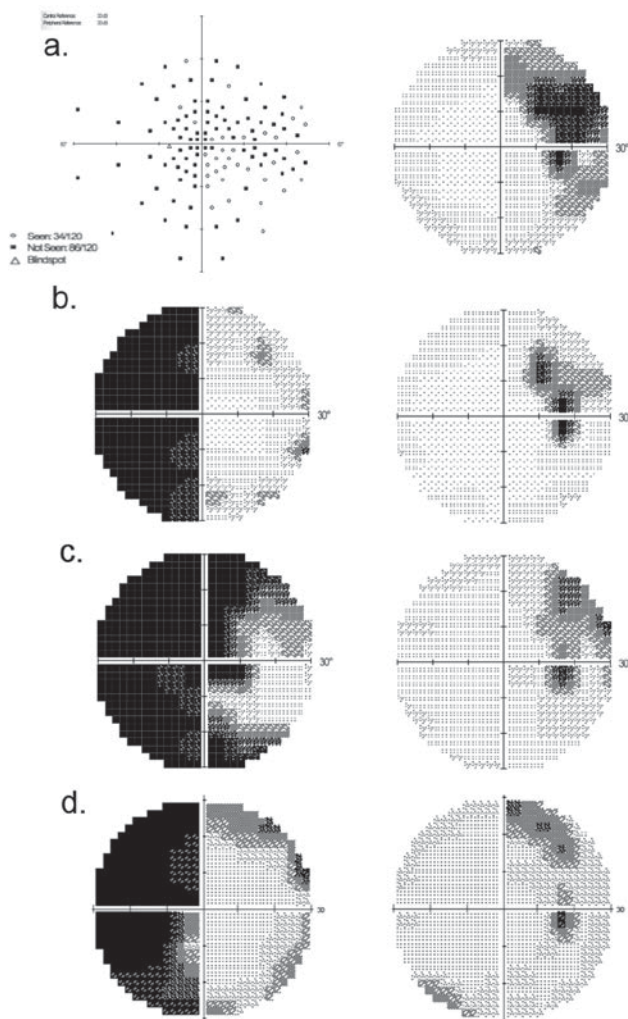


FIGURE 1 Serial automated perimetry. (a) Right eye showed an incomplete temporal hemianopic defect and a full-field 120-point screening test for the left eye showed a temporal hemianopia with spillover. (b) Right eye showed enlargement of the blind spot with defects in the superotemporal quadrant; the left eye showed a temporal hemianopia. (c) The visual fields in the right eye showed enlargement of the blind spot with an increase in the defect size. The left eye showed a temporal hemianopia with spillover defects crossing the vertical midline. (d) Post-operative perimetry showed an improvement in the depth and size of the defects in both eyes.

sella with chiasmal prolapse; however, no recurrence of the tumour was noted (Figures 2b, a, b). An endoscopic endonasal transsphenoidal chiasmectomy with reconstruction of the diaphragma sellae was performed. Intraoperatively, an empty sella with a large arachnoid sac filled with cerebrospinal fluid was demonstrated. The arachnoid was pushed upwards and the diaphragma sellae was reconstructed with a rostral layer of fascia lata and the sella was packed with muscle; both harvested from the right thigh. Fascia lata was placed caudally in layers to construct the sella floor. Post surgery, the BCVA was 6/6 N6 in the right eye and 6/18 N36 in the left eye. HVF charting showed only a few scattered temporal defects in the right eye and temporal hemianopia with no spillover in the left eye (Figure 1d). A

post-chiasmectomy MRI scan, however, was unavailable for comparison.

DISCUSSION

Pituitary tumour is one of the most common primary intracranial neoplasms, accounting for approximately 15% of all intracranial neoplasms.¹ Almost a third of all pituitary adenomas present with vision deterioration.² Being the most crucial sign of a pituitary adenoma, early recognition of visual findings may help bring a successful outcome.³ Visual improvement is often achieved in patients by decompression surgery with transsphenoidal hypophysectomy.⁴

The empty sella refers to a peculiar anatomical condition showing a sella turcica only partially filled by a pituitary gland severely flattened against the sellar floor. The term primary empty sella is used to refer to the condition when it is unrelated to previous surgical intervention, radiotherapy or pharmacological form of therapy.⁵ Secondary empty sella syndrome (ESS) occurs following surgical, medical or radiation treatment of pituitary or other sellar lesions.⁶ One study estimated the incidence of empty sella syndrome secondary to tumour to be as high as 79.2%.⁵ Ophthalmic manifestations are known to be more common in secondary ESS than primary ESS.⁶ Visual field changes are believed to occur as a result of downward herniation of the optic apparatus into the pituitary fossa. The different types of visual field losses described in this clinical entity include bitemporal hemianopsia, unilateral temporal defect, arcuate scotoma, central scotoma, and binasal field defects.⁵ However, the exact cause of vision loss is unclear as there exist in literature examples that fall in every subset: those who have normal vision in the presence of a prolapsed chiasma, those who have defective vision in the presence of a clearly demonstrable chiasma prolapse, and finally, those who have visual field defects in the absence of any obvious chiasmal prolapse.^{3,7,8} This downward prolapse of the optic chiasma is reported to occur after removal of large adenomas with considerable suprasellar extension, as was the case in our patient.³

The mechanism that leads to chiasmal herniation is still not known. Many factors, including traction, vascular compression, and kinking of the chiasma, have been found to be responsible, either singly or combined.⁹ The size of the opening through the diaphragma varies among individuals and the dura of the diaphragm may be very thin.¹⁰ These conditions may be further exacerbated by expansive growth of a pituitary macroadenoma such that, following its reduction in size by treatment, the overlying neural structures are able to herniate into the sella. Furthermore, it has also been hypothesized that the diaphragma sellae is susceptible to erosion following radiation therapy.¹¹

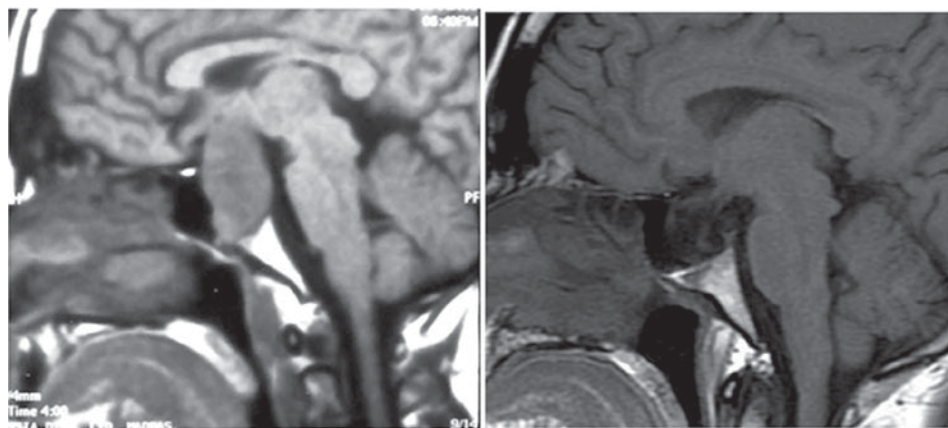


FIGURE 2 (a) Sagittal T1-weighted MR image showing the large pituitary adenoma expanding the bony sella and compression of the chiasm. (b) Sagittal T1-weighted image, 6 months post tumour resection.

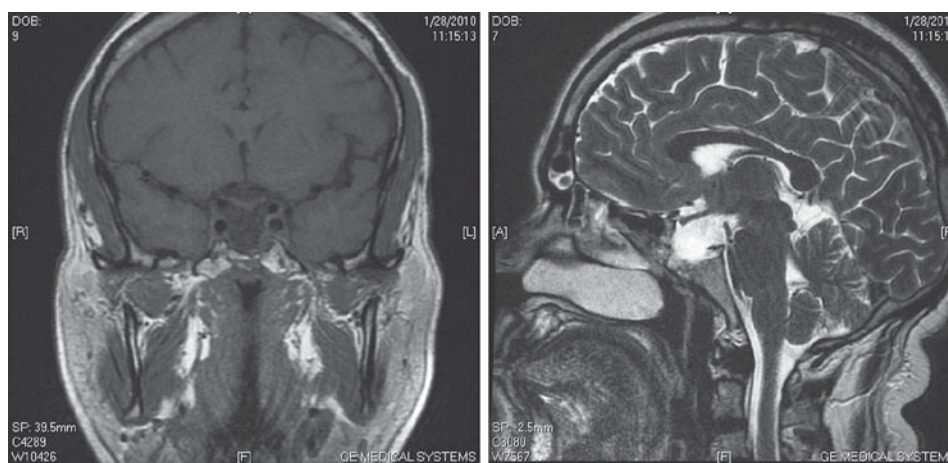


FIGURE 3 (a) Post tumour resection, coronal T1 MRI (6 months post tumour resection). (b) Sagittal T2-weighted image. Both images show complete excision of the tumour resulting in secondary empty sella; however, the chiasm is low placed and kinked.

The mechanism of the vision loss secondary to chiasmal prolapse has been thought to be due to a mechanical process (kinking, tethering, or compression) and an ischaemic one. Chuman et al. discussed a case of delayed visual loss following dopaminergic treatment of prolactinoma.¹² Twelve months following the reduction in the dose of pergolide, visual loss had recovered and MRI at the time of visual recovery showed no change in the position of the prolapsed chiasm. They believe that as visual recovery occurred without a visible change in the position of the chiasm, traction is unlikely to be a cause of delayed visual loss. It has also been suggested that the reduction in dopaminergic agonist dosage could have caused a regrowth of tumour that was simply too small to detect radiologically but enough to release tethering of the optic chiasm or its vascular supply.^{13,14}

Guinto et al. have demonstrated in their study that visual field defects may occur in cases of primary empty sella syndrome even without any radiological evidence of herniation of the visual system.⁷ They concluded from the two cases discussed in their paper, which improved after surgery, that in this syndrome, traction

on the infundibular stalk may actually cause some microscopic anatomic changes in the visual system or in its vascular supply that may not be demonstrable on imaging studies.

Also, there is disagreement among authors whether the visual deterioration occurs due to adhesions between the optic apparatus and tumour capsule.³ Radiotherapy is known to induce the formation of adhesions; however, visual deterioration has been noted even after medical treatment of prolactinomas without any surgical procedure, or radiotherapeutic intervention.⁹

Thomé and Zevgaridis have commented on the effect of the intimate relationship between anterior cerebral arteries and the optic nerves on the pathophysiology of reversible disturbance of chiasmal function. The authors have hypothesised the concept of vascular compression by describing the case of a 61-year-old female who reported a rapid visual deterioration, 8 months following surgery to remove a haemorrhagic pituitary adenoma. Imaging showed no downward displacement of the optic nerve and chiasm; however, the left optic nerve was found to be closely in contact

with the anterior cerebral artery. Furthermore the patient consistently claimed an improvement of vision after on-demand medication with short-acting anti-hypertensive medication, which prompted a surgical decompression. The authors mention that the intimate relationship between artery and nerve with consequent pulsatile pressure may constitute a causative factor in delayed visual dysfunction after pituitary surgery.¹⁵

Kaufman and associates reported no relationship between the degree of chiasmal herniation and the severity of visual loss in seven patients with empty sella syndrome secondary to surgical or radiation treatment of pituitary adenomas.¹⁰

Landolt also mentions in his commentary of a technical note by Zona et al. on transsphenoidal treatment of empty sella by means of a silastic coil that he found no correlation between the surgical result (improvement, no change or a slow deterioration) and any radiological or clinical factor that would allow a prediction of the final outcome or a selection of patients who might experience a benefit from the intervention.¹⁶ Landolt also mentions that the surgically induced change in the chiasmal contour could have a beneficial effect by altering the cerebrospinal fluid flow; this, however, has not been unequivocally demonstrated.

Surgical management of chiasmal prolapse is chiasmopexy, which can be attempted either through a transsphenoidal or transcranial route.^{17–20} The surgery involves releasing adhesions, relieving traction on the optic system by elevating the sellar floor extradurally with various materials, the most common being autologous tissues such as muscle and fascia lata, as in our case. Other materials used include bone, balloon, and silastic coil.^{6,19,20} It is reported that the volume of fat or muscle packing decreases over time as a result of scar retraction. Chiasmopexy involving extradural packing has also been achieved by using a Silastic (Dow Corning, Auburn, MI) coil, fashioned by means of a ventricular catheter arranged as a spiral.¹⁶ Silastic, being inert, has a lower risk of post-operative shrinkage, and is not associated with inflammatory complications and owing to its elasticity, it does not cause excessive compression of sellar, parasellar, or suprasellar structures.¹⁶

Sekhar and Oliveira also have described a case of a 53-year-old woman who presented with diminution of vision in both eyes 2 years after a transsphenoidal resection of a large pituitary tumour.²¹ MRI showed a chiasmal prolapse into the empty sella. Right optic canal decompression was performed and the chiasma was freed from adhesions via a right frontotemporal craniotomy and an orbital osteotomy. Vision post-operatively improved from 20/800 (right eye) and 20/200 (left eye) to 20/200 in both eyes.

Cases such as ours underscore the need for serial perimetry in all pituitary tumours, through the entire duration of therapy. In cases where the tumour has been

surgically managed, perimetry might give the first clue to pathologies such as tumour recurrence or as was in our case, chiasmal prolapse, thus ensuring prompt treatment.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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