Case Report

Intrachiasmatic craniopharyngioma: Assessment of visual outcome with optical coherence tomography after complete surgical removal

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Abstract

Background: Optic chiasm invasion by a craniopharyngioma (CP) is exceptional. Surgical treatment of intrachiasmatic CPs associates a high risk of chiasm injury, which should be properly addressed before surgery.

Case Description: We present a 46-year-old woman admitted to the hospital with low visual acuity (0.1 in the right eye and 0.5 in the left) and a severe defect in her visual fields, in addition to headaches, diabetes insipidus, and a long-term depressive disorder. Her visual deficit progressed from a right homonymous temporal inferior quadrantanopia to an almost complete loss of vision in both eyes that only spared the upper nasal quadrants. Brain MRI showed a rounded third ventricle tumor with a potbelly expansion of the optic chiasm, suggesting chiasm invasion by the tumor. Optical coherence tomography (OCT) showed the thinning of the retinal nerve fiber layer (RNFL) in the superior and temporal wedges of the right eye and in the temporal wedge of the left one. The tumor was completely removed by employing a frontotemporal craniotomy and a translamina terminalis approach. Histological analysis showed a squamous-papillary CP. Postoperatively, a significant worsening of the visual defect was evidenced on the perimetry, which was related to a marked RNFL atrophy measured with OCT, as compared to the preoperative study. The poor long-term visual outcome in this patient correlated well with the results of postoperative OCT.

Conclusions: Preoperative analysis of retinal atrophy with optic coherence tomography allows a reliable assessment of the patient’s visual outcome in CPs involving the optic chiasm.

Key Words: Craniopharyngioma, optic chiasm, optical coherence tomography, third ventricle, visual outcome

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INTRODUCTION

Craniopharyngiomas (CPs) represent a complex group of epithelial benign (WHO I) tumors presumably originated from cell remnants of the Rathke’s pouch.\(^\text{[6,16]}\) CPs may develop at any point along the pituitary–hypothalamus axis, from the sella turcica to the third ventricle (3V). Very often, CPs cause progressive decrease of visual acuity and/or constriction of the visual fields related to the anatomical distortions these lesions cause on the optic chiasm.\(^\text{[21]}\) The primary development of a CP within the optic chiasm or the invasion of this structure by the tumor is, however, an exceedingly rare event. Only few verified intrachiasmatic CPs have been reported in medical literature to date.\(^\text{[2,5,11]}\) This subgroup of CPs may cause a severe swelling of the optic chiasm similar to that observed in optic chiasm gliomas.

In the present report, we describe a new case of an intrachiasmatic CP diagnosed in an adult woman who showed a progressive worsening of her vision. The tumor was totally removed using a perional translamina terminalis approach. This study presents, in a comprehensive way, the correlation between the patient’s postoperative visual outcome and the structural injury caused to her optic apparatus by this intrachiasmatic CP, evaluated through optical coherence tomography (OCT), a useful tool for quantitative assessment of the structural damage of the retinal nerve fiber layer (RNFL).

CASE DESCRIPTION

This 46-year-old woman was initially admitted to our hospital with headache, progressive decrease in visual acuity, and a fever above 38.5°C for the last month, in addition to polyuria and polydipsia for the last 2 weeks. Upon admission, her neurological exploration was unremarkable. Visual exploration disclosed a reduced visual acuity of both eyes, 20/200 in the right eye and 20/40 in the left one. The initial perimetry evidenced a homonymous left inferior quadrantanopia, whereas the fundoscopic exam did not reveal any abnormality. All her baseline endocrine tests were also normal at this time. The brain MRI displayed a 1.5-cm mixed solid-cystic lesion occupying the lower 3V and adjacent hypothalamic–chiasmatic region [Figure 1a–c]. As the patient was seropositive for HIV infection and had suffered from lung tuberculosis several years before, this lesion was presumed to correspond to a cerebral tuberculoma related to the HIV infection, and specific therapy with isoniazid and rifampicin was initiated.

Six months later, the patient was admitted again due to visual deterioration and symptoms of panhypopituitarism and diabetes insipidus. She was treated with hormone replacement therapy and also received psychiatric replacement therapy and also received psychiatric evaluation for a depressive disorder which had worsened in the last year.

Preoperative neuro-ophthalmological examination

At this point, a thorough neuro-ophthalmological and neuroradiological evaluation was performed. The patient could only see light with her right eye and could distinguish hand movements from 2 m distance with the left one. Visual fields were impaired severely in both eyes with a generalized loss of sensitivity in the right one and sparing of the upper nasal quadrant of the left one (Figure 2a, perimetry on 07/16/2012). A mild blurred temporal border in the left macula was observed on funduscopic exam. OCT (SD-OCT SPECTRALIS, Heidelberg Engineering, Heidelberg, Germany, Software 5.3) was used to reveal slight thinning of the RNFL in the superior and temporal wedges of the right eye (90 µm on average) and atrophy of the temporal wedge in the left eye (Figure 3, preoperative OCT). An MRI study showed a significant enlargement of the 3V lesion, which had a basal cauliflower-like nodule and a cystic upper component [Figure 1d–f]. The chiasmatic (suprasellar) cistern was tumor free,
and both the pituitary stalk and the pituitary gland were anatomically intact. The optic chiasm was grossly distorted, showing a potbelly downward deformation on coronal sections, due to tumor infiltration [Figure 1e]. In addition, on T2-weighted axial images, the optic tracts displayed a “moustache-like” appearance, characterized by a hyperintense signal of edema extending along both optic tracts and into the adjacent hypothalamus [Figure 1d].

**Surgical procedure**

The tumor was approached through a right frontotemporal craniotomy. After opening the Sylvian fissure, the optic chiasm was exposed. It was displaced against the tuberculum sellae and presented a quite swollen and shortened appearance [Figure 4a]. The tumor occupying the 3V was approached through the lamina terminalis, which was found to have a ballooned and thinned aspect protruding above the optic chiasm. The lesion could be dissected out from the 3V walls, except at its solid basal portion where a solid tongue of the tumor infiltrated the right posterolateral region of the chiasm and extended into the swollen proximal portion of the right optic nerve [Figure 4a]. After severing the intrachiasm tumor attachment with careful sharp dissection under microscope magnification, the tumor was totally removed. Pathological diagnosis identified a CP of the squamous-papillary variety [Figure 4b1 and b2].

**Postoperative course and visual outcome**

The patient recovered successfully from the surgical procedure, although her panhypopituitarism, diabetes insipidus, and subjective visual disturbance remained unchanged. Complete removal of the lesion was confirmed on postoperative MRI [Figure 5a], with a small defect or breach at the 3V floor that extended to the junction of the optic chiasm and the right optic tract, just at the site where the tumor infiltrated the chiasm [Figure 5b and c].

Follow-up neuro-ophthalmological assessment 1 week after surgery showed some improvement in the patient’s visual acuity and visual fields. Visual acuity returned to its prior initial values, from only light perception to 20/60 in the right eye and 20/40 in the left one. A marked improvement of vision in the nasal hemi-field in the left eye was observed as well, although only a partial recovery at the temporal-superior quadrant of the right eye (perimetry on 1/26/2012), a result congruent with the invasion of the chiasm’s posterior aspect and right optic tract’s medial aspect (top scheme in B). As the tumor enlarges, it encroaches upon a wider area of the posterior and central chiasm, including both optic tracts, thus damage resulting in a severe visual deficit (second perimetry on 07/16/2012) that only spared the uncrossed temporal inferior fibers from the left retina along the chiasm’s left edge (second scheme from the top in b and brown tract of fibers in c)). After removal of the CP, the persistence of the visual deficit in the lower quadrants of the right eye and in the nasal quadrants of the left eye (immediate postoperative perimetry on 8/6/2012) was congruent with irreversible injury to the chiasm’s posterior aspect and the medial aspect of the right optic tract (third scheme from the top in B and blue tract of fibers in C). The late postoperative perimetry exam (postoperative perimetry on 01/21/2013) showed a significantly worsening of the visual defect, probably due to degenerative changes in retinal fibers associated with gliotic scarring (bottom scheme in B).
A significant worsening of the visual defect was evidenced on the perimetry (Figure 2, fourth perimetry on 01/21/2013), probably due to degenerative changes of retinal fibers associated with gliotic scarring. This damage was congruent with the postoperative retinal atrophy disclosed on the OCT. No tumor recurrence was demonstrated on the last postoperative MRI, performed 1 year after surgery.

DISCUSSION

Intrachiasmatic CPs: An intraventricular topographical variant

CPs with either a primary intrachiasmatic development or true anatomical invasion of the optic chiasm represent a quite exceptional topographical variant, with very few cases recorded in the literature.[2,5,11,19] In all these cases, the tumor largely occupied the 3V and presented the tightest and widest adhesions to the posterior edge of the optic chiasm, which was infiltrated by a tongue-like tumor protrusion.

Therefore, all these lesions corresponded to strictly 3V CPs, which were developed above an anatomically intact 3V floor.[14,15] CPs with a primary intra-3V topography usually push the optic chiasm downward against the tuberculum sellae, causing the chiasm to displace to a prefixed position, presenting a much shorter and wider appearance.[21] In our case, the right hemi-chiasm and the right optic tract...
Chiasm pathways injured by intrachiasmatic CPs: Correlation with visual field deficits

Over 2 million nerve fibers pass through the optic chiasm, in which a spatial segregation of crossed and uncrossed fibers can be evidenced. The majority of fibers that form the uncrossed pathway arise from the ipsilateral temporal hemiretina and the crossed pathway from the contralateral nasal hemiretina. In the early 1960s, Hoyt et al. traced the retinal projections in the chiasm of primates and revealed a rostral-caudal (antero-posterior) spatial segregation of retinal fibers within the chiasm, in which the crossed fibers from the dorsal retina (upper quadrants) cross the chiasm at a caudal (or posterior) position, whereas those from the ventral retina (lower quadrants) tend to cross at a more rostral (or anterior) position [Figure 2c].

Fibers from the macula represent the major bulk of fibers within the chiasm and are found more centrally and caudally (within the central-posterior portion of the chiasm) than the rest of fibers.

Visual disturbances associated with CPs are mainly related to the particular type of anatomical distortion to the optic chiasm caused by the tumor. In the case of intrachiasmatic CPs, a topography centered at the chiasm–infundibulum junction, the tumor first encroaches upon the posterosuperior and central portion of the optic chiasm, the area where the crossed fibers from macular and upper nasal retinal quadrants follow their course to the opposite optic tract. Consequently, the chiasm injury associated with this topographical variant produces the worst visual field defect in the temporal lower quadrants.

The sequential stages of visual field impairment in our patient were congruent with the topography and the anatomical region of the chiasm injured by the tumor. At the initial perimetry, the patient presented a homonymous left inferior quadrantanopia, worse in the right eye, a deficit explained by a damage to the posterior edge of the optic chiasm, and the medial aspect of the right optic tract [Figure 2a]. This is the region of the chiasm initially infiltrated by the tumor [Figure 1], in which the tongue-like intrachiasm tumor extension damaged the axonal bundles coming from the upper nasal quadrant of the left retina and the upper temporal quadrant of the right one [Figure 2c]. The right eye is obviously the one suffering the most severe loss of vision, as the damage involves a significant amount of the bundles from the foveal and perimacular areas of the right retina, including the uncrossed fibers from the temporal quadrants.

The rapid worsening of patient’s visual deficit over the following 6 months [Figure 2a, perimetry on 07/16/2012] was in correlation with encroachment or invasion of the central chiasm and right optic tract by the tumor, which spared the uncrossed temporal inferior fibers from the left retina. [Figure 2b, c] At this time, the OCT
proved an overall thinning of the RNFL that involved mainly the superior wedges in both retinas, a result in agreement with the tumor invasion of the posterior-upper chiasm region [Figure 3, preoperative OCT]. The OCT also showed atrophy in the left eye’s temporal wedge which is not well explained by the tumor extension into the chiasm, although this finding has been frequently reported for chiasm compressions by macroadenomas.⁹

**Surgical removal and visual outcome of intrachiasmatic CPs: The predictive value of OCT**

Intrachiasmatic CPs essentially represent a particular variant of 3V CPs which are infiltrating the optic chiasm.¹⁷,²¹-²³ Consequently, the surgical removal of these lesions requires the use of an approach to the 3V providing an optimal view of the tumor attachment to the optic chiasm. The frontotemporal or pterional approach, besides providing an optimal view of the optic chiasm and adjacent cisternal spaces, also permits the opening of the lamina terminalis for the removal of the intra-3V component of the tumor.¹⁰,¹⁷ The use of a translamina terminalis approach allowed us to remove the intrachiasmatic CP described here, through the release of the tumor attachment to the chiasm under microscopy magnification.

One week after the procedure, the first postoperative perimetry examination showed a significant improvement in visual acuity in the nasal field of the left eye but only a partial recovery of vision in the temporal superior quadrant of the right eye (Figure 2a, perimetry on 08/06/2012). These findings are concordant with an irreversible lesion of the axonal pathways at the chiasm-right optic tract junction, the area infiltrated by the tumor, where a residual anatomical defect is evident on the postoperative MRI [Figure 5]. Apparently, the inferior nasal fibers from the right retina that cross the chiasm through its anterior edge, in addition to the temporal uncrossed fibers of the left eye, seemed to have been spared following CP removal [Figure 2b]. The results of the postoperative OCT performed around this time showed, however, that the degree of atrophy in the RNFL had progressed as compared to the prior study, especially at the superior area of the right retina and the temporal one in the left eye [Figure 5]. Six months later, a marked deterioration of the visual status was confirmed in the perimetry, with preservation of islands of vision only in the upper temporal quadrants of both eyes, the result congruent with a severe bilateral RNFL atrophy (Figure 2a, perimetry on 01/21/2013). Progressive degeneration of the retinal pathways occurred within a few months after the surgical procedure, a process probably linked to the development of a glial scar at the site of the intrachiasmatic wound.

No conclusive explanations regarding the initial incongruence between the early postoperative visual improvement but worsening OCT results in this patient can be stated. The greater atrophy in the RNFL immediately following the surgical procedure would logically reflect the injury to the chiasm associated with the surgical maneuvers employed to dissect the intrachiasmatic tumor attachment. RNFL thinning reflects irreversible retinal ganglion cell damage, which usually occurs after prolonged compression of the visual pathways.¹⁴,²²,²⁴ Hence, any structural damage of the retina detectable with OCT predicts the existence of irreversible injury at the optic chiasm/optic tracts that will associate visual loss to be confirmed on standard automated perimetry.¹¹,¹²,²⁴ Despite the apparent partial visual recovery observed on our patient’s initial postoperative perimetry, the OCT already reflected structural damage in the retinal ganglion cells, which had advanced with respect to the preoperative study and anticipated a long-term worsening of visual function.

The final question to consider is the degree of removal to be attempted when dealing with a CP that infiltrates the optic chiasm. A generalized surgical attitude cannot be recommended, and the most appropriate, judicious decision should be established on an individual basis. The high risk of irreversible chiasm injury associated with radical removal of a CP must be pondered with the rate of tumor recurrence, taking into consideration the preoperative visual status, the patient’s age, and, above all, the extent and strength of tumor attachment. The impossibility of applying any type of radiosurgery on the optic chiasm for the treatment of CP remnants adds an extra level of complexity to this issue. Preoperative OCT assessment of the structural damage to the visual pathways may help to predict the potential reversibility of visual impairment and provides helpful insight to guide the intraoperative surgical decisions.

**CONCLUSIONS**

Intrachiasmatic CPs represent an exceptional variant of 3V CPs characterized by the invasion of the postero-central region of the optic chiasm. A characteristic potbellied deformation of the optic chiasm visible on preoperative MRI points to CP infiltration of this structure. Tumor encroachment of the chiasm’s posterior aspect and adjacent optic tracts usually cause a visual deficit involving initially the temporal lower quadrants. The pterional approach combined with the opening of the lamina terminalis provides an optimal exposure of any CP extension into the optic chiasm. Preoperative analysis of retinal atrophy with OCT allows a reliable assessment of the patient’s visual outcome and it may help to decide about the degree of CP removal to be performed.

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**Abbreviations**
CP: Craniopharyngioma; MRI: Magnetic resonance imaging; OCT: Optical coherence tomography; RNFL: Retinal nerve fiber layer. 3V: Third ventricle; 3VF: Third ventricle floor.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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