PITFALLS IN THE DIAGNOSIS OF CRANIOPHARYNGIOMA: 2 CASE REPORTS

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SUMMARY

Two cases of craniopharyngioma with delayed diagnosis are presented.
Patient 1 had mild visual loss that initially had been attributed to pigment epithelial detachment in the macular area. Patient 2 had blurred vision in the left eye, although visual acuity was 10/10 at both eyes. She had a history of a posttraumatic neurosurgical procedure and was treated for blepharospasm. Both patients had initially negative CT imaging. Visual field defects suggested a chiasmal lesion and incited to additional neuroradiological investigation. Magnetic resonance imaging revealed a craniopharyngioma in both cases.

RÉSUMÉ

Deux cas cliniques de craniopharyngiome diagnostiqué tardivement sont présentés. Le 1er patient avait une baisse modérée de l'acuité visuelle, qui avait été attribuée initialement à un décollement de l'épithélium pigmentaire maculaire. La seconde patiente se plaignait d'une vision floue à l'oeil gauche. L'acuité visuelle était de 10/10 aux deux yeux. Cette patiente avait des antécédents neurochirurgicaux posttraumatiques et avait été traitée pour blépharospasme. La tomodensitométrie computérisée était négative dans les deux cas. Les champs visuels étaient très suggestifs pour une lésion chiasmatique justifiant des examens neuroradiologiques plus précis. L'IRM a confirmé le diagnostic de craniopharyngiome dans les 2 cas.

SAMENVATTING

Twee gevallen met een uitgestelde diagnose van craniopharyngioom worden voorgesteld.
Patiënt 1 had een matig visusverlies dat initieel werd toegeschreven aan een pigmentbladloslating in de maculaire zone. Patiënte 2 kloeg van een wazige visus in het linker oog, alhoewel de gezichtsscherpte beiderzijds 10/10 bedroeg. Deze patiënte had antecedenten van een neurochirurgische ingreep na trauma en was behandeld voor blefarospasme.
Beide patiënten hadden aanvankelijk een negatief CT-scan onderzoek.
De gezichtsvelddefecten suggereerden echter een letsel thv het chiasma en zetten aan tot bijkomende diagnostische beeldvorming. Kernspintomografie beeldvorming bracht in beide gevallen een craniopharyngioom aan het licht.

KEY-WORDS

Craniopharyngioma, visual field, delayed diagnosis, MRI

MOTS-CLÉS

Craniopharyngiome, champ visuel, diagnostic tardif, IRM

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INTRODUCTION

Bitemporal hemianopia is the most typical ophthalmological presentation of tumors in the chiasmal region. Nevertheless there are many exceptions to this rule and the most constant and reliable ophthalmological finding in chiasmatic lesions is an unilateral temporal visual field defect confined to the vertical meridian. Craniopharyngiomas are rare but fast-growing tumors, presenting frequently with atypical progressive visual acuity loss and atypical visual field defects (3).

There appears to be a bimodal age incidence, with one peak in childhood and a second peak

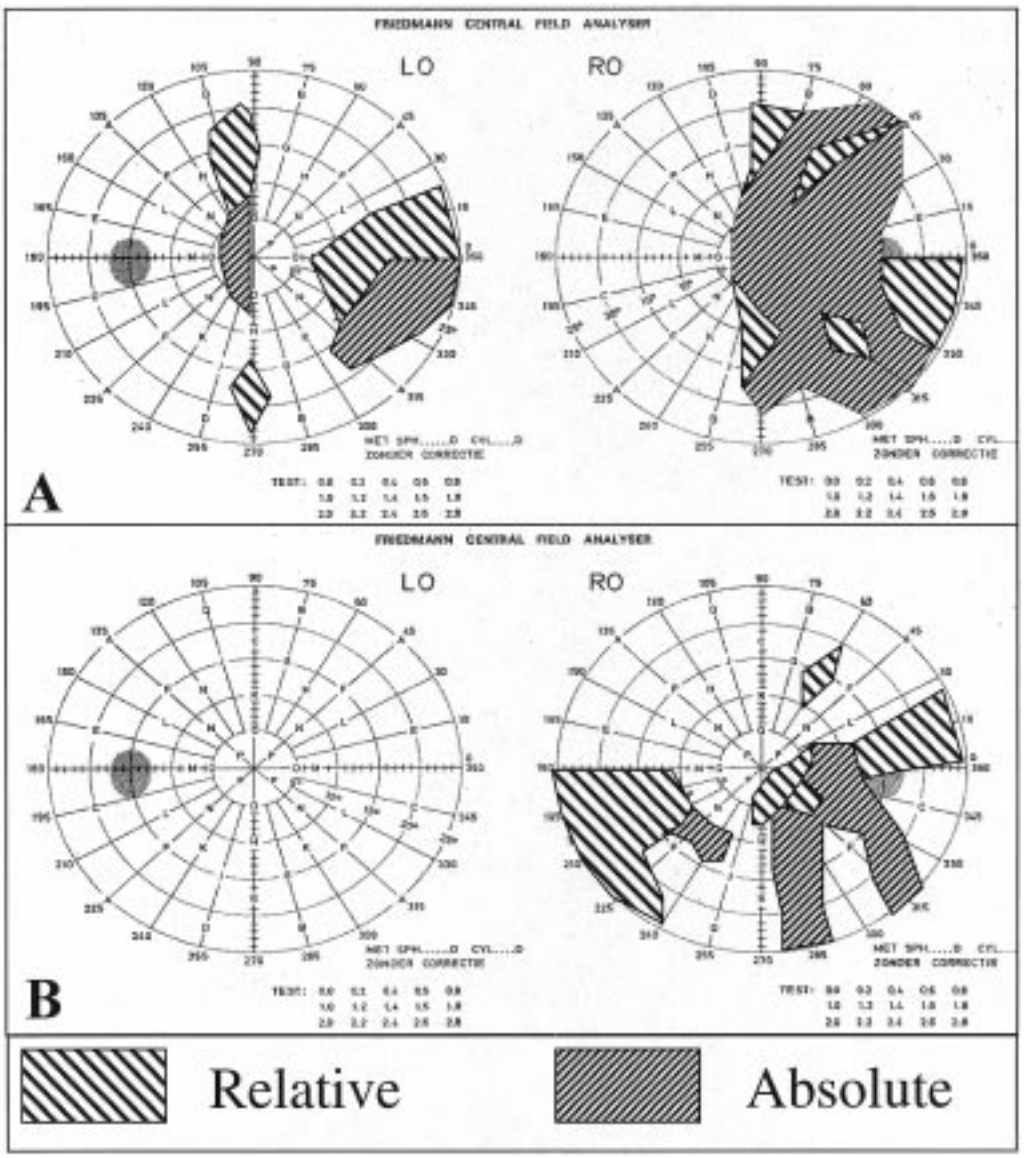

Fig 1. Central visual field of patient 1
A. pre-operative
B. post-operative (1 month after neurosurgery and radiotherapy)

in adults 40-70 years of age. Both sexes are equally affected. Due to their cystic appearance these tumors may be easily overlooked on routine neuroradiological imaging.

CASE REPORTS

CASE 1

A 60-year-old male patient presented with a 2 months history of deteriorating vision in the left eye and more recently also in the right eye.
When first seen by an ophthalomologist vision was 7/10 in the right eye and 6/10 in the left eye. Amslergrid showed temporal metamorphopsia in both eyes. There was no afferent pupillary defect. Slit lamp examination was normal. Fundoscopy and fluorescein-angiography showed a nasal paracentral pigment epithelial detachment in both eyes without underlying choroidal new vessels. Optical coherence tomography showed a normal foveolar depression in both eyes. The patient was advised to use visual aids and extra light while reading.
CT imaging without contrast enhancement was negative.

When seen 3 months later, the patient mentioned a further deterioration of vision in both eyes and was aware of ''skipped areas''. Visual acuity was counting fingers in the right eye and 4/10 in the left eye. Amslergrid showed both metamorphopsia and temporal scotoma in both eyes. Fundoscopy was identical to the examination 3 months earlier. The optic disc appeared normal in both eyes.

Central visual field testing revealed a temporal defect confined to the vertical meridian in the right eye. In the left eye a milder temporal defect confined to the vertical meridian and a less relevant nasal defect were present (fig. 1a).

Electroretinography was normal, but pattern visual evoked responses showed a grossly reduced amplitude and delayed response in both eyes.

The visual field results incited to additional neuroradiological investigation.

MR imaging revealed a cystic mass situated at the midline in the suprasellar cistern, highly suggestive of a craniopharyngioma (fig. 2). The size of the lesion was 2 × 2.5 × 2.7 cm and the cyst's wall was enhanced with contrast injection.

An endocrinological evaluation was normal.

The patient was referred to the department of neurosurgery where a neurosurgical procedure revealed a suprasellar mass compressing the chiasm and the right optic nerve. Dissection of

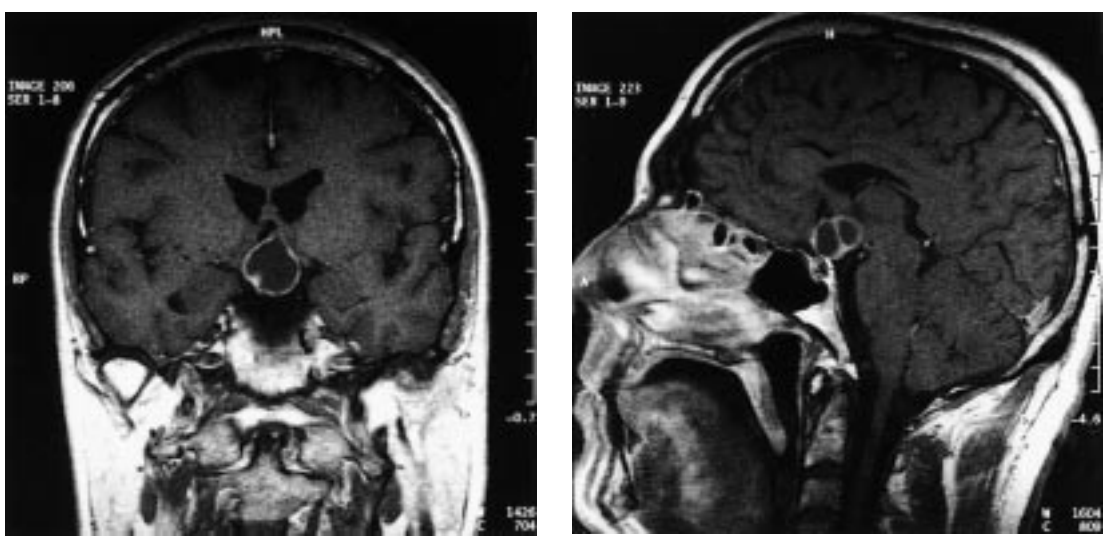

Fig 2. MRI of patient 1: cystic mass at the midline in the suprasellar cistern
A. coronal view B. sagittal view

the mass appeared to be very difficult and a complete removal was not possible.
The diagnosis of craniopharyngioma was confirmed by anatomopathological examination.
Neurosurgical subtotal resection of the craniopharyngioma was followed by radiotherapy. One month later the patient had recovered a visual acuity of 10/10 in both eyes and a visual field testing was normal in the left eye and showed a mild temporal defect in the right eye (fig 1b).

CASE 2

A 52-year-old female patient underwent trepanation after a skullbase fracture 33 years ago. She was treated for blepharospasm with botulinum toxin injections. She noticed blurred vision in the left eye for one month and complained of headache. Vision in both eyes was 10/10 with optimal correction. She had moderate corneal astigmatism in the left eye. Amslergrid was normal in both eyes. Color vision was normal in both eyes. There was no afferent pupillary defect. The optic discs appeared normal. Visual field testing showed a defect in the temporal inferior quadrant in the left eye (fig. 3a).
Pattern visual evoked responses showed normal amplitudes and latencies in both eyes.
CT scan showed a hypodens area in the left temporal hemisphere, that could be attributed to the trauma and surgery 33 years ago. There was no evidence of an expanding lesion.

Two months later the patient complained of persisting headache and mentioned reading problems with frequently missing the next line when reading. Vision was 3/10 in the right eye and 8/10 in the left eye. Amslergrid was normal in the right eye and in the left eye the temporal half of the grid was missed. Visual field examination again showed a denser temporal inferior field defect confined to the vertical meridian in the left eye (fig. 3b). There was no afferent pupillary defect. The optic discs appeared normal.
The pattern visual evoked responses showed reduced amplitudes and delayed answers at the right eye.
MR imaging showed a cystic mass in the chiasm, without involvement of the optic nerves

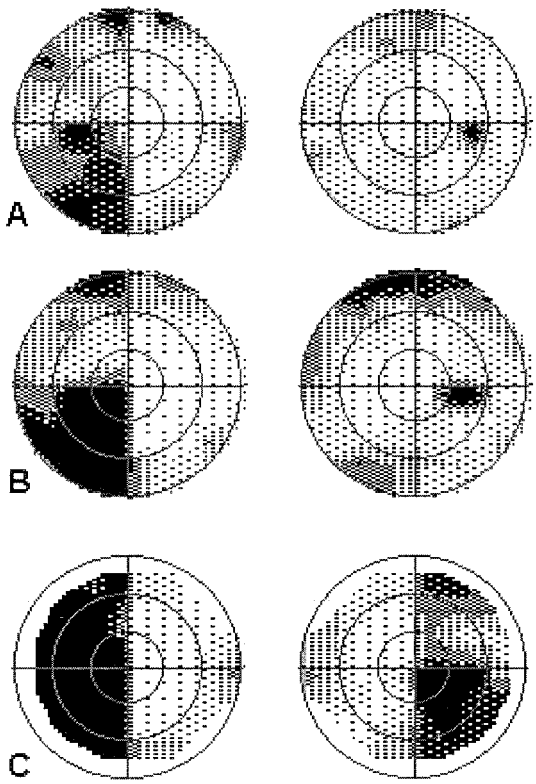

Fig 3. Automated perimetry of patient 2: evolution of the temporal defect in both eyes
A. initial visual field
B. 2 months later
C. 6 months later

(fig. 4). The differential diagnosis of craniopharyngioma, glioma or cyst of Rathke's pouch was put forward. The old (posttraumatic) lesion in the left temporal lobe was confirmed and appeared stable.
An endocrinological evaluation was normal.

The patient was referred to the department of neurosurgery. Because the preferential neuroradiological diagnosis was optic glioma and because the clinical situation was stable the neurosurgeons decided to a non-invasive follow-up with clinical and neuroradiological examinations.
Six months later the patient presented with an acute deterioration of vision in the right eye since 2 weeks. Visual acuity was 1/10 in the right eye and 10/10 in the left eye. Central visual field examination showed a manifest worsening in both eyes. The temporal defect in the

left eye progressed to a total temporal hemianopia. In the right eye a temporal defect confined to the vertical meridian now appeared (fig. 3c).

Because of the deterioration of visual acuity and visual field the patient was immediately referred to the department of neurosurgery. During the operative procedure a retrochiasmal lesion was found. The anatomopathological examination confirmed the diagnosis of craniopharyngioma.

One month after surgery visual acuity was 10/10 in both eyes. There was a homonymous left inferior quadrantanopia post-surgically.

DISCUSSION

Technical developments have provided the ophthalmologist with powerful new tools for the investigation of lesions affecting the visual pathways, but over-reliance on one technique alone may have serious consequences. It should be emphasized that a negative CT scan does not unequivocally rule out a space occupying lesion and more detailed neuroradiological imaging is mandatory where this is suspected on clinical grounds (6).

Craniopharyngiomas are benign intracranial tumors which arise from embryonic squamous cells thought to be remnants of Rathke's pouch. Although they are not invasive, they are difficult to remove surgically because they originate in close apposition to the hypothalamus, pituitary stalk, optic chiasm, and carotid arteries and frequently have adhesions to these structures.

Craniopharyngiomas are always located, at least in part, in the suprasellar cistern, where they compress the intracranial portion of the anterior visual sensory system. The tumor may compress the optic nerves, optic chiasm and optic tract from any direction.

Some craniopharyngiomas may project into the third ventricle to such an extent that they obstruct the cerebrospinal fluid flow and produce hydrocephalus.

The tumor is always surrounded by a capsule with variable consistency that may be partially calcified. The body of the tumor may be cystic, solid or both.

The symptoms and signs produced by craniopharyngiomas are determined not only by the size and position of the tumor but also by the age of the patient.

Adults with craniopharyngiomas most often experience slowly progressive visual loss, over many months or even years. In addition to visual loss some adult patients also develop symptoms related to dysfunction of the hypothala-

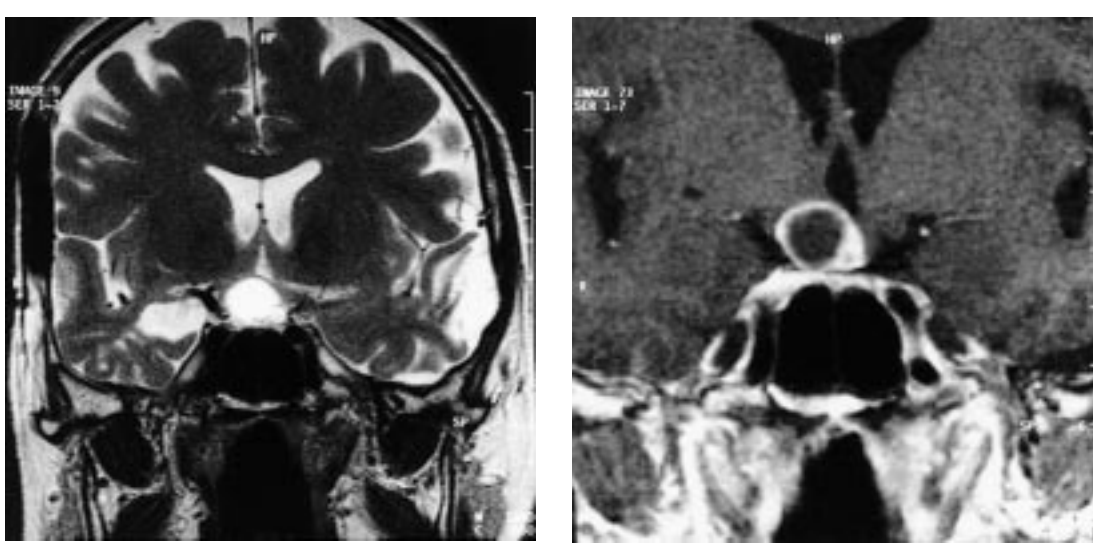

Fig 4. MRI (coronal view) of patient 2: cystic mass in the chiasm, without involvement of the optic nerves
A. with contrast injection B. without contrast injection

mus, anterior pituitary or both. Such patients may complain of impotence, galactorrhea, reduced energy, weight gain or symptoms of diabetes insipidus (3,10).

Unlike adults, children and adolescents often develop symptoms and signs of increased intracranial pressure and abnormalities that occur from involvement of the hypothalamic-pituitary axis, including growth failure, retarded sexual development, infantilism, obesity, diabetes insipidus (11).
Although the majority of children and adolescents with craniopharyngiomas suffer loss of vision, the extent of visual loss is usually not appreciated until very late or when other abnormalities become apparent. Thus, by the time such patients are first examined, they may already have severe optic atrophy.

The diagnosis of craniopharyngioma is often suspected from the patient's clinical presentation.
CT scanning and MR imaging, when performed properly, virtually always show the lesion with its cardinal features: location in the suprasellar region and composition of solid and cystic portions with frequent calcification (1). In some cases, however, the tumor is primarily cystic and fills the suprasellar cistern. Such a lesion may appear isodense with CSF on CT scan, and as such may remain undiagnosed unless it is recognized that the cistern is enlarged and that its normal pentagonal shape is distorted.
The CT technique is superior to MR imaging in the detection of calcification. The MR imaging technique is superior to CT for determining tumor extent and provides valuable information about the connections of the tumor with the surrounding structures. As a consequence CT and MR imaging have complementary roles in the diagnosis of craniopharyngiomas (4,12).

The treatment of craniopharyngioma is somewhat controversial (2, 7,8,9,13). In all patients total removal of the tumor should be attempted. If total removal is deemed too dangerous because of adhesions to surrounding structures, a subtotal resection should be performed, followed by radiotherapy. In many cases a subtotal resection that controls the visual symptoms without causing endocrine dysfunction (panpituitary insufficiency) is preferred. Patients should then be monitored by clinical examination and neuro-imaging for evidence of tumor growth.

Morbidity is due to visual, hypothalamic, endocrine and intellectual disturbances. Treatment has often a negative impact on the patient's quality of life (11). Survival for children diagnosed with craniopharyngioma is excellent even with relapse, although postoperative visual and endocrinological morbidity are high. Failures occur, both radiographically and clinically, typically in the first 3-4 years after surgery, suggesting a need for close surveillance with neuro-imaging and clinical examination, particularly in younger children. Lack of calcification at the moment of diagnosis has been associated with a lower relapse rate (5).

At post-surgical follow-up most patients have an improved level of visual function. Adequate hormonal supplementation and regular pharmacological adjustments are often necessary. Small stature, obesity, headache, and emotional and sexual disturbances are frequent causes of long-term disability notwithstanding adequate drug regimens. The majority of patients resume normal social life and are fully reintegrated (13).

Both patients presented in this paper showed a progressive visual loss. Hemianopic temporal visual field defects were suggestive for tumors in the chiasmal region. In both cases nonrelated ocular signs and negative CT imaging delayed the diagnosis. Notwithstanding negative CT imaging, hemianopic temporal visual field defects, even when unilateral, should be investigated by additional and detailed MR imaging.

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