Neuro-ophthalmic manifestation of suprasellar craniopharyngioma

Abstract

Introduction :

Craniopharyngiomas are histologically benign brain tumours arising from the remnants of Rathke's pouch. Despite their benign appearance, their clinical behaviour is aggressive, causing serious morbidity by damaging the optic chiasm. Epidemiological studies suggest a bimodal age distribution with a peak in childhood and a second peak in the sixth decade. Major clinical symptoms are progressive decline of visual acuity and constriction of visual fields and bilateral optic atrophy. The classic field abnormality associated with optic chiasma disorders is a bitemporal hemianopia. Magnetic Resonance Image is particularly useful for the topographic and structural analysis of the tumor and is the most important imaging modality used to plan the surgical approach.

Purpose :

To report a neuro-ophthalmic manifestation of suprasellar craniopharyngioma.

Case Report :

A 31 year old female came to Cicendo Eye Hospital with the left eye cannot see anything and blurred vision in the right eyes since 2 weeks ago. This patient complains, with history of severe headache, nausea and seazure since 2 months ago and body weakness 1 month after delivery in 6 years ago. General physical examination was within normal limits. Visual acuity was no light perception in left eye and finger counting in right eye. Both eyes deviated inward and limitation lateral eye movement. Decreased pupillary reflex occurred in both eyes. Relative afferent pupillary defect (RAPD) was grade 4 in left eye. The anterior segment examinations were within normal limits. Hemianopia temporal visual field defect in the right eye by confrontation test. Both optic discs were pale, and the rest of posterior segment examinations were within normal limits. Optic Disc Optical Coherence Tomography of both eyes revealed decreased optic nerve fiber layer thickness. Magnetic Resonance Image (MRI) showed suprasellar craniopharyngioma. This patient was diagnosed with bilateral optic disc atrophy, sixth nerve palsy and right hemianopia temporal caused by suprasellar craniopharyngioma. The patient was revered to Neurosurgery department.

Conclusion :

Ocular symptoms in suprasellar craniopharyngioma can manifest as visual field defect and eye movement abnormalities. Proper ocular examination and evaluation can determine early therapy and therefore produce a better prognosis for the patient.

I. Introduction

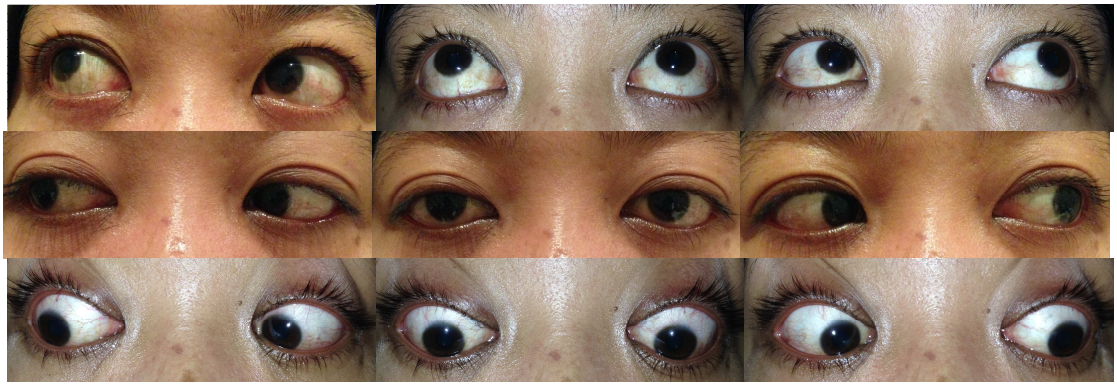
Craniopharyngiomas are benign but aggressive tumours arising along the path of the craniopharyngeal duct and most frequently located in the sellar or suprasellar region. Studies report a bimodal age distribution with one peak before age 15 and another in the sixth and seventh decades. The tumour are close relationship to the anterior visual pathway gives rise to a variety of visual signs and the classical presentation is with bitemporal hemianopia or inferior quadrantanopia due to chiasmal compression and bilateral optic atrophy.[1,2]

Systemic symptoms include effects of raised intracranial pressure and disruption of the hypothalamic pituitary pathway. Computed tomography (CT) classically shows a heterogeneous suprasellar mass with calcification and extension into the chiasm or third ventricle. However, calcification may be absent in adults and magnetic resonance imaging (MRI) is more sensitive in detecting cystic components and delineating tumour extension into the chiasm or third ventricle. The treatment usually combines total or subtotal resection with adjunctive radiotherapy. The tumour has a strong propensity for recurrence. [1,2]

II. Case Report

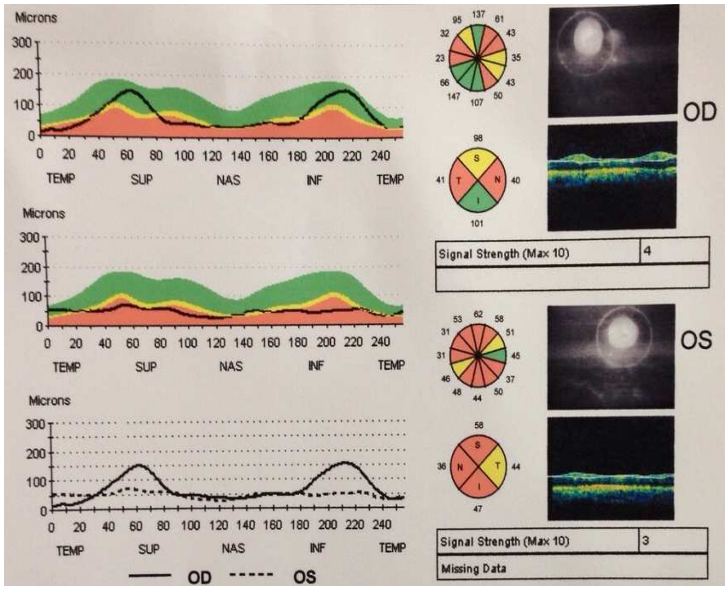
A 31 year old female came to Cicendo Eye Hospital with the left eye cannot see anything and blurred vision in the right eyes since 2 weeks ago. This patient complains, with history of severe headache and nausea since 2 months ago. She got seizure in several time 2 months lately and body weakness 1 month after delivery in 6 years ago. The patient's period was regularly each month. She never got head trauma, ocular movement pain, and redness in eye.

General physical examination was within normal limits. Visual acuity was no light perception in left eye and finger counting in right eye. Both eyes deviate inward and limitation lateral eye movement. Decreased pupillary reflex occurred in both eyes. Relative afferent pupillary defect (RAPD) was grade 4 in left eye. Rest of anterior segment examinations were within normal limits. Hemianopia temporal visual field defect in the right eye by confrontation visual field test. Papil pallor was determined by direct funduscopy on both eye, and the rest of posterior segment examinations were within normal limits.



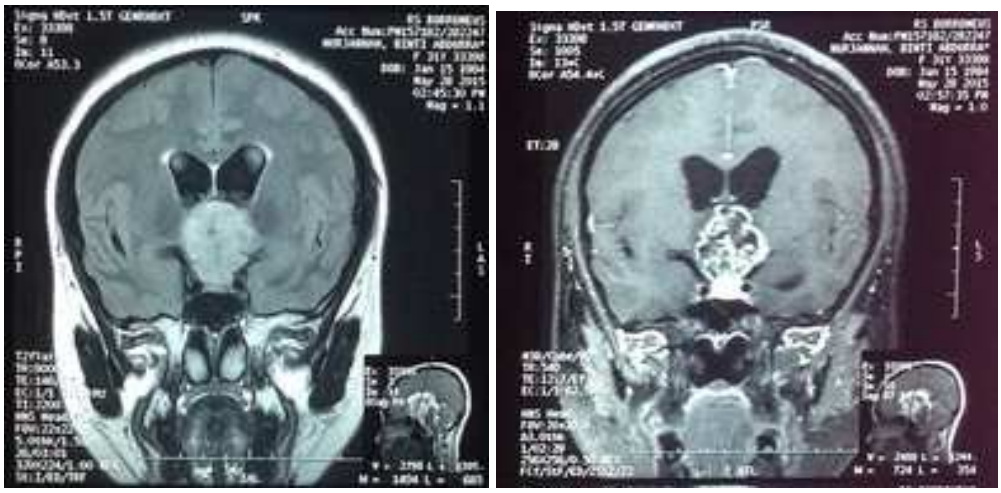
Picture 1. Nine cardinal eye movement shows limited movement to lateral.
Source : National Eye Center Cicendo Eye Hospital.

Optic Disc Optical Coherence Tomography of both eyes revealed decreased optic nerve fiber layer thickness. Average retinal nerve fiber layer was 69,97µ and 46,34µ in the right and the left eye respectively.



Picture 2. Optic Disc Optical Coherence Tomography revealed decreased optic nerve fiber layer thickness
Source : National Eye Center Cicendo Eye Hospital.

Magnetic resonance imaging was requested, and it revealed complex cystic mass and calsification within the tuberculum sella and clivus occipital, 30 x 40 x 45 mm in size. The mass impinging on third ventricle and foramen monro induces hydrocephalus and lateral ventricle widen. Lateral displacement of optic nerve, anterior displacement of chiasma and optic tract compression. Compression of the hypothalamus, tuber sinerium, corpora mamilaria, fornix, left frontomedial frontal lobe and left temporal medial lobes, fossa interpeduncularis, pedunculus cerebri and pons from the mass. Neurohypophyse and adenohypophyse on sella tursica seen normal. Complex cystic mass suggest a craniopharyngioma.



Picture 3. Coronal image of magnetic resonance imaging shows a pituitary adenoma with elevation and compression of the optic chiasm and the mass impinging on third ventricle and foramen monro.
Source : National Eye Center Cicendo Eye Hospital.

Picture 4. Magnetic resonance imaging scan patient shows showing suprasellar lesion with chiasmatic displacement (Left). Lateral displacement of optic nerve, anterior displacement of chiasma and optic tract compression (right).
Source : National Eye Center Cicendo Eye Hospital.

Based on the clinical features and radiologic imaging finding, this patient was diagnosed with bilateral optic disc atrophy and bilateral sixth nerve palsy and right temporal hemianopia caused by suprasellar craniopharyngioma. The prognosis of this patient is *quo ad vitam dubia ad bonam* and visual outcome is *quo ad functionam dubia ad malam.* The patients was reffered to Neurosurgery department, though the patient choose the traditional alternative treatment.

III. Discussion

Craniopharyngiomas account for 1–4% of all intracranial tumours and 20% of the tumours of the sellar and chiasmatic region. Craniopharyngiomas most frequently arise in the pituitary stalk and project into the hypothalamus. They can extend anteriorly, posteriorly, superiorly or laterally. These are predominantly suprasellar tumours, although they may be found within the sella tursica. In their usual location they may originate from squamous epithelial remnants of the Rathke pouch and sac. There is a possibility that craniopharyngiomas can originate anywhere along the tract of the obliterated craniopharyngeal duct.[1-4]

Craniopharyngiomas are approximately equally common in males and females, the male to female ratio is 1.1–1.4:1. There is a bimodal age distribution, with one peak in children between 5 and 14 years, and a second peak in adults between 50 and 75 years of age, but the majority of the patients are children. The patient in this case is adult women, similar in one case report that a 34 years-old woman infrasellar craniopharyngioma.[5-6]

It is generally accepted that two different subtypes of craniopharyngiomas exist. The classic adamantinomatous type appears to be present in more than 95% of paediatric cases, and is found most often in the suprasellar space, where they adhere to the adjacent vessels and indent the floor of the third ventricle. The papillary squamous variant is almost exclusively seen in adults and represents close to 30% of all the craniopharyngiomas seen in this population.[2,3,7,8]

These are typically slow growing tumors. Symptoms usually develop insidiously and become obvious only after it attains a size of 3 cm. The tumour in this patient is 30 x 40 x 45 mm. The presentation of craniopharyngioma varies with the size, location and extension of the tumour. The most common symptoms include headache, hormonal imbalance and visual impairment. Most women complain of amenorrhea.[1,2,9]

Headache has been documented as the most common systemic symptom of craniopharyngioma, reported in 42–68% of adults. Other systemic symptoms include endocrine failure. Diabetes insipidus was the most common pituitary deficiency. Symptoms of raised intracranial pressure such as nausea and vomiting were often dismissed as non-specific. [1,2,9,10]

This patients had a history of seizure. The convulsions have been observe in 20 to 50 percent of all patients with brain tumour. A first seizure during adulthood is always suggestive of

brain tumour and has been the most common initial manifestation of primary and metastatic neoplasm.[2]

The most common presentation in many studies is blurred vision. Craniopharyngioma grows from the squamous epithelium on the infundibulum between the under surface of the brain and the upper surface of the pituitary. Hence, the tumour tends to compress the chiasm classically from behind and above. Posterosuperior compression of the chiasm produces the classic field abnormality is a bitemporal hemianopia. The peripheral temporal visual fields usually are involved first. Parasellar lesions that involve the chiasm whether compressing or infiltrating this area produce gradually progressive, bilateral, often asymmetric visual loss. Quadrantanopia is usually of the inferior quadrant because of the superior location of the fibres. Temporal hemianopic scotomas are common due to compression of the posterior chiasmal angle.[1-3,5-10]

Craniopharyngioma thus behave differently from other sella and suprasellar tumours such as pituitary adenoma. Craniopharyngioma is characterized by pleomorphism due to its high recurrence tendency and a wider variety of field defects. In contrast, pituitary adenoma classically has a bitemporal field defect that post-decompression tends to show rapid and marked improvement in visual acuity and visual field, which often continues in the long term. [2,8-10]

The afferent visual system is affected at presentation in most cases of craniopharyngioma. The pastient has a manifestation of bilateral abducens nerve palsy. In Baskin and Wilson's series of childhood and adult patients with craniopharyngiomas, 72% had visual deficits and 7% had third or sixth cranial nerve palsies. One case report patient with a Rathke's cleft cyst who presented with rapidly progressive bilateral 6th nerve palsy. Magnetic resonance imaging showed a hypodense mass that filled the sella and compressed the cavernous sinus. The increased intrasellar pressure may have been the cause for the bilateral abducens nerve palsy, which improved following surgical resection. [8,11]

In chiasmal syndromes, the optic discs may show no visible abnormalities initially, even in the face of significant visual field loss. More commonly, there is subtle evidence of optic neuropathy, such as peripapillary retinal nerve fiber layer dropout and mild disc pallor. With more damage, the optic discs show typical atrophy, often in the temporal portion of the disc

corresponding to the papillomacular bundle of retinal nerve fibers and the nasal fibers resulting in band atrophy. Cupping of the disc may increase. [1,10]

Optic atrophy occurs in up to 60% of patients and is commoner than disc swelling. One study reported 52% patients had bilateral optic atrophy, which was asymmetric in almost 50% cases. Some patient had papilloedema or had normal fundi. Long-standing chiasmal compression produces bitemporal hemianopia with loss of half of the maculopapillary bundle on the temporal side of the disc and the nasal hemiretinal fibres passing into the nasal side of the disc: so-called 'bow-tie atrophy'. An affected optic nerve may produce more central loss, with impaired visual acuity. dyschromatopsia and an RAPD on the affected side.[1,10]

Should raised intracranial pressure occur, papilloedema occurs but only in the remaining temporal retinal nerve fibres streaming into the top and bottom of the disc from the intact temporal retina. This swelling of the superior and inferior nerve fibre bundles on the disc was named 'twin-peaks papilloedema' by Hoyt. Most tumors that produce a chiasmal syndrome do not cause increased intracranial pressure and thus are not associated with papilledema.[1,10]

The suspicion of a craniopharyngioma is based on clinical and radiological findings. By MRI examination, the tumor is of variable T1 signal, often hyperintense. The T1 hyperintensity is usually secondary to high protein content in the cyst fluid. However, other causes of T1 hyperintensity in craniopharyngiomas have been described fat, cholesterol, hemorrhage, or even mild calcification. The mixed solid and cystic nature of the tumor is clearer on MR than on CT. Occasionally with small cysts, even on postcontrast study, the heterogeneous nature of the tumor may not be apparent on CT. This problem does not occur with MR. Even small cysts are identifiable and the smaller solid portion of the tumor is usually heterogeneous or even isointense with brain.[12]

The magnetic resonance is the imaging modality of choice. A magnetic resonance examination should include thin T1 sagittal and coronal sections both pre- and postcontrast through the sella and suprasellar region. It is useful to include a precontrast fat saturation T1sequence as it will help to identify the posterior pituitary. The diagnosis is finally proven by histology. [5,10,12]

Currently, craniopharyngioma is treated primarily by trans-sphenoidal or transcranial surgery, whereas post-surgical radiotherapy is not routinely applied in all patients. Chen et al

reported of the 36 patients, seven had total resection of the craniopharyngioma. Twenty-five had subtotal resection, 18 of these underwent radiotherapy postoperatively. Six patients required shunt insertion for associated hydrocephalus. Recurrent disease is treated by repeat surgery and or radiotherapy. The endoscopic endonasal approach allows improved visualization, avoids brain retraction, and there are no external scars. [4,5,10]

Patients with craniopharyngioma tend to have poorer visual outcomes, probably as a result of delayed diagnosis, with long-standing compression of visual structures. Visual outcome in this patients is *quo ad functionam dubia ad malam.* Craniopharyngioma are also associated with more severe anatomic distortion and infiltration leading to increased difficulties in surgical removal. [1,10]

Early and accurate diagnosis may lead to a better visual function. The survival prognosis of patients treated for craniopharyngioma is favourable, with reported 10-year survival rates of approximately 90% in both adults and children. Unfortunately, adult patients treated for craniopharyngioma show persistent impairment in quality of life, especially in the physical subscales. The prognosis of this patient is *quo ad vitam dubia ad bonam.*[13,14]

Tumour recurrence is common and thought to occur in 20–40% of cases. The literature documents examples of craniopharyngioma recurrences along the surgical tract, as well as remote ipsi and contralateral metastases via cerebrospinal fluid seeding. One study has report of a craniopharyngioma of papillary type to exhibit metastatic behavior. The tumor spread opposite the side of craniotomy.[14-16]

IV. Conclution

Craniopharyngiomas are rare benign tumors of the sella rare a with low grade histological malignancy. Despite high survival rates quality of life is frequently impaired in long-term survivors due to sequel becaused by the anatomical proximity of the tumor to the optic nerve, pituitary gland, and hypothalamus. But multi center approach with surgical expertise in such extensive cases can save the patient from the associated morbidity and mortality. [9]

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