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ATYPICAL OPHTHALMIC MANIFESTATIONS OF CHIASMAL TUMOURS

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Abstract :

Bitemporal hemianopic visual field loss, headache, endocrinopathy and other signs of raised intracranial tension are the most common presenting features of chiasmal tumours. We report two patients with chiasmal tumours presenting with atypical clinical features. One patient presented with bilateral optic disc cup ratio of 0.6 simulating normotensive glaucoma was later diagnosed with craniopharyngioma. Another patient presented with features of retrobulbar neuritis with sudden onset unilateral defective vision was subsequently diagnosed as pituitary adenoma. With a strong clinical suspicion and help of appropriate neuro imaging one can accomplish early detection of chiasmallesions among patients with atypical ophthalmic manifestations unexplained by obvious pathological conditions within the eye may prevent disastrous blindness and other neurological complications.

Keyword :chiasmal tumours, Normotensive glaucoma, retrobulbar neuritis Though they comprise only 3 per cent of all intracranial tumours, they represent > 9 per cent of intracranial neoplasms in childhood and 30 per cent of all new growths in the hypophyseal area. Craniopharyngioma has a bimodal presentation. The first peak being in children aged 5-10 years and a second peak in adults aged 50-60 years. Pituitary adenomas are tumours that occur in the pituitary gland, and account for about 15% of intracranial neoplasms. Tumors which exceed 10 mm in size are defined as macroadenomas, and those smaller than 10 mm are referred to as microadenomas. Most pituitary adenomas are microadenomas, which often remain undiagonosed, and have an estimated prevalence of 16.7%Three major clinical syndromes have been described which relate to the anatomic location of the chiasmal tumours namely, Prechiasmal localization typically presents as progressive decline of visual acuity and constriction of visual fields due to optic atrophy.
Retrochiasmal location commonly manifests as papilledema, abducens palsy due to increased intracranial pressure, and hydrocephalous in children.
Intrasellar tumour manifests with headache and endocrinopathy.

Case reports:

Case 1:

A 47 yrs old female presented with complaints of progressive progressive diminution of vision OU over 6 months and occasional episodes of generalized headache not associated with vomiting. On examination of OD, vision was 6/12 NIP. Anterior segment examination revealed, pupil reacting normally to direct but sluggishly to consensual light reflex. On examination of OS, vision was 2/60 NIP. Pupillary assessment revealed grade II RAPD. Other findings OU were within normal limits. On examination of fundus, media was clear, there was bilateral symmetrical cupping of the disc with C:D ratio of 0.6 and macula appeared normal (Fig-1.1). Tension by Gold manappla nation tonometer in OD was 14 mm Hg and OS was 16 mm Hg. Aprovisional diagnosis of Normotensive Glaucoma was considered. Colorvision was normal in OD but unreliable in OS due to poor vision. Visual field testing of OD revealed temporal hemianopia (Fig: 1.2), while that of OS could not be assessed
On radiographic evaluation, X - Ray skull, lateral view showed noenlargement of the sella turcica but bony erosions were noted retrospectively (Fig: 1.3). CT Scan of the brain showed a large well defined lobulated hypodense mass measuring 5.4 × 5.1 × 3.4 cm in size with central specks of calcification, insinuating along the sellar, suprasellar and parasellar spaces suggestive of craniopharyngioma. Mild enhancement of the solid component was seen after contrast. There was mass effect on IIIventricle with midline shift, subfalcine and uncal herniation (Fig: 1.4). MRI brain confirmed the above findings (Fig: 1.5).
The patient was immediately referred to a Neurosurgeon and was successfully operated subsequently. The patient returned to us after 2months. On review, the vision was 6/12 in OD, and 6/18p in OS. Fundus examination revealed optic disc pallor OU. Visual field changes persisted.

Case 2: A 26 yrs old female presented with complaints of sudden loss of vision OS for 7 days and occasional episodes of generalized headache not associated with vomiting. There was no history of any trauma, fever, pain or any other systemic complaints. On further interrogation patient gave history of secondary amenorrhea for last 6 months for which she did not seek any medical consultation. On examination of OD, vision was 6/24 with pH 6/12NIG. Anterior segment examination revealed, pupil reacting normally to direct but sluggishly to consensual light reflex. On examination of OS, vision was CFCF. Pupillary assessment revealed grade III RAPD. Other findings OU were within normal limits. On examination of fundus, media was clear, disc, vessels and macula appeared normal. Tension by Goldma napplanation tonometer in OD was 18 mm Hg and OS was 15 mm Hg. Aprovisional diagnosis of retrobulbar neuritis OS was made. Color vision was normal in OD but unreliable in OS due to poor vision. With this diagnosis in mind, patient was started with injection i.v Methyl Prednisolone 1 gm in two divided doses for three days followed by tab. Prednisolone 50 mg once daily, according to the Optic Neuritis Treatment Trial along with neuro vitamin supplementation. Haematological work up of the patient was within normal limits After a period of one week, vision in OS improved to6/60 NIP and was subjected to visual field testing by Bjerrum's screen, which revealed bitemporal hemianopia (Fig: 2.1).Urgent imaging was then advised for the patient. On radiographic evaluation, X - Ray skull, lateral view showed no enlargement of the sellaturcica (Fig: 2.2). MRI of the brain showed a mass measuring: 2.5 x 1.4 x 1.8 cm with edema showing as T1 hypo and T2 hyper, focal haemorrhage /inspissated cystic degen-eration within superior pole of adenoma impinging and elevating optic chiasma. All the above features were diagnostic of intra
– supra sella PITUTARY MACROADENOMA. The initial improvement ofvision with steroid therapy was attributed to the decrease in inflammationaround the mass, thus reducing the pressure effect on the optic chiasma,and tract (Fig: 2.3).
The patient was immediately referred to a Neurosurgeon and was successfully operated subsequently using the transsphenoidal approach. The patient returned to us after 6 months. On review, the vision was 6/9 in OD, and 6/12 in OS. Fundus examination was normal OU. Visual field changes regressed.

Discussion:
Primary brain tumours constitute a major reason for seeking neurological consultations worldwide. The location and type of tumour influences the clinical presentation and the management option. The clinical features in patients with brain tumours may be caused by their mass or irritating effect, influence on hormone secretion/depression or their mass related pressure effect on the surrounding structures or development of hydrocephalus[9]. Radiological images aid in the diagnosis of chiasmal syndromes. X- ray of the skull is diagnostic of craniopharyngioma in children. In adults a normal x ray does not exclude the diagnosis. 80-87% of craniopharyngiomas are calcified and 70-75% are cystic. Calcifications are more common in children (90%) than in adults (50%).CT scan is the most sensitive method to demonstrate calcifications which appear as high-density areas. CT has replaced the plain radiograph. It is useful in defining both calcified and cystic parts of the lesion. Fluid content of the cyst has the same density as CSF; contrast administration enhances the capsule of the cyst. MRI, with its multiplanar capability, is essential for defining the local anatomy and is the most important imaging modality used to plan the surgical approach. MRA is used for visualizing the major cerebral vessels and their relation to the tumor; it has largely replaced the 4-vessel angiogram.
Foroozen[5] divides the etiologies of chiasmal syndromes into intrinsic and extrinsic causes. Intrinsic implies thickening of the chiasm itself, like glioma and multiple sclerosis and extrinsic implies compression by another structure in case of meningioma, neurofibroma, pitutary adenoma and craniopharyngioma. Other less common causes of chiasmal syndrome are metabolic, toxic, traumatic or infectious in nature.Neuro ophthalmic manifestations depend on the location of the compressing mass. Visual field testing is a good screening test for chiasmal involvement. Lee has divided optic chiasmal syndromes into anterior, middle and posterior locations[4]. Anterior chiasmal syndrome affects the junction of the optic nerve and chiasm. Middle chiasmal syndrome relates to thedecussating fibers in the body of the optic chiasm while posterior chiasmal syndrome involves the caudal fibers.
The classic anterior chiasmal lesion affects the optic nerve fibers and the contralateral inferonasal fibers located in Wilbrand's knee. This will produce an ipsilateral optic neuropathy, often manifested as a central scotoma, and a defect involving the contralateral superotemporal field. This is also known as a junctional scotoma. An alternative explanation for the contralateral field deficit has been provided by Horton[3].
Middle lesions affecting the uncrossed temporal fibers are rare. These can result in a nasal or binasal hemianopia. Lesions in the body of the chiasm most commonly disrupt the crossing nasal retinal fibers. This leads to a bitemporal hemianopia. The field of vision may still be full when both eyes are open but stereovision will not be possible. However, if fusion of the images is lost, perhaps due to a preexisting phoria, binocular diplopia may result[5].
The macular fibers cross more posteriorly in the chiasma, hence they are damaged in posterior chiasmal syndrome. This leads to a smaller, paracentral bitemporal field loss. As the temporal macular fibers are not damaged, color vision and visual acuity are preserved. Posterior lesions may also involve the optic tract and cause a contralateral homonymous hemianopia.
Other eye signs of chiasmal syndromes include[1],
1 Optic disc pallor rather than papilledema may be apparent with an ophthalmoscope if the insult is longstanding. If the lesion does not affect the lateral uncrossed fibers, the pallor may take on a bowtie configuration. This is due to loss of retinal ganglion cells nasal to the macula in the papillomacular bundle.

2 Anomalous optic discs can be seen due to close proximity between the anterior visual pathway and the cells of origin of the tumour.

3 Concomitant squint or paralytic strabismus due to abducens palsy as a result of raised intracranial pressure or sensory exotropia due to early on set primary optic atrophy.

4 Acute loss of vision which may mimic retrobulbar neuritis can occur due to sudden haemorrhage or inflammation of the mass.

5 Amblyopia in absence of amblyogenic factors.

6 See saw mystagmus.
7 Diminished stereopsis, hemifield slide - bitemporal hemianopia → lossof the temporal visual field → loss of sensory fusion→ binocular vertical or horizontal diplopia.

8 Post fixational blindness i.e inability to recognize objects beyond the point of fixation.

9 Compressive lesions often cause headache and may compress the third ventricle leading to hydrocephalus and papilledema.

Our first patient presented with bilateral symmetrical 0.6 cupping of the disc, which mimicked normotensive glaucoma. The differential diagnosis of NTG includes compressive lesions of the optic nerve and tract amongst other congenital disorders and acquired causes like methyl alcohol poisoning, optic neuritis, AION, NAION, etc. Hence the indications to perform neuro – imaging evaluation in NTG, include General: age < 50 years, new onset or increased severity of headaches, localising neurological symptoms and neurologic visual abnormalities. Ocular: colour vision abnormalities, pallor of remaining neuroretinal rim, highly asymmetric cupping, lack of disc and visual field correlation and visual field defect respecting midline[7].Our second patient presented with sudden loss of vision in one eye and was diagnosed with pituitary macroadenoma. In general, there is equal distribution of pituitary tumors between men and women. Corticotrophin secreting tumors are an exception. These tumors occur mainly in women, with a female-to-male ratio of 4:1.In general, pituitary adenomas are diagnosed more frequently in women of childbearing age than in men probably because of the association of these tumors with menstrual irregularities. Amenorrhea is common in women with macroadenomas; this finding suggests a pituitary lesion. Tumors affect individuals of all ages, but the incidence increases with age, peaking between the third and sixth decades.

Clinical masquerades of pituitary adenoma include chronic retrobulbar optic neuritis, nutritional amblyopia, uncorrected refractive error, normal-tension glaucoma, and age-related maculopathy. Bilateral tilted-disc syndrome can result in a superior bitemporal field defect similar to that observed in pituitary adenoma. However, the field defect in tilted-disc syndrome is unchanging and does not respect the vertical midline, whereas the field defects in chiasm-compressive lesions are progressive and do respect the vertical midline. Morbidity related to macroadenomas is associated with expansion of the tumor into the optic tracts and the cranial nerves adjacent to the cavernous sinus and may include permanent visual loss, ophthalmoplegia, and other neurologic complications. Some tumors recur after radiation therapy and surgery. Pituitary apoplexy is rare complication of pituitary tumors caused by sudden bleeding into the tumor or infarction of the pituitary gland. Headache of sudden and severe onset is the main symptom, associated with visual disturbances or ocular palsy. Signs of meningeal irritation or altered consciousness may be present. Secondary adrenal failure due to ACTH deficiency may be life threatening.

In rare cases, a pituitary adenoma may invade the orbit, and devastating consequences to the integrity of the globe and ocular structures may result. Therefore, early recognition of this complication is of the utmost importance to begin appropriate treatment to minimize ocular and orbital damage.

Surgery by means of the transsphenoidal approach is considered the technique of choice when pituitary surgery is indicated. Surgery has the advantage of rapidly lowering hormone levels. For microadenomas, the cure rate is greater than 50%. Tumors larger than 1 cm can recur and may require additional treatment. Infection, cerebrospinal leakage, vascular injury, double vision, visual loss, and pituitary deficiency are rare postoperative complications. A false aneurysm of the cavernous carotid artery and a carotid cavernous fistula have been reported as complications after transsphenoidal surgery

Since ocular signs are frequently the presenting feature of these tumours, many of the patients being first seen at an eye clinic, their recognition by the ophthalmologist is clearly of great importance. Certainly early diagnosis is desirable, since modern advances in treatment have resulted in better prospects for successful eradication of the growth, if its detection is timely and its location favorable.

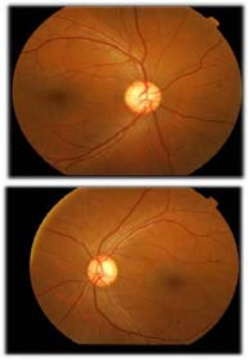

Fig 1.1 – B/L symmetrical 0.6 cupping

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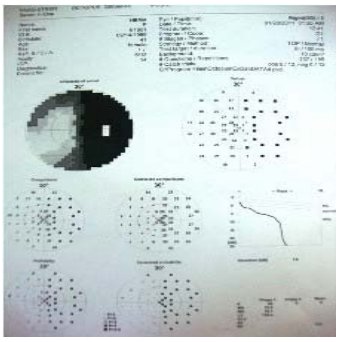


Fig 1.2 – OD: visual field showing temporal hemianopia

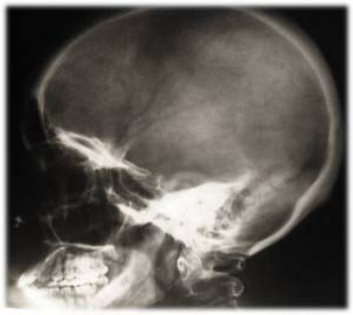


Fig 1.3 – normal sella turcica in lateral X – ray of skull

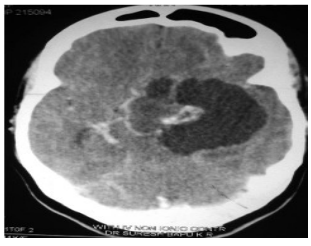


Fig 1.4 – CT scan brain – showing hypodense mass with calcification, supra sellar , parasellar extension and midline shift.

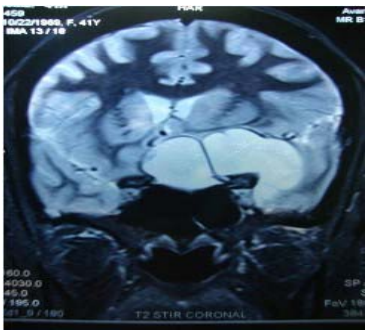


Fig 1.5 - MRI brain T 1 W images confirms CT findings

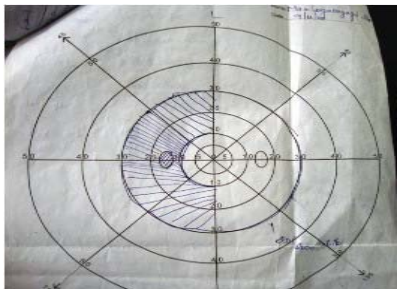
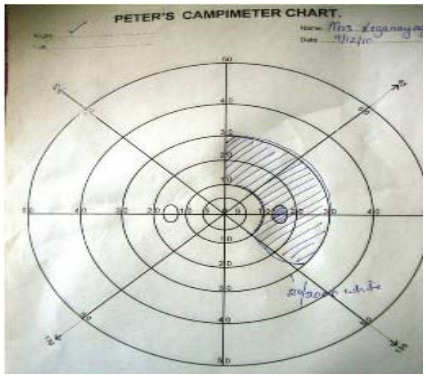


Fig 2.1: Bjerrum's visual field showing incongruous bitemporal hemianopia.



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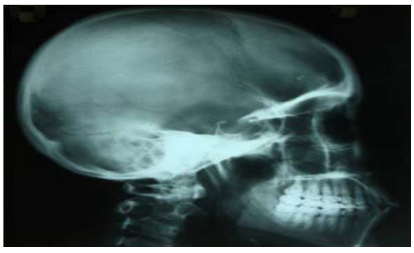

Fig 2.2: X-Ray Skull lateral view – normal sella turcica.

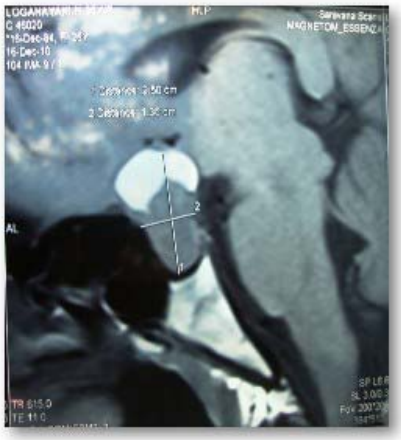

Fig 2.3: MRI Brain - mass measuring: 2.5 x 1.4 x 1.8 cm with edema showing as T1 hypo and T2 hyper, focal haemorrhage / inspissated cystic degeneration within superior pole of adenoma impinging and elevating optic chiasma.

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