Unusual Visual Manifestations of Pituitary Tumours

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Abstract
A retrospective study of 57 cases with pituitary tumours was done. The visual fields [VF] were analysed and pattern of visual field was noted. 16 patients had normal visual fields. 41 patients had VF defects [VFD] in the temporal quadrant. They were variants of the classical bitemporal field defect, the characteristic finding of pituitary lesions. Unusual field defects like arcuate scotomas [n=1], binasal VFD [n=1], bilateral superonasal quadrantanopia [n=1] were found. 13 patients had unusual manifestations: Motor manifestations in 8 patients (1 patient also had Superior orbital fissure syndrome), 7 had pituitary apoplexy, papilloedema in 1 patient (due to obstruction of Foramen of Monro). An attempt was made to explain these.

Key Words: Pituitary Tumours, Apoplexy, Visual Field Defects.

Introduction
Pituitary adenoma is the most common tumour to affect pituitary accounting for 10 to 15 % of intracranial neoplasms. The clinical manifestations are varied depending on the cell type within the tumour, hypo or hypersecretion of hormones, direction of local spread and invasion of adjacent structures. Both nonvisual and visual manifestations are of extreme importance in the diagnosis, management and prognosis of patients with pituitary adenomas.

Pituitary adenomas are diagnosed earlier nowadays due to availability of radioimmunoassay techniques for the hormones and increasing use of CT scanning and MRI imaging, done at times for indications unrelated to suspicion of pituitary tumours (like after head injury, for evaluating headache). The percentage of patients presenting with visual loss or from the effect of tumour has decreased dramatically over last 50 years and now, the most common presentation is endocrine dysfunction. However there remain patients in whom visual sensory or motor dysfunction of extraocular muscles occur either as presenting manifestation or as an associated feature of a pituitary adenoma.

Bitemporal field defect is the most common visual abnormality produced by pituitary adenomas. It can be uni or bitemporal hemianopia usually involving superior field first, and more densely than inferior field. Apart from these, pituitary adenomas can also have unusual ophthalmic manifestations like arcuate field defects, isolated nasal hemianopias, papilloedema, ocular motor dysfunction (of which pituitary apoplexy being the most common cause), CSF rhinorrhea etc.

In this case series, we present a few of such findings in our series of patients.
Material and Methods:

57 patients were enrolled in the study.

Type of study: Retrospective analysis of the case records of patients with pituitary adenoma who visited the ophthalmology outpatient department over the last 2 years.

Inclusion criteria: An ophthalmic evaluation was done including assessment of visual acuity and visual fields for all patients of pituitary tumour. Visual field defects were noted in every case. Any degree of bitemporal fields was taken as the expected type of field defect i.e. whether complete/incomplete, quadrantanopic or hemianopic, relative or absolute. All other visual abnormalities, if any, were noted including visual loss, papilloedema, relative afferent pupillary defect [RAPD]. III, IV, V, VI nerve palsies were noted. Visual field defects other than temporal were noted as atypical: Nasal field defect without temporal field defect, arcuate field defects, central or centrocaecal field defects.

Neuroimaging films were reviewed and an attempt was made to find reasons for the unusual field defects. Normal visual acuity for distance was defined as 6/6 on Snellen’s chart with appropriate refractive correction. Diagnosis of visual field defects was done according to the system suggested by Ravi Thomas et al as follows: Quadrantanopia was diagnosed if either of the following criteria were fulfilled:

1. Depression of thresholds by 5 db or more, in 3 or more contiguous points adjacent to the vertical meridian in the involved quadrant as compared to their mirror image points across the vertical meridian.

2. The pattern deviation plot showed 3 or more points adjacent to the vertical meridian in the involved quadrant depressed to the 1% probability level with normal mirror image points across the vertical meridian. For the diagnosis of hemianopia, the diagnostic criteria for quadrantanopia had to be applicable to both quadrants comprising the hemifield.

3. Advanced field defects were considered hemianopic if comparison of the least involved quadrant across the vertical meridian, met the threshold depression criteria for the diagnosis of quadrantanopia. Here the pattern deviation plot criterion on its own was not considered diagnostic.

4. Atypical field defect was defined as a defect that did not fit into any characteristic diagnostic pattern considered typical of pituitary adenomas.

Observations

Out of 114 eyes of 57 patients, 62 eyes had normal vision. 11 patients had normal vision in both the eyes. (Table 1). 3 patients were bilaterally blind with only light perception. 5 others were blind in one eye with temporal hemianopic field defect in the other eye.

<table>
<thead>
<tr>
<th>Visual acuity</th>
<th>No of eyes</th>
</tr>
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<tbody>
<tr>
<td>6/6</td>
<td>62</td>
</tr>
<tr>
<td>6/9- 6/18</td>
<td>28</td>
</tr>
<tr>
<td>6/24- 6/60</td>
<td>7</td>
</tr>
<tr>
<td>&lt; 6/60</td>
<td>17</td>
</tr>
<tr>
<td>Total</td>
<td>114</td>
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Out of 57 patients studied, 13 patients showed atypical manifestations (Table 3)
Table 3. Atypical manifestations in patients with pituitary adenoma.

<table>
<thead>
<tr>
<th>Manifestation</th>
<th>No of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arcuate scotoma</td>
<td>1</td>
</tr>
<tr>
<td>Isolated nasal U/L or B/L field loss</td>
<td>2</td>
</tr>
<tr>
<td>Papilloedema</td>
<td>1</td>
</tr>
<tr>
<td>Motor abnormalities</td>
<td>2</td>
</tr>
<tr>
<td>Pituitary apoplexy</td>
<td>7</td>
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**Field defects:**

1. **Right nasal hemifield loss** (Figure 1a): In one of the patients the tumour was encasing the cavernous portion of the right carotid artery compressing the optic chiasma from right lateral side. (Figure 1b). This might have affected the optic nerve fibres coming from the ipsilateral temporal retina of right eye leading to right nasal hemianopia. Other eye was normal.

Arcuate scotoma: One patient had double arcuate scotoma in both eyes (Figure 2a). Here the tumour was encasing the intracranial portion of both optic nerves more so on right optic nerve (Figure 2b). This compression may have produced these scotomas characteristic of optic nerve involvement. This is also substantiated by the presence of optic nerve head (ONH) pallor in both eyes.

Incidentally this patient also had left hyperemic optic disc. This patient had been diagnosed elsewhere as, optic neuritis in view of the field defects and optic disc hyperemia and treated with intravenous dexamethasone.

Bilateral superonasal quadrantanopia (Fig 3): This implies compression from below affecting the temporal fibers first causing nasal visual field defect.

Motor abnormalities:

VIth nerve paresis: A 13 year old boy presented with vomiting, headache and double vision since 1 month. Double vision was maximum on looking to the right side. He was found to have right VIth nerve paresis. On MRI there was a sellar lesion with suprasellar extention, invading right cavernous sinus. (Figure 4) This paresis and diplopia completely disappeared after the surgical removal of the tumour.

Superior orbital fissure syndrome with partial III nerve palsy: This patient presented with headache for 2 weeks duration and on examination showed partial third nerve palsy on the right side. The vision, pupils and visual fields were normal, and on MRI, had enlarged pituitary and diagnosed as incidentaloma.
He also had a soft tissue enhancing mass in the right superior orbital fissure (Figure 5). A diagnosis of right superior orbital fissure syndrome was made and nerve palsy recovered well with oral steroids. Probably the two masses were unrelated.

1. Pituitary apoplexy with total ophthalmoplegia: One of the patients of apoplexy presented with left complete IIIrd, IVth and VIth nerve palsy. After transnasal endoscopic resection of the tumour, over a period of 7 months, these recovered completely. Another patient with apoplexy had restricted elevation in both eyes.

Pituitary apoplexy: Out of the 58 cases reviewed, we found 7 cases (10.53%) of pituitary apoplexy. Their manifestations included sudden onset of severe visual loss, altered sensorium, altered behaviour headache etc. are summarized in Table 4. The time period between onset of symptoms and diagnosis of apoplexy ranged between 2 weeks to seven months. All these patients had very good outcome after the surgical resection of tumour, except for 2 patients (one had residual tumour, and the other had gross compression of optic nerve by the tumour).
Table 4. Clinical features of patients with pituitary apoplexy

<table>
<thead>
<tr>
<th>Clinical feature</th>
<th>No of patients (%)</th>
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<tbody>
<tr>
<td>Decreased vision</td>
<td>5 (60%)</td>
</tr>
<tr>
<td></td>
<td>[total loss 5 eyes]</td>
</tr>
<tr>
<td>Motor nerve palsies</td>
<td>1 (20%)</td>
</tr>
<tr>
<td>Headache</td>
<td>7 (100%)</td>
</tr>
<tr>
<td>Vomiting</td>
<td>3 (50%)</td>
</tr>
<tr>
<td>Altered sensorium</td>
<td>3 (50%)</td>
</tr>
<tr>
<td>Altered behaviour</td>
<td>1 (20%)</td>
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In one patient of Stevens Johnson syndrome, vision did not improve inspite of good recovery of cornea and tear film. Best corrected visual acuity was 6/12, with Optic Nerve Head pallor in right eye. He was advised to continue tear substitutes and review after 2 weeks for visual field assessment when his systemic condition would be better. On the 3rd day after being discharged from the hospital, he came to the casualty with sudden complete loss of vision in right eye and was found to have pituitary apoplexy (Figure 6).

One patient had papilloedema with blurring of disc margins and a history of frank papilloedema as per her records.

Fig. 3. Bilateral superonasal quadrantanopia

Fig. 4. Pituitary tumour invading right cavernous sinus
in height, and width. Enlargement of the pituitary < 10 mm is called Microadenoma. An enlargement more than this is called Macroadenoma. Because of its anatomical relation with the chiasma [Fig. 7], the pituitary tumors cause VF defects by compressing the ON fibres.

**Discussion**

Tumours of pituitary gland are extremely important for an ophthalmologist due to their various ophthalmic manifestations.

Pituitary gland is situated in the sella tursica. The pituitary adenoma is its most common involvement [90 %]. The normal size of pituitary is 5.4±0.9 mm in height, and width. Enlargement of the pituitary < 10 mm is called Microadenoma. An enlargement more than this is called Macroadenoma. Because of its anatomical relation with the chiasma [Fig. 7], the pituitary tumors cause VF defects by compressing the ON fibres.

*The VF defects are the mirror images of the ON defects i.e. since compression is from below scotomas first appear in the temporal hemi field as the crossed fibres are most susceptible, especially the inferior ones. Thus the VF seems to progress from above – below i.e. clockwise in the RE and anticlockwise in the LE. These defects may be complete / incomplete/ partial/ total dense. Chiasma*
Fig. 8. Picture of pre and post fixed chiasma

can be prefixed [10%] or postfixed [10%] [Fig. 8]. VF loss patterns are different in these cases. In prefixed chiasma, optic tract would get affected first, i.e. homonymous hemianopic pattern of VF loss and in postfixed either one of the optic nerves may be affected more, with VFD of one eye more, and an altitudinal pattern.

There is a significant change in the way pituitary tumours come to attention. Hollenhorst and Young reviewed 1000 cases of pituitary tumours between 1940 to 1962 and found that 70% of these patients had either loss of visual acuity or visual defects or both, but in a subsequent study of the patients between 1971 to 1982 at the same hospital, only 20% patients had reduced visual acuity and 32% had visual field defects. In the case series studied by Anderson et al (200 consecutive cases of pituitary tumours between 1971 to 1982) only 16% patients had decreased visual acuity and 32% had visual field defects. So there is a decline in the number of patients with visual problems in pituitary adenomas.

In our case series, 66 eyes out of 114 [57 patients] had normal VA. Also only 16 patients (28.1%) had normal visual fields in both eyes. This reflects the need of vigilance by the ophthalmologists in our country to consider the possibility of pituitary tumour in dealing with patients of unexplained visual loss.

Bitemporal hemianopia has been described as the classical field defect in pituitary tumours. It was found in only 10 patients (17.5%). Depth perception difficulties may be seen in patients with bitemporal field defects like difficulty in clipping nails, threading, catching a ball because of the blind triangular area just beyond fixation in these patients [Fig. 9]. None of our patients had these complaints. Different kinds of field defects were found like one eye hemianopia and contralateral superotemporal quadrantanopia; one eye blind and contralateral temporal hemianopia; bilateral superotemporal quadrantanopia etc. This emphasizes that pituitary tumours cause not only bitemporal hemianopia but other kind of field defects also and an ophthalmologist should be aware of these. As the tumor grows the visual field defects keep extending. Nonsecreting tumors are detected later when the size is larger, as only visual symptoms may be the presenting feature.

One of our cases had an arcuate scotoma. Earlier arcuate field defects have been reported in patients with pituitary tumours. These have been ascribed to the damage to intracranial portion of the optic nerve, with postfixed chiasmas. Superior arcuate field defect can occur with a postfixed chiasma as, an optic nerve [ON] rather than chiasma, lies over the sella turcica.
compressing the ON from below and causing a superior arcuate defect.

Walsh & Hoyt\(^1\) write, that in rare cases when tumour grows between two optic nerves shifting them laterally, it may compress optic nerves against the anterior clinoid process and internal carotid arteries, affecting the temporal nerve fibers first, giving rise to bilateral nasal field defects. We propose a similar explanation for our patient with right nasal hemianopia where tumour was found to encase right internal carotid artery, which might have compressed the temporal fibres in right optic nerve giving rise to a nasal hemifield loss.

Many explanations are given to explain the reason for bitemporal hemianopia as the first field defect in pituitary lesions. Fibers from inferior parts of retina are situated inferiorly in optic chiasma which get compressed first by the pituitary tumour. So it should produce a superior altitudinal field loss, but actually it affects temporal field preferentially. This has been explained by the susceptibility of nasal fibers to effects of compression, or it may be related to blood supply of crossing fibres at chiasma.\(^1\) In our case which showed bilateral superonasal quadrantanopia, we could not offer any explanation by the neuroimaging study. The cause could either be the fragile blood supply of the crossing fibres (as demonstrated by Lao and Gao in cases where bitemporal hemianopia was present without any chiasmal compression)\(^9\) or fine mechanics of compression selectively affecting the crossing fibres.

Papilloedema can occur as a part of Foster Kennedy Syndrome [very rarely seen with pituitary lesions] or due to hydrocephalus with compression of the foramen of Monro. Our patient had only blurred disc margins. She had come after excision of the pituitary mass to our institution for radiotherapy and carried reports that indicated papilloedema prior to surgery, due to hydrocephalus with compression of the foramen of Monro as per the CT scans available.

Pituitary tumours causing abnormalities of extraocular movements are rare. In Hollenhorst and Young’s review, they found 4.6 % patients \(^4\) while Trautman et al found 1.4 % patients with abnormalities of extraocular movements.\(^10\) We found 4 such patients (7.2 %) in our study with a smaller sample size. This number is slightly higher compared to the earlier studies. Isolated involvement of VIth nerve is also rare with very few cases reported so far.\(^11,12,13\) We found one such patient. It was ascribed to invasion of cavernous sinus by the tumour affecting the VIth nerve.

Other unusual presentations not seen in our series are hemifeild slide phenomena, visual hallucinations, junctional syndrome, homonymous hemianopia and see saw nystagmus.

The patient who had superior orbital fissure syndrome, had only a microadenoma and probably his motor manifestations were not related to the pituitary tumour. But there was a soft tissue mass in superior orbital fissure. This emphasizes the need for a careful examination of the neuroimaging, which may reveal other coexisting abnormalities which might be responsible for some of the clinical features of the patient and not get biased by the tumour itself.

Pituitary apoplexy is a rare, major clinical event with neurological, neuro-ophthalmological, cardiovascular and hormonal consequences, resulting from an acute infarction of pituitary adenoma. There has been confusion about the exact definition of the entity. Walsh and Hoyt suggest that this term should be used for cases with acute haemorrhage into pituitary adenoma producing, not only infarction of the gland but also damage to adjacent structures particularly optic nerves, optic chiasma, ocular motor nerves and hypothalamus.

We found 7 (12.3 %) cases with apoplexy according to the above definition. About 5 % patients are said to suffer from this complication.\(^2\) Headache was the commonest symptom followed by decreased vision. This finding is similar to the earlier reports.\(^14-17\) But compared to these studies, the incidence of motor palsies was less in our series. We had a 13 year old boy with apoplexy, although it is rare in children.\(^1\)

Except for 3 patients with apoplexy, others had a good outcome after trans-sphenoidal resection of the tumour. One of these cases with poor postoperative outcome, had residual tumour, and two had direct compression of optic nerve by the tumour. Histopathologically these 3 cases were found to have haemorrhagic infarction. According to a study by Semple PL, De Villiers JC et al, patients who presented with histological features of pituitary tumor infarction alone had less severe clinical features at the time of presentation, a longer course prior to presentation and a better outcome than those presenting with hemorrhagic infarction or frank
hemorrhage. Our observation of poor outcome in these patients also corroborates with this.

4 of these patients with apoplexy were treated earlier for altered consciousness, abnormal behaviour, hyponatremia etc. and later were found to have apoplexy. Agrawal D and Mahapatra AK in their study have shown that even completely blind eyes may have remarkable improvement in vision if surgical decompression of the optic apparatus is undertaken early.

**Conclusion**

Still significant numbers of patients with pituitary tumours either present with or have associated decrease in visual acuity or visual field defects. Although bitemporal hemianopia is the classical visual field defect caused by pituitary tumours, other variants of field defects also occur. Field defects like arcuate scotomas, isolated nasal field loss, although uncommon, can occasionally be seen and their presence does not rule out pituitary tumours per se. Looking at the need for early surgical intervention for a better outcome, it is very important both for an internist and an ophthalmologist to keep in mind the entity- pituitary apoplexy (in patients with headache, visual loss and altered sensorium), in the appropriate clinical setting.

**Acknowledgements**

We are grateful to the Department of Neurosurgery at Amrita Institute of Medical Sciences, Cochin for referral of patients.

**References**