Case Report

Pituitary Macroadenoma with Apoplexy, a Mimicker of Normal Tension Glaucoma (NTG): A Case Report

Thinley [1], Keepa Vaidya[2], Sandip Tamang[3]

[1,3]Department of Ophthalmology, Eastern Regional Referral Hospital, Mongar, Bhutan.
[2]Department of Neuro-ophthalmology, Tilganga Institute of Ophthalmology, Gaushala, Kathmandu, Nepal.

ABSTRACT

Background: Asymmetric visual field defects with a high cup-to-disc ratio resembling glaucomatous damage can be the only clinical features of a pituitary macroadenoma with apoplexy, posing diagnostic challenges.

Case: A 39-year-old man presented with a history of gradual onset, progressive visual obscuration in the right visual field for one month without any other accompanying neurological symptoms. On examination, his intraocular pressure (IOP) was 16 mmHg in both eyes. Posterior segment examination revealed a cup-to-disc ratio (CDR) of 0.8 in the right and 0.7 in the left eye, with concentric neuroretinal rim thinning and mild temporal disc pallor. These findings led to the diagnosis and treatment of normal tension glaucoma (NTG) elsewhere. Visual field examination showed field defects featuring a junctional scotoma. The MRI scan of the brain revealed a well-defined mass lesion situated in the sella suggestive of pituitary macroadenoma with possible apoplexy confirmed later through histopathological examination. The neurosurgery team successfully removed the tumor through the endoscopic endonasal trans-sphenoidal approach and the patient reported significant improvement in vision and visual field defects.

Observations: In our case, visual signs and symptoms were the only presenting features, caused by pituitary macroadenoma with apoplexy, which can be a potentially life-threatening condition. Although pituitary apoplexy is an acute condition with numerous neurological signs and symptoms, visual symptoms with high CDR and neuroretinal rim thinning with mild temporal disc pallor were the only features seen in our case, posing a diagnostic challenge. There were no associated systemic manifestations. However, the patient had a favorable outcome because of prompt diagnosis and multidisciplinary management.

Conclusion: The spectrum of clinical manifestations of pituitary macroadenoma with apoplexy should encompass a gradual onset, progressive asymmetric visual field defect with temporal disc pallor to facilitate timely diagnosis and effective management.

Keywords: Junctional scotoma; normal tension glaucoma; pituitary apoplexy; pituitary macroadenoma.

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Corresponding Author
Dr Thinley
Department of Ophthalmology,
Eastern Regional Referral Hospital, Mongar, Bhutan
Email: droduelhamo@gmail.com

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INTRODUCTION

Pituitary adenomas are benign tumors characterized by slow growth that originate from the secretory cells of the anterior pituitary gland, with a prevalence rate of approximately 20%. These tumors are the most common among the masses involving the sella and they usually occur in the third decade of life. About half of these tumors are hormone-secreting. (Ezzat et al., 2004) Prolactinomas account for the majority, around 70-80% of all endocrine-secreting pituitary adenomas. The annual diagnosis rate of prolactinoma is about 3-5 new cases per 100,000 people. (Daly and Beckers, 2020) Prolactinoma is the primary cause of hyperprolactinemia, which can lead to erectile dysfunction in men and amenorrhea, galactorrhea, and infertility in females. (Molitch, 2017 and Lim and Korbonits, 2018)

Pituitary macroadenomas can cause various symptoms which can be attributed primarily to the mass effects of the tumor or its endocrinological effects. These symptoms may include headache, vomiting, nausea, visual field defects, ophthalmoplegia, and cranial nerve palsy.(Ezzat et al., 2004 and Hornyak et al., 2009) Pertaining to the ocular symptoms, bitemporal visual field defects are the most commonly associated with pituitary macroadenomas (Dubuisson, Beckers and Stevenaert, 2007). Nonetheless, junctional scotoma is also one of the common manifestations of pituitary macroadenomas. (Borgman, 2019)

Here, we present a case of an asymmetric visual field defect caused by pituitary macroadenoma with apoplexy. Timely recognition and multidisciplinary team management led to the recovery of the visual field defect and a favorable prognosis.

CASE REPORT

A 39-year-old male, otherwise healthy and a monk by profession, presented to a Tertiary Eye Hospital with a complaint of gradual onset, progressive visual obscuration in his right visual field for a month's duration. Besides this, the patient denied having any other symptoms such as headaches, nausea, vomiting, or any other complaints. He has been on antihypertensive medication (Tab Losartan 25 mg BD) for the last four years. There was no history of diabetes, family history of blindness, or trauma. He had previously been evaluated by ophthalmologists in another country and was managed as a case of NTG with topical anti-glaucoma medication (Gtt. Timolol 0.5% BD in both eyes). However, despite the treatment, his visual symptoms were worsening, which prompted him to seek a second opinion at our center. At the time of his presentation, he was experiencing difficulty performing binocular functions. The patient reported that he had to tilt his head to one side to walk, and he was unable to aim properly while serving tea or coffee into a cup.

On initial examination, his best corrected visual acuity was 6/60 and 6/6 in the right eye and left eye, respectively. Color vision was defective in the right eye and normal in the left eye. On the Hirschberg test, his eyes were orthotropic, and his extraocular motility test was normal in all gaze positions. Further, a positive relative afferent pupillary defect (RAPD) was detected in the right eye during anterior segment examination. The intraocular pressure taken by Goldmann applanation tonometry was 16

mmHg in both eyes. However, the posterior segment examination revealed a cup-to-disc ratio (CDR) of 0.8 in the right eye and 0.7 in the left eye. Also of note was the presence of a concentric neuroretinal rim thinning and mild temporal disc pallor in the right eye. The disc margins were well-defined, and there were no signs of papilledema. The rest of the fundus appeared normal (Figure 1). The visual field examination with the help of Humphrey Field Analyzer (HFA) 30-2 test, depicted a total field defect in the right eye and a superior temporal field defect in the left eye suggesting a junctional scotoma (Figure 2). The patient was referred for neuroimaging (MRI brain), which showed a well-defined T1 intermediate to high intensity and predominantly T2 low signal intensity mass lesion, measuring approximately 3.5 x 2.1 x 2.6 cm, located in the sella with suprasellar extension, with a few foci of T1 and T2 high signal within the lesion, patchy areas of heterogeneous post-contrast enhancement, which tended to compress the optic chiasm anteriorly, suggestive of pituitary macroadenoma with possible apoplexy (Figure 3).

Thus, it was the compression of the optic nerve by the tumor that caused the patient's visual symptoms of field defects in both eyes. The patient was subsequently referred to a neurosurgery hospital for further evaluation and management. Blood investigations performed at the neurosurgery hospital displayed an increased level of prolactin at 5000 mIU/mL (normal range: 244–454).

Other investigation reports were as follows: cortisol: 123.6 ng/mL(normal range 82.5-138 ng/mL); Adrenocorticotropic hormone(ACTH): 26.5 pg/mL(normal range 10-60pg/ml); Insulin-like growth factor-1: 65.9 ng/mL(normal range 68-220 ng/mL); Triiodothyronine (FT3): 3.12 pg/mL(normal range 2.0-4.4pg/mL); Thyroxine (FT4): 0.98 ng/dL(normal range 0.9-2.3 ng/dl); Thyroid- stimulating hormone (TSH): 2.21 mIU/mL(normal range 0.35-4.50 mIU/mL); Serum total testosterone: 17.41 ng/dL(normal range 300 -1000ng/dL); and serum growth hormone: 0.22 ng/mL(normal range 0.1-6.5ng/dl). The serum sodium and potassium levels were 142 mEq/L (normal range 135-145 mEq/L) and 3.8 mEq/L (normal range 3.5-5.5 mEq/L), respectively.

The patient underwent an uneventful tumor resection through an endoscopic endonasal trans-sphenoidal approach. Histopathological examination of the excised tissue was found to be compatible with a pituitary adenoma with apoplexy.

Postoperatively, the patient recovered well and reported a drastic improvement in his visual symptoms. He was able to perform binocular activities. The patient was discharged on tablet Cabergoline 0.25 mg twice a week and was advised to continue with his antihypertensive medication.

At 2 months postoperatively, the patient did not complain of any worsening visual symptoms. His best corrected visual acuity was 6/36 in the right eye and 6/6 left eye. There was a grade 1 RAPD in the right eye. However, his visual field report showed significant improvement compared to his preoperative status (Figure 4).

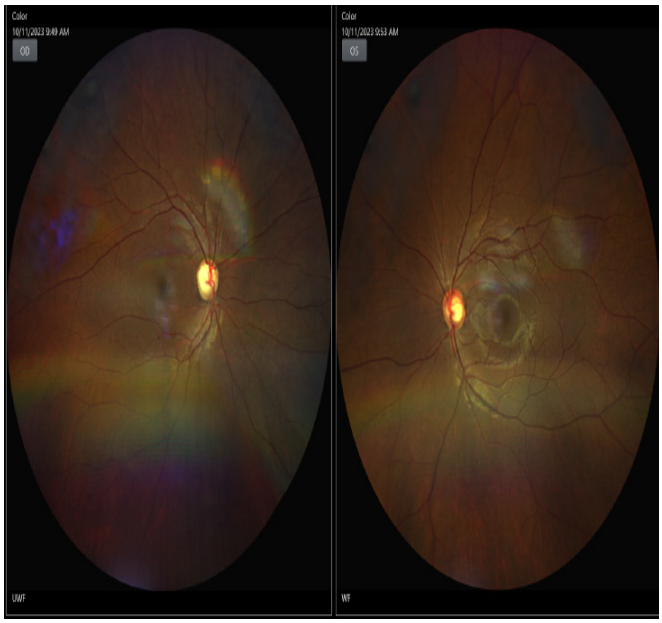

Figure 1: Fundus photograph showing high cup-disc-ratio with mild temporal disc pallor more in the right eye.

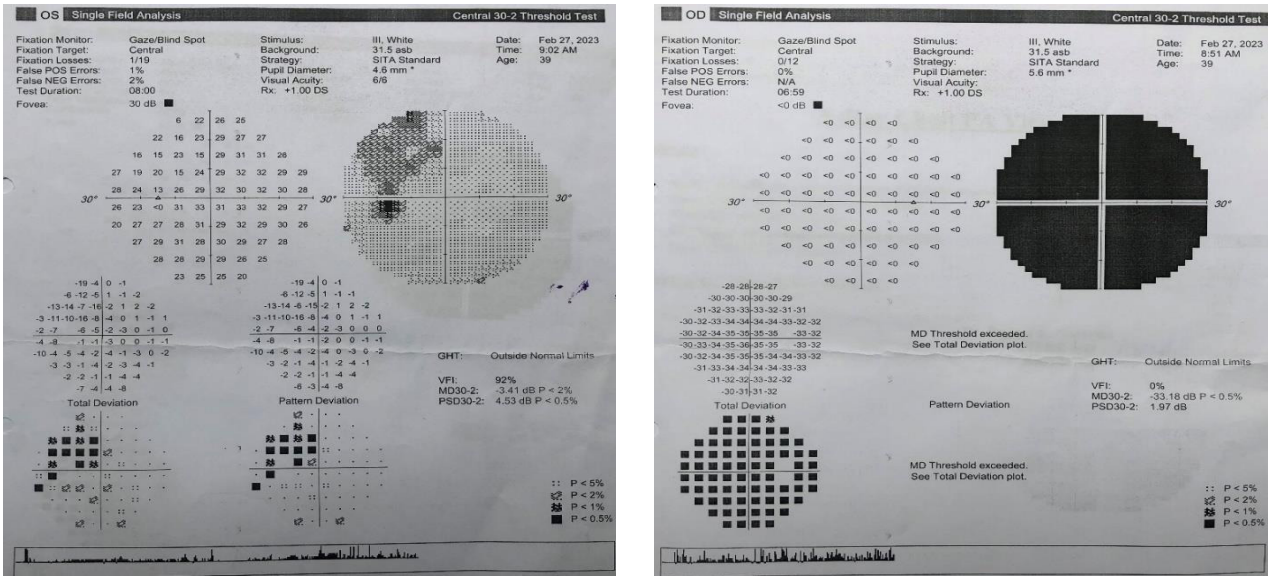

Figure 2: Visual field test reports showing total field defect of the right eye and supero-temporal defect of the left eye.

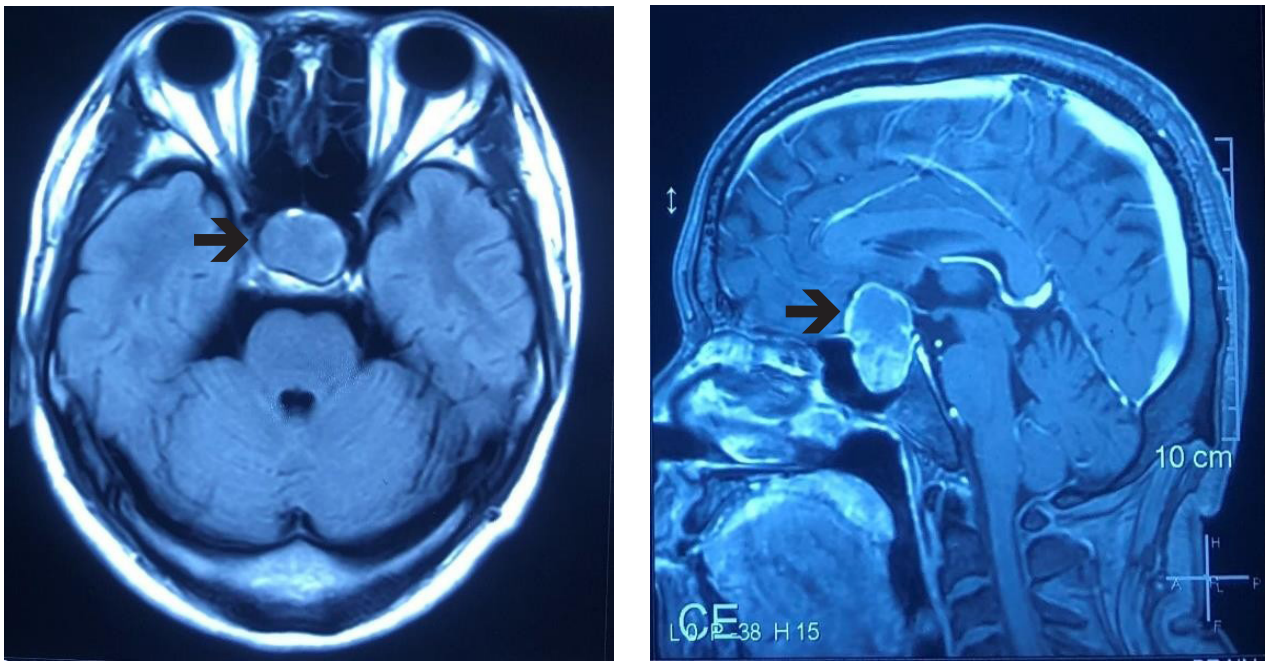

Figure 3: A. Axial (T1 W / FLAIR) magnetic resonance imaging (MRI) brain scan showing a tumor(arrow) measuring approximately 3.5 x 2.1 x 2.6 cm, compressing the chiasmal segment of the optic nerve, more onto the right optic nerve than the left. B.Sagittal MRI scan (post-contract) showing large pituitary tumor with suprasellar extension (arrowhead) with heterogeneous contrast enhancement within the tumor indicating hemorrhagic infarction.

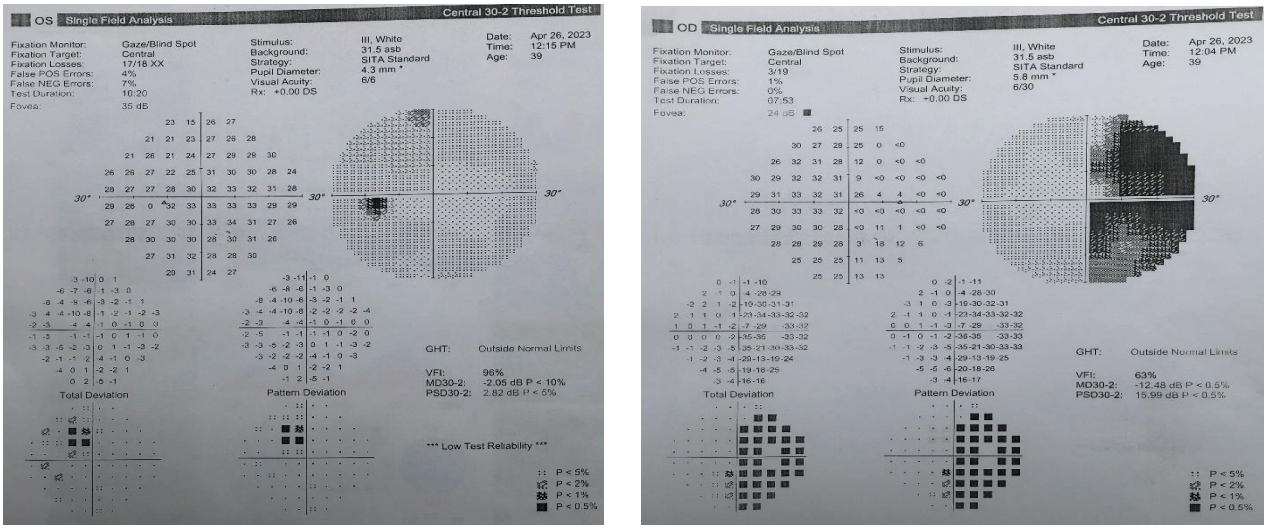

Figure 4: Improvement in post-operative visual field test showing right eye temporal hemi-field defect and left eye small supero-temporal defect.

DISCUSSION

Pituitary apoplexy is not a very common condition but it is potentially a life-threatening condition. It can manifest with various presentations, such as sudden onset of headache, nausea, vomiting, diplopia, visual abnormalities, and altered consciousness. Headaches, nausea, and vomiting primarily occur as a result of meningeal irritation, increased intracranial pressure, and adrenal insufficiency. Visual abnormalities are mainly caused by compression of the optic nerve and chiasm by the tumor. (Hornyak, Digre and Couldwell, 2009) In over 50% of cases, no precipitating factors are identified. However, there are some risk factors such as hypertension, medications, major surgeries, coagulopathies, etc., among which our patient exhibits hypertension as a risk factor.(Biagetti & Simò, 2022)

Our patient had pituitary macroadenoma with apoplexy, which presented with a visual field defect in the right eye in the absence of any other accompanying symptoms. The patient's blood investigations revealed a markedly elevated prolactin level, suggesting a prolactinoma, which was confirmed by brain MRI imaging and histopathological examination. Timely referral and surgical interventions contributed to a favorable visual outcome without any further complications.

The principal objective of this case report is to highlight the significance of ocular symptoms in raising suspicion of pituitary macroadenoma with apoplexy and its importance of early detection. Although pituitary apoplexy by definition is of acute nature with myriad neurological and visual signs and symptoms, it can also present in a subtle manner with only visual symptoms as in our case. Visual field defects, along with increased cup-to-disc ratio on fundus examination, can lead to incorrect diagnosis and management. These, in turn, can cause delays in the diagnosis of a pituitary macroadenoma, which may potentially pave the way for severe complications including life-threatening situations. Therefore, when patients present with gradual onset progressive asymmetrical visual field defects representing a junctional scotoma accompanied by suspicious optic cup-to-disc changes along with temporal disc pallor, it can be crucial to consider the possibility of intracranial lesions such as pituitary adenoma. It is of great importance that such patients are subjected to further neurological assessments.

Junctional scotoma can be caused by compressive lesions specifically at the junction of the optic nerve and the optic chiasm injuring the anterior visual pathway. Our patient presents with a right central scotoma with a left superior visual field defect (junctional scotoma) which is caused by the pituitary adenoma with apoplexy compressing the ipsilateral optic nerve at the junction with the chiasm and contralateral crossing infero-nasal retinal fibers.

The patient's blood investigations revealed a markedly elevated prolactin level, suggesting that the culprit was prolactinoma, which was confirmed by brain MRI imaging and histopathological examination. Timely referral and surgical interventions contributed to a favorable visual outcome without any other complications.

CONCLUSION

Pituitary macroadenoma with apoplexy can manifest subtly as an asymmetrical visual field defect accompanied by optic disc changes that

resemble glaucomatous damage. A lesion that causes compression at the junction of the optic nerve and the chiasm may cause cupping of the optic disc and junctional scotoma which can be misleading in diagnosis. This can present diagnostic challenges and may result in mismanagement of the condition, potentially leading to serious complications. It is crucial to maintain a high level of suspicion for timely diagnosis and effective management.

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Consent for publication: Written informed consent was taken from the patient for the publication of this case report along with the images.

Declaration of competing interests: None declared.

REFERENCES

Daly, A.F. and Beckers, A. (2020) 'The Epidemiology of Pituitary Adenomas', *Endocrinology and Metabolism Clinics of North America*, 49(3), pp. 347–355. doi:10.1016/j.ecl.2020.04.002.

Dubuisson, A.S., Beckers, A. and Stevenaert, A. (2007) 'Classical pituitary tumour apoplexy: Clinical features, management and outcomes in a series of 24 patients', *Clinical Neurology and Neurosurgery*, 109(1), pp. 63–70. doi:10.1016/j.clineuro.2006.01.006.

Ezzat, S. *et al.* (2004) 'The prevalence of pituitary adenomas: A systematic review', *Cancer*, 101(3), pp. 613–619. doi:10.1002/cncr.20412.

Hornyak, M., Digre, K. and Couldwell, W.T. (2009) 'Neuro-ophthalmologic manifestations of benign anterior skull base lesions', *Postgraduate Medicine*, 121(4), pp. 103–114. doi:10.3810/pgm.2009.07.2036.

Lim, C.T. and Korbonits, M. (2018) 'Update on the clinicopathology of pituitary adenomas', *Endocrine Practice*, 24(5), pp. 473–488. doi:10.4158/EP-2018-0034.

Molitch, M.E. (2017) 'Diagnosis and treatment of pituitary adenomas: A review', *JAMA - Journal of the American Medical Association,* 317(5), pp. 516–524. doi:10.1001/jama.2016.19699.