

Neurologic Disorder Masquerading as Postpregnancy Progression of Keratoconus

Rohit Shetty, Sharon D'Souza, Vardhaman P Kankariya, Samaresh Srivastava, Viraj Vasavada, Kareeshma Wadia

ABSTRACT

A 28-year-old woman with bilateral stable keratoconus for 2 years came with a complaint of recent blurring of vision 4 weeks postpartum. Her best-corrected visual acuity had dropped by three lines in the right eye and two lines in the left eye with the same manifest refraction. Corneal topography and Scheimpflug imaging revealed mild progression of the keratoconus, but not enough to explain the drop in visual acuity. However, fundus examination revealed temporal disk pallor in both eyes. Visual field analysis revealed classic bitemporal hemianopia. Subsequently, magnetic resonance imaging (MRI) of brain was performed which revealed a pituitary macroadenoma with pituitary apoplexy that was impinging on the pons. Based on these findings, urgent neurosurgery was performed, and the patient regained her visual acuity and fields 2 weeks later. Neuro-ophthalmologic disorders should always be kept as a differential diagnosis in pregnant and postpartum women. A high index of suspicion from the ophthalmologist can often avert life-threatening problems.

Keywords: Pituitary apoplexy, Keratoconus, Masquerade.

Key message: Neuro-ophthalmologic disorders can masquerade as a variety of clinical presentations in pregnant and postpartum women. A high index of suspicion from the ophthalmologist can often avert life-threatening problems.

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INTRODUCTION

Pregnancy is often associated with ocular changes which may be more commonly transient but occasionally, permanent. The ocular effects of pregnancy may be physiological or pathological or may be modifications of pre-existing conditions.¹ Physiological changes include changes in the cornea and refractive status. Pre-existing conditions, such as keratoconus may progress during pregnancy.^{2,3} Moreover, neuro-ophthalmological disorders should be kept in mind in pregnant women presenting with visual acuity or field loss.

We report the case of a 28-year-old female with keratoconus who presented in the early postpartum period with what seemed like progression of keratoconus, but was detected to have pituitary adenoma on detailed investigations.

CASE REPORT

A 28-year-old female, known to have keratoconus for the last 2.5 years, presented to us with complaints of worsening

of vision, and difficulty viewing her mobile phone, progressively worsening since 4 months. She was 4 weeks postpartum at this time. Her pregnancy had been uneventful, with no history of ocular complaints, health problems and had a full-term normal delivery.

She had presented to us for the first time 2.5 years back. Her uncorrected distance visual acuity (UDVA) was 20/200 in the right eye and 20/30 in the left eye, that corrected to 20/20 in the right eye (+1.5 D Sph/-5.0 D Cyl@60) and 20/15 in the left eye (+0.5 D Sph/-1.0 D Cyl@90). Placido disk and scanning slit-based videokeratography (Orbscan, Bausch and Lomb), rotating Scheimpflug imaging (Pentacam, Oculus, Germany), slit-lamp biomicroscopy and funduscopy was performed in both eyes. She was diagnosed to have keratoconus in the right eye (Stage II, Amsler-Krumeich classification) and forme fruste keratoconus in the left eye (Figs 1A to D). She was given contact lenses and followed up. At 1-year follow-up, corrected distance visual acuity (CDVA) in the right eye was 20/20 (+1.5 D Sph/-5.0 D Cyl@60) and in the left eye was 20/20 (+1.0 D Sph/-2.0 D Cyl@100) with minimal progression. She remained stable subsequently for the next year as well.

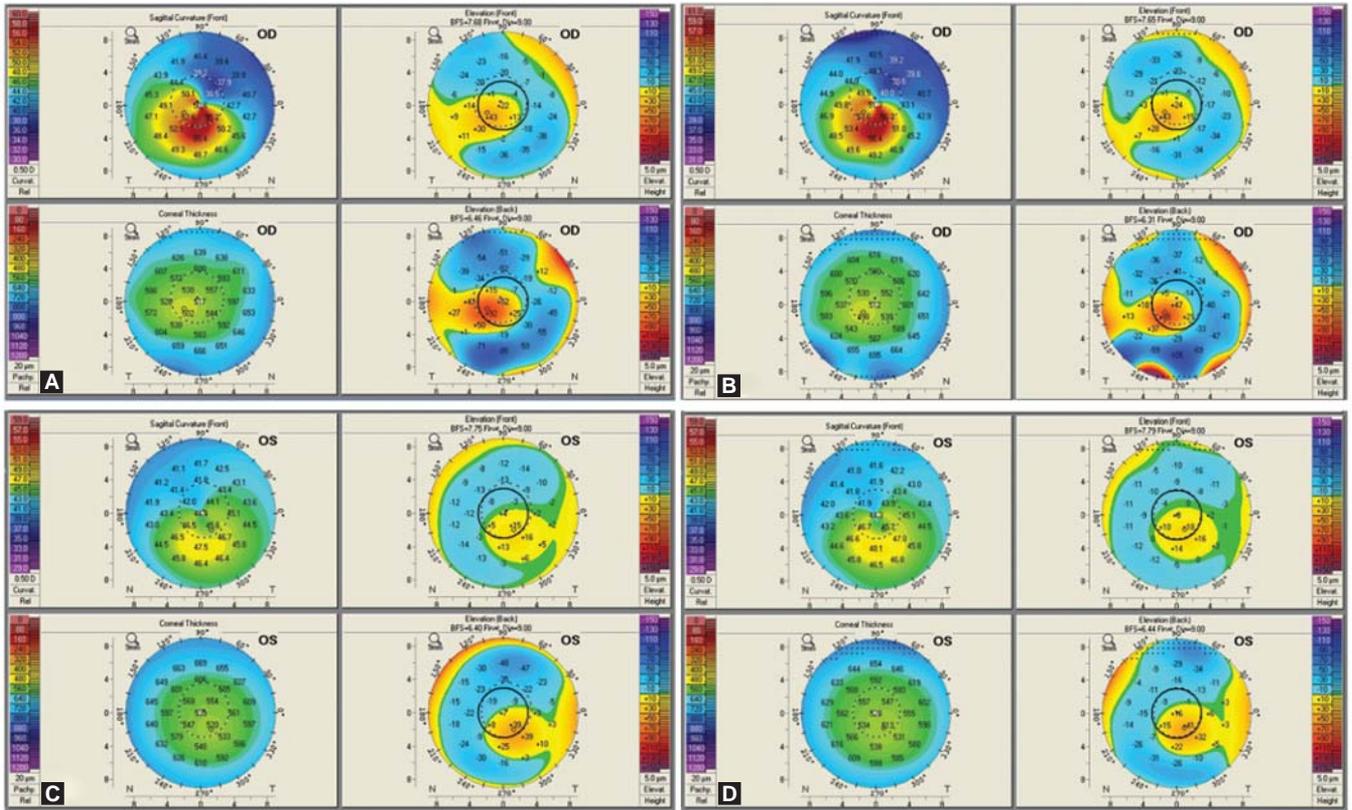
Currently, her visual acuity had dropped to 20/63 (+2.0 D Sph/-5.5 D Cyl@60) in the right eye and 20/32 (+1.5 D Sph/-2.5 D Cyl@100) in the left eye, with just a minor change in the refraction. Topography and Scheimpflug imaging showed minimal progression in both eyes, with the predominant change seen on the posterior elevation float (Figs 1A to D).

However, her specific complaint of difficulty in reading her mobile phone could not be correlated with this degree of progression alone. Fundus evaluation revealed temporal disk pallor in both eyes. Humphrey visual field analysis revealed bitemporal, congruous hemianopia (Fig. 2). She was immediately referred to a neurologist and magnetic resonance imaging (MRI) of brain revealed a nonfunctioning pituitary macroadenoma with hemorrhage and fluid levels (Fig. 3).

She was advised and underwent urgent surgical trans-sphenoidal decompression on the same day. Postoperatively, at 3 weeks, her vision improved to 20/30 in both eyes and visual field analysis showed partial resolution of the field defects (Fig. 4).

DISCUSSION

Visual acuity loss during or after pregnancy needs a high index of suspicion. In this case, the first differential



Figs 1A to D: Scheimpflug imaging of right eye at baseline (A) and presentation (B) left eye at baseline (C) and presentation (D) showing some progression of the keratoconus in both eyes

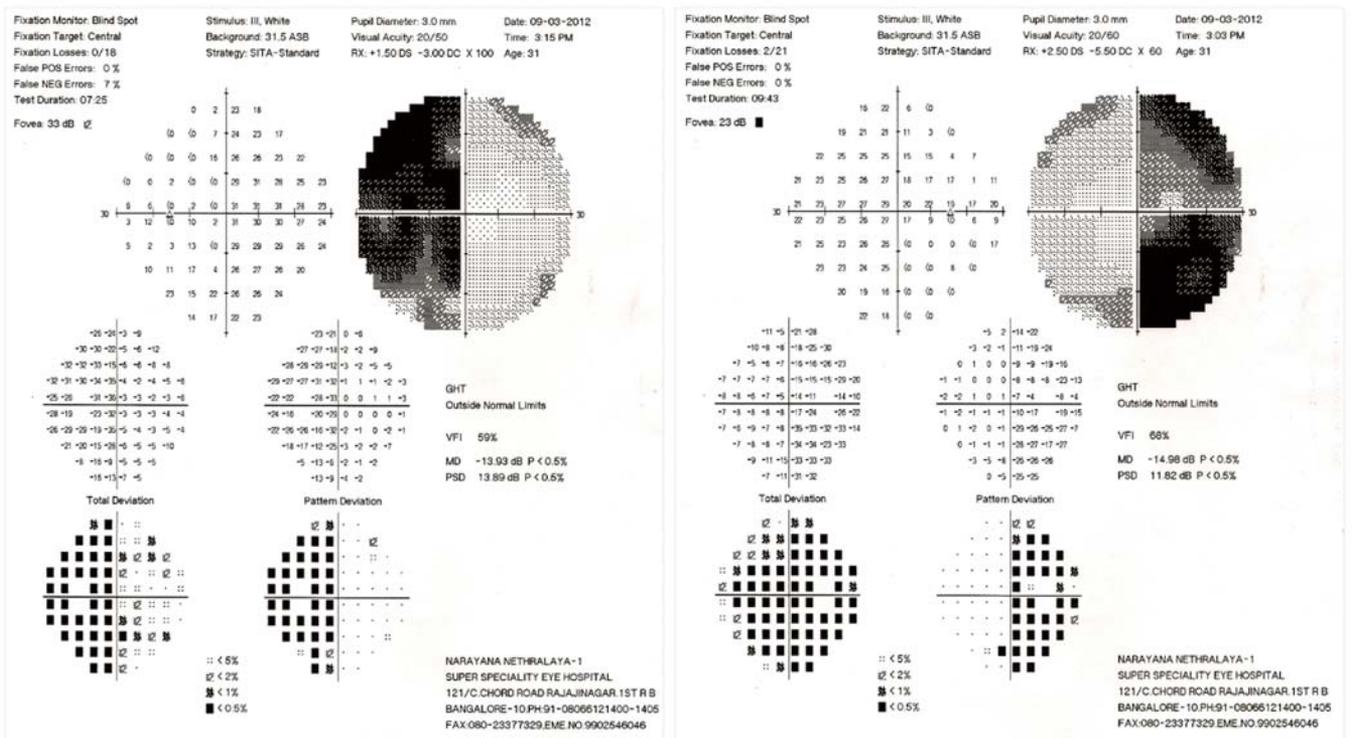


Fig. 2: Humphrey visual field analysis revealing classic bitemporal hemianopia

diagnosis to come to mind would be a progression of the keratoconus. Hormonal changes during pregnancy may affect corneal biomechanics negatively, and pregnancy may be an underestimated risk factor for progression of

keratoconus.² In a previous case series,² the authors showed pregnancy-induced keratoconus progression in patients with no accompanying disease. Changes in estrogen levels may play a role in corneal ectasia. A significant stiffness-reducing

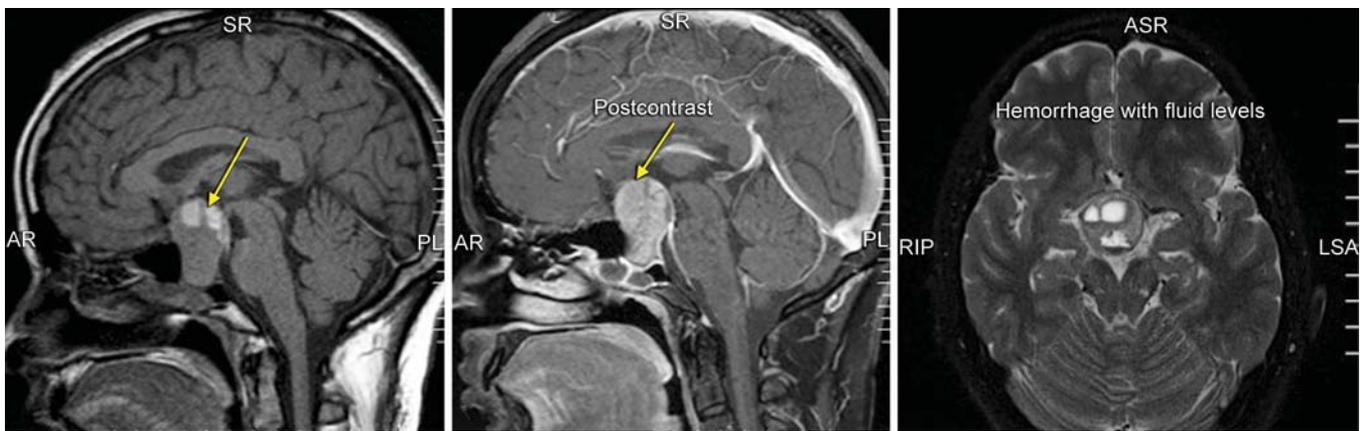


Fig. 3: MRI brain showing pituitary adenoma with fluid level, suggestive of hemorrhage

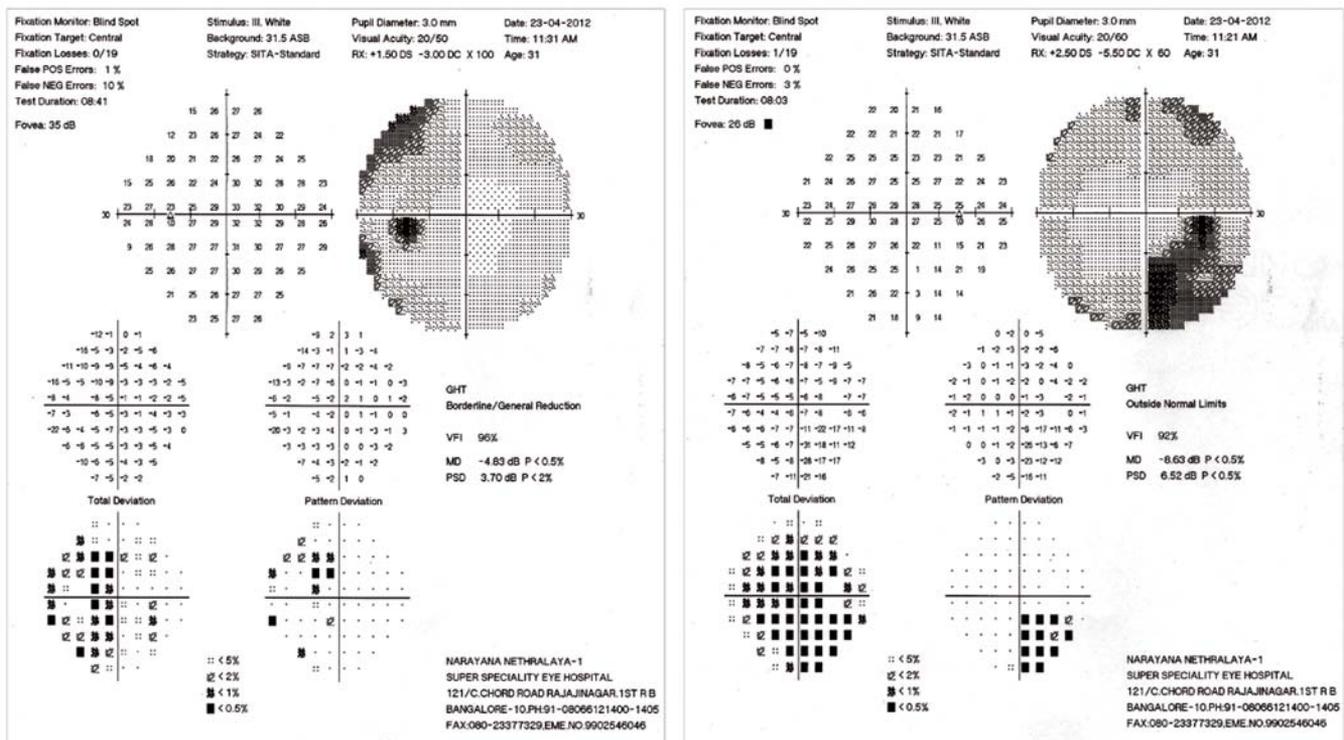


Fig. 4: Resolution of visual field defects following resection of the adenoma

effect of estrogen on cornea has been shown, which suggests that high estrogen states, such as pregnancy may predispose biomechanically weak corneas to ectasia development or progression.^{4,5} In addition, various studies have demonstrated that serum levels of matrix metalloproteinases (MMPs) are increased and serum levels of tissue inhibitors of MMPs (TIMPs) are decreased during pregnancy.⁶ A case of hypothyroxinemia-induced acute corneal hydrops during pregnancy,⁷ has also been reported. The authors suggested that the progression was related to thyroid gland dysfunction.

In this case, CDVA had significantly deteriorated despite a stable refraction. Moreover, the complaint of visual loss in a particular direction in a patient with longstanding compensated keratoconus could not be explained by mild

progression of the cone. A careful fundus evaluation and visual field analysis helped us clinch the diagnosis at a very crucial point.

This was an atypical presentation of pituitary apoplexy masquerading as a progressive keratoconus. Neuro-ophthalmological disorders, such as venous sinus thrombosis, benign intracranial hypertension, pituitary adenoma, meningioma and optic neuritis should be kept in mind as differential diagnosis in pregnant and early postpartum women.¹ Physiologic changes in pregnancy make vascular conditions more frequent, including retinal artery occlusion and pituitary apoplexy. Therefore, women presenting with visual acuity loss, visual field loss, persistent headaches or oculomotor palsies should be examined for the above possibilities.

Pituitary apoplexy is a potentially life-threatening emergency. Pituitary apoplexy occurs when an existing pituitary adenoma undergoes acute hemorrhage, infarct or both. The reported incidence of pituitary tumor apoplexy is usually less than 5% with a range between 0.6 and 10%. Common predisposing factors include closed head trauma, hypotension, hypertension, history of pituitary irradiation, cardiac surgery, anticoagulant therapy and pregnancy.^{8,9} Clinical features of pituitary apoplexy include sudden onset of headache, nausea, vomiting, visual symptoms, ptosis, altered mental status and endocrinologic dysfunction. The tumor and intrasellar contents may expand superiorly, compressing the optic chiasm, optic tracts, and optic nerves, producing decreased visual acuity in 52% and visual field defects in 64% of patients.

Although rare, visual symptoms could be the early presenting features, as in the case here. If the ophthalmologist does not maintain a high index of suspicion, timely diagnosis of such cases can be missed. Sellar MRI is a sensitive study for imaging the pituitary gland.¹⁰ Management includes emergently stabilizing patient's symptoms. Many patients will require intravenous fluids and blood transfusions. It is crucial that high dose corticosteroids are also administered to replace endogenous hormone deficiency and to prevent edema on parasellar structures. Often, urgent trans-sphenoidal surgical decompression is required as definitive treatment.¹⁰ Improvement in visual field, visual acuity and diplopia is typically observed after therapy. Preoperative preservation of at least light perception vision predicts better visual outcomes postoperatively compared with patients with absence of light perception. Surgery within the first week is associated with better visual outcome when compared with delayed surgery.

CONCLUSION

Neuro-ophthalmic conditions can masquerade themselves into varied presentations. Therefore in pregnant and postpartum women, a high index of suspicion and a thorough ophthalmic evaluation can often be vision and life saving.

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