Medical Management of Glaucoma Secondary to Idiopathic Elevated Episcleral Venous Pressure

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ABSTRACT

Idiopathic elevated episcleral venous pressure (IEEVP) is a rare cause of secondary open-angle glaucoma. The authors report a case of a 36-year male who presented with dilated episcleral vessels in the right eye. Gonioscopy revealed bilateral open angles with blood in Schlemm's canal in the right eye. Dilated retinal examination showed advanced glaucomatous optic neuropathy in the affected eye. Notably, the patient was also a known case of Gilbert syndrome. A thorough multidisciplinary evaluation, including orbital and neuroimaging as well as laboratory work-up, was undertaken to rule out secondary causes of elevated episcleral venous pressure (EVP) such as carotid-cavernous fistula, Sturge-Weber syndrome, or thyroid eye disease. The patient has been managed conservatively with topical antiglaucoma medications for the past two years, maintaining stable intraocular pressure and visual fields. The plan is to proceed with augmented trabeculectomy if any deterioration occurs, keeping in view the possibility of choroidal effusions.

Key Words: *Glaucoma, Open-angle glaucomas, Antiglaucoma agents, Intraocular pressures*.

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INTRODUCTION

Idiopathic elevated episcleral venous pressure (IEEVP), also known as Radius-Maumenee syndrome, was first described by Minas and Podos in 1968 and is an uncommon cause of secondary open-angle glaucoma.[1] International literature reports limited cases of the IEEVP. According to a case report published in 2020, there were 55 documented cases of IEEVP.[2] It is diagnosed after ruling out other causes of increased episcleral venous pressure (EVP), such as carotid-cavernous fistula, orbital varices, Sturge-Weber syndrome, retrobulbar tumour, and thyroid eye disease.[3] None of the reported cases have been associated with Gilbert syndrome. The pathogenesis involves unclear mechanisms, with genetic predisposition and congenital vascular abnormalities being probable contributing factors. The onset varies from the late teens to the sixth decade and affects both genders equally. It can be unilateral or asymmetrically bilateral.[4] To the best of the author's knowledge, IEEVP has not yet been reported in the Pakistani population.

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In this case report, we present a patient with IEEVP associated with Gilbert syndrome in a 36-year male patient from Pakistan.

CASE REPORT

A 36-year male patient, known case of hypertension and asymptomatic Gilbert syndrome, presented with a 5-month history of a decrease in light brightness and periocular heaviness in the right eye. The patient did not have a significant past ocular history.

On examination, the best corrected visual acuity (BCVA) was 6/6 in the right eye with -0.75 DS, and 6/6 in the left eye with a plano lens. Slit lamp examination showed dilated and tortuous episcleral veins in the affected eye only (Figure 1A). The right eye relative afferent pupillary defect was positive.

The patient had central corneal thicknesses of 584 µm and 595 µm, with corrected intraocular pressure (IOP) of 36 mmHg and 18 mmHg in the right and left eyes, respectively. Gonioscopy showed bilateral open angle with blood in the Schlemm's canal in the right eye only (Figure 1B). On fundus examination, the right cup to disc ratio (CDR) was 0.9 with a deep cup, laminar dot sign, nasalisation of blood vessels, pink neuroretinal rim, clear disc margins, and healthy macula. The left fundus examination was unremarkable with CDR of 0.2. The patient was diagnosed with secondary open-angle glaucoma in the right eye. Optical coherence tomography (OCT) retinal nerve fibre layer (RNFL) (Figure 1C), ganglion cell complex (GCC), and Visual Fields 24-2 (VF 24-2, Figure 1D), showed glaucomatous damage in the right eye and normal in the left eye. The patient was started on topical timolol (0.5%) eye drops in the right eye.

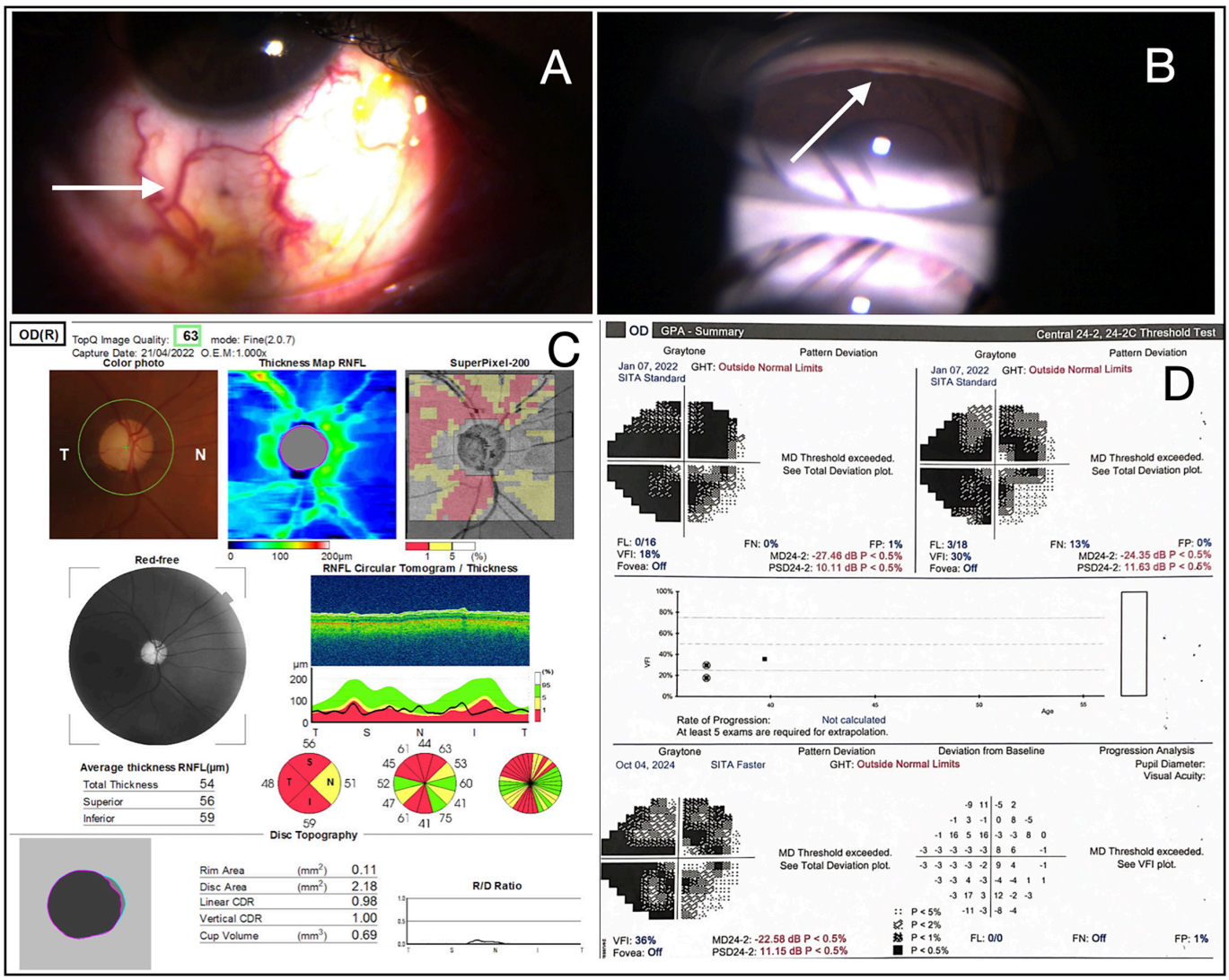

Figure 1: Right eye (A) Anterior segment photograph showing dilated and tortuous episcleral vessels (white arrow). (B) Blood in the Schlemm's canal on gonioscopy (white arrow). (C) Optical coherence tomography (OCT) retinal nerve fibre layer (RNFL) marked nerve fibre loss. (D) Visual field progression analysis.

He was investigated for the cause of episcleral venous abnormalities. The MRI brain and MRA brain were unremarkable. A neurologist was consulted to rule out the carotid-cavernous fistula. Thyroid function tests were within normal limits, ruling out the Grave's disease.

The patient had three follow-ups at 2-week intervals, and anti-glaucoma medications were added in a stepwise manner. A corrected IOP of 19 mmHg was achieved with a full topical anti-glaucoma treatment in the right eye (dorzolamide, timolol, brimonidine 0.2%, and latanoprost).

To monitor the disease, investigations were repeated at 3-month intervals. The patient was followed for a duration of 3 years, from December 2021 to January 2025, and remained stable on the maximum tolerable topical anti-glaucoma treatment.

DISCUSSION

In patients presenting with dilated episcleral veins and increased IOP, a suspicion of systemic involvement should be raised, and clinicians should look beyond ocular causes in such cases. This patient had been diagnosed with Gilbert syndrome three years prior to presenting with ocular complaints. Gilbert syndrome is characterised by elevated levels of the unconjugated bilirubin, which acts as a natural antioxidant and may reduce arteriolar stiffness.[5] The authors were unable to find any empirical or anecdotal evidence of an association between IEEVP and Gilbert syndrome.

In a case series by Rhee *et al*., six cases of IEEVP were reported, five of whom were males. All developed conjunctival injection in early adulthood and underwent glaucoma surgery in their sixth or seventh decade of life.[3] Another case series by Alam *et al*. included 13 patients, six of whom required trabeculectomy.[6]

The authors did not find any gender predilection. However, many reported cases are unilateral and exhibit similar clinical features.

Diagnostic evaluation should include a comprehensive ophthalmic assessment with OCT and automated perimetry for diagnosis and monitoring of RNFL damage and visual field defects. In addition, radiological imaging such as MRI and vascular imaging such as MRA or digital subtraction angiography should be performed to rule out carotid-cavernous fistula.[1,3]

As with primary open-angle glaucoma (POAG), medical management is the first-line therapy in IEEVP. A case report by Breazzano *et al*., supported by another case report published in 2016, documented the spontaneous resolution of presumed IEEVP with medical treatment.[6,7]

The efficacy of laser trabeculoplasty in this population remains unclear, with studies suggesting minimal IOP reduction from argon laser trabeculoplasty.[2]

Surgical treatment is indicated when medical treatment is ineffective, and the choice of procedure is crucial due to the high risk of complications such as expulsive haemorrhage and choroidal effusions. Trabeculectomy with slow decompression using anterior chamber maintainers, tight trabeculectomy, and prophylactic sclerostomies are debatable options.[3] According to a case reported from India, a 40-year patient with IEEVP underwent trabeculectomy and despite non- hypotonus IOP, developed suprachoroidal detachment.[8]

Non-penetrating deep sclerectomy, despite being challenging, is a safer option as it precludes sudden hypotony.[1]

In the present case, there was adequate IOP response to medical therapy, with a stable optic disc, OCT, and visual fields. The future management plan is to proceed with medical management, keeping in view the possibility of spontaneous resolution, but with a low threshold for surgery if automated perimetry shows progression.

In conclusion, IEEVP is a rare cause of secondary open-angle glaucoma. It can be managed by observation (spontaneous resolution), medically, or surgically, keeping in view the possible risks associated with both treatment options.

PATIENT'S CONSENT:
Written informed consent was obtained from the patient.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
AS: Contributed to the idea, planning of the case report, wrote the initial draft, and helped improve the manuscript through critical review.
AK: Interpreted the clinical findings, reviewed and revised the manuscript, and ensured the accuracy and integrity of the work.
Both authors approved the final version of the manuscript to be published.

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