Case Report

Radius-Maumenee Syndrome, A Rare Entity: A Case Report

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ABSTRACT

Background: Radius–Maumenee syndrome (RMS) is a rare idiopathic condition characterized by the enlargement of episcleral vessels and an elevation in intraocular pressure (IOP) leading to secondary glaucoma.

Case: In this report, we present a case of RMS in a 23-year male who experienced redness in Right Eye (RE) and intermittent headaches for a duration of 10 years. He had a best corrected visual acuity of 6/6 in both eyes.

Observations: During slit lamp biomicroscopy, episcleral venous engorgement was observed in both eyes. IOP was 28 mmHg in the RE and 14 mmHg in Left Eye (LE). In fundus photographs of RE and LE, cup asymmetry and retinal nerve fiber layer defects in superotemporal and inferotemporal regions of RE were present. Optical coherence tomography (OCT) Optic Nerve Head with hood report of RE showed loss of double hump pattern with asymmetric and severe retinal nerve fiber layer thinning in superior and inferior quadrant. Visual field testing of RE showed superior and inferior arcuate scotoma. Brain and orbit magnetic resonance angiography (MRI Angiogram) revealed no abnormal voids indicating cavernous fistula or other orbital lesions. The presence of secondary open angle glaucoma with episcleral venous engorgement and negative test results for other potential conditions led to the diagnosis of RMS in the patient. The patient is now under combination of aqueous suppressants and uveoscleral outflow increasing drugs.

Conclusion: This case serves as a reminder to ophthalmologists about potential association of glaucoma if there's enlargement of episcleral vessels in eyes without inflammation.

Key words: Episcleral veins dilatation, Idiopathic, Radius Maumenee syndrome, Raised intraocular pressure, Secondary glaucoma.

Financial Interest : Nil
Conflict of Interest : Nil

Received : 17.11.2022
Accepted : 29.12.2022

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Access this article online
Website: www.nepjol.info/index.php/NEPJOPH
DOI: https://doi.org/10.3126/nepjoph.v15i1.49617

ISSN: 2072-6805, **E-ISSN:** 2091-0320

INTRODUCTION

Elevated episcleral venous pressure (EVP) is a clinical observation that may be linked to increased intraocular pressure (IOP) and glaucoma if not treated over an extended period. According to the Goldmann equation: IOP = F/C + EVP (F-aqueous humor inflow, C-outflow facility). The normal episcleral venous pressure is 8-10 mm Hg; variability depends on the measurement technique (Brubaker, 1967). In the acute setting, a linear relationship exists, where a 1 mmHg increase in episcleral venous pressure corresponds to a 1 mmHg rise in intraocular pressure. However, the understanding of this relationship in chronic cases is not as clear. The causes of elevated EVP can be categorized into three groups:1) **Venous obstruction** involves conditions like thyroid associated ophthalmopathy, retrobulbar tumor, thrombosis in cavernous sinus or orbital veins, inflammation in episcleral or orbital veins, and blockage of the superior vena cava, 2) **Arteriovenous anomalies** including carotid artery—cavernous sinus fistula, orbital varices, dural shunts and Sturge-Weber syndrome; and 3) **Idiopathic**. Idiopathic Elevated Episcleral Venous Pressure (IEEVP) with secondary open angle glaucoma, also known as Radius-Maumenee syndrome, is an uncommon clinical condition characterized by elevated intraocular pressure and subsequent damage of the optic nerve by glaucoma (El Alami et al, 2021). IEEVP was first documented by Minas and Podos in 1968 (Minas and Podos, 1968). In 1978, Radius and Maumenee reported four additional cases with idiopathic dilated episcleral vessels and secondary open-angle glaucoma (Radius and Maumenee, 1978). Only 46 cases have been published since then (El Alami et al, 2021; Marques et al, 2018). Our aim of reporting this case is because of the rarity of the condition which is diagnostically and therapeutically very challenging. RMS is an infrequent condition and represents an unusual cause of secondary glaucoma. As far as we are aware, this is the first reported case in Nepal.

CASE REPORT

A 23-years-male presented to our outpatient department (OPD) with chief complaints of redness in RE for 10 years and headache on and off for 10 years. He did not have a history of eye disease and systemic disease. There is no injury or trauma to the head or neck, no history of proptosis – either persistent or intermittent, no history of anterior neck swelling or symptoms suggestive of dysthyroid orbitopathy, no complains of experiencing diplopia, pulsing sounds in ears, and no pulsing sensation in the eye socket. His visual acuity was 6/6 in both eyes. The adnexa examination revealed no notable findings, ocular motility was within the normal range, and there were no signs of relative afferent pupillary defect. There was no visible proptosis or chemosis and retropulsion was normal. Neurological exam showed no focal deficits. No carotid or ocular bruits were detected during the examination. The anterior segment evaluation showed numerous tortuous and engorged episcleral vessels in the right eye, while the conjunctival vessels appeared normal with no signs of chemosis (Figure 1). Cornea was clear. Pupil was round, regular and reactive with 3 mm on slit lamp examination, anterior chambers were deep and quiet without iris alterations and had clear lens in both eyes. IOP was 28mm Hg on RE 14 mm Hg on LE.

Gonioscopy showed open iridocorneal angles without pigment or neovascularization and no visible blood in Schlemm's canal. Fundoscopy of RE revealed a cuptodisc ratio of 0.8:1 with superior and inferior neuro-retinal rim (NRR) thinning of optic disc, but no dilation or tortuosity of the retinal vessels were observed and LE had round, sharp optic disc of 0.3:1 CDR with healthy NRR (Figure 2). Optical coherence tomography (OCT) of optic nerve head and retinal nerve fiber layer (RNFL) with hood report were indicative of glaucomatous damage of the right eye (Figures 3 and 4) and normal LE. Time Of Flight (TOF) brain MR Angiogram axial image showed normal intracranial arterial circle of Willis with no aneurysm or other vascular malformation (Figure 6).

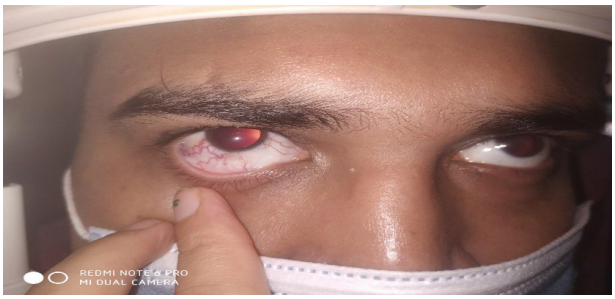

Figure 1: Anterior segment photo of RE and LE showing episcleral vessel dilation and tortuosity in RE.

Figure 2: Fundus photo of the RE and LE showing cup-disc asymmetry of 0.8:1 RE, 0.3:1 LE.

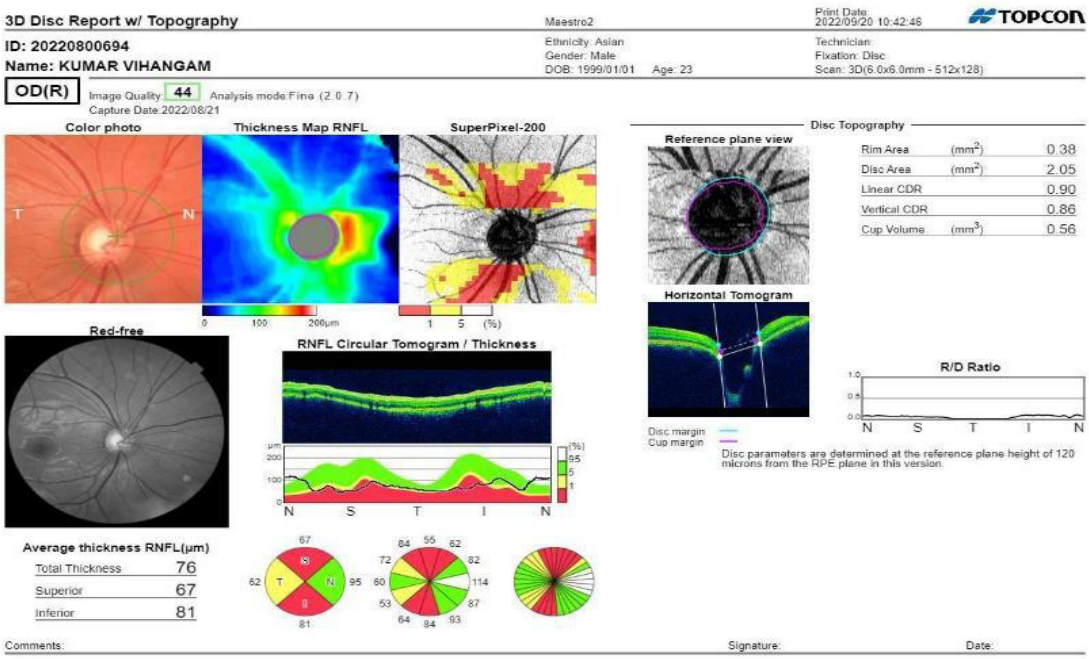

Figure 3: Optical coherence tomography of optic nerve head and retinal nerve fiber layer RE) showed loss of double hump pattern with asymmetric and severe retinal nerve fiber layer thinning in superior and inferior quadrant.

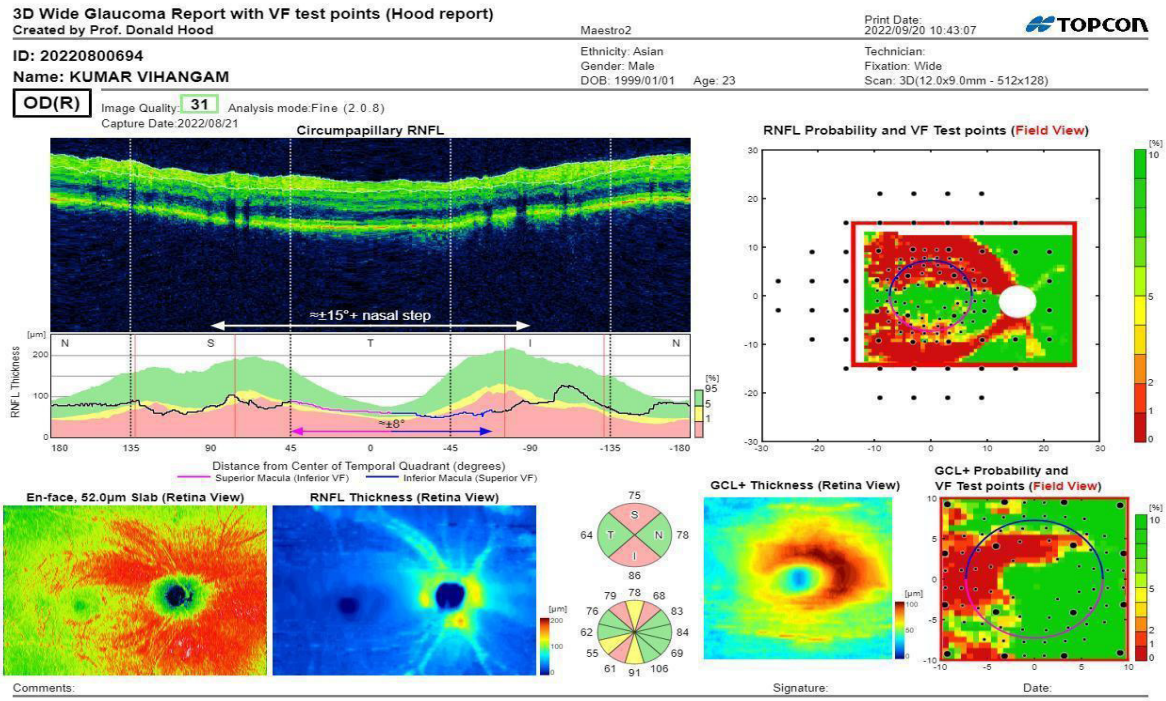

Figure 4: Hood report with visual field and GCL complex RE - Perimetry showed right-sided superior and inferior arcuate scotoma while GCL thickness map showed ganglion cell thinning in RE.

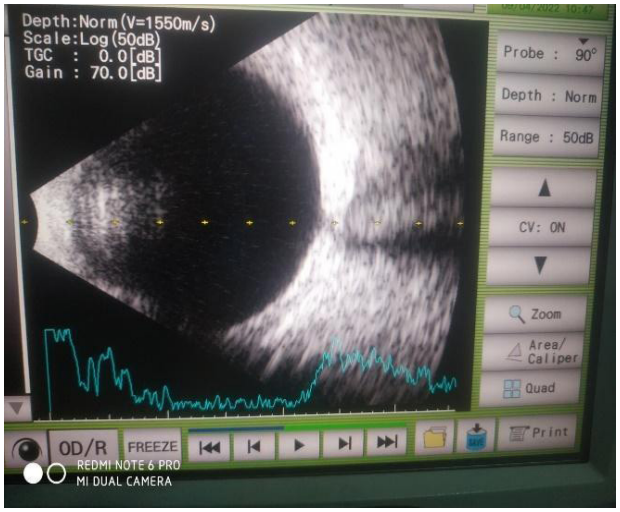

Figure 5: B scan and A scan of RE showing normal findings.

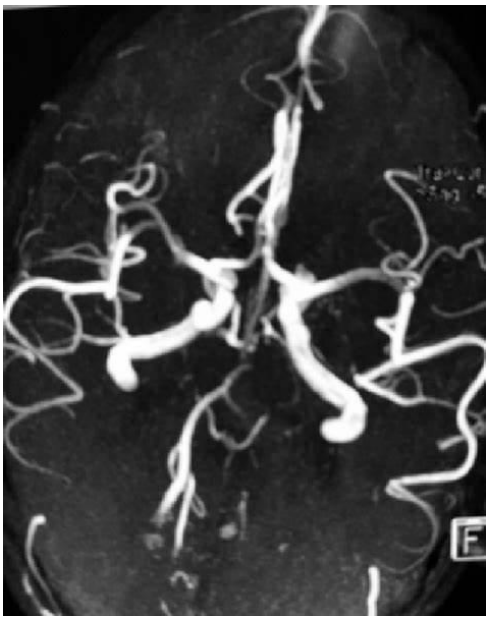

Figure 6: TOF brain MRA MIP axial image within the normality.

The patient also had MRI brain and orbit with techniques of Brain- axial FLAIR and DW1, Orbit -Axial T1 SE and T2 FRFSE, Coronal T1 SE and T2 FRFS Axial FIESTA and DW1 showing no abnormal flow voids in the cavernous sinus with no significant abnormality in brain parenchyma and orbit. This also excluded hypertrophy of the extraocular muscles, orbital or intracranial space-occupying lesions and presented no indirect signs of vascular abnormalities. Chest x-ray and color doppler echocardiography showed no abnormalities. After ruling out the intracranial and intraorbital causes for raised episcleral venous pressure, we came to the final diagnosis of RMS.

The patient was prescribed a combination of brimonidine 0.2 mg/ml and timolol 0.5 mg/ml as an antiglaucoma medication to suppress aqueous flow. A subsequent appointment was arranged for two weeks later, during which the patient's intraocular pressure (IOP) was 26 mm Hg in RE while the LE measured 18 mm Hg. Since the target pressure was not achieved, the patient was given additional treatment with Dorzolamide hydrochloride 2% and bimatoprost 0.1mg/ml in the RE. If the topical therapy is unsuccessful in preventing the progression of the disease, the patient will be recommended for filtering surgery in the RE.

DISCUSSION

The intraocular pressure is influenced by the speed at which aqueous humor is produced, drainage and episcleral venous pressure. Normal episcleral venous pressure is from 8 to 10 mm of Hg and for every 1 mm Hg increase in episcleral venous pressure, there is a corresponding 1 mm Hg rise in intraocular pressure. The aqueous humor is directed into the episcleral venous plexus, then into anterior ciliary vein, and enters the superior ophthalmic vein. Beyond the annulus of Zinn, it enters the cavernous sinus, then proceeds through the internal jugular vein to reach the superior venacava, eventually draining into the right atrium. Consequently, any obstruction in this drainage pathway could lead to an increase in episcleral venous pressure. Prolonged elevation of episcleral venous pressure often causes reduction in aqueous outflow facility and this consequently causes secondary glaucoma. The primary indicator of increased EVP is the dilatation of episcleral and conjunctival vessels, which is commonly observed. Such diseases include venous obstructions: examples of these are dysthyroid ophthalmopathy, anterior and posterior scleritis, tenonitis, superior vena cava syndrome and those obstructing the orbital venous drainage like retrobulbar tumors and cavernous sinus thrombosis, arteriovenous fistulas from carotid-cavernous fistulas, orbital varices, Sturge Weber syndrome and idiopathic. Arteriovenous fistulas are the most frequent cause of raised IOP due to increased EVP. These usually present with associated features such as chemosis, pulsatile proptosis and diplopia and clinical diagnosis is confirmed by CT or MRI angiography showing dilated superior ophthalmic vein or cavernous sinus. Sturge-Weber syndrome mostly has facial cutaneous angiomatosis, and also might have a history of seizure along with eye signs. Thyroid ophthalmopathy was ruled out with relevant history, clinical examination and specific blood tests.

RMS diagnosis relies on clinical features such as episcleral vessel dilation and elevated IOP,

resembling the presentation of open angle glaucoma. While the exact pathogenesis of this condition remains unclear, it is believed to result from a congenital abnormality in the vasculature, leading to abnormal pressure in blood drainage (Grieshaber et al, 2007). One hypothesis explaining the difficulty in venous return points to the hyalinization of schlemms's canal, which causes obstruction in the distal part of anterior chamber (Lanzl et al, 1996). It can occur as a unilateral or a bilateral condition and at any age, although all the cases reported have been in persons over 20 years of age (de Keizer, 1983). There is no known sex predilection, but the majority of cases reported were among women (de Keizer, 1983). There is familial predisposition in RMS, and it has been described in a mother and her daughter (Minas and Podos, 1968). There is currently limited research on the genetic component of this disease. The treatments for RMS are comparable to those used for primary open angle glaucoma (POAG) (Stock et al, 2013). Due to the compromised aqueous outflow in RMS, aqueous suppressants are more efficacious in the management of increased IOP caused by elevated episcleral venous pressure (EVP). Meanwhile medications or techniques that aim to increase aqueous humor outflow through the trabecular drainage pathway, such as pilocarpine, Selective Laser Trabeculoplasty (SLT) and Micro pulse Laser trabeculoplasty (MLT) have limited efficacy. Apraclonidine has demonstrated effectiveness in reducing intraocular pressure in conditions of elevated episcleral venous pressure by inducing arterial vasoconstriction, which subsequently reduces blood flow to the eye. In cases of uncontrolled intraocular pressure by drugs, surgical interventions such as trabeculectomy, sclerectomy and sinusotomy may become necessary. There have been reports indicating that eyes undergoing filtering surgery are at a heightened risk of postoperative uveal effusion (Parikh et al, 2011).

CONCLUSION

This case reminds the ophthalmologist about the possibility of glaucoma in cases of episcleral vessel dilation in non-inflamed eyes. A diagnosis of RMS should be considered in cases of chronically red eye without symptoms of allergy, discomfort or discharge, especially when there is asymmetrical or unilateral elevation of intraocular pressure. It is the diagnosis of exclusion which requires ruling out the causes of increased EVP.

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