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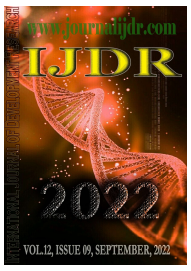


CASE STUDY OPEN ACCESS

# 20 YEAR OLD MALE WITH IDIOPATHIC ELEVATED EPISCLERAL VENOUS PRESSURE: A CASE REPORT

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## ABSTRACT

High episcleral venous pressure results in IOP elevation and optic nerve glaucomatous damage. Any obstruction or communication with the arterial system, like a cavernous sinus thrombosis or a carotid cavernous fistula, can impair drainage of the episcleral venous system and cause retrograde IOP elevation. In rare cases, no cause for the high venous pressure is found. Such cases are diagnosed as having Idiopathic Elevated Episcleral Venous Pressure (IEEVP). So far only 27 cases of IEEVP have been published in the English literature, none of them with a follow up of more than one year. We present the case of a 20 year old male with IEEVP that was diagnosed in 2009 and has been under follow up with good IOP control since then.



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# INTRODUCTION

Intraocular pressure (IOP) and its relationship with episcleral venous pressure was described long time ago by the Goldman equation, $P_0=(F/C)+P_v$, and it depends on three factors, the rate of aqueous production(F), the facility of aqueous outflow from the eye (C) and episcleral venous pressure($P_v$). As can be seen by this equation any elevation in scleral venous pressure will directly lead to an elevation in IOP due to impeded aqueous outflow. [1] Patients with elevated episcleral venous pressure may present with a variety of clinical findings including dilated episcleral veins without inflammation, open iridocorneal angle, blood in Schlemm's canal and elevated IOP, which may lead to glaucomatous optic nerve damage.[2] In most cases elevated episcleral pressure can be attributable to an underlying cause like a carotid-cavernous-sinus fistula, cavernous sinus thrombosis, superior vena cava syndrome, dural arteriovenous shunt, Sturge–Weber syndrome, thyroid ophthalmopathy, orbital obstructive lesions or orbital varices. In rare cases, no cause for the elevated episcleral pressure can be found. Such rare cases are diagnosed as having Idiopathic Elevated Episcleral Venous Pressure (IEEVP)[2-3]. Around 40 cases of IEEVP have been reported in the literature, none of them reporting on the long term follow up and prognosis of this condition. We present a case of bilateral IEEVP with 10 years of follow up.

## CASE PRESENTATION

A 20 years old male was referred to a tertiary center from a rural area with a history of progressive bilateral visual loss that started 5 years prior to referral. One year after symptoms developed the patient was diagnosed with glaucoma and treated with topical latanoprost. Besides the visual symptoms the patient referred only occasional moderate headaches. A detailed family history of the patient could not be obtained because the patient was adopted in a confidential manner. Both eyes had an uncorrected visual acuity (UCVA) of hand movement (HM) which improved to 20/30 on right eye (RE) and did not improved in the left eye (LE) with refraction. A LE 30° exotropia without any movement restrictions was also found. On slit lamp biomicroscopic examination bilateral engorged episcleral veins were found(Figure1), as well as mid dilated non-reactive pupils.On gonioscopy an open iridocorneal angle (Schaffer grade IV) with blood present in Schlemm´s canal (Figure 2) was observed. Measured IOP was 33mmHG in RE and 42mmHG in the LE. Fundus examination revealed bilateral optic disc pallor with a 0.9 excavation in the RE and total optic disc cupping in the LE (Figure 3) and retinal blood vessel tortuosity. A visual field (VF) test (SITA standard white/white, stimulus II 24-2) showed severe glaucomatous damage on RE (DM -34.30dB,DSM 1.83dB) and could not be completed on the LE due to low visual acuity. A stimulus V, central 10-2 VF was

then done in the RE which showed a small 5° visual island (Figure 4). In the fluorescein angiography (FANG) slow venous filling and peripheral retinal and choroidal hypoperfusion where noted. An orbit ultrasonography (US) demonstrated bilateral dilated supraorbital veins (Figure 5). Due to the severity of the case a bilateral Ahmed valve was implanted. The patient developed hypotony in the LE during the immediate postoperative period that got complicated with a sub macular choroidal detachment that resolved with conservative management. IOP on discharge day was 10mmHg in RE and 8mmHg in LE.

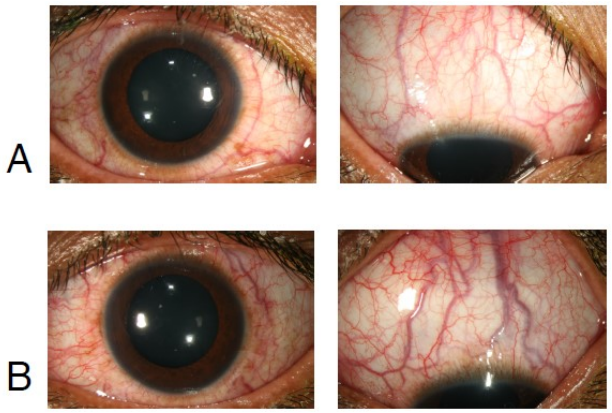


**Figure 1. Dilated episcleral vessels in RE (A) and LE (B)**

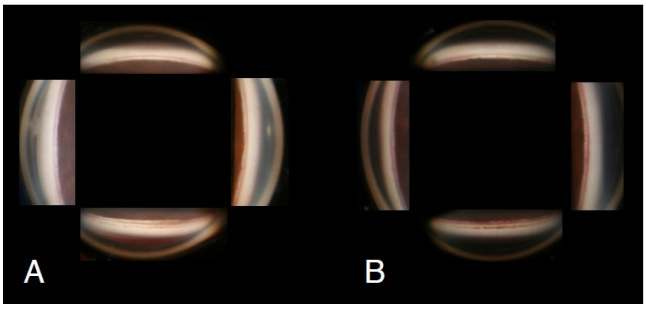


**Figure 2. Gonioscopic view of Iridocorneal angle of RE (A) and LE (B), note the presence of blood in Schlemm´s canal, mainly in LE**

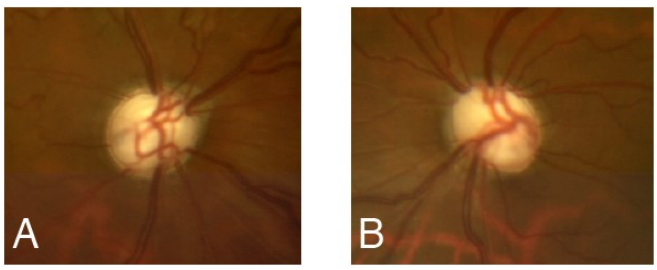


**Figure 1. Optic nerve RE (A) and LE (B)**

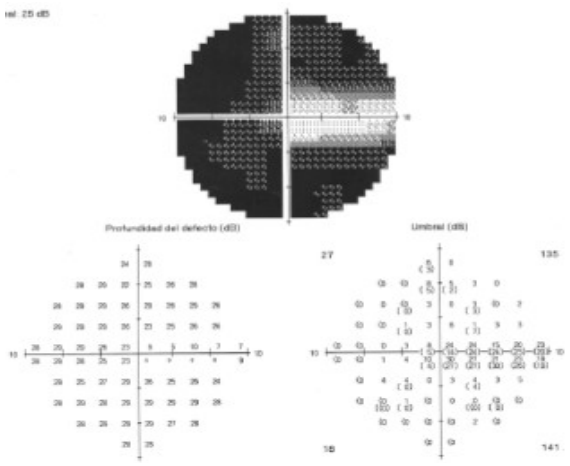

**Figure 4. RE central 10-2 white/white visual field, note the advanced glaucomatous visual field damage**

Patient underwent further examination as an outpatient once IOP was stabilized, a CT angiography and an MRI angiography were done, but showed only hypoplasia of the right transverse sinus, enlargement of the left transverse sinus and a generalized dilatation of the venous intracranial circulation that more pronounced in the left side. Due to a dilatation of the jugular bulb, a CT angiography of the neck and chest was done to rule out any obstruction of the venous drainage at this level without any pathologic findings. Since the patient had no retrobulbar pulsations or murmurs a low flow arteriovenous dural fistula was deliberately looked for and ruled out. 3 months after surgery patient presented with an IOP of 30mmHg in RE and 34mmHg in LE, so topical treatment with timolol, dorzolamide and brimonidine was started with partial response, so a topical bimatoprost was added. Since then the patient has been under follow up with controlled IOP, ranging from 16 to 18 mmHg and stable visual field during the last 10 years without any change in treatment.

# DISCUSSION

The first cases of IEEVP were published by Minas and Podos in 1968 and were members of the same family [4]. Not much was published until 1978 when Radius and Maumenee published 4 cases of open angle glaucoma associated with dilated episcleral blood vessels that did not present any known causes to have elevated episcleral venous pressure and thus coined the termIdiopathic Dilated Episcleral Veins (IDEV) [5]. Both terms, IEEVP and IDEV are used indifferently. Since then only 27 cases have been published in the English medical literature, and none of the cases have more than one year of follow up [2,3,6] The onset of the disease varies from the late teens to the 4$^{th}$ decade of life. IEEVP is a diagnosis of exclusion, and only after a detailed physical examination and proper imaging techniques have ruled any other cause for elevated episcleral venous pressure, a diagnosis of this entity can be done [2,3,6]. Aqueous humorexits the eye via one of the two described pathways (trabecular or uveoscleral) and enters the episcleral venous plexus, flows through the anterior ciliary veins which drain to the superior ophthalmic vein. It then exits the orbit via the superior orbital fissure to the cavernous sinus and follows the drainage pathway via the internal jugular vein into the systemic circulation. Any obstruction at any level of this drainage pathway can cause a retrograde pressure elevation which will ultimately result in IOP elevation [1]. In our patient hypoplasia of the right transverse sinus, enlargement of the left transverse sinus and a generalized dilatation of venous intracranial circulation more pronounced in the left side and dilatation of the jugular bulb where found on CT and MRI angiography. These are known to be normal anatomical variants and were not associated with any neurological findings. A neck and chest CT angiography ruled out any obstruction on the drainage pathway up to the right atrium. No other published case describes these intracranial vascular findings. Since no exact pathophysiologic cause has been discovered for IEEVP, treatment is aimed at reducing IOP to limit optic nerve damage in a similar way as in primary open angle glaucoma. Since our patient had significant optic nerve damage an Ahmed valve implantation was done and since then he has been on maximal topical therapy, this treatment has been effective in maintaining a stable IOP and thus conserving his visual field on the RE. Our patient's surgery was complicated by a sub macular choroidal detachment, and even thought the location of such detachment is unusual, choroidal detachments have been described before in patients with dilated episcleral veins who underwent filtrating surgery [8], It is important to be aware of such possibility in these patients and take all possible measures to avoid it.

# CONCLUSIONS

IEEVP is an uncommon condition and not much information has been published about it, this case demonstrates that with proper treatment and follow up, IOP management and preservation of useful vision is feasible in these cases. The authors certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation

in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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