CASE REPORT

Radius-Maumenee syndrome: a rare cause of glaucoma

Sara Homem de Melo Marques,^{1,2} Cláudia Farinha,^{1,3} Amélia Martins,¹ Pedro Faria^{1,3}

¹Centro de Responsabilidade Integrado de Oftalmologia, Centro Hospitalar e Universitário de Coimbra EPE, Coimbra, Portugal

²Ophthalmology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

³Faculdade de Medicina da Universidade de Coimbra, Coimbra, Portugal

Correspondence to

Dr Sara Homem de Melo Marques, saramelomarkes@gmail.com

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SUMMARY

Case presentation of a 41-year-old woman with long-standing bilateral eye injection whose clinical findings included bilateral episcleral vessel engorgement and tortuosity, raised intraocular pressure with open iridocorneal angles and left optic disc cupping. All remaining objective examination was normal. She underwent cranial and orbits CT and CT-angiography scans which were unremarkable. Optical coherence tomography of the peripapillary nerve fibre layer and automated perimetry were compatible with advanced glaucomatous damage on the left eye. The patient was diagnosed with idiopathic elevated episcleral venous pressure or Radius-Maumenee syndrome. If medical therapy is unable to prevent disease progression, left eye filtering surgery will be proposed.

BACKGROUND

Episcleral venous pressure (EVP) is difficult to measure and, to date, physiological factors that control it are still poorly understood. Despite this, several pathologies are known to cause EVP increase with consequent increase in intraocular pressure (IOP). Arteriovenous fistulas are the most frequent cause of raised IOP due to increased EVP.¹ Other possible causes are venous sinus thrombosis, Sturge-Weber syndrome, thyroid ophthalmopathy or space-occupying lesions.^{2,3} Idiopathic elevated episcleral venous pressure (IEEVP) or Radius-Maumenee syndrome is a rare clinical entity characterised by increased EVP that leads to increased IOP and glaucomatous damage of the optic nerve.⁴ Since its first description in 1968,⁵ only 44 cases have been published.

CASE PRESENTATION

We present a case of a 41-year-old woman who was sent to our ophthalmology emergency department with the suspicion of carotid-cavernous fistula. She was previously healthy and was not on any chronic systemic medication but had chronic bilateral conjunctival hyperaemia for over a year and intermittent subacute headaches. She had recently been seen in an ophthalmic private practice with an IOP of 25 mm Hg in the right eye (OD) and 28 mm Hg in the left eye (OS), and was started on a fixed combination eye-drops containing travoprost and timolol in both eyes daily.

Her first clinical assessment showed 20/20 visual acuity bilaterally (OU), preserved and symmetric pupillary reactions, full ocular motility with no

evidence of proptosis or eye misalignment. Biomicroscopy presented engorged and tortuous episcleral vessels OU, more evident on the left, normal conjunctival vessels, clear anterior chambers, no iris alterations and clear lenses (figure 1). IOP measured by Goldmann applanation tonometry was 14 mm Hg OD and 19 mm Hg OS. Funduscopy revealed an apparently healthy disc (cup:disc ratio 0.4) and posterior segment OD, and severe disc cupping with pallor (cup:disc ratio 0.9) with otherwise normal posterior segment OS (figure 2). Gonioscopy showed open iridocorneal angles without pigment or neovascularisation and no visible blood in Schlemm's canal.

INVESTIGATIONS

The patient had CT scans of the head and orbits, which excluded hypertrophy of the extraocular muscles, orbital or intracranial space-occupying lesions and presented no indirect signs of vascular abnormalities.

Optical coherence tomography (OCT) of the peripapillary nerve fibre layer showed asymmetric and severe retinal nerve fibre layer thinning on the left side (figure 3) and automated perimetry showed left-sided superior and inferior arcuate scotomata (figure 4A). These examinations presented no significant alterations on the right side (figures 3 and 4B).

To definitely exclude intracranial vascular abnormalities, a CT angiography was performed. It had no evidence of arteriovenous fistulas, venous sinus thrombosis, aneurismatic dilatations, stenosis or irregularities.

DIFFERENTIAL DIAGNOSIS

Several diseases can lead to intraocular hypertension due to increased EVP, the most frequent being intracranial arteriovenous fistulas, namely carotid-cavernous fistulas.^{1,2} These usually present associated features such as chemosis, proptosis, pulsatile proptosis and diplopia and clinical diagnosis is confirmed by CT or MRI angiography.⁶

Other possible causes are venous sinus thrombosis, Sturge-Weber syndrome, thyroid ophthalmopathy or orbital space-occupying lesions.^{2,3,7}

A rare aetiology for the association of intraocular hypertension and episcleral venous dilatation is IEEVP. This syndrome leads to chronically increased IOP and glaucomatous damage of the optic nerve.⁴ It is a diagnosis of exclusion that cannot be made without investigations.

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Figure 1 Anterior segment photos of the right (A) and left (B) eyes showing episcleral vasculature engorgement and tortuosity.

TREATMENT

Therapy with antiglaucoma medication was maintained, and a re-evaluation visit was scheduled in 2 weeks for additional complementary examinations.

OUTCOME AND FOLLOW-UP

On the first re-evaluation visit, the patient had suspended the antiglaucoma drops without previous notice. Objective examination was globally similar, except for IOP which had risen to 21 mm Hg OD and 24 mm Hg OS. The fixed combination of travoprost 40 µg/mL and timolol 5 mg/mL was reintroduced with IOP normalisation. However, 3 months later, OS automated perimetry showed decrease in global sensitivity and larger scotoma size. OS therapeutic options were discussed with the patient and topical brinzolamide 10 mg/mL was added. If topical therapy is unable to avoid disease progression, the patient will be proposed filtering surgery OS.

DISCUSSION

IEEVP (Radius-Maumenee syndrome) is characterised by arteri-
alised, engorged and tortuous episcleral veins,² with raised IOP
and open angle glaucoma (after the exclusion of other causes for
raised EVP).^{4,5} To our knowledge, this is just the 45th published
case of IEEVP since it was first described by Minas and Podos⁵
in 1968. Age of onset is variable, with cases being diagnosed
between late teens and the sixth decade of life,^{3,8} it seems to
affect females and males almost equally, and most cases are
bilateral with asymmetric involvement.⁸ Congenital vascular
abnormalities and a familial predisposition have been proposed

for its pathogenesis but, so far, no clear mechanism has been
recognised.⁷

As already mentioned, this syndrome is an exclusion diag-
nosis and can only be established after other more common
and potentially life-threatening conditions have been discarded.
Among these are the causes of elevated EVP described in the
"Differential diagnosis" section. Imaging examinations such
as CT or MRI scans should be done to exclude intraorbital
and intracranial pathologies, while vascular imaging tech-
niques such as angiography and CT or MRI angiography
are used to exclude cerebrovascular abnormalities. Orbital
colour Doppler is a non-invasive method to examine orbital
and ocular blood flow. In cases of carotid-cavernous fistula, it
shows dilated superior ophthalmic veins with reversal of the
normal blood flow.⁶ Despite being useful, this examination is
neither routinely performed nor generally accessible, and has
only been used in IEEVP suspects by Foroozan *et al*⁷ and Lanzl
et al.⁹ Both groups reported normal calibre of superior
ophthalmic veins with normal retrograde blood flow.

Complementary study should be associated with an exhaus-
tive ophthalmological evaluation. Gonioscopy typically
features an open iridocorneal angle with blood in Schlemm's
canal. This is not pathognomonic, and blood may sometimes
not be seen,⁸ as was the case in our patient. Gonioscopy is
also important to help exclude other causes of glaucoma. Since
symptoms have sometimes been present for years at the time
of diagnosis, OCT and automated perimetry should be done to

Figure 2 Retinography of the right (A) and left (B) eyes showing left eye disc cupping (cup:disc ratio 0.9) and pallor.

Figure 3 Optical coherence tomography (OCT) scan showing asymmetric nerve fibre layers on the right eye (OD) and left eye (OS) with severe retinal nerve fibre layer thinning on the left side.

assess the level of optic nerve damage and document possible progression.

Medical management with aqueous suppressants is the first-line therapy. A single publication by Breazzano *et al*¹⁰ reports a case of spontaneous resolution of presumed IEEVP that persisted after discontinuation of medical therapy. However, these patients appear to respond less favourably to antiglaucoma drops compared with patients suffering from primary open-angle glaucoma, and a great majority requires filtering surgery.^{3 11} Trabeculectomy is the preferred choice, but intraoperative and postoperative choroidal effusions seem to be

frequent even at IOPs not usually considered hypotonic.^{8 12} A slow decompression is recommended which might require prophylactic sclerostomies, anterior chamber maintainers and a tight trabeculectomy. Knowing that trabeculectomy is potentially more hazardous when the EVP is raised, a primary tube implant such as the Ahmed valve can present as a surgical option to prevent complications associated with decompression. In our case, there was good IOP response to medical therapy. Despite this, due to the young age of the patient, if automated perimetry and optic nerve OCT continue to show likely progression, early surgical treatment will be proposed.

Figure 4 Automated perimetry—first examination. Left eye examination (A) with global sensitivity loss, nasal step and arcuate scotoma. Right eye examination (B) within normal range with no significant field defects.

In conclusion, IEEVP is an infrequent syndrome that courses with engorgement and tortuosity of episcleral vessels, increased IOP and glaucomatous optic nerve damage. Medical treatment is usually insufficient with most patients needing filtering surgery. This case highlights the necessity of having a big clinical suspicion when patients present with long-standing eye injection. Before establishing a final diagnosis, other conditions that present with dilated episcleral vessels must be excluded, and these investigations may prove both sight-saving and life-saving.

Learning points

- Long-standing episcleral venous dilatation can be caused by intracranial or intraorbital pathologies such as carotid-cavernous fistulas, venous sinus thrombosis, Sturge-Weber syndrome, thyroid ophthalmopathy or space-occupying lesions.
- Idiopathic elevated episcleral venous pressure (IEEVP) is rare and courses with dilated episcleral vessels, intraocular hypertension and glaucomatous optic nerve damage.
- IEEVP is an exclusion diagnosis.
- IEEVP usually responds poorly to medical therapy, frequently requiring filtering surgery to control intraocular pressure.

Contributors SHdMM was responsible for writing the manuscript. CF acquired patient data and designed the work. AM collected and organised all patient iconography. PF critically revised patient data and manuscript content.

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