

# Congenital isolated idiopathic episcleral arteriovenous malformation

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## ABSTRACT

**Purpose:** To report the occurrence of an episcleral AVM and to discuss the gravity as well as management of dilated tortuous episcleral vasculature.

**Observation:** A 16-year-old Asian male presented with redness in the left eye since birth and a history of gradual painless progressive increase in size since the past two years, causing a cosmetic concern for the patient. There was no history of any diminution of vision, diplopia, headache, trauma or any similar lesions anywhere in the body.

On examination, unaided visual acuity (UCVA) was 6/6 Snellen in both eyes. Hirschberg's reflex was central and extraocular movements were full and free in both the eyes.

On slit-lamp examination, dilated tortuous episcleral vessels were found in the temporal aspect of the palpebral aperture, shaped like a single continuous vascular frond, vertically measuring 10–11mm in extent and horizontally commencing 6mm from the temporal limbus but the posterior limit could not be discernible. The episcleral plane was freely mobile from the overlying conjunctiva and underlying sclera. The dilated episcleral vasculature remained unchanged on Valsalva maneuver and on bending forward.

Intraocular pressure was 14 mmHg by Goldmann's appplanation tonometer in both eyes. Iris was normal in configuration and gonioscopy revealed open angles without any evidence of any neovascularisation in both eyes. Fundus examination was unremarkable.

USG B-scan was grossly normal. MR Angiography of head and orbit with using thin high-resolution cuts (1mm using 3T machine) was done and was later followed up with a Contrast Enhanced MRI. Both imaging studies revealed an isolated episcleral AVM with a possible feeder vessel from the anterior ciliary artery to the lateral rectus and without any intraorbital or intracranial lesion.

A diagnosis of isolated episcleral AVM was made. The episcleral plane was dissected and dilated tortuous vessels were excised. After cautery, conjunctival autograft was obtained from the fellow-eye was used to cover the defect. Histopathological examination revealed both thick and thin walled vessels of varying calibre corresponding to arteries and veins confirming the diagnosis of isolated episcleral AVM. Ref. Fig. 2. No recurrence has been observed over a follow-up period of 1 year.

**Conclusion:** AND IMPORTANCE: Isolated idiopathic episcleral AVM should be a diagnosis of exclusion. A thorough systemic and ophthalmological evaluation including a glaucoma workup is necessary for both diagnosis and management. UBM and MRI/MRA may be required in select cases to rule out any associations or complications. Management includes observation, embolization and excision.

## 1. Introduction

Orbital arteriovenous malformations (AVM) are variants of congenital hamartomas and symptoms are generally precipitated by incidental

trauma.<sup>1</sup> Anterior segment AVMs occur very infrequently despite the orbit being a common site of AVMs.<sup>1</sup> We report a case of congenital idiopathic isolated episcleral AVM mimicking orbital varices.

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## 2. Case report

A 16-year-old Asian male presented with redness in the left eye since birth. There was history of gradual painless progressive increase in size since the past two years, causing a cosmetic concern for the patient. There was no history of any diminution of vision, diplopia, headache, trauma or any similar lesions anywhere in the body. Patient did not have any symptoms suggestive of any thyroid disorders or any past hospital admissions.

On examination, unaided visual acuity (UCVA) was 6/6 Snellen in both eyes. On pen-torch examination, Hirschberg's reflex was central and extraocular movements were full and free in both the eyes. Pupils were equal in size and bilaterally reactive to light.

On gross examination, Nafziger's test (checking asymmetry in antero-posterior position of the eyes by viewing from above the head after aligning the superior orbital rims with bilateral frontal bossing) was negative. On slit-lamp examination, dilated tortuous episcleral vessels were found in the temporal aspect of the palpebral aperture, shaped like a single continuous vascular frond, vertically measuring 10–11mm in extent and horizontally commencing 6mm from the temporal limbus but the posterior limit could not be discernible. Fig. 1. The surrounding conjunctiva and episclera appeared unremarkable. The overlying conjunctiva could be easily moved over the dilated episcleral vasculature and the episcleral plane containing dilated vasculature could be easily moved over the sclera. The vessels could not be blanched by a 5% solution of phenylephrine. There were no aggravating or relieving factors and the dilated episcleral vasculature remained unchanged on Valsalava maneuver and on bending forward. The orbital margins were freely and fully palpable while Hertels's exophthalmometer read 21mm bilaterally.

Intraocular pressure was 14 mmHg by Goldmann's appplanation tonometer in both eyes. Iris was normal in configuration and gonioscopy revealed open angles without any evidence of any neovascularisation in both eyes. Fundus examination was unremarkable.

Ultrasound B-scan was grossly normal. Ultrasound biomicroscopy revealed normal iris-ciliarybody configuration and ciliary processes appeared normal. Magnetic Resonance Angiography (MRA) of head and orbit with using thin high-resolution cuts (1mm using 3T machine) was done and was later followed up with a Contrast Enhanced Magnetic Resonance Imaging (MRI). Both imaging studies revealed an isolated episcleral AVM with a possible feeder vessel from the anterior ciliary artery to the lateral rectus and without any intraorbital or intracranial lesion.

A diagnosis of isolated episcleral AVM was made. The episcleral plane was dissected and dilated tortuous vessels were excised. The posterior limit could easily been seen once the globe was adducted using



Fig. 1. Slit lamp image of the isolated episcleral AVM under diffuse illumination.

a Lim's forceps. After cautery, conjunctival autograft was obtained from the fellow-eye was used to cover the large defect for better cosmesis. Histopathological examination revealed both thick and thin walled vessels of varying calibre corresponding to arteries and veins confirming the diagnosis of isolated episcleral AVM. Fig. 2. No recurrence has been observed over a follow-up period of 1 year.

## 3. Discussion

Isolated orbital arteriovenous malformations (AVM) are not common and hence have been described in literature in various locations. Depending on their size, location, fixity to structures, arterial component and inflow-outflow volume, they may result in conjunctival and/or episcleral congestion, chemosis, proptosis and limitation of extra ocular movements.<sup>1</sup> Anterior segment AVMs are, however, not so common and have been reported in the ciliary body and iris.

There are 3 reasons why we intended to highlight a congenital isolated episcleral AVM is this patient:

1. The gravity of dilated episcleral vasculature
2. The necessity and type of imaging required
3. Management

### 3.1. Gravity of dilated episcleral vasculature

Episcleral congestion could indicate simpler etiologies like viral conjunctivitis to grave etiologies like ciliary body melanomas. The differential diagnoses can be classified on the basis of etiology and on the basis of the vascular structure involved (predominantly venous or arteriovenous). Table 1.

However innocuous as it may seem, dilated episcleral vasculature may actually be a harbinger of any serious intraocular retrolental pathologies like chronic intermediate uveitis and ciliary body melanoma, both of which may not be apparent on a slit lamp and may demand a thorough dilated indirect ophthalmoscopy. Lee et al. reported a patient with dilated episcleral vessels that was diagnosed as a ciliary body AVM.<sup>2</sup> Mozaffarpour M. described a patient with dilated episcleral congestion that was reported to have intracranial AVM.<sup>3</sup> Gregory et al. reported a case where in an occipital AVM presented with unilateral signs mimicking a carotico-cavernous fistula (CCF).<sup>4</sup> Isolated dilated episcleral vessels need to be differentiated from orbital varices which show unilateral intermittent non-pulsatile proptosis with manoeuvres that increase venous pressure. In essence, it is imperative to undertake a thorough ophthalmic and systemic examination in every patient with dilated episcleral vasculature.

Secondly, it would be prudent to keep in mind the cause-effect relationship of episcleral venous congestion. Literature has numerous

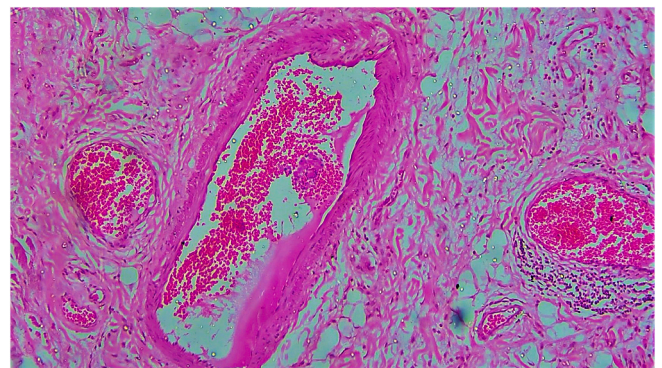


Fig. 2. Histopathological examination H&E section depicting both arteries and veins in excised specimen.

**Table 1**  
Etiological and morphological classification of dilated episcleral vasculature.

ETIOLOGY	VASCULAR STRUCTURE INVOLVED	
	PREDOMINANTLY VENOUS	ARTERIO-VEINUS
INFECTIVE	Conjunctivitis, Cavernous sinus thrombosis	
INFLAMMATORY	Episcleritis, Scleritis, Anterior uveitis, Intermediate uveitis	
ENDOCRINE	Thyroid Eye Disease	
TRAUMA	Dural fistula	Carotico-Cavernous fistula, Episcleral AVM,
NEOPLASIA	Superior venacava syndrome secondary to retrobulbar tumors	Ciliary body melanoma, Iris melanoma
CONGENITAL	Sturge-Weber Syndrome	
IDIOPATHIC	Orbital Varices	Episcleral AVM, Radius-Maumenee syndrome

reports of bilateral idiopathic dilated episcleral venous congestion leading to bilateral glaucoma<sup>5,6,7</sup> as well as unilateral idiopathic dilated episcleral venous congestion leading to ipsilateral glaucoma.<sup>8,9</sup> Groh MJ et al.<sup>10</sup> even reported a case of the Radius-Maumenee syndrome. Recently, Greslechner R. et al.<sup>11</sup> reviewed the pathophysiology, clinical findings and differential diagnosis of secondary open-angle glaucoma due to elevated episcleral venous pressure. The present AAO guidelines<sup>12</sup> also dictate that all efforts must be made to decrease the intraocular pressure (IOP) in such patients and apraclonidine<sup>13</sup> is the drug of choice in such cases. However we would also like to point out that the dilated episcleral vessels in all the above patients were secondary to venous congestion and hence present diffusely 360° circumcorneal whereas the dilated episcleral vessels in our patient were well defined and clearly isolated sectorally in the temporal aspect. Nevertheless a thorough glaucoma workup is a must in all cases of dilated episcleral vasculature.

### 3.2. Necessity and type of imaging required

Baseline ophthalmic examination including visual acuity, pupillary reaction, dilated slit lamp examination under high magnification, peripheral fundal examination, exophthalmometry and extraocular movements would be sufficient to give an indication to the etiology of the dilated episcleral vasculature.

Imaging is indicated when the ophthalmologist is suspecting an intraocular or extraocular AVM, an intraocular neoplasia or an intracranial pathology.<sup>14</sup> An MRI or MR Angiography may be able to delineate the size, location and extent of the AVMs and CCF as well as intracranial lesions. Intraocular pathologies like ciliary body AVM or ciliary body melanomas may require the use of UBM to demarcate the retro-irideal structures. While investigating the patient, we had ordered an MRI Head-Orbit with two objectives in mind:

- i. To decipher and delineate a feeder vessel if any
- ii. To rule out any concurrent intraocular, retrobulbar, intracranial AVM

When we expressed these motives to the Dept. of Radiology, they conducted a plain MRI first and ruled out any concurrent vascular pathology. Since an MR Angiography gives better definition of the arterial tree, the MRI was followed up with an MRA<sup>15</sup> but still the feeder vessel could not be discernible. Finally when a contrast enhanced MRI was done, the episcleral AVM was hyperintense in the same time of flight (TOF) as the ophthalmic artery and this gave an indication that the possible feeder vessel could be a branch of the anterior ciliary artery supplying the lateral rectus but was of very small calibre and hence would not pose a problem for achieving hemostasis during surgery.

### 3.3. Management

Depending on the size, location and extent of the extraocular AVM, treatment includes observation, embolization with or without sclerotherapy and surgical excision. Management obviously would involve a multidisciplinary approach in cases of intracranial AVM and/or CCFs.

Akritisidou reported a case of limbal episcleral AVM and had maintained the patient on follow-up because the lesion was benign stationary without any apparent associations and complications.<sup>16</sup> Since the primary concern of our patient was cosmetic, we excised the lesion and placed a conjunctival autograft after applying cautery to the base.

### 4. Conclusion

Isolated idiopathic episcleral AVM should be a diagnosis of exclusion. A thorough systemic and ophthalmological evaluation including a glaucoma workup is necessary for both diagnosis and management. UBM and MRI/MRA may be required in select cases to rule out any associations or complications. Management includes observation, embolization and excision.

### Contributorship

All the authors were involved in the concept and design of the study, data acquisition, data analysis and interpretation, drafting manuscript, technical support and final review of the manuscript.

### Patient consent

Consent to publish this case report has been obtained from the patient in writing.

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### Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

### Declaration of competing interest

None of the authors have any conflicts of interest.

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