Spontaneous hyphema from iris microhemangioma in Eisenmenger syndrome

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ABSTRACT

Purpose: We describe a patient with Eisenmenger syndrome and spontaneous hyphema from iris microhemangioma, two rare entities with a plausible pathophysiological connection.

Observations: A 56-year-old Caucasian female with a background of cyanotic congenital heart disease complicated by Eisenmenger syndrome presented with non-traumatic hyphema and blurred vision. Multiple vascular tufts consistent with iris microhemangiomas were observed around the pupil margins bilaterally, with no iris or retinal neovascularization. In the affected eye, there was active bleeding from one lesion at 12 o’clock generating a macrohyphema. Additional findings included prominent episcleral injection and retinal venous tortuosity in both eyes. The active microhemorrhage and hyphema resolved with local medical management.

Conclusions and importance: Chronic hypoxemia and erythrocytosis are known to induce dilation of the retinal and episcleral blood vessels in Eisenmenger syndrome. Corresponding dilation of iris stromal vessels may contribute to the formation and prominence of iris microhemangiomas.

1. Introduction

Iris microhemangiomas (IMH), also known as Cobb’s tufts or iris vascular tufts (IVT), consist of tightly coiled blood vessels at the iris pupil margin. Histologically, they have been demonstrated to be a true hamartoma composed of thin-walled iris stromal blood vessels.\textsuperscript{1} Although rare, IMH are important clinically because they are an uncommon cause of spontaneous hyphema and can cause transient blurred vision and elevated intraocular pressure. Their etiology is unclear but they have never been reported in children, suggesting they are acquired, and usually found bilaterally, suggesting a systemic abnormality or bilateral ocular condition underlies their presence.\textsuperscript{2}

Eisenmenger syndrome is characterized by congenital cardiac defects causing left-to-right shunting of blood with subsequent pulmonary hypertension leading to reversal of the shunt into a cyanotic right-to-left shunt. Retinal vascular dilation and tortuosity in Eisenmenger syndrome has been well described and correlates with the degree of hypoxemia and erythrocytosis.\textsuperscript{3} These patients also have increased branching of retinal vessels leading to an overall complex vascular bed. This report describes a patient with Eisenmenger syndrome and prominent iris microhemangioma, highlighting two rare entities with a plausible pathophysiological connection.

2. Case report

A 56-year-old Caucasian female presented to the Emergency Department with a two-day history of atrumatic blurred vision in the right eye with visible macrohyphema. Previous medical history was significant for cyanotic congenital heart disease (dextro-Transposition of the Great Arteries with unrestricted ventricular septal defect and patent ductus arteriosus) complicated by Eisenmenger syndrome and secondary erythrocytosis. Other medical history included atrial flutter treated with ablation, iron-deficiency and gout. She was taking oral Macitentan for pulmonary hypertension but no blood-thinning agents. The patient reported several prior minor episodes of spontaneous bleeds which affected each eye on separate occasions but would self-resolve without seeking treatment.

Distance corrected visual acuity on presentation was 20/20 and 20/15 in the right and left eyes, respectively. Intraocular pressure was slightly elevated in the right eye (23 mmHg), and within normal limits in the left eye (16 mmHg). There was marked right ciliary flush. Multiple vascular tufts consistent with iris microhemangiomas were observed...
around the pupil margins bilaterally. There was no visible iris or retinal neovascularization. In the right eye, there was active bleeding from one lesion at 12 o’clock with a macrohyphema <1mm height (Fig. 1). Blood pressure was normal (118/72 mmHg). Topical atropine twice daily and dexamethasone four times daily was commenced and at review two days later the active bleeding had ceased and macrohyphema resolved (Fig. 2). The visual acuity was unchanged while intraocular pressure had reduced slightly to 20 mmHg in the right eye and 15 mmHg in the left eye. Subsequent review 2 weeks later showed return to equal visual acuity of 20/15 in each eye, normal intraocular pressure of 15 mmHg in both eyes and clear anterior chambers. Mild episcleral injection and diffuse dilation and tortuosity of the retinal veins remained in both eyes along with quiescent iris microhemangiomas at the pupil margin.

3. Discussion

Iris microhemangiomas have previously been associated with diabetes mellitus and myotonic dystrophy, as originally described by Cobb et al. However, a recent systematic review notes a range of reported comorbidities, particularly cardiorespiratory disease. There has been one previous reported case of bilateral IMH and recurrent microhyphema in a patient with a background of congenital cyanotic heart disease in 1977. This case hypothesized that IMH was a consequence of iris neovascularization, however subsequent histopathology including...
electron microscopy demonstrates that IMH represents a separate entity.\(^1\)

Our patient has typical features of Eisenmenger syndrome with chronic hypoxemia (resting oxygen saturation 78%) leading to secondary erythrocytosis (hemoglobin 22.5g/dL). Comparable to the dilated episcleral veins and retinal venous tortuosity that have previously been described in Eisenmenger syndrome,\(^3\) we hypothesize that chronic hypoxemia and erythrocytosis induces dilation of the iris stromal vessels contributing to the formation and prominent appearance of IMH in this patient. This is supported by the finding of IMH in association with other cardiorespiratory conditions.\(^5\) The active hemorrhaging and hyphema in this case resolved with local medical management, as is usual, although argon laser photocoagulation has previously been used to successfully treat refractory cases.\(^7\)

This report identifies a rare case of spontaneous hyphema from an iris microhemangioma in a patient with Eisenmenger Syndrome which responded well to local medical management. Identifying this association may improve our understanding of the etiology of this rare condition, as well as highlight a potential long-term ocular manifestation of congenital heart disease.

**Patient consent**

The patient consented in writing to the publication of this case including the associated images.

**Authorship**

All listed authors meet the ICMJE criteria for Authorship.

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