Long-Term Follow-Up of Recurrent Spontaneous Hyphema Caused by Ruptured Persistent Fetal Vasculature

DOI 10.1055/a-1739-0462
Klin Monatsbl Augenheilkd 2022; 239: 556–558

ISSN 0023-2165

Thieme

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Langzeitbeobachtung eines rezidivierenden Hyphämas hervorgerufen durch persistierende fötale Gefäße

Introduction

Persistent fetal vasculature (PFV) is a failure of regression of fetal intraocular vasculature during intrauterine life. Its clinical manifestation is variable among patients, depending on the subtype of PFV – anterior, posterior, or combined PFV [1, 2].

In particular, persistent pupillary membranes are part of anterior PFV. These vessels are remnants of the anterior tunica vasculosa lentis and are variably perfused from the minor arterial circle of the iris at the level of the collarette. Rarely, persistent pupillary membranes can cause recurrent hyphema in adults [3 – 8].

We report the medical management and the long-term follow-up of an adult woman with recurrent spontaneous hyphema caused by a ruptured remnant of a persistent pupillary membrane.

History and Signs

A 28-year-old female patient was referred to the vitreoretinal surgery department of the Jules Gonin Eye Hospital for recurrent spontaneous hyphema in her left eye. The patient's general past medical history was unremarkable; in particular, there was no coagulation disorder, no high blood pressure, no systemic medication, and no previous ocular blunt trauma.

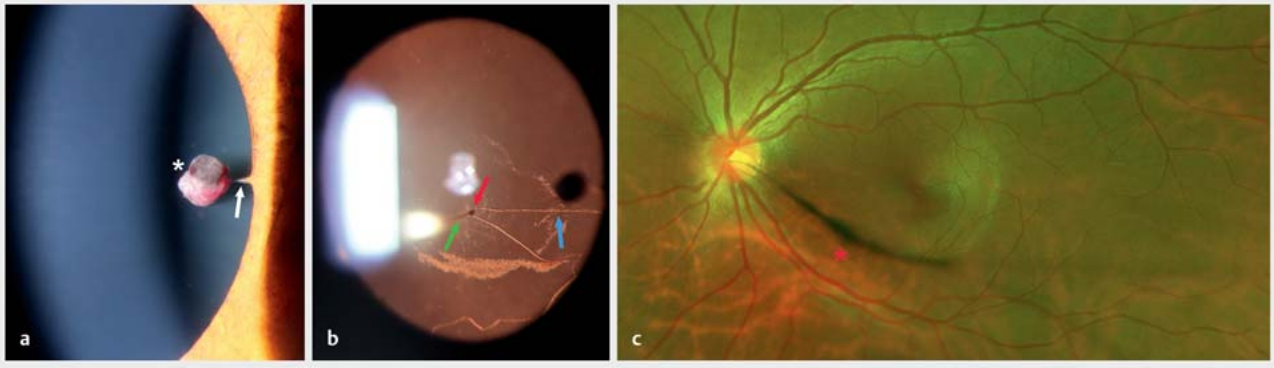
The patient reported five episodes of discomfort and blurred vision in her left eye during the last 2 months, caused by recurrent hyphema, which had prompted an ophthalmic examination [9]. The first episode occurred getting out of the shower, without any eye injury.

At presentation, the entire ophthalmic examination of the right eye was normal. The left eye had a reduced best-corrected visual acuity of 20/300. Intraocular pressure was 36 mmHg. Biomicroscopy revealed a 1-mm hyphema and a blood clot enmeshed in a vascular malformation temporally on the anterior lens capsule, connected by one small vessel to the iris margin at 3 o'clock. On the posterior lens capsule, a Mittendorf dot connected posteriorly to a persistent hyaloid artery and anteriorly to the remnant of the posterior tunica vasculosa lentis. Fundus examination was normal, except for the persistent hyaloid artery inserting at the level of the optic disc (▶ **Fig. 1**).

Anterior segment fluorescein angiography of the left eye showed an intact posterior and anterior vascular perilental network, with a patent residual vessel connecting to the vascular loop on the lens surface. Posterior segment fluorescein angiography did not show patent posterior fetal vasculature. Anterior segment OCT showed the blood clot connected to the perfused vascular loop (▶ **Fig. 2**). Ultrasound biomicroscopy revealed no large vascular mass posterior to the iris.

Therapy and Outcome

Topical treatment with corticosteroid (dexamethasone), cycloplegic (scopolamine), antihypertensive drops (timolol, dorzolamide, brimonidine), and oral acetazolamide permitted resorption of the hyphema, normalization of the intraocular pressure (11 mmHg), and total recovery of



▶ **Fig. 1** **a** Color photograph of the left eye anterior chamber showing the blood clot enmeshed in a vascular malformation (white asterisk), connected by one small vessel to the iris margin at 3 o'clock (white arrow) corresponding to the persistent pupillary membrane. **b** Color photograph in retro illumination of the left eye, focused on the posterior lens capsule, showing the Mittendorf dot (red arrow) connected posteriorly to the persistent hyaloid artery (green arrow) and anteriorly to the remnant of the posterior tunica vasculosa lentis (blue arrow). **c** Fundus photograph of the left eye showing the persistent hyaloid artery (red asterisk).

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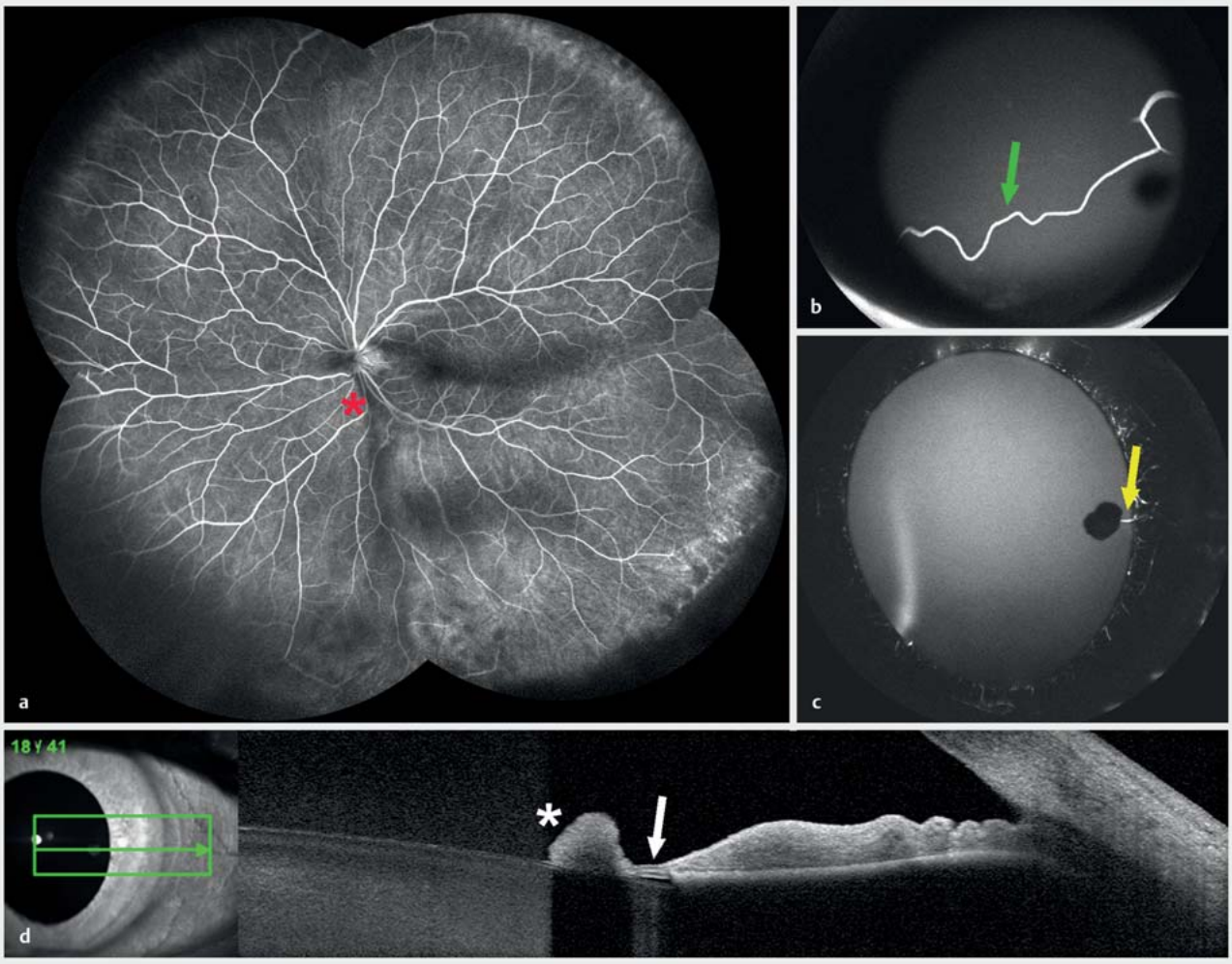

▶ **Fig. 2** **a** Fundus fluorescein angiography of the left eye, showing normal retinal vasculature, and a non-patent posterior hyaloid artery (red asterisk). **b, c** Anterior segment angiography of the left eye, showing a patent persistent posterior tunica vasculosa lentis (green arrow), and patent persistent pupillary membrane (yellow arrow). **d** Anterior segment OCT of the left eye, showing the blood clot (white asterisk) connected by the bridging persistent pupillary membrane (white arrow).

the best-corrected visual acuity (20/16) after a 5-month follow-up.

Image comparison over several months of the vascular system between the loop on the anterior crystalline surface and the iris margin showed that originally two vessels were present [9], one of which ruptured and retracted, leaving one single vascular connection (▶ **Fig. 1 a**).

The potential indication for a surgical procedure with coagulation of the remaining vessel in the anterior chamber was discarded due to the potential adverse side effects, the unnecessary perturbation of the residual ramified fetal vascular network around the crystalline lens, the excellent visual acuity, and the young age of the patient.

Discussion

PFV is typically sporadic, mostly unilateral (10% are bilateral), and non-heritable [2]. The clinical spectrum of PFV depends on the location of the vasculature remnants. Anterior PFV encompasses a persistent pupillary membrane, anterior tunica vasculosa lentis, irido-hyaloid blood vessels, and posterior tunica vasculosa lentis. Clinical manifestations of anterior PFV include iris or pupil deformation, shallow anterior chamber, hyphema, cataract, congenital subluxation of the lens, and retrolental membrane. Posterior PFV is represented by the persistence of the vasa hyaloidea propria and/or the hyaloid artery. In case of combined PFV, clinical features of both anterior and posterior PFV are present [1, 2].

Isolated remnants of the persistent pupillary membrane are often seen in eyes as thin white filaments (17.6 to 31.9% of the eyes), but persistent blood-containing vessels are found only in 0.3% of the eyes [3].

We describe a case of combined PFV with persistent pupillary membrane, posterior vasculosa lentis, and non-perfused hyaloid artery. The patient presented as an adult with blood-filled vessels of a persistent pupillary membrane causing recurrent hyphema. Only six cases of bleeding from a strand of the persistent pupillary membrane in adults have been reported in the literature [3–8].

Investigations for PFV include fluorescein angiography and B-scan ultrasonography. Fluorescein angiography is used to determine the boundaries of the patient's vessels, which is crucial to confirm PFV, but also to exclude other differential diagnoses of neovascularization [1, 2]. B-scan ultrasonography is indicated to investigate the extent of PFV, as a retrolental membrane can form in the presence of persistence irido-hyaloid blood vessels. Furthermore, ultrasonography is useful to exclude an intraocular mass, which can cause abnormal iris neovascularization [1, 2].

In our case, fluorescein angiography confirmed the persistent pupillary membrane and posterior tunica vasculosa lentis, and ultrasonography excluded a large vascular mass posterior to the iris.

In the majority of previously reported cases, bleeding of the anterior PFV was related to other predisposing factors such as aneurysmal dilatation on the vessel [4, 5], minor trauma [6], high blood pressure [3, 5], or strenuous effort [7]. Our patient did not present these factors, but image comparison over several months showed that one vessel [9] retracted, leaving only one single vascular connection on the anterior crystalline surface at the iris margin after resolution of the hyphema. Therefore, we hypothesize that bleeding was secondary to the rupture of one of the strands, which spontaneously clotted, stopping the recurrent bleeding.

Previously reported therapy for hyphema secondary to a persistent pupillary membrane includes topical drugs (antihypertensive drops, corticosteroid, cycloplegics) [4, 6, 8] and argon laser photocoagulation of the vessel [3]. In our patient, conservative medical management with topical drops and systemic acetazolamide was successful in normalizing intraocular pressure and visual acuity. Further intervention with coagulation of the remaining vessel was not indicated in our opinion, due to the potential risk of cataract formation and/or rebleeding.

In conclusion, recurrent spontaneous hyphema can rarely appear secondary to the rupture of a residual anterior persistent fetal vasculature. We recommend conservative management initially, as the bleeding appears to be self-limiting following the contraction of the ruptured vessel and resorption of the hyphema.

Conflict of Interest

The authors declare that they have no conflict of interest.

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DOI 10.1055/a-1739-0462
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Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany