Eye (2005) 19, 1336–1352

www.nature.com/eye

CORRESPONDENCE

Sir,

Spontaneous hyphaema from pupillary vascular tufts in a patient with branch retinal vein occlusion

Pupillary vascular tufts are clusters of convoluted fine vascular loops that are rarely seen at the pupillary margin and can occasionally cause spontaneous hyphaema[1–3] and rise of intraocular pressure.[4,5] They typically occur bilaterally in otherwise healthy individuals above the age of 50 years[1,2,6,7] as well as in patients with acquired systemic diseases such as hypertension,[2] diabetes,[6,8,9] and myotonic dystrophy.[8,10] They have also been reported in some congenital conditions such as hereditary haemorrhagic telangiectasia,[11] orbital cavernous haemangioma[3] and with remnants of pupillary membrane.[12] Few reports described vascular tufts as a coincidental finding in patients with retinal venous occlusions.[6,9]

We describe a case of pupillary vascular tufts causing spontaneous hyphaema in the context of acute branch retinal vein occlusion. We discuss its clinical implications and propose an explanation that links the occurrence of the hyphaema from the tufts to the venous occlusion.

Case report

A 66-year-old Caucasian female was referred to the ophthalmic casualty with a 1-day history of blurred vision in the left eye. She had no history of ocular abnormality or trauma but gave a medical history of a well-controlled hypertension and an obsessive-compulsive disorder.

External ocular examination was normal and visual acuity was 6/6 and 6/9 in the right and left eye, respectively. On slit-lamp examination of the anterior segment, there was a blood clot of approximately 2-mm size attached to a vascular tuft at the pupillary margin of the left eye at 12 o'clock position (Figure 1a). The pupils were normally reactive, however, and the rest of the iris stroma was normal in both eyes. In the anterior chamber of the left eye, there was a faint flare and red blood cells but the intraocular pressure was normal in both eyes. Gonioscopy revealed a trace of blood in the lower part of the left angle but there was no neovascularisation.

Dilated fundus examination of the left eye showed signs of a recent superotemporal branch retinal vein occlusion: superficial flame-shaped and deep retinal haemorrhages as well as cotton–wool spots in the area tributary to the occluded vein.

After 2 days, the blood clot was absorbed and detailed examination of the irides showed vascular tufts protruding from the pupillary margins of both eyes (Figure 1b) but were conspicuously larger and more prominent on the upper half of the left pupil (Figure 1c).

After 4 weeks, fluorescein angiography of the left eye exhibited areas of capillary dropout and venous wall staining indicative of retinal ischaemia. Blood investigations revealed activated protein C resistance and mildly elevated cholesterol (6.4 mmol/l) but fasting blood sugar and glucose tolerance tests were normal. The patient was prescribed aspirin 75 mg a day and was referred to a dietitian for control of her cholesterol level.

After 6 months, visual acuity improved to 6/6 in the left eye but was still slightly blurred in comparison to the right eye. A repeat fluorescein angiography revealed no retinal neovascularisation (Figure 1d), and late leakage from the vascular tufts at the pupillary margin (Figure 1e).

At 9 months, spontaneous vitreous haemorrhage occurred from retinal neovascularisation and scatter laser photocoagulation to the ischaemic retina was performed. No recurrent bleeding occurred over the period of her follow-up, which lasted 18 months to date but the tufts that had bled in the left eye exhibited greyish discolourations and reduction in size (Figure 1f). No similar changes in size or colour of any of the other tufts elsewhere were seen.

Comment

Although pupillary vascular tufts have been reported before in patients with retinal venous occlusion,[6,9] this is, to our knowledge, the first case report of spontaneous hyphaema from pupillary vascular tufts that coincided with the onset of an ischaemic branch retinal vein occlusion.

Our patient presented with symptoms and signs of a BRVO and had signs of a fresh hyphaema on the corresponding side, which strongly suggested that both happened either simultaneously or in close succession. Although the presence of the tufts bilaterally excludes a causal relationship to the development of the vein occlusion, the greater number and larger tufts on the same side and even on the corresponding part of the iris to the area of the vein occlusion indicate a likely influence to the vein occlusion on the tufts that made them bleed at the time of its happening. Clinical and experimental studies have shown that dilatation, increased circulation time and increased permeability of the iridal vasculature occur early after the development of retinal venous occlusions.[13–15] Driven mainly by the retinal ischaemia, these haemodynamic changes could have led in our patient to increased intravascular pressure of the tufts and consequently to the development of hyphaema. As such, we postulate that these dilated tufts can perhaps be considered as the iridal counterparts of intraretinal microvascular abnormalities

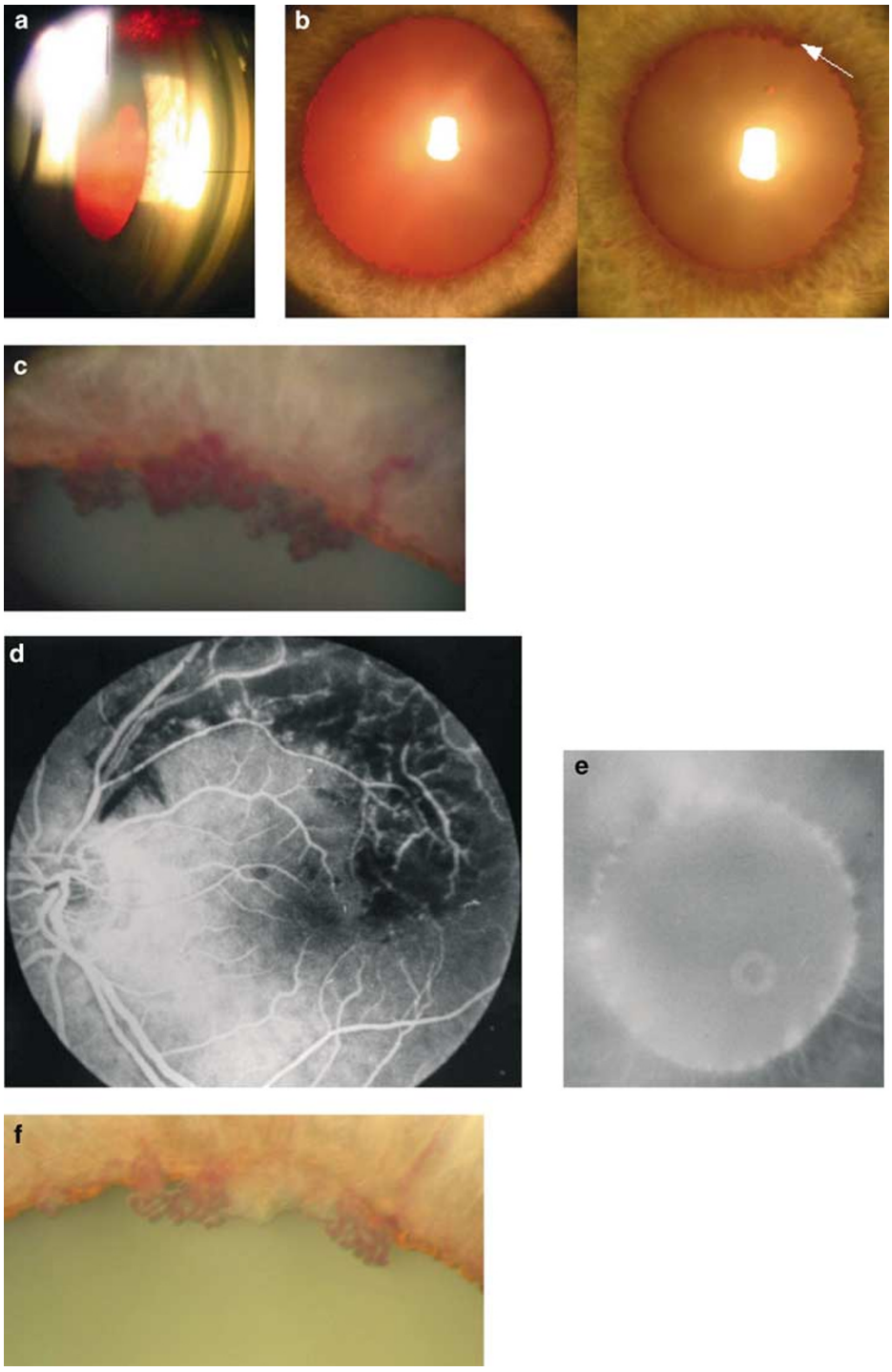

Figure 1 (a) A blood clot at the pupillary margin at presentation. (b) Colour photographs of the pupils showing bilateral pupillary vascular tufts that are more prominent on the upper half of the pupillary margin of the left eye (arrow). (c) A magnified picture of the vascular tufts that had bled, arrow in (b). (d) and (e) Fluorescein angiography of the fundus and pupil of the left eye. It shows capillary dropout and venous wall staining of the blocked retinal vein; as well as leakage from the pupillary margin. (f) A colour photograph of the pupillary tufts in (c) showing reduction in size and greyish discolouration.

or disc collateral vessels in that they are preformed capillaries and are affected by the ischaemic changes in the retinal vasculature.

Laser photocoagulation of the tufts was suggested prior to intraocular surgery[16] or in cases of recurrent hyphaema.[1,17] In our case, greyish discolouration and

reduction in size of the tufts that had bled, were observed 6 months after retinal laser photocoagulation in the absence of similar changes in the other tufts. These were most probably signs of involution induced either by the retinal photocoagulation or by the bleeding, which have not been reported before.

We conclude that spontaneous hyphaema from pupillary vascular tufts may indicate a recent onset of a retinal venous occlusion and that their presence can be a risk factor for the development of hyphaema during the acute stage of an ischaemic retinal venous occlusion. Also, we believe that, as much as the ischaemia could lead to iridal vascular tuft engorgement, bleeding from the tufts or retinal laser photocoagulation of the ischaemic retina might initiate their involution.

Acknowledgements

The authors would like to thank Mr Alan Finch, the Medical photographer at Essex County Hospital, for the fluorescein angiography photographs.

References

1 Coleman SL, Green WR, Patz A. Vascular tufts of pupillary margin of iris. *Am J Ophthalmol* 1977; **83**: 881–883.
2 Podolsky MM, Srinivasan BD. Spontaneous hyphema secondary to vascular tuft of pupillary margin of the iris. *Arch Ophthalmol* 1979; **97**: 301–302.
3 Savir H, Manor RS. Spontaneous hyphema and vessel anomaly. *Arch Ophthalmol* 1975; **93**: 1056–1058.
4 Blanksma LJ, Hooijmans JM. Vascular tufts of the pupillary border causing a spontaneous hyphaema. *Ophthalmologica* 1979; **178**: 297–302.
5 Akram I, Reck AC, Sheldrick J. Iris microhaemangioma presenting with total hyphaema and elevated intraocular pressure. *Eye* 2003; **17**: 784–785.
6 Cobb B. Vascular tufts at the pupillary margin: a preliminary report on 44 patients. *Trans Ophthalmol Soc UK* 1969; **88**: 211–221.
7 Dahlmann AH, Benson MT. Spontaneous hyphema secondary to iris vascular tufts. *Arch Ophthalmol* 2001; **119**: 1728.
8 Mason GI. Iris neovascular tufts. Relationship to rubeosis, insulin, and hypotony. *Arch Ophthalmol* 1979; **97**: 2346–2352.
9 Mason G. Iris neovascular tufts. *Ann Ophthalmol* 1980; **12**: 420–422.
10 Cobb B, Shilling JS, Chisholm IH. Vascular tufts at the pupillary margin in myotonic dystrophy. *Am J Ophthalmol* 1970; **69**: 573–582.
11 Cota NR, Peckar CO. Spontaneous hyphaema in hereditary haemorrhagic telangiectasia. *Br J Ophthalmol* 1998; **82**: 1093.
12 Brusini P, Beltrame G. Spontaneous hyphaema from persistent remnant of the pupillary membrane. A case report. *Acta Ophthalmol (Copenh)* 1983; **61**: 1099–1103.
13 Laatikainen L, Blach RK. Behaviour of the iris vasculature in central retinal vein occlusion: a fluorescein angiographic study of the vascular response of the retina and the iris. *Br J Ophthalmol* 1977; **61**: 272–277.
14 Nork TM, Tso MO, Duvall J, Hayreh SS. Cellular mechanisms of iris neovascularization secondary to retinal vein occlusion. *Arch Ophthalmol* 1989; **107**: 581–586.
15 Hayreh SS, Rojas P, Podhajsky P, Montague P, Woolson RF. Ocular neovascularization with retinal vascular occlusion-III. Incidence of ocular neovascularization with retinal vein occlusion. *Ophthalmology* 1983; **90**: 488–506.
16 Winnick M, Margalit E, Schachat AP, Stark WJ. Treatment of vascular tufts at the pupillary margin before cataract surgery. *Br J Ophthalmol* 2003; **87**: 920–921.
17 Bandello F, Brancato R, Lattanzio R, Maestranzi G. Laser treatment of iris vascular tufts. *Ophthalmologica* 1993; **206**: 187–191.

MA Elgohary and JH Sheldrick

Essex County Hospital,
Lexden Road,
Colchester CO3 3NB,
UK

Correspondence: MA Elgohary,
Tel: +44 1206 852166;
Fax: +44 1206 744742.
E-mail: m.elgohary@doctors.org.uk

Conflicting interest: None

Eye (2005) **19,** 1336–1338. doi:10.1038/sj.eye.6701756; published online 12 November 2004

Sir,

Cytomegalovirus in aetiology of Posner–Schlossman syndrome: evidence from quantitative polymerase chain reaction

Posner–Schlossman syndrome (PSS) (glaucomatocyclitic crisis) was characterized in 1948 by recurrent episodes of hypertensive iridocyclitis.[1] The aetiology is unknown but current theories favour an infective origin especially herpes simplex virus (HSV).[2] We present a case in which CMV DNA was identified following an acute relapse of PSS.

Case report

A 35-year-old immunocompetent Chinese man with a history of recurrent bilateral Posner–Schlossman syndrome (PSS) presented with acutely raised intraocular pressure (IOP) in the right eye. Prior to this, IOP was controlled with topical brimonidine to both eyes. He was diagnosed with a relapse of PSS and treated