(Figure 1). Follow-up has been stable since (18 months post-treatment).

Comment

Observation may be warranted for early onset post-traumatic CNV (less than 6 months after trauma) as neovascularization appears to be part of the normal healing process, and may regress.[1] Argon laser treatment has been successful in extra-foveal post-traumatic CNV.[2,3] If subfoveal, visual prognosis is poorer.[4,5] Surgical removal has been reported in subfoveal cases,[1] but proof of one treatment over another or over observation has not yet been shown.

PDT reduces major visual loss secondary to CNV in diseases other than AMD: myopia, presumed ocular histoplasmosis syndrome, angioid streaks and idiopathic causes.[6] It appears more effective in cases where CNV is not related to AMD.[6] To our knowledge, this is the first report to document the use of PDT in treating CNV secondary to choroidal rupture. In this case, good VA prior to CNV, major exsudation with rapid aggravation and young patient age prompted us to offer PDT. In selected cases, we feel that PDT with veteporfin injection may be offered to patients presenting post-traumatic subfoveal CNV.

References

1 Gross JG, King LP, de Juan E, Powers T. Subfoveal neovascular membrane removal in patients with traumatic choroidal rupture. *Ophthalmology* 1996; **103**: 579–585.
2 Hilton GF. Late serosanguineous detachment of the macula after traumatic choroidal rupture. *Am J Ophthalmol* 1975; **79**: 997–1000.
3 Fuller B, Gitter KA. Truamatic choroidal rupture with late serous detachment of macula. *Arch Ophthalmol* 1973; **89**: 354–355.
4 Luxenberg MN. Subretinal neovascularization associated with rupture of the choroid. *Arch Ophthalmol* 1986; **104**: 1233.
5 Wyzeynski RE, Grossniklaus HE, Frank KE. Indirect choroidal rupture secondary to blunt ocular trauma. A review of eight eyes. *Retina* 1988; **8**: 237–243.
6 Sickenberg M, Schmidt-Erfurth U, Miller JW, Pournaras CJ, Zografos L, Piguet B *et al*. A preliminary study of photodynamic therapy using verteporfin for choroidal neovascularization in pathologic myopia, ocular histoplasmosis syndrome, angioid streaks and idiopathic causes. *Arch Ophthalmol* 2000; **118**: 327–336.

J Conrath, O Forzano and B Ridings

Ophthalmology Department
Timone Hospital Marseille, France

Correspondence: J Conrath
Ophthalmology Department
Timone Hospital
264, rue Saint Pierre
F-13385 Marseille, France
Tel.: +33 491385470
Fax: 033 491387079
E-mail: johnconrath@yahoo.fr

The authors have no proprietary interest in any technology or drugs discussed in this article.

Eye (2004) **18,** 946–947. doi:10.1038/sj.eye.6701357
Published online 5 March 2004

Sir,

Ophthalmic presentation of hereditary haemorrhagic telangiectasia

Ocular abnormalities in hereditary haemorrhagic telangiectasia **(**HHT) have been described in the literature, but generally as incidental findings. We report a patient who presented with ocular abnormalities who was subsequently diagnosed as suffering from a mild variant of HHT.

Case report

A 55-year-old lady presented to the ophthalmology unit with a 4-week history of a red, painless left eye. There was no previous history of eye problems and visual acuity was 6/6 in both eyes. On examination of the left eye, there was injection of the medial bulbar conjunctiva. There was no abnormality of her palpebral conjunctiva and the intraocular pressure and fundus examination were normal. A diagnosis of episcleritis was made and she was commenced on a short course of prednisolone 0.5% eye drops. At her review 4 weeks later, there was no improvement in the injection of the left eye. The dilated vessels were mobile over the underlying episclera. It was then noted that there was also some injection of the medial limbal conjunctiva of the right eye resembling a small bulbar conjunctival telangiectasia (Figure 1). The right eye otherwise had no abnormality.

On questioning, the patient admitted frequent nosebleeds throughout her life ceasing spontaneously without medial assistance. She also reported occasional bleeding from her mouth and on examination she had several telangiectasia on her hard palate (Figure 2) and on the buccal mucosa. She had a small telangiectatic lesion on her chin but none visible elsewhere on her skin.

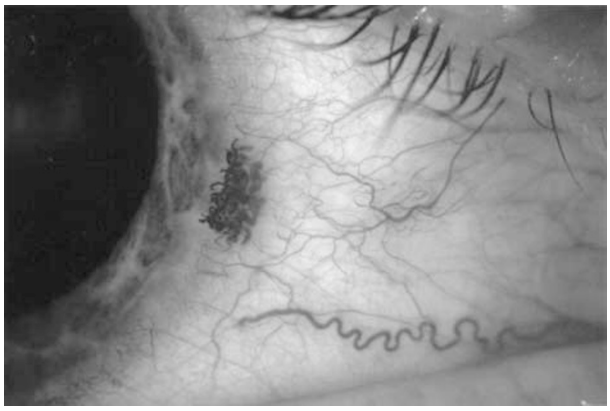

Figure 1 Bulbar conjunctival telangiectasia in the right eye.

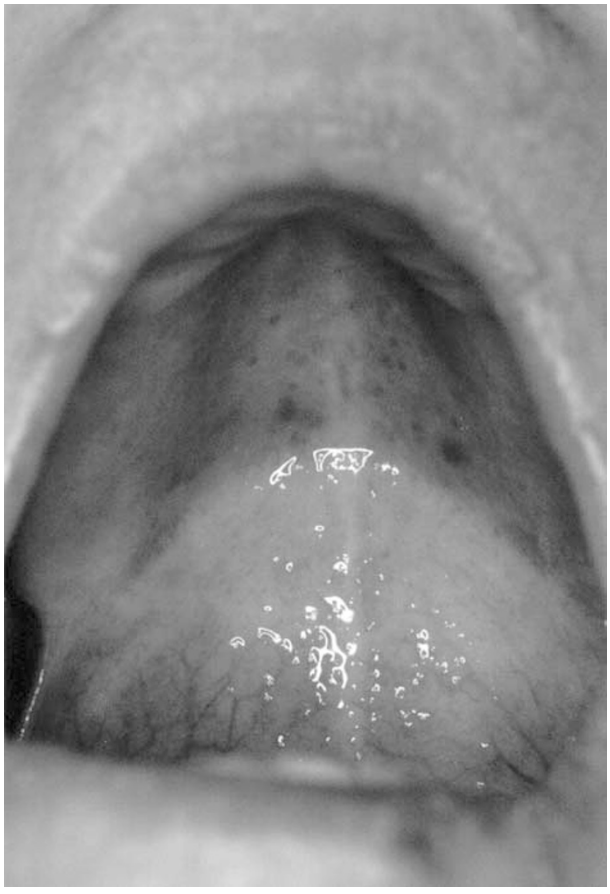

Figure 2 Telangiectasia on patient's hard palate.

She had no rectal bleeding and was otherwise healthy. Autoimmune screening and full blood picture were normal.

She stated that her son also suffers from frequent nosebleeds. He has no known ocular abnormality and is otherwise healthy.

Following these developments, the diagnosis of episcleritis was revised to HHT and the topical steroid treatment stopped. The conjunctival appearance was unchanged after the cessation of treatment.

Comment

HHT is a genetically determined disorder affecting blood vessels throughout the body. The inheritance is autosomal dominant with a high degree of penetrance but variable expression. Approximately 20% of all cases are unaware of a family history. The disease is characterised by dilated thin-walled vascular anomalies of the skin and mucous membranes, and recurrent epistaxis is the usual presenting symptom. In the literature, eye involvement has been documented in 45–65% of patients with HHT, with the most common lesions being conjunctival telangiectasias usually of the palpebral conjunctiva.[1,2] Retinal arteriovenous malformations, retinal telangiectasia and choroidal haemorrhage during intraocular surgery have also been seen rarely.[1–3]

Diagnosis of the disease is based on clinical findings of mucosal and/or skin lesions and bleeding from the lesions. Our patient had typical features of HHT with lesions of the skin and buccal mucosa, bleeding from the mouth and a personal and family history of epistaxis. In the vast majority of sufferers, HHT becomes manifest before the age of 21 years. We propose that our patient has a mild variant of HHT presenting late in life. Interestingly, the literature has reported that low doses of oestrogens in contraceptive pills may aggravate the condition.[4] Our patient had been commenced on hormone replacement therapy (HRT) a few months prior to the appearance of the lesions and the oestrogen administration may have played a part in the manifestation of the disease in the eye.

To our knowledge, there is only one previous report in the literature of HHT presenting initially as an ocular abnormality.[5] In addition, it is very unusual for the lesions to be located on the bulbar conjunctiva and for the patient to develop the disease so late in life.

The case illustrates the value of systemic examination when a patient presents with an ocular problem.

References

1 Brant AM, Schachat AP, White RI. Ocular manifestations of hereditary hemorrhagic telangiectasia (Rendu–Osler–Weber disease). *Am J Opthalmol* 1989; **107**: 642–646.
2 Vase I, Vase P. Ocular lesions in hereditary haemorrhagic telangiectasia. *Acta Ophthalmol* 1979; **57**: 1084.
3 Mahmoud TH, Deramo VA, Kim T, Fekrat S. Intraoperative choroidal hemorrhage in the Osler–Rendu–Weber syndrome. *Am J Opthalmol* 2002; **133**: 282–284.

4 Rowley PT, Kernick J, Cheville R. Hereditary haemorrhagic telangiectasia: aggravation by oral contraceptions? *Lancet* 1970; **1**(7644): 474–475.
5 Soong HK, Pollock DA. Hereditary hemorrhagic telangiectasia diagnosed by the ophthalmologist. *Cornea* 2000; **19**(6): 849–850.

FA Knox and DG Frazer

Royal Victoria Hospital, Belfast, Northern Ireland

Correspondence: FA Knox
Tel: +2890 240503 Ext 3152
Fax: +2890330744
E-mail: angelaknox@ireland.com

Eye (2004) 18, 947–949. doi:10.1038/sj.eye.6701360
Published online 5 March 2004

Sir,

Sight threatening complications in porphyria cutanea tarda

The porphyrias are a group of disorders of haem metabolism with deficiency in the enzymes of the haem biosynthetic pathway resulting in excess porphyrin production.[1] Porphyria cutanea tarda (PCT) is the most common of eight subtypes with a predilection among black Southern Africans.[1] Clinical manifestations are predominantly dermatological with photoactive porphyrins depositing in the skin causing bullae, hyper- and hypo-pigmentation, pseudoscleroderma, and hypertrichosis in s un-exposed areas.[2] Sight-threatening ocular manifestations are rare and we describe a case of PCT presenting with corneal perforation and scleromalacia perforans.

A 54-year-old black female presented with a 6-week history of pain and loss of vision in the right eye, and progressive darkening and coarseness of her skin.

Examination revealed dark skin and sclerodermatous-like facial features (Figure 1). Visual acuity was hand movements right and 6/5 left. There were bilateral, symmetrical areas of punched out scleral thinning with choroidal show temporally in the interpalpebral fissures. The sclera was moderately inflamed in the right with a thin cornea, central perforation, and flat anterior chamber (Figure 2).

She had a tender hepatomegally and no systemic features suggestive of collagen-vascular disease.

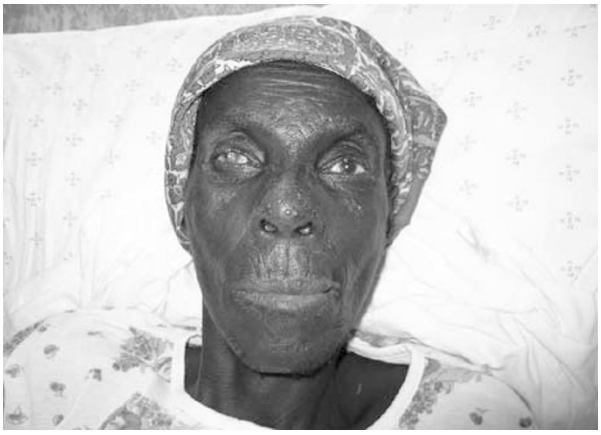

Figure 1 Sclerodermatous features in porphyria cutanea tardia.

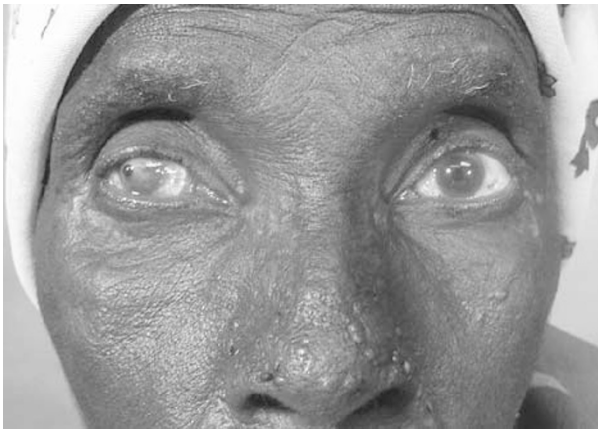

Figure 2 Perforated right cornea and bilateral scleromalacia.

Urine porphyrins were 14 035 nmol/l, serum iron 58.3 μmol and ferritin 1790 ng/l confirming a diagnosis of PCT and iron overload.

Treatment included oral prednisolone 60 mg daily and cyclosporine 2% drops QDS, avoiding topical steroids because of the risk of scleral perforation. Oral steroids were tapered and topical cyclosporine and lubricants continued. There has been steady improvement in scleral thickness and no sign of disease progression. The right eye is comfortable with a sclerosed cornea. She has undergone serial phlebotomy and is avoiding alcohol and sun exposure.

PCT is a hepatic porphyria characterised by deficient uroporphyrinogen decarboxylase activity and may be autosomal dominantly inherited or may occur sporadically.[1] The predilection in black Southern Africans may be due to the increased incidence of haemochromatosis caused by ingestion of traditional tribal beer brewed in iron pots.[1] Hereditary PCT is characterised by enzyme deficiency in all tissues while in acquired PCT, the deficiency is isolated to the liver and