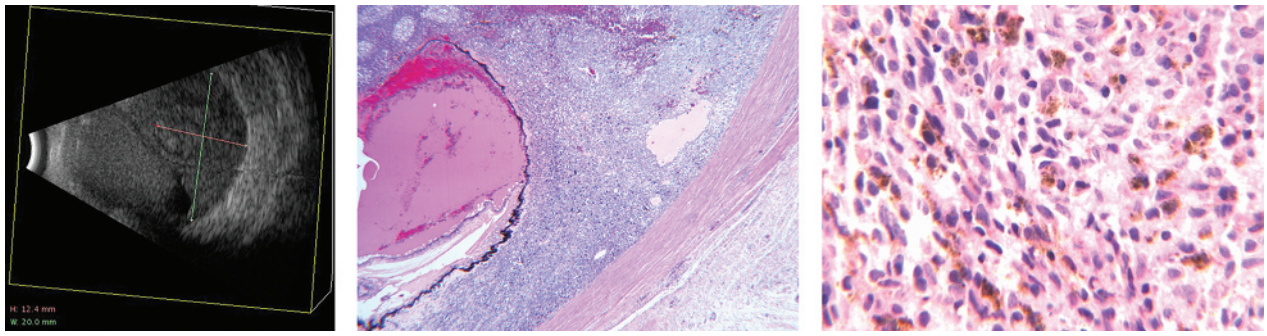

Figure 2 (Left) Three dimensional B-scan ultrasound reconstruction reveals a AJCC-T3 mushroom-shaped choroidal melanoma surrounded by a dense vitreous haemorrhage. (see *BJO* website (www.bjophthalmol.com/supplemental) 20 MHz dynamic B-scan ultrasound examination demonstrates blood flow ''twinkling'' within the tumour). (Middle) Histopathological evaluation of the exenteration specimen revealed no evidence of residual conjunctival melanoma and an intact sclera at the base of the predominantly epithelioid cell (pT3) choroidal melanoma (haematoxylin and eosin ×10).[4] (Right) In contrast, histopathological evaluation of the original conjunctival melanoma specimen revealed plump spindle cells and small epithelioid cells with moderate pigment production characterise this malignant melanoma of conjunctival origin. There is an attendant inflammatory response (haematoxylin and eosin ×40).

P T Finger, C E Iacob, S A McCormick
The New York Eye and Ear Infirmary, New York City, New York, USA

Correspondence to: Paul T Finger, The New York Eye Cancer Centre, 115 East 61st Street, New York City, New York 10021, USA; pfinger@eyecancer.com

doi: 10.1136/bjo.2006.093799

Accepted for publication 1 May 2006

Financial disclosure: No author has any financial interest in the subject matter presented. Research Supported by The Eye Care Foundation, Inc, New York City, New York, USA.

See *BJO* website (www.bjophthalmol.com/supplemental) for a further figure.

References

1 **Lommatzsch PK**, Werschnik C, Gutz U. [Intraocular invasion of melanoma of the conjunctiva]. *Klin Monatsbl Augenheilkd* 1999;**215**:370–2.
2 **Gow JA**, Spencer WH. Intraocular extension of an epibulbar malignant melanoma. *Arch Ophthalmol* 1973;**90**:57–9.
3 **Tinning S**. Malignant melanoma of the conjunctiva and iris in a case of primary acquired melanosis of the conjunctiva. *Acta Ophthalmol (Copenh)* 1978;**56**:837–41.
4 **Haik B**, Ainbinder DJ, Finger PT, *et al*. Part X: Ophthalmic sites (carcinoma of the eyelid, carcinoma of the conjunctiva, malignant melanoma of the conjunctiva, malignant melanoma of the uvea, retinoblastoma, carcinoma of the lacrimal gland, sarcoma of the orbit). In: Greene FL, Page DL, Fleming ID, *et al*. *AJCC cancer staging manual*. 6th ed. New York: Springer-Verlag, 2002:347–84.
5 **Finger PT**. Do you speak ocular tumor? *Ophthalmology* 2003;**110**:13–14.
6 **Finger PT**. ''Finger-tip'' cryotherapy probes: treatment of squamous and melanocytic conjunctival neoplasia. *Br J Ophthalmol* 2005;**89**:942–5.
7 **Kurli M**, Finger PT. Topical Mitomycin chemotherapy for conjunctival malignant melanoma and primary acquired melanosis with atypia: 12 years' experience. *Graefes Arch Clin Exp Ophthalmol* 2005;**243**:1108–14.
8 **Romero JM**, Finger PT, Rosen RB, *et al*. Three-dimensional ultrasound for the measurement of choroidal melanomas. *Arch Ophthalmol* 2001;**119**:1275–82.
9 **Reddy S**, Kurli M, Tena LB, *et al*. PET/CT imaging: detection of choroidal melanoma. *Br J Ophthalmol* 2005;**89**:1265–9.

Combination photodynamic therapy and juxtascleral triamcinolone acetonide for the treatment of a peripapillary choroidal neovascular membrane associated with papilloedema

Peripapillary choroidal neovascularisation is an uncommon entity that can be associated with significant visual loss. It has been noted with papilloedema resulting from pseudotumour cerebri.[1] Treatment for peripapillary choroidal neovascularisation has involved observation, thermal laser photocoagulation, or surgical excision.[2] We report a case of peripapillary choroidal neovascularisation from papilloedema secondary to pseudotumour cerebri that was successfully treated with combination photodynamic therapy and juxtascleral triamcinolone acetonide.

Case report

A 27 year old woman presented to the neuro-ophthalmology clinic for headaches and transient visual obscurations in both eyes. Visual acuity was 20/20 in both eyes and fundus examination was significant for optic nerve oedema in both eyes. After undergoing diagnostic testing, which included visual field analysis, lumbar puncture, and magnetic resonance imaging, a diagnosis of pseudotumour cerebri was firmly established. The patient was started on oral acetazolamide and noticed improvement of symptoms. At a follow up visit 4 months later, she presented with progressive vision loss in the left eye. Visual acuity was 20/30 in the left eye and fundus examination revealed optic nerve oedema, in addition to subretinal fluid and blood adjacent to the nerve. Fluorescein angiography was performed and confirmed the presence of a peripapillary choroidal neovascular membrane (fig 1). The patient underwent combination treatment with photodynamic therapy with verteporfin (Visudyne, Novartis Pharmaceuticals, East Hanover, NJ, USA) and juxtascleral triamcinolone acetonide on the same day. The spot size for photodynamic therapy was determined by measuring the greatest radial distance from the optic nerve edge to the border of the area of leakage on the fluorescein angiogram and a total dose of 18 J/cm^2 was delivered. Triamcinolone acetonide, 40 mg, was injected into the juxtascleral space after a conjunctival cutdown was performed 8 mm posterior to the limbus. Six months after combination therapy, visual acuity in the left eye improved to 20/20 and there was resolution of the subretinal fluid and no evidence of leakage from the choroidal neovascular membrane (fig 2). In addition, intraocular pressure remained within normal limits at all office visits.

Comment

The association of choroidal neovascularisation with papilloedema is not completely understood. It has been suggested that the physical deformation of the peripapillary tissues may create the required path for the growth of the choroidal neovascular membrane.[3] Laser photocoagulation for peripapillary choroidal neovascularisation causes damage to the overlying retina and has been associated with vitreous haemorrhage and branch arteriole occlusion. Surgical removal of peripapillary lesions is invasive and can often lead to defects in the retinal pigment epithelium.[2] Photodynamic therapy has recently been reported to be safe and successful in the treatment of peripapillary choroidal neovascularisation from age related macular degeneration and presumed ocular histoplasmosis syndrome.[4] The advantage of photodynamic therapy is the reduced risk of collateral damage to surrounding tissue, compared to surgical excision and thermal laser photocoagulation. As it has been used in the treatment of circumscribed choroidal haemangioma, photodynamic therapy does not appear to have an adverse affect on the optic nerve.[5] However, photodynamic therapy can cause a release of angiogenic factors. Triamcinolone acetonide has antiangiogenic and antipermeability effects and combination therapy with photodynamic therapy and juxtascleral triamcinolone acetonide has been shown to be beneficial in treating subfoveal choroidal neovascular membranes related to age related macular degeneration.[6]

Peripapillary choroidal neovascularisation can cause visual loss in patients with papilloedema from pseudotumour cerebri.

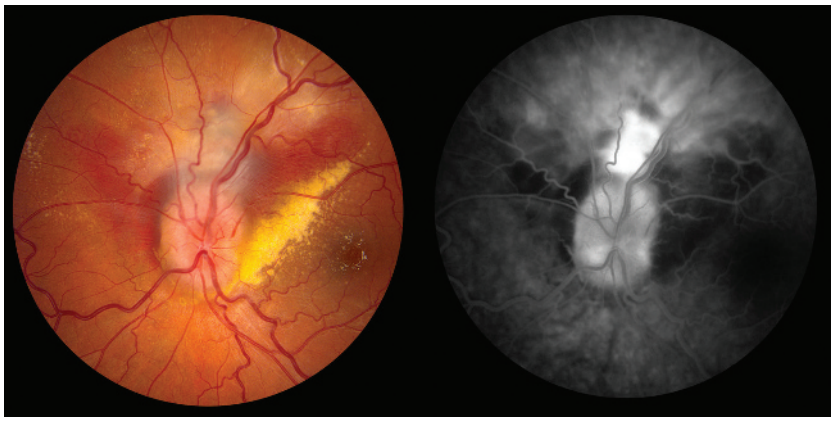

Figure 1 Left eye of a patient with papilloedema and a peripapillary choroidal neovascular membrane. There is prominent subretinal haemorrhage and exudation near the peripapillary choroidal neovascular membrane in the colour photograph (left), and a fluorescein angiogram reveals late leakage (right).

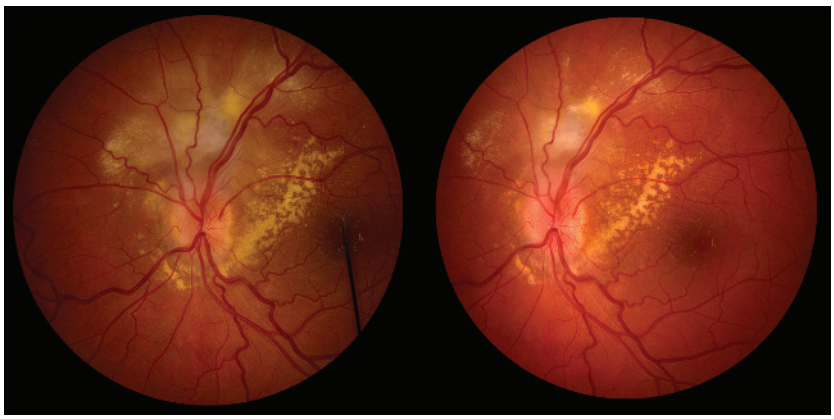

Figure 2 Colour photographs after treatment with combination photodynamic therapy and juxtascleral triamcinolone acetonide. At 2 months after treatment (left), there is a resolving edge of exudation and at 6 months after treatment (right), the choroidal neovascular membrane has decreased in size and there is resolution of subretinal blood.

Although a longer follow up may be necessary, we report the successful treatment of a peripapillary choroidal neovascular membrane related to papilloedema with combination photodynamic therapy and juxtascleral triamcinolone acetonide.

A Tewari, G K Shah, M S Dhalla, J B Shepherd
Barnes Retina Institute and Washington University Department of Ophthalmology and Visual Science, St Louis, MO, USA

Correspondence to: Gaurav K Shah, MD, Barnes Retina Institute, 1600 S Brentwood, Suite 800, St Louis, MO 63144, USA; gkshah1@gmail.com

doi: 10.1136/bjo.2006.096172

Accepted for publication 18 May 2006

References

1 **Morse PH**, Leveille AS, Antel JP, *et al*. Bilateral juxtapapillary subretinal neovascularization associated with pseudotumor cerebri. *Am J Ophthalmol* 1981;**91**:312–17.
2 **Castellarin AA**, Sugino IK, Nasir M, *et al*. Clinicopathological correlation of an excised choroidal neovascular membrane in pseudotumour cerebri. *Br J Ophthalmol* 1997;**81**:994–1000.
3 **Morse PH**, Leveille AS, Antel JP, *et al*. Bilateral juxtapapillary subretinal neovascularization associated with pseudotumor cerebri. *Am J Ophthalmol* 1981;**91**:312–17.
4 **Rosenblatt BJ**, Shah GK, Blinder K. Photodynamic therapy with verteporfin for peripapillary choroidal neovascularization. *Retina* 2005;**25**:33–7.
5 **Robertson DM**. Photodynamic therapy for choroidal hemangioma associated with serous retinal detachment. *Arch Ophthalmol* 2002;**120**:1155–61.
6 **Van de MA**, Sandhu SS, Kak R, *et al*. Effect of posterior juxtascleral triamcinolone acetonide on choroidal neovascular growth after photodynamic therapy with verteporfin. *Ophthalmology* 2005;**112**:1896–903.

Advanced bilateral persistent fetal vasculature associated with a novel mutation in the Norrie gene

Unilateral cases of persistent fetal vasculature (PFV) are sporadic,[1] while bilateral cases may be associated with X linked Norrie disease.[2] Associated sensorineural deafness and mental retardation in males[3] may be absent,[4] and genetic testing can be useful in separating sporadic PFV from Norrie disease as the following report illustrates.

Case report

A male proband (III:1, fig 1), the first child of healthy non-consanguineous white parents, was born by normal vaginal delivery, at term, following normal pregnancy and weighed 3.64 kg. Bilateral PFV was noted (fig 2) with a normal systemic examination including hearing test. During bilateral vitreo-lensectomy, severe retinal and optic nerve dysplasia was found. The four generation family history was negative for eye problems. Development at 15 months was normal. The karyotype was normal. A missense mutation in the Norrie disease gene (NDP) was identified in the proband. This was 123G>T, leading to an amino acid substitution of arginine to serine at codon 41 (R41S), which is a non-conservative change. As this sequence change had not been previously reported in Norrie disease, it was unclear whether it was a pathogenic mutation or a polymorphism in the gene, unrelated to the patient's phenotype. Genetic testing of the mother (II:2, fig 1) identified the same mutation in the heterozygous form, indicating that it had not arisen de novo in the proband. In the absence of signs of PFV in the mother, genetic testing of other family members was performed. The mutation was absent from the proband's maternal grandparents, indicating that it had probably arisen on one of the maternal X chromosomes. It is therefore highly likely to be a pathogenic mutation.

Comment

Norrie disease is an X linked disease causing bilateral blindness as a result of the primitive

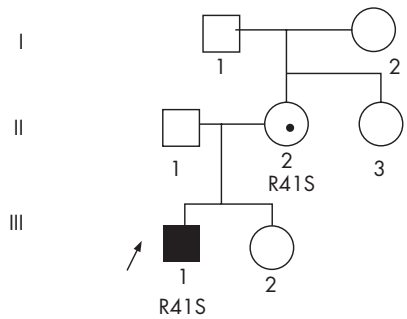

Figure 1 Three generation pedigree. Males are shown as squares and females as circles. The proband is indicated with an arrow. The solid symbols represent affected individuals and open symbols represent unaffected individuals. A dot in the centre of a symbol represents an unaffected carrier.

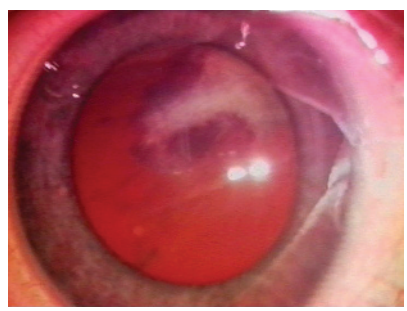

Figure 2 Lenticular vascularisation of the left eye.