plasma viscosity, prothrombotic screen and protein electrophoresis were normal. Free protein S was 56% (normal values 23–63%), protein C activity 87% (normal values 70–130%), anti-thrombin 92% (normal values 75–125%) and activated protein C resistance 3.6 (normal values 2.6–3.8). No factor V Leiden test was performed as the screening test (activated protein C resistance) was negative. Her alcohol intake of 30 units per week resulted in a mild thrombocytopenia ($121 \times 10^9/l$) and a raised gamma-glutamyl transpeptidase (419). Her vision at present is counting fingers in the right eye and 6/18 in the left eye.

Comment

Activated protein C resistance has been described as the most common known cause of CRVO in young patients under 50 years of age.¹ This view has been challenged recently.² The combined oral contraceptive pill (COCP) has been implicated as major risk factor for the development of CRVO.³ The combination of smoking and COCP may have contributed to this patient's bilateral CRVOs. Antibodies to synthetic sex hormones^{4,5} such as cyproterone and ethinyloestradiol have been noted in patients with deep vein thromboses. These antibodies may act as prothrombotic factors. This is the first report of a young transsexual with bilateral CRVOs. It raises issues with respect to the long-term treatment modalities used in gender reassignment.

References

1. Larsson J, Olafsdottir E, Bauer B. Activated protein C resistance in young adults with central retinal vein occlusion. *Br J Ophthalmol* 1996;80:200–2.
2. Gottlieb JL, Blice JP, Mestichelli B, Konkle BA, Benson WE. Activated protein C resistance, factor V Leiden and central retinal vein occlusion in young adults. *Arch Ophthalmol* 1998;116:577–9.
3. Kirwan JF, Tsaloumas MD, Vinall H, Prior P, Kritzing EE, Dodson PM. Sex hormone preparations and retinal vein occlusion. *Eye* 1997;11:53–6.
4. Leroy O, Beuscart C, Senneville, Visticot F, Tillie I, Brion M, Beaucaire G. Deep venous thrombosis and antibodies to cyproterone acetate. *Lancet* 1990;336:509.
5. Beaumont V, Lemort N, Beaumont JL. Oral contraception, circulating immune complexes, antiethinyloestradiol antibodies and thrombosis. *Am J Reprod Immunol* 1982;2:8–12.

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Sir,

Vasoproliferative tumour of the ocular fundus associated with Waardenburg's syndrome

The vasoproliferative tumour of the ocular fundus is an uncommon, benign vascular mass that can lead to exudation, retinal detachment and ultimate visual loss. It can occur as an isolated finding or be associated with a number of ocular conditions including intermediate uveitis, retinitis pigmentosa, toxoplasmic chorioretinitis, ocular toxocariasis and trauma.^{1–3} We present a case of such a tumour occurring in a patient with Waardenburg's syndrome.

Case report

A 47-year-old woman with known Waardenburg's syndrome was found on routine ocular examination to have an intraocular mass in her left eye. Her visual acuity was 6/6 in both eyes. She showed several of the classic features of Waardenburg's syndrome, including white forelock, mild telecanthus and unilateral congenital deafness. Her father and son were similarly affected. Ocular features included iris heterochromia, the left iris being a uniform blue colour (Fig. 1) and the right iris brown in colour apart from the superior third which was blue (Fig. 2). Both fundi showed diffuse retinal pigment epithelial (RPE) loss with areas of pigment clumping (Fig. 3).

The pertinent finding was in the inferior quadrant of the left eye where a $9 \times 7 \times 2.5$ mm ill-defined pink retinal mass with surrounding subretinal exudation and overlying serous retinal detachment was found (Fig. 4). There were attenuated feeder vessels, but no dilated, tortuous feeder vessels as seen with the retinal capillary haemangioma associated with Von Hippel's disease. As the patient was asymptomatic, cautious observation was advised.

Comment

Vasoproliferative tumours of the ocular fundus were reported by J. Shields *et al.* in 1983 in a report of 12 cases.¹ A subsequent series by C. Shields *et al.* reported 103 cases of similar lesions and categorised them into primary and

Fig. 1. Slit-lamp photograph of the hypochromic left iris.

Fig. 2. Slit-lamp photograph of the right iris showing sectoral hypochromia.

secondary types.² There have been several reports detailing associations with a number of ocular conditions including intermediate uveitis, toxoplasmic chorioretinitis, ocular toxocariasis, ocular trauma, retinitis pigmentosa, Coats' disease and following retinal detachment surgery.^{2,3} These conditions are often associated with RPE alterations and such alterations were noted in 79% of the secondary tumours in our prior series.² This association has led to the suggestion that vasoproliferative tumours may represent excessive vascularisation of pigment epithelial proliferation or reactive gliosis.²

Waardenburg's syndrome is an autosomal dominant condition characterised by white forelock, prominent root of the nose, congenital deafness, telecanthus and iris heterochromia. In addition, Goldberg in 1996 described pigmentary abnormalities of the fundus, in particular RPE atrophy on the side corresponding to the hypochromic iris.⁴

In our patient, the vasoproliferative tumour developed in the hypopigmented left eye. It is speculated that the RPE changes apparent in this eye (diffuse atrophy with pigment clumping) may have predisposed

Fig. 3. Fundus photograph of the left macula showing retinal pigment epithelial loss and sparse RPE clumping.

Fig. 4. Fundus photograph of the vasoproliferative tumour. Note the surrounding subretinal exudation, RPE hyperplasia and lack of dilated feeder vessels.

the patient to the development of this mass, similar to those eyes with retinitis pigmentosa. Waardenburg's syndrome should be added to the list of conditions associated with vasoproliferative tumours of the ocular fundus.

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References

1. Shields JA, Decker WL, Sanborn GE, Augsberger JJ, Goldberg RE. Presumed acquired retinal hemangiomas. *Ophthalmology* 1983;90:1291-300.
2. Shields CL, Shields JA, Barrett J, De Potter P. Vasoproliferative tumours of the ocular fundus: classification and clinical manifestations in 103 patients. *Arch Ophthalmol* 1995;113:615-23.
3. Medlock RD, Shields JA, Shields CL, Yarian DL, Beyrer CR. Retinal hemangioma-like lesions in eyes with retinitis pigmentosa. *Retina* 1990;10:274-7.
4. Goldberg MF. Waardenburg's syndrome with fundus and other abnormalities. *Arch Ophthalmol* 1996;76:797-810.

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