Extensive Myelination with Retinal Detachment: A Diagnostic Challenge

S Ho[1], L Mowatt[2], B Patil[1], RA Scott[3], GR Kirkby[3]

INTRODUCTION

Myelinated retinal nerve fibres have been associated with various conditions such as myopia, amblyopia, retinal vascular anomalies and vitreomacular traction syndrome (1, 2, 3). We present two patients with extensive macular myelination associated with retinal detachment and one of them posed a diagnostic challenge.

Case Reports

Case 1

A 35-year old Caucasian female presented with a three-week history of deteriorating vision, floaters and a shadow in the inferior nasal quadrant of her right eye.

The right eye had a best corrected visual acuity of hand movements due to a congenital cataract, amblyopia and high myopia. The left eye was normal with an unaided visual acuity of 6/6. On examination, the right fundus showed a subtotal rhegmatogenous retinal detachment involving the macula, with a superotemporal horseshoe retinal tear. There were tobacco dust and vitritis in the vitreous. The retina was immobile and showed a peculiar white area over the macula and peripapillary region obscuring the retinal vessels. B scan ultrasonography suggested severe retinal fibrosis. A diagnosis of rhegmatogenous retinal detachment with possible retinal infiltration was made. Right vitrectomy, lensectomy, endolaser with intra-ocular gas (25% SF_6) tamponade was performed. The vitreous sample was sent for culture and cytopathology and the results were negative. Two-weeks postoperatively, the retina was flat and the white macular appearance was identified as a large area of myelinated nerve fibres. However, her vision never improved beyond 1/60 due to dense amblyopia.

Case 2

A 50-year old female presented with a one-week history of a field defect in the outer aspect of her right eye. Past ocular history revealed ipsilateral anisometropic amblyopia with a myopia of minus 12 diopters. Her best-corrected right visual acuity was count fingers (CF). The left eye was normal with an unaided visual acuity of 6/6. Fundal examination demonstrated an inferonasal, rhegmatogenous, macula-sparing retinal detachment with extensive myelinated nerve fibres involving the entire macula (Figure). The detached posterior hyaloid face was thickened and opaque. She underwent a scleral buckling procedure with cryopexy and was discharged. At her six-month postoperative visit she had a best corrected visual acuity of count finger and her retina was flat.

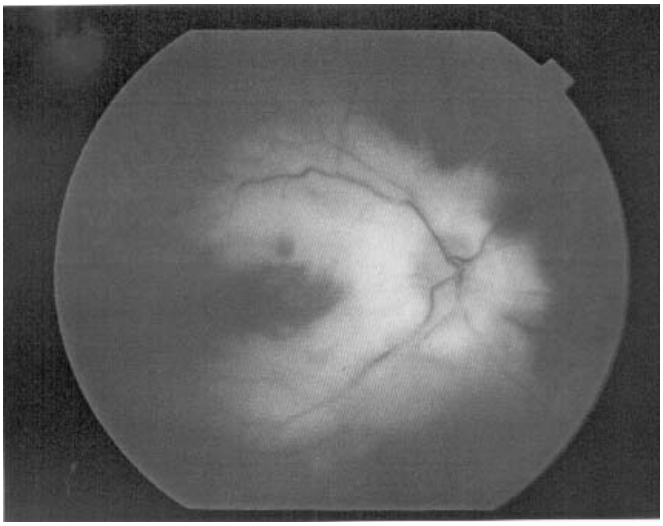

Figure: Extensive retinal myelination in the macula of this patient's right eye with a peripheral retinal detachment (not included in this photograph) for Case 2.

DISCUSSION

Retinal detachment in association with extensive retinal myelination can pose diagnostic problems. A few of the differential diagnoses to be considered are proliferative vitreoretinopathy (PVR), exudative retinal detachment and detachments secondary to inflammatory conditions. Proliferative vitreoretinopathy is characterized by the proliferation of membranes on one or both surfaces of the retina making the retina rigid and immobile. It generally takes several months for eyes without previous surgery to develop PVR (4). The fibrosis of PVR is predominantly preretinal or subretinal but not intraretinal. Proliferative vitreoretinopathy characteristically results in increased retinal stiffness on kinetic ultrasonography (5). Case one appeared to have PVR on presentation with apparent peripheral fibrosis and reduced mobility on kinetic ultrasonography.

Another possible differential diagnosis was retinal detachment in association with inflammatory or malignant infiltration (6). This was excluded by the absence of a history of uveitis, any vascular sheathing or papillitis. Diagnostic vitreous biopsy also yielded a negative result. There was no evidence of infectious uveitis.

In conclusion, if the area of detachment involves myelination of the retinal nerve fibers, the appearance may mimic PVR or inflammatory or infiltrative retinal disease. Retinal detachments with associated PVR require more

From: [1]Ophthalmology Department, Queen's Medical Centre, Nottingham NG7 2UH, UK, [2]Department of Surgery, Radiology, Anaesthesia and Intensive Care, The University Hospital of the West Indies, Kingston 7, Jamaica, West Indies and [3]Birmingham and Midland Eye Centre, Dudley Road, Birmingham B18 7QH, UK

Correspondence: Dr S Ho, Ophthalmology Department, Queen's Medical Centre, Derby Road, Nottingham, NG7 2UH, United Kingdom. Fax: (44) 115 970 9749), e-mail s_ho2@ya hoo.com.

complex vitreoretinal procedures, hence correct pre-operative diagnosis is important in planning surgical intervention. A diagnosis of myelinated retinal nerve fibers is important as these detachments in the absence of PVR can be managed in most cases with simple primary vitreoretinal surgical techniques. When faced with a retinal detachment associated with an opaque macula and peripapillary area, retinal myelination should be included in the differential diagnosis as it has an impact on the surgical management.

REFERENCES

1. Lee MS, Gonzalez C. Unilateral peripapillary myelinated retinal nerve fibres associated with strabismus, amblyopia and myopia. Am J Ophthalmol 1998; **125:** 554–6.
2. Leys AM, Leys MJ, Hooymans JM, Craandijk A, Malenfant M, van Germeersch D, et al. Myelinated nerve fibres and retinal vascular abnormalities. Retina 1996; **16:** 89–96.
3. Hubbard GB, Thomas MA, Grossniklaus HE. Vitreomacular Traction syndrome with extensively myelinated nerve fibres. Arch Ophthalmol 2002; **120:** 670–1.
4. Thompson JT. Proliferative vitreoretinopathy. Ryan SJ, ed. Retina. St Louis, MO: Mosby Company Inc 2000: 2289–316.
5. Fuller, DG, Laqua H, Machemer R: Ultrasonographic diagnosis of massive periretinal proliferation in eyes with opaque media (triangular retinal detachment). Am J Ophthalmol 1977; **83:** 460–4.
6. Nagpal A, Biswas J. Pseudouveitis – analysis of cases misdiagnosed as posterior uveitis. Ocul Immunol Inflamm 2006; **14:** 13–20.