

Sir,

Familial myelinated retinal nerve fibres *Eye* (2003) **17**, 96–97. doi:10.1038/sj.eye.6700266

Myelinated retinal nerve fibres occur in 1% of the population.^{1,2} Although myelination has been extensively studied, the cause of abnormal retinal myelination is subject to debate.^{3,4} Here, we describe two cases of myelinated retinal nerve fibres, in a patient and her mother, suggesting that an inherited defect may be responsible in certain cases.

Case report

A 22-year-old female presented to our department with symptoms of ocular irritation. She was previously fit and well. On examination, visual acuity was 6/5 in both eyes with +1.50 RE and +2.00 LE. External inspection showed anterior blepharitis, consistent with her symptoms. Routine fundoscopy revealed peripapillary myelinated nerve fibres in both eyes. The right was more

significantly affected, with myelination of the nasal peripapillary, up to half a disc diameter from the optic nerve margin. The left eye had a small area of myelination on the inferior disc margin (Figure 1). The ophthalmic examination was otherwise unremarkable including normal pupil reactions and full fields.

Subsequent examination of the patient's mother revealed extensive bilateral peripapillary nerve fibre myelination. The appearance was similar in both eyes, forming a confluent pattern of myelination up to two disc diameters from the normal optic nerve margin (Figure 2).

Comment

Retinal myelination is usually observed as an asymptomatic, isolated finding following routine examination. It is usually considered to be nonprogressive, although there are two cases in which progression has been confirmed.^{5,6} Rarely regression or disappearance of the aberrant myelin has been noted following optic neuritis or ischaemia.⁷ Myelinated retinal



Figure 1 Fundus photographs of daughter showing peripapillary myelinated nerve fibres. These are up to half the disc diameters in the nasal peripapillary area in the right eye and small area of myelination to the disc in the left eye.



Figure 2 Fundus photographs of mother showing extensive bilateral peripapillary nerve fibre myelination, in confluent pattern, up to two disc diameters from the normal optic nerve margin.

nerve fibres occur more commonly in association with neurofibromatosis, Down's syndrome and craniofacial dysostosis.^{1,8} There are, however, very few reports of familial myelination of retinal nerve fibres.^{9,10} The findings of myelinated fibres, in both mother and daughter, suggest that a genetic factor could play a role.

Normal myelination of optic nerve axons is thought to occur in three phases. Firstly, the oligodendrocyte lineage disseminates, via migratory oligodendrocyte progenitors, along large axons. The progenitors are thought to end migration when a local axonal signal falls below a critical level. Secondly, progenitors generate oligodendrocytes responsible for early myelination. The third phase involves consolidation of myelin, via mature progenitors. These migrate only short distances, slowly producing oligodendrocytes.⁴

The formation of aberrant myelinated retinal nerve fibres is thought to be due to failure to prevent oligodendrocyte lineage cells from passing through the lamina cribrosa or optic nerve head. It has been postulated that astrocytes in the lamina cribrosa are specialised to act as a barrier via the orientation and number of their glial filaments.⁴ The mechanism by which an inheritable defect could affect this process is still unknown.

It is possible that familial myelinated retinal nerve fibres are more common than stated as they are generally asymptomatic and family members are not regularly screened.

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Sir,

Post-chemotherapy premacular subhyaloid haemorrhage

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We read with interest Rennie's series of premacular subhyaloid haemorrhage treated successfully with Nd:YAG laser in the August 2001 issue.¹ The causes of the haemorrhage in their series included Valslava retinopathy, macroaneurysm, branch vein occlusion, proliferative diabetic retinopathy and idiopathic. We describe here a case of bilateral premacular subhyaloid haemorrhage as a result of chemotherapy-induced pancytopenia.

Case report

A 59-year-old Asian man was referred to the Eye Department 2 weeks after developing sudden deterioration of vision in both eyes. A diagnosis of Stage IV B Mixed Cellularity Hodgkin's lymphoma had been made 6 months previously and chemotherapy had been commenced using the ChIVPP/PABLOE regime (chlorambucil, vinblastine, carbazine, prednisoline/prednisolone, adriamycin, bleomycin, vincristine, etoposide). The second chemotherapy treatment was given 10 days before the onset of his ophthalmic symptoms. This was complicated