Retinal Myelinated nerve Fibres and their association with anomalous Retinal vasculature and Vitreous hemorrhage

Author’s Affiliation

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Purpose: Background/Purpose: Anomalous retinal vessels may develop in a region of myelinated nerve fibers, and these vessels may cause vitreous hemorrhages.

Methods: The clinical histories of five patients with retinovascular abnormalities in or around a patch of myelinated nerve fibers are presented. None of the reported patients had other evidence of systemic disease. The cases were traced by a retrospective study conducted at the Department of Ophthalmology, SIMS and Services Hospital, Lahore between December 2004 and November 2012.

Results: Retinal vascular abnormalities ranged from mild telangiectasia to frank neovascularization. Age at diagnosis ranged from 18 to 62 years. Vitreous hemorrhages occurred in all five patients. Laser photoagulation was applied in all five patients.

Conclusion: We suggest that the abnormal structure of the myelinated nerve fibers and the thickened nerve fiber layer of the affected portions of retina may play a role in the onset of retinal vascular abnormalities and eventually cause telangiectasis, neovascularization, and vitreous hemorrhages. This suggestion is based on the absence of other causes of neovascularization or vitreous hemorrhage in all five patients, and on the relatively young age of the patients.

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**Introduction:**

Myelinated nerve fibres may occur in up to 0.98% of patients and are generally considered a benign developmental abnormality. They are seen as white, opaque, striated patches with feathery margins. They are present at retinal nerve fibre layer and may be discrete or, as occurs more commonly, adjoin the optic disc. They may be associated with ocular disorders such as axial myopia and refractory amblyopia. Occasionally they are a part of congenital multi-system disorders such as Gorlin's syndrome and autosomal dominant vitreoretinopathy with skeletal malformations. Minning has reported abnormal blood vessels in patches of myelinated nerve fibres along with repeated vitreous hemorrhages, but his observations have not been emphasized in the literature.

Five patients (mean period of follow-up, 6 years; range, 6 months to 9 years). The patient characteristics are summarized in Table 1 and are presented in more detail in the following case reports.

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<th>Follow-up (yrs)</th>
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<th>Laser</th>
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Table 1: Showing type of retinal vascular abnormality; Neov, neovascularization and presence or absence of axial myopia.

(Figure 1) Fundus photograph of right eye showing dense myelinated nerve fibres in the paramacular and perimacular area.

**Case 1**

The patient at age 28yrs presented with blurred vision in the right eye. His medical history did not reveal anything remarkable. Visual acuity was correctable to 6/6 in the left but only up to 6/12 in the right. Examination of the right fundus showed a mild vitreous hemorrhage, and dense myelinated nerve fibres in the peripapillary, paramacular area as well as within the arcades. The left fundus was normal. Eight years prior to his presentation, a similar episode occurred in the same eye for which he required laser treatment. Fluorescein angiography revealed areas of capillary dropout and abnormal blood vessels with leakage in the area of myelinated nerve fibres indicative of neovascularization. Focal laser photocoagulation was performed. In the last 9 years, no recurrence of vitreous hemorrhage has been noted (Figure 1).

The fundus of the right eye was normal. A small vitreous hemorrhage, extensive patches of myelinated fibres, and vascular tufts confined to these areas were noted on ophthalmoscopy of the left eye. Fundus examination of the left eye (Figure 2) showed myelinated nerve fibres along the upper and inferior temporal arcades. On fluorescein angiography, in the upper patch of myelin, telangiectatic vessels with staining were noted. In the inferior patch, retinal vascular changes were more pronounced, and the late phase angiogram showed mild leakage indicative of neovascularization.

The macula appeared normal. Focal laser photocoagulation was performed in the area of neovascularization. The condition remains stable to date.

**Case 2**

A 24-year-old healthy young man presented with a history of sudden onset vitreous floaters in the left eye. Corrected visual acuity was 6/6 in the right eye and 6/9 in the left eye. The fundus of the right eye was normal.
Funduscopy and fluorescein angiography revealed a small neovascular tuft at the inferotemporal quadrant and patches of myelinated nerve fibres (Figure 4). Sectoral laser photocoagulation was performed and the vitreous hemorrhage resolved. The patient returned 5 years later because of recurrent vitreous hemorrhage in the left eye. Laser photocoagulation was applied to and around the area of the neovascular tuft. No recurrence of the hemorrhage has been reported. This patient also had no significant medical history.

(Figure 4). Fundus photograph of the inferotemporal retinal periphery of the left eye showing a neovascular tuft and mild vitreous hemorrhage (Arrow) in an area of Myelinated nerve fibre.

Case 5

A 48-year-old man, otherwise medically fit, presented with sudden visual loss in his left eye. 3 years ago, he had a similar episode in the same eye and required three sessions of laser treatment. Examination revealed a mild vitreous haemorrhage and myelinated nerve fibres with retinal vascular anomalies (Figure 5) in the inferotemporal quadrant of his left eye. His vision was reduced to 6/12 in the left eye and 6/6 in the right A fluorescein angiograph demonstrated telangiectasies in the region of myelinated nerve fibres with leakage. Sector (Temporal) Argon laser photocoagulation was performed and the vision returned to 6/6 after 2 months. It has stayed stable to date.

(Figure 5). Fundus photograph of the left inferotemporal retina showing anomalous blood vessels (Arrows) in a patch of Myelination.
Discussion:

Myelinated nerve fibres have been reported with various ocular conditions including strabismus, macular aplasia, anamalopia, nystagmus, keratoconus, colobomas and epiretinal membranes. Other associations are neurofibromatosis, Down's and Gorlin's syndromes, autosomal dominant vitreoretinopathy with skeletal malformations. Most cases of myelinated nerve fibre are not associated with retinal vascular abnormalities. In his paper on myelinated retinal nerve fibres, Straatsma reported no associated vascular anomalies in a series of 42 eyes from autopsies and 37 eyes from a clinical group.

Rarely, myelinated nerve fibres can be associated with various retinal vascular abnormalities. These range from telangiectasias, collaterals to frank neovascularization. The exact cause of neovascularization in MNF is not known yet but some researchers suggest mechanical disruption of the blood vessels by myelin. Others suggest a localized ischemic process (owing to increased diameter of myelinated nerve fibres) and release of local angiogenic factors such as vascular endothelial growth factor (VEGF) similar to diabetic retinopathy and other vaso-obstructive disorders such as Branch vein occlusion, sickle cell disorder etc.

Literature review shows only one paper described by Leys at al, a case series of (seven patients), exhibiting vascular complications associated with myelinated nerve fibres.

There are a handful of case reports of co-existence of retinal vascular abnormalities and myelinated nerve fibres. Minning and Davidsorf observed neovascular growth in the center of a patch of myelinated nerve fibres in a 47-year-old man whose medical history was unremarkable. These vessels caused repeated vitreous hemorrhage and required panretinal photocoagulation. Minning and Davidoff postulated that in their case, thickened nerve fibres caused a sequence of events resulting in local ischemia or mechanical damage and blockage of the retinal blood vessels. Berry-Brincat and Shafquat also described recurrent vitreous hemorrhage as an unusual presentation with myelinated nerve fibres.

Due to rare occurrence of the condition, treatment modalities are limited too. Only few of the reported symptomatic patients had experienced laser treatment. In some patients, vitreous hemorrhage was controlled as well as without regression of the neovascularization; while in others, recurrent vitreous hemorrhages persisted in spite of repeated laser sessions. Vitrectomy was needed in only one patient in order to clear the vitreous hemorrhage. The role of anti-VEGF in the treatment in this condition certainly need to be addressed. Our findings indicate that the disturbed anatomy of the myelinated nerve fibres and the thickening of the affected retinal portion trigger the beginning of abnormalities in the retinal microvasculature, leading eventually to telangiectasias, arteriolar venular blockage, neovascularization and, ultimately, vitreous hemorrhages. We have based this hypothesis on the fact that other causes of neovascularization or vitreous hemorrhage in all five patients were absent, and also that the patient with this abnormal coexistence were all relatively young.

References: