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

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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 photocoagulation 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

Introduction:

Myelinated nerve fibres may occur in upto 0.98% of patients and are generally considered a benign developmental abnormality1. 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 2,3. Minning has reported abnormal blood vessels in patches of myelinated nerve fibres along with repeated vitreous hemorrhages10, 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.

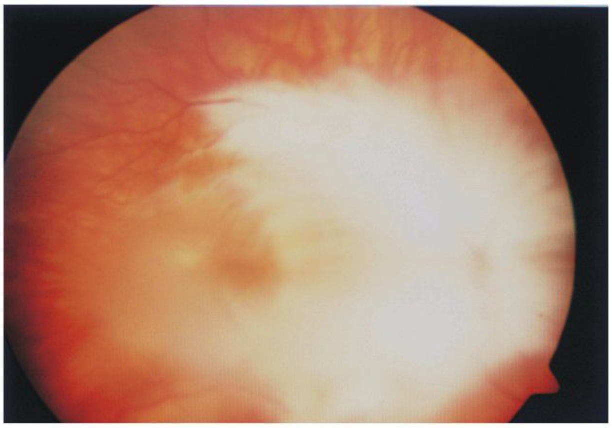
left eye. The fundus of the right eye was normal.

Case I

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 angiograph 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 (Figure1).

Case	Sex	Age (yrs)	Vessels	Follow-up (yrs)	Vitreous Hemorrhage	Laser	Vitrectomy	Myopia
1	M	28	Neov	10	+	Laser	-	-
2	M	24	Neov	8	+	Laser	-	-
3	F	33	Neov	7	+	Laser	-	-
4	F	43	Neov	3	+	Laser	-	+
5	M	48	Neov	0.5	+	Laser	-	-

Table1; Showing type of retinal vascular abnormality; Neov, neovascularization and presence or absence of axial myopia.



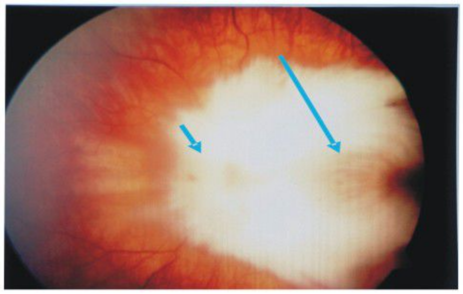
(Figure1) Fundus photograph of right eye showing dense Myelinated nerve fibres in the paramacular and perimacular area.

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

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.

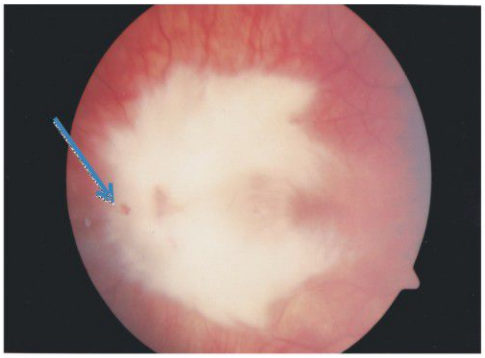
The macula appeared normal. Focal laser photocoagulation was performed in the area of neovascularization. The condition remains stable to date.



(Figure2). Fundus photograph of the left eye showing Myelinated nerve fibres involving the Optic disc (Short arrow), macula (Long arrow) as well as the whole region between the arcades

Case 3

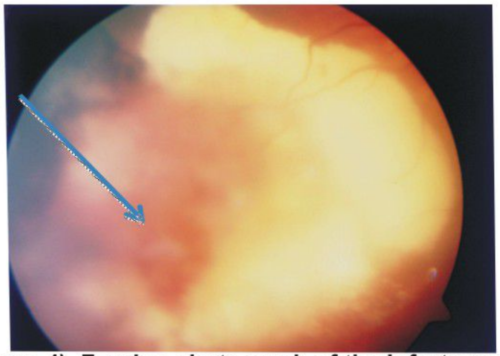
This 33-year-old woman had history of three episodes of recurrent vitreous haemorrhage in her left eye. Visual acuity was 6/6 in both eyes. Ophthalmoscopy of the right eye was unremarkable; in the left eye, extensive patches of myelinated nerve fibres surrounding the optic disc and extending along the main vascular arcades in the posterior pole and midperiphery were revealed in the area of myelinated nerve fibres, the retinal vasculature was abnormal with numerous dilated segments of small caliber vessels. The vascular anomalies were most pronounced in the peripapillary area (arrow in Fig 3). Fluorescein angiography showed leakage of these vessels, indicative of neovascularization. Panretinal laser photocoagulation was performed, and subsequently at that time, hemorrhages recurred. However, the hemorrhages cleared completely after 6 weeks, a further top-up laser photocoagulation was performed and visual acuity remained 6/6.



(Figure 3). Fundus photograph of the left eye showing dense Myelination in the posterior pole with anomalous vessels (arrow).

Case 4

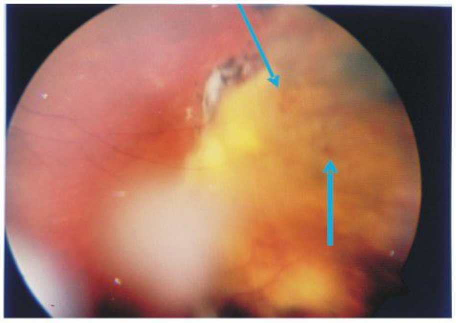
A 43-year-old woman was referred after two episodes of vitreous hemorrhage in her left eye. She denied any medical problems. Visual acuity and examination of the emmetropic right eye were normal. The left eye was amblyopic and myopic, with a refractive error of -5 diopters. Funduscopy and fluorescein 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 date. This patient also had no significant medical history.



(Figure 4). Fundus photograph of the Infrotemporal 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 telangiactesia 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 infro-temporal 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, amblyopia, nystagmus, keratoconus,[6,10] colobomas and epiretinal membranes[5]. Other associations are neurofibromatosis, Down's and Gorlin's syndromes[2], autosomal dominant vitreoretinopathy with skeletal malformations[3]. Most cases of myelinated retinal nerve fibres are not associated with retinal vascular abnormalities. In his paper on myelinated retinal nerve fibres, Straatsma[1] 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[4-10]. These range from telengiactesias, 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[7], 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 Davidorf[10] 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 Davidorff[10] 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[9] 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[6-8]. In some patients, vitreous hemorrhage was controlled with[7] as well as without[6] regression of the neovascularization; while in others, recurrent vitreous hemorrhages persisted in spite of repeated laser sessions[8]. Vitrectomy was needed in only one patient in order to clear the vitreous hemorrhage.[3] 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 telangiectasiae, 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.

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