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Case Report

An Unusual Case of Peripheral Retinoschisis Complicated by Vascular Malformations: A Case Report

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Abstract

Senile retinoschisis (SR) is an uncommon condition in which the neurosensory retinal layers abnormally separate, usually at the level of outer plexiform layer. Here, we report a rare case of bilateral senile retinoschisis with aneurysmal vascular lesions in the schitic region that led to intraschitic bleeding and later vitreous haemorrhage. We discuss the possible nature of the disease and the management course.

Keywords: Peripheral Retinoschisis; Vitreous Haemorrhage; Retinal Detachment

Introduction

Acquired or degenerative retinoschisis is an idiopathic condition characterized by gradual peripheral splitting of the retinal layers, typically between inner nuclear and outer plexiform layers. It mostly affects individuals above the age 50 years with equal predisposition to both the genders. The disease remains asymptomatic in majority of cases with bilateral occurrence reported up to 80% of cases.

Several vascular abnormalities in the setting of senile retinoschisis have been reported. Sheathed blood vessels on the inner retinal layer and white 'snowflakes' may be noted. Cases with telangiectatic retinal vessels overlying areas of senile retinoschisis and associated vitreous hemorrhage and neovascularization have been reported in literature [1]. Coats like response associated with intraschitic vessels and reactive vascular response has also been described [2].

Here we report an unusual case of bilateral senile retinoschisis with aneurysmal vascular lesions within the schitic area of the right eye which caused intraschitic and subsequently vitreous bleed. We discuss the possible nature of the disease and the management course.

Case Report

A 78-year-old male with no systemic co-morbidities presented to our retina clinic for routine evaluation. On ophthalmic examination, his best-corrected visual acuity was 6/6p, N6 in both eyes. Anterior segment examination was unremarkable. Dilated fundus examination revealed multiple berry-like orange-red retinal lesions seen within an area of retinoschisis in the infero-temporal periphery of right eye (Figure 1 A,B). It was associated with minimal adjacent sub-retinal fluid. Left eye examination showed shallow retinoschisis and chorioretinal degeneration in infero-temporal peripheral quadrant. There was no evidence of inner or outer schitic wall break in either eye. No vasculitis or inflammatory changes were noted. Macula was normal in both the eyes (Figure 2).

Optical coherence tomography (SD-OCT) through schitic area in the right eye showed globular hyper-reflective lesions attached

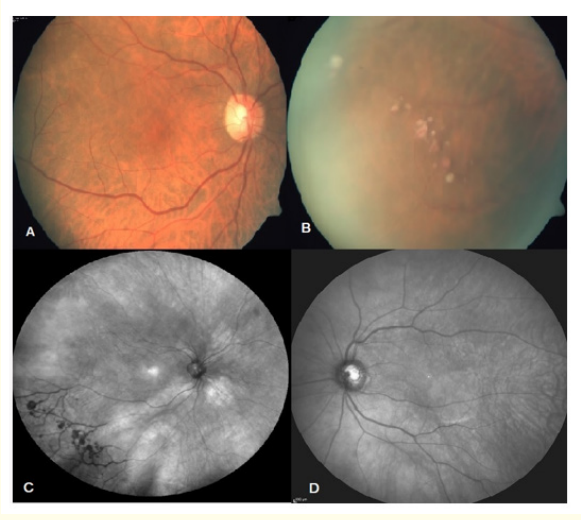

Figure 1: A,B. Color fundus photograph of the right eye showing multiple retinal lesions within the area of retinoschisis in inferotemporal quadrant C. IR image of the right eye showing well demarcated vascular lesions. D. IR image of left eye.

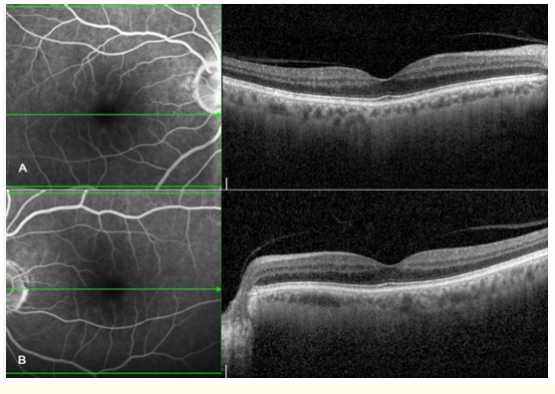

Figure 2: A,B : SD-OCT line scan showing normal fovea in both eyes.

to the undersurface of inner layer of retinoschisis (Figure 3). Fluorescein angiography indicated early hyper fluorescence coupled with late leakage from these vascular lesions. Patient was regularly followed up. As vision remained stable with self-resolution of sub retinal fluid, no active intervention was made.

On review after 2 years, right eye showed increase in number of vascular malformations with associated subretinal hemorrhage.

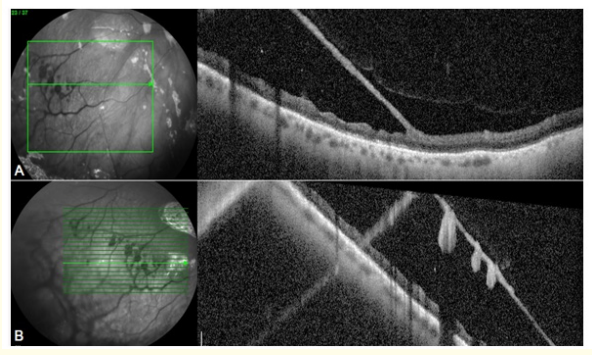

Figure 3: SD-OCT line scan of right eye through A. infero-temporal periphery showing retinoschisis. B. showing globular hyper-reflective lesions attached to undersurface of inner schitic layer.

Combined FFA and ICGA was performed (Figure 4 and 5). Fluorescein angiography confirmed the increase in number of vascular lesions with intense late leakage. Hyper-fluorescent lesions were also evident in Indocyanine green angiography. Owing to the progression of vascular activity, patient was managed with three intravitreal ranibizumab injections. Patient remained stable in post injection follow ups.

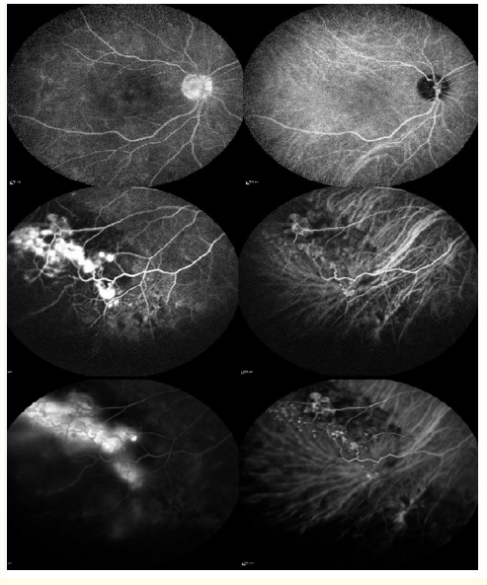

Figure 4: Combined FFA and ICGA in right eye showing early filling of vascular lesions with profuse late leakage in FFA. Lesions were evident in ICGA also.

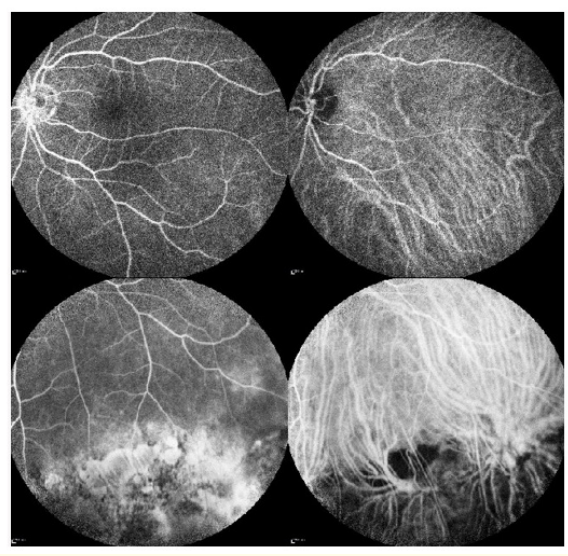

Figure 5: Infero-temporal peripheral quadrant of left eye in dye angiography showing staining of chorioretinal degeneration and settled retinoschisis..

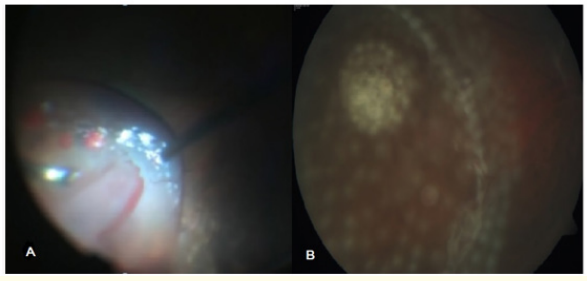

Figure 6: A. Intra-operative video-image of the vascular lesions in inferotemporal quadrant of the right eye. B. Postoperative color fundus image showing the stable endodiathermised deeper vascular lesion and surrounding endolaser marks in the affected area in silicone oil filled eye.

9 months later, patient presented with sudden diminution of vision in right eye with vision deteriorating to CF at 3 m. Examination revealed vitreous hemorrhage in the right eye. B scan ultrasonography in inferotemporal quadrant showed persistent retinoschisis with intraschitic bleed. The patient was managed surgically. After thorough pars plana vitrectomy, large retinal vessels at the schisis margins were treated with diathermy, which allowed us to perform the inner wall retinectomy. Vascular lesions were endo-diathermized and subsequently excised along with the inner wall of the peripheral schisis cavity. One sessile lesion was deeply situated with attachment to outer retinal layers. This lesion was endodiathermised and lasered (Figure 6). Following endolaser of schitic area, silicone oil tamponade was done. Postoperative examination at 1 week revealed stable retina with good visual recovery to 6/12.

Discussion and Conclusion

Senile typical retinoschisis is an asymptomatic benign degenerative condition of the peripheral retina. Patients often become symptomatic due to complications like associated rhegmatogenous retinal detachment and progressive scotoma. Even though rare, vascular abnormalities associated with senile retinoschisis have also been previously reported [1,2]. Rupture of poorly supported retinal vessels in inner schitic layer can lead to intra-cavity bleed or cause other vision threatening complications like vitreous hemorrhage as noted in our case [1,3,4].

Various pathologies of such vascular malformations have been described. There have been case reports suggesting the aneurysmal microvascular capillary terminals could be a result of initial compensation for schitic process [6]. Campo., *et al*. [1] in his case series reported similar vascular abnormalities due to ischemia, secondary to long standing inner retinal elevation. They proposed these lesions are of neovascular origin as evident by the profuse dye leakage and patchy capillary filling in FFA. Durkin., *et al*. reported retinal microvascular abnormalities appearing to be "dilated capillary terminals" without fluorescein leakage secondary to myopic retinoschisis, and hypothesized that capillary remodeling occurred as the inner and outer retinal leafs separated, which may induce bleeding if the schitic cavity progressed more rapidly, mimicking leakage of neovascularization in FFA [7]. Our case was a non-myope with intense fluorescein leakage corresponding to the vascular abnormality on FFA. However, Ong., *et al*. [5]. refuted the notion as capillary non perfusion was not demonstrated on OCT-Angiography. They proposed that the atypical vascular changes and leakage pattern could be of tractional origin rather than due to neovascularization. These vascular changes of inner retina were similar to our case except for the fact that we demonstrated an intra-operative deeper vascular lesion extension which questions the tractional changes being the only reason for these aneurysmal changes. We believe that ischemia played more significant role

in our case as evident from the progressive nature of the disease, leakage pattern in FFA and initial stabilization of the disease process with anti-VEGF treatment. But it is difficult to interpret the nature of these vascular lesions with angiography alone, as it gives an equivocal result since leakage occurs from stretched vessels also and the fact that CNP areas of extreme peripheral retina is difficult to assess with FA.

Retinoschisis and vascular abnormalities have been described in pars planitis [8,9]. Gelliskin., *et al*. have described a case of blood filled senile retinoschisis with coats like response and associated cystoid macular edema, optic disc staining and vitritis [2]. Absence of inflammatory ocular signs in our case made this diagnosis unlikely. Patel., *et al*. [10] have described a similar case of vaso-proliferative changes secondary to X linked retinoschisis with good response to anti-VEGF therapy. Secondary Vaso-proliferative tumors (VPT) are seen as a reactive vascular response to an ocular pathology and are frequently bilateral. They present with or without feeder vessel and are commonly seen in infero-temporal quadrant. Aneurysmal nature, variable amount of gliosis and associated peripheral retinal detachment associated with these lesions have also been described in the literature. Lack of gliosis in this long-standing disease expels VPT as a possibility in our case.

Our patient was surgically managed with pars plana vitrectomy and complete excision of the inner schitic layer along with the vascular lesions. Surgery was concluded with endolaser treatment and silicone oil tamponade. Vitrectomy with inner schitic wall retinectomy through diathermized vessels in surgical management of complicated congenital retinoschisis has been previously described by Ferrone., *et al*. [11]. We also agree that in such cases, complete removal of posterior cortical vitreous is enabled only by the removal of the inner schitic wall. If this cortical vitreous is left behind, later this collagen framework can act as a substrate for the cell proliferation aggravating the retinal traction.

Our case reinforces the necessity of proper follow up in such patients with retinal microvascular abnormalities associated with retinoschisis which may be vision threatening, if not timely and properly managed.

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