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Clinical outcome in acquired retinoschisis

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Abstract
Introduction: Retinoschisis is defined as splitting of the neurosensory layer of retina. It can be congenital, acquired or secondary type. Clinically, the patients show typically a cystic space stellate maculopathy or a foveal schisis.
Methods: This study was a retrospective case series. The inclusion criteria were either sex, no systemic disorder, and ready for subsequent follow ups. Through clinical history of the patient was taken. Visual acuity, intraocular pressure measurement, anterior segment examination and fundoscopy were performed. Optical Coherence Tomography of the suspected eye was done.
Results: Based on clinical and Optical Coherence Tomography findings, the diagnosis of acquired retinoschisis was made.
Conclusion: The clinical presentation of the patient and understanding of retinal anatomy is important in diagnosis of retinoschisis. Early diagnosis helps in management and reduces the risk of permanent visual loss.

Keywords: Retinoschisis, Retinal detachment, Optical coherence tomography

Introduction

The retina is microscopically divided into ten distinct layers. They are grouped into the pigmented layer (outermost layer) and the neurosensory layer (nine inner layers). Retinoschisis is defined as splitting of the neurosensory retinal components.[1] It can be congenital, acquired (also known as degenerative or senile), or secondary (associated with myopia, trauma, sickle cell anemia, and other diseases) [2].

Congenital Retinoschisis (XLRS) is a rare bilateral vitreoretinal disease transmitted as an X linked recessive trait associated with a mutation of the XLRS1 gene located on the short arm of the X chromosome, Xp22 [3]. The estimated prevalence is up to 1 in 20,000 with a bilateral appearance in 40% of the patients [4]. Senile retinoschisis (SR) was first described in 1933 by Bartels [5]. In most cases, it is an asymptomatic bilateral disease with a prevalence between 1.65% and 7.00% among individuals aged >40 years, and it usually affects the peripheral retina [6].

Clinically, the patients show typically a cystic space stellate maculopathy or a foveal schisis and peripheral retinoschisis of various degrees of severity on fundoscopy [7]. Other authors described this condition in Optical Coherence Tomography (OCT) and OCT angiography (OCTA) as splitting of inner layers of retina and perifoveal microvascular changes, more evident in the deep vascular plexus [8]. The splitting of the inner retinal layers, mainly involving Inner Nuclear Layer (INL) followed by Outer Plexiform Layer (OPL) and ganglion cell layer (GCL), determines the formation of foveal schisis that leads to early bilateral visual loss [9]. In addition, retinoschisis may be complicated by retinal detachment, vitreous or intraretinal hemorrhage and vascularized vitreous veils. In general, the management of the disease and its complications is conservative because macular involvement is rare, spontaneous progression is not frequent in the natural history of the disease, and prophylactic treatment can pose risks [10].

Materials and Methods

This study was a retrospective case series. The inclusion criteria were either sex, no systemic disorder, and ready for subsequent follow ups. Through clinical history of the patient was taken. Visual acuity, intraocular pressure measurement, anterior segment examination and dilated fundoscopy were performed. Optical Coherence Tomography (OCT) of the suspected eye was done when there was no improvement in visual acuity with pin hole or glasses.

Results

The following cases were studied and analysed

Case 1

A 22 year old female, resident of rural area of Jharkhand, India presented in outpatient department (OPD) of Regional Institute of Ophthalmology (RIO), Rajendra Institute of Medical Sciences (RIMS), Ranchi with chief complaints of diminution of vision in right eye since 2 years which was gradual in onset and progressive in nature. On examination her Visual acuity was 6/36 in right eye and 6/6 in left eye respectively. There was no improvement in right eye with pinhole or glasses. Intraocular pressure recorded by non-contact tonometer was 12 and 10 mm Hg in right and left eye respectively. There was no family history of any other eye diseases.

Anterior segment examination was within normal limit. Fundoscopy revealed a cartwheel-like appearance near the macula in right eye, while peripheral schisis was apparent. Optical Coherence Tomography (OCT) of right eye showed cystic lesions in the inner nuclear and outer plexiform layers of the retina, while the macular thickness was normal. The left eye was within normal limits. Based on clinical and OCT findings, the diagnosis of acquired retinoschisis was made. The patient was put on oral acetazolamide (Diamox 250 mg twice daily) for 2 weeks after which her visual acuity improved to 6/18 in right eye.

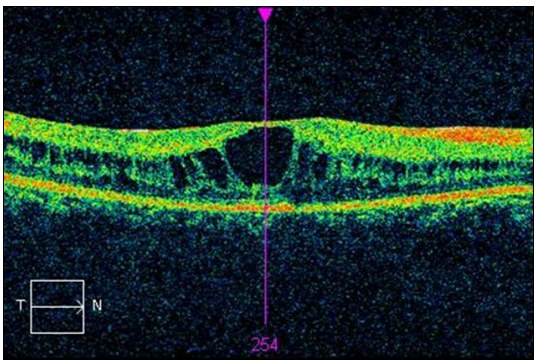

Fig 1: Optical coherence topography showing cystic spaces in both inner and outer macular retina in right eye.

Fig 2: Inferior retinoschisis of the right eye involving the macula.

Case 2

A 45 year old male patient was examined in outpatient department (OPD) of Regional Institute of Ophthalmology (RIO), Rajendra Institute of Medical Sciences (RIMS), Ranchi with chief complaints of gradual diminution of vision in left eye since 1 year and 6 months. His best corrected visual acuity was 6/6 in right eye and 6/60 in left eye. Colour vision assessed with Ishihara plates, intraocular pressure measured by non-contact tonometry, and anterior segment examined by slit-lamp biomicroscopy was normal in both eyes. There was no history of ocular trauma or associated secondary disorder. On dilated fundoscopy, left fundus demonstrated cartwheel-like appearance near macula and dendritic filariform structures in the inner layer of the schisis. OCT of the left eye showed dilated cystic spaces in the inner nuclear and outer plexiform layers of the retina. On the basis of above findings, acquired retinoschisis was diagnosed.

Discussion

Clinical diagnosis and disease progression monitoring has been greatly improved using Spectral Domain OCT. An OCT study has shown that visual acuity is correlated with outer segment thickness and not the total retinal thickness. Other investigation modalities such as fundus autofluorescence, fluorescein angiography, ERG, electro-oculography, colour vision and visual field test is useful but has limited diagnostic value. The use of carbonic anhydrase inhibitor (CAI) has been reported in a few studies. The results show that CAI was able to promote resolution of cystic fluid in the fovea and maintain stability of the disease. However the respond is not seen in all patients and the mechanism of action is still unclear. CAI may be used as a non-invasive management; however more studies are needed to determine its role as a treatment option.

Literature suggests that acquired retinoschisis progresses to a more serious retinal concern in about 15% of cases.[1] If retinoschisis does progress to a retinal detachment, it is difficult to repair. Clinical presentation of the detachment determines which type of retinal procedure is used, but a scleral buckle, and/or pars plana vitrectomy are most commonly performed. Up to 40% of the cases require a second surgical repair.

Apushkin *et al.* [11] postulated that a reduction in visual acuity was not associated with the size of the cystic areas or macular thickness; this is in line with Gerth *et al.* [12] who found that retinal layer abnormalities did not correlate with visual acuity. On the other hand, Yang *et al.* [13], in a retrospective study of 20 eyes with XLRS, concluded that microstructure defects of the outer plexiform layer are frequent in XLRS and may be closely related to poor visual acuity in such patients. Other authors supported a potential mechanism for visual loss similar to age-related macular degeneration.

Conclusion

During the past two decades, there have been extensive advancements in the understanding of all types of retinoschisis at the clinical, molecular, genetic and cellular level. The role of OCT in early diagnosis is crucial. The clinical presentation of this patient and understanding of retinal anatomy is what prompted the diagnosis. Patients who are not managed appropriately are at risk for permanent visual loss. Therefore, it is important that ophthalmologist be well-versed in the newest treatments and theories surrounding this pathology.

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