

Isolated foveo-macular Schisis: A series of 4 cases

Somak Mazumdar, Tushar K Sinha, Shubhendu K Boral, Arnab Das

Abstract: Congenital retinoschisis is characterised by foveo-macular schisis with peripheral retinoschisis in 50% patients. Isolated foveo-macular schisis is an entity which is not so common. These patients need thorough retinal evaluation along with systemic evaluation. They also require proper management as they frequently suffer from significant visual loss which is often uncorrectable. Here we report 4 (four) cases of bilateral congenital foveo-macular schisis without any peripheral retinoschisis with various modes of presentations. Spectral (fourier) domain Optical Coherence Tomography (SD-OCT) was done at primary presentation and all subsequent follow ups. The purpose of reporting these cases is to find out the different anatomical changes of macula and managements accordingly.

Key words: foveo-macular schisis, spectral (fourier) domain Optical Coherence Tomography, central foveal thickness

Case 1

An 18 year old male presented with dimness of vision of both eyes since childhood which is progressive in nature. No systemic illness was associated. On examination, his best corrected visual acuity (BCVA) was 6/12, N6 and 6/24, N8 in right and left eye respectively. Intra ocular pressure (IOP) was 15 & 14 mm Hg in right and left eye respectively. Retina was attached all throughout. Cystic changes observed at both maculas including fovea. SD-OCT showed cystic spaces at fovea extending up to total macular region. The cysts appeared at outer plexiform (OP) to outer nuclear (ON) layers. Central foveal thickness was 426 and 482 micron respectively in right and left eye. No peripheral retinoschisis was detected. The patient was advised glasses, short term topical NSAID and follow up at 3 months interval.



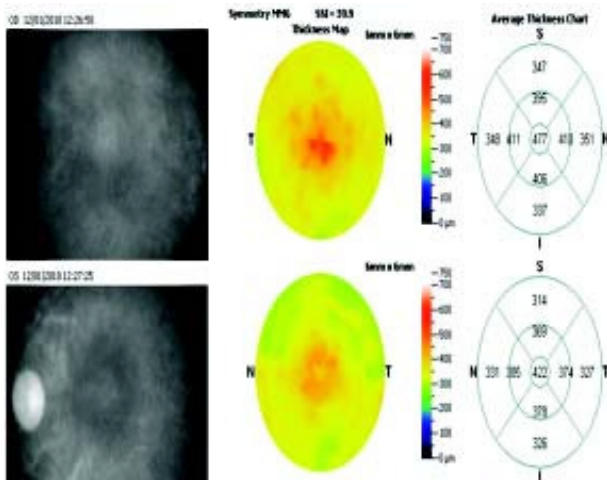
Address for correspondence:

Dr. Somak Mazumdar,
Disha Eye Hospitals & Research Centre,
Barrackpore, Kolkata - 700120, India.

Case 1: Right and left eye Fundus photographs

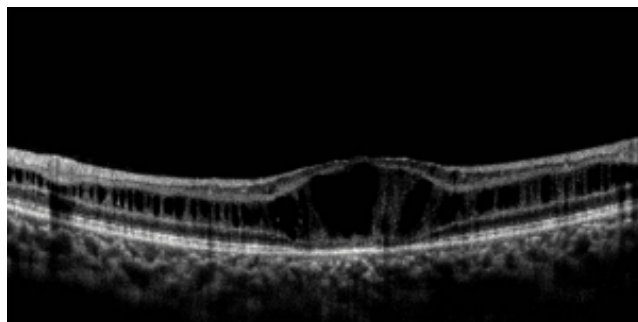
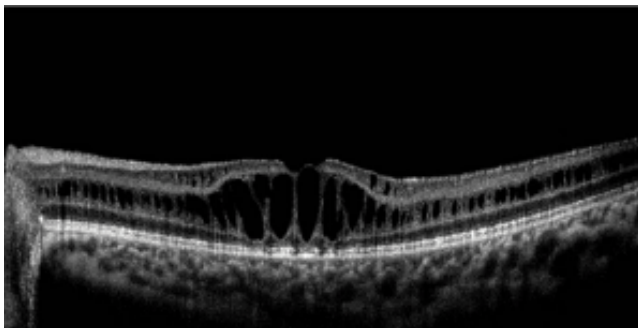
Case 2

A 12 year old male presented with profound dimness of vision in the right eye since childhood which is progressive in nature. No systemic illness was associated. On examination, BCVA was 6/60, N10 and 6/12, N8 in right and left eye respectively. IOP was 14 & 10 mm Hg in right and left eye respectively. Retina was attached all throughout.



Case 2: Central Foveal thickness

Diffuse cystic changes observed at both maculas including fovea. Additionally vitreous floater was detected in the left eye. No peripheral retinoschisis was detected. SD-OCT showed large cystic spaces at fovea involving OP to



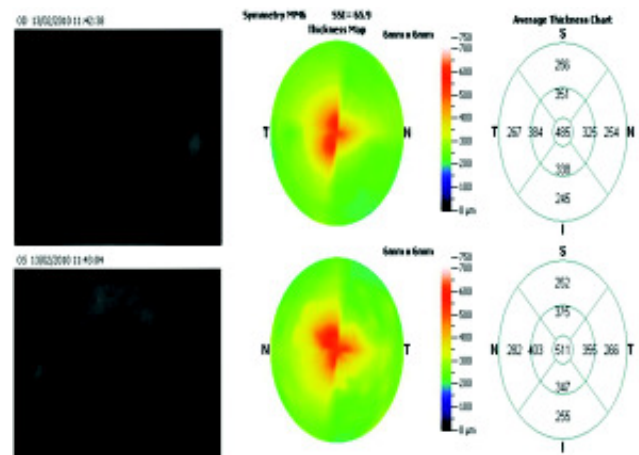
Case 2: OCT right eye and left eye

photoreceptor layer. Parafoveal involvement was less severe, confined at inner nuclear (IN) layer only. Central foveal thickness was 477 and 422 micron respectively in right and left eye. But interestingly, though anatomical changes were almost same in both the eyes, vision impaired markedly in the right eye.

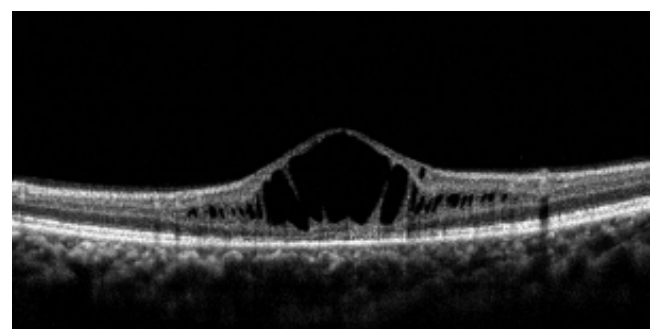
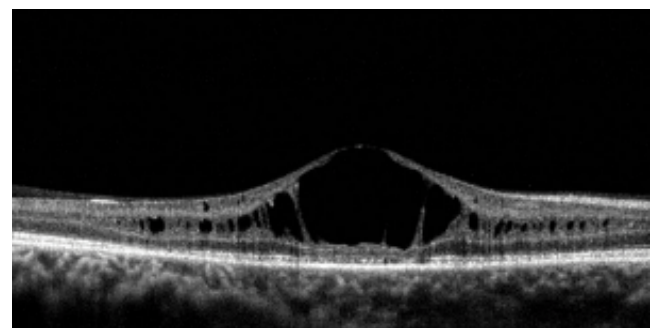
This patient was also advised short term topical NSAID with antioxidants and follow up at 3 months interval.

Case 3

A 38 year old male presented with dimness of vision in the both eyes since childhood which is progressive in nature.



Case 3: Central Foveal thickness



Case 3: OCT right eye and left eye

No systemic illness was associated. On examination, BCVA was 6/18, N8 in both eyes. IOP was 14 & 13 mm Hg in right and left eye respectively. Retina was attached all throughout. RPE changes observed at both maculas including fovea. Shiny foveal reflex was detected in both the eyes. This case was diagnosed as bilateral Stargardt's disease.

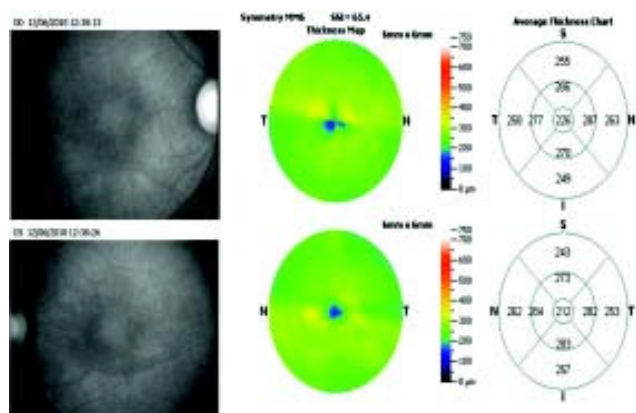
SD-OCT showed a very large cyst at fovea with impending macular hole. No vitreomacular traction was detected. Parafoveal small cystic changes extended hardly beyond 500 micron from the centre of the fovea and involved IN-OP layer. CFT was 485 and 511 micron respectively in right and left eye.

This patient was advised glasses as he was a school teacher. Short term topical NSAID with antioxidants and follow up at 3 months interval was also advised.

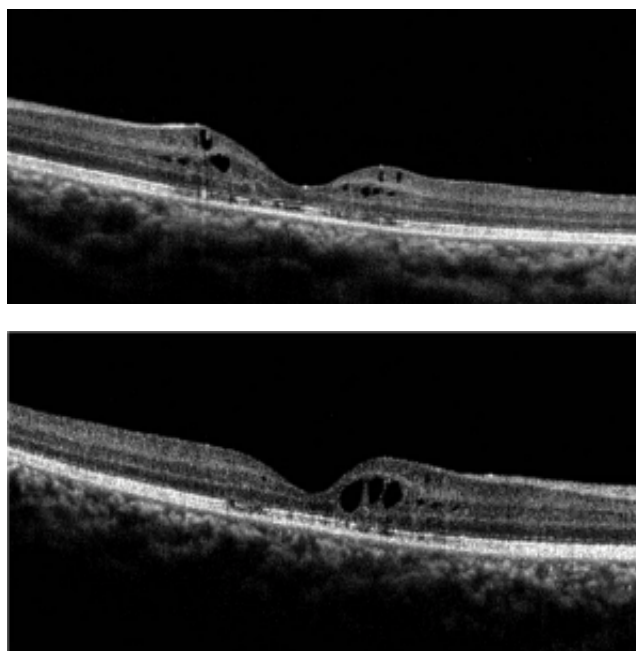
Case 4

The last case was a 19 year old male presented with dimness of vision in the left eye since childhood which is progressive in nature. No systemic illness was associated. On examination, best corrected visual acuity (BCVA) was 6/9, 6/12 and N6 in right and left eye respectively. IOP was 19 mm Hg in both the eyes. Retina was attached all throughout. Foveal reflex was good in both the eyes. No peripheral retinoschisis was detected. To know the cause of visual loss, SD-OCT was done. It showed few cysts at ganglion cell layer-ON layer at parafoveal region. Proper foveal architecture was maintained. Mild parafoveal disruption of photoreceptors detected. CFT was rather less, 226 and 212 micron respectively in right and left eye.

This patient was advised glasses, short term topical NSAID and follow up at 3 months interval.



Case 4: Central Foveal thickness



Case 4: OCT right and left eye

Discussion

Retinoschisis was not noted in any case. All patients were male with age ranged 12-38 years. In our series, isolated foveo-macular schisis presented with mild to severe dimness of vision. Previous literatures lack to produce visual disturbances. In our series visual acuity ranged from 6/9 (log MAR 0.2) to 6/60 (log MAR 1.0) with an average of 6/20 (log MAR 0.52). Near vision ranged from N6 to N10. IOP was normal in all four cases. Schisis cavity was almost universally found in all cases previously reported and involved RNFL to outer nuclear layer. In our series, spoke like schisis involved ganglion cell layer to outer nuclear layer. Both diffuse and localised pattern were observed. Retinal thickening and atrophy both were observed previously. In current series, 75% eyes have retinal and macular thickening. The central foveal thickness ranged from 422-511 micron with an average of 475.4 micron. Only 25% cases have early foveal thinning. Photoreceptors were found disrupted and irregular. But RPE was within normal limit in all 4 cases. Retinal detachment was not found in any case.

Vitreous floater was detected in one case which may be a sign of degeneration. More retinal thickening definitely produced more visual impairment, but not directly correlated with visual acuity.

Vision impairment is probably due to photoreceptor damage and further augmented by retinal thickening with

loss of foveal architecture. SD-OCT clearly finds out the anatomical changes and this is a very important tool which perhaps rules out the use of fundus angiography. Management consists of glasses, short term topical NSAID and regular followup. More advanced stages may need low visual aids.

To conclude, our series highlights isolated foveo-macular schisis with different anatomical changes at macula. This may be partially treatable and require long term follow-ups preferably under SD-OCT findings.

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