Stellate Nonhereditary Idiopathic Foveal Retinoschisis: A Case Report

Abstract:
59 year-old male presents with asymmetric bilateral stellate foveal schisis with a mild reduction in visual acuity. Further optometric testing elucidates a rare case of stellate nonhereditary idiopathic foveal retinoschisis (SNIFR).

Case History:
Patient demographics:
59 year-old African American male
Chief complaint:
Distorted and blurred vision that is worse in the right eye compared to the left eye. Patient reports that his vision appears as if he is looking through a “drop of water” for the last six weeks.

Ocular History:
The ocular history is significant for nuclear sclerotic cataracts.

Medical History:
The medical history is positive for a stroke in 2003 and a pituitary adenoma in 2003.

Medications:
Amlodipine, Carvedilol, Lisinopril, Omeprazole

Pertinent findings:
Clinical:
Best-corrected visual acuity is 20/30 OD, OS. Slit lamp exam showed bilateral 3+ nuclear sclerotic cataracts. Dilated fundus exam revealed a stellate macular appearance OD>OS.

Diagnostic Testing:
Fundus Photography OU: Shows a stellate macular appearance OD>OS.

Fundus Auto-Fluorescence (FAF) OU: Enhances the stellate macular appearance OD>OS.

Spectral Domain Optical Coherence Tomography (SD-OCT) OU: Shows separation of the neurosensory retina at the level of the outer plexiform layer (OPL) OD>OS.

Differential Diagnosis:
SNIFR (primary), congenital X-linked retinoschisis (CXL), tractional retinoschisis, degenerative retinoschisis, and enhanced S-cone syndrome.¹

Diagnosis:
SNIFR
Treatment, management:
Referral to a retinal specialist to clear the patient for cataract surgery. Patient education and close observation with SD-OCT for the development of subretinal fluid and potential serous retinal detachment.\(^1,9\)

Discussion:
SNIFR is a unilateral and less commonly asymmetric bilateral stellate foveomacular retinoschisis that presents with little to no symptoms. With limited case examples there appears to exist a female predilection, with increased susceptibility possibly attributable to gender differences in retinal thickness.\(^11\) Visual acuity is relatively spared ($\geq 20/40$) unless subretinal fluid develops.\(^1,9\) All known cases of SNIFR involve the separation of the neurosensory retina at the level of the OPL with lesser involvement of the outer nuclear layer (ONL).\(^1\) Fluorescein angiography (FA) reveals no leakage or staining.\(^1\)

There are many other causes of macular retinoschisis, each with their own subtle nuances. These include CXL\(^2\), tractional retinoschisis\(^3\), degenerative retinoschisis\(^4\), and enhanced S-cone syndrome\(^5-7\). CXL\(^2\) is a hereditary retinoschisis linked to a defect in the \(RS1\) gene and presents as foveal retinoschisis with approximately half of patients developing concurrent peripheral retinoschisis. Visual acuity often begins to deteriorate in the first to second decade of life and stabilizes between 20/60 to 20/120.\(^2\) Although genetic testing for defects in the \(RS1\) gene was not pursued in this case, there was no known hereditary or acquired predisposition warranting testing. In CXL\(^2\) SD-OCT shows that the neurosensory retina is typically split at the level of the inner nuclear layer (INL), with less common involvement of the OPL.\(^8\) Further, the significant asymmetry of the findings in this report make the likelihood of a hereditary component low.

Tractional retinoschisis can result from multiple mechanisms, including vitreomacular traction after incomplete posterior vitreous detachment (PVD), epiretinal membrane, and residual cortical vitreous after PVD.\(^10\) This would be obvious on SD-OCT as changes would be seen at the ILM/posterior hyaloid interface and these changes are often associated with a myopic fundus. In this case the myopic status is attributable to lenticular changes and not axial length, with the patient nearly emmetropic prior to his lenticular changes.

Degenerative retinoschisis involves a split of the neurosensory retina at the OPL but is almost always isolated to the inferior temporal retina with rare cases involving the posterior pole.\(^4,13\) Further, bilateral retinal involvement is common in degenerative retinoschisis but there are no known reports of bilateral macular involvement.

Enhanced S-cone syndrome is an autosomal recessive disorder with foveomacular retinoschisis but is also characterized by progressive night blindness, complicated cataracts, and retinal pigment epithelial (RPE) changes.\(^5-7\) Further, SD-OCT shows severe degeneration with a splitting just above the photoreceptor layer.\(^6\)

SNIFR usually requires no active management as patients commonly have only a mild reduction in visual acuity and present with no symptoms. Reported cases of subretinal fluid and subsequent serous retinal detachment have been reported, however.\(^1,9\) Unsurprisingly visual acuity was markedly reduced in these cases. Pars plana vitrectomy with internal limiting membrane (IML) peel has shown success in a limited sample size of patients with reduced visual acuity from SNIFR.\(^9\) This efficacy is
attributable to acceleration of the diffusion of serous fluid from the retina to the vitreous. Caution should be employed for those who have retinoschisis and are seeking cataract surgery. Reported cases of complications, including retinal detachment, following cataract surgery have been noted.

**Conclusion:**

SNIFR is a unilateral and less commonly an asymmetric bilateral stellate foveomacular retinoschisis that presents with near normal visual acuity (≥20/40) and splitting of the neurosensory retina at the OPL. Primary eye care providers should differentiate SNIFR from other hereditary and nonhereditary forms of retinoschisis as they have different presentations and visual outcomes. Individuals should be monitored regularly with OCT to rule out development of subretinal serous fluid and subsequent serous retinal detachment.

**References:**