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Case report

Stellate nonhereditary idiopathic foveomacular retinoschisis resolution after vitreomacular adhesion release

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STRUCTURED ABSTRACT

Purpose: To present a case of stellate nonhereditary idiopathic foveomacular retinoschisis (SNIFR) resolution associated with vitreomacular adherence (VMA) release and propose a potential contributing association between SNIFR and vitreomacular interactions.

Observations: A 67-year-old female patient was diagnosed and followed for SNIFR in OD with spectral-domain optical coherence tomography (SD-OCT) scans at presentation and subsequent visits at 3, 6, 16 and 22 months. VMA and foveomacular retinoschisis remained unchanged on SD-OCT during the first 6 months of the follow-up. At 16-month follow-up visit, SD-OCT revealed VMA release and an important improvement of the macular schisis. At 22 months of follow-up, SNIFR cavities completely resolved in the presence of posterior hyaloid separation from the macular area without any adjunct treatment. The authors could not identify any other possible cause to justify the resolution of SNIFR other than VMA release in this case. Patient did not undergo any treatment for OD other than phacoemulsification 3 months after initial visit.

Conclusion: The present case illustrates with SD-OCT scans a possible association between SNIFR resolution and VMA release, highlighting a potential tractional component of the posterior vitreous on the internal limiting membrane and consequent glial cells stretching with schisis formation.

Summary statement

Vitreomacular adhesion release may be associated to the anatomical resolution of stellate nonhereditary idiopathic foveomacular retinoschisis.

Ethics

Ethics Committee advice is not required for the routine medical care of patients.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Introduction

Stellate nonhereditary idiopathic foveomacular retinoschisis (SNIFR) was first described in 2014 by Ober et al.[1] and its pathogenesis remains to be clarified. Previous reports suggested that macular retinoschisis could be associated to vitreous traction in eyes with fragile nerve fiber layer[2] and that the stellate pattern observed in SNIFR could be associated with the lack of structural support provided by local blood vessels in the Henle fiber layer.[3]

We report a case of SNIFR with spontaneous resolution after vitreomacular adherence (VMA) release.

Case report

A 67-year-old white woman was referred to Ophthalmology Department of Maisonneuve-Rosemont Hospital, University of Montreal (Montreal, QC, Canada) in March 2016 for foveomacular retinoschisis in OD and vitreomacular traction (VMT) syndrome in OS. She had a

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previous medical history remarkable for type 2 diabetes diagnosed 2 years before the referral, hypertension and hypothyroidism. Patient refraction was +4.50 in OD and +3.00 + 0.50 × 150° in OS. Best corrected visual acuity was 20/40 in OD and 20/25 in OS. Slit-lamp biomicroscopy revealed a grade 2 nuclear cataract in OD and mild nuclear sclerosis in OS. Fundus evaluation showed radial spoking around the fovea in OD and suspicion of VMT in OS. No signs of diabetic retinopathy were identified.

On SD-OCT scan, OD presented retinal splitting at the level of Henle fiber layer (HFL) with bridging tissue separating cystic spaces in the fovea and broad vitreomacular adhesion (Fig. 1A). HFL en face slab of OD evidenced the stellate appearance of the macula with alternating radial hyporeflective and hyperreflective areas (Fig. 2A). SD-OCT scan of OS confirmed the presence of VMT which was treated with combined phacoemulsification and pars plana vitrectomy during the follow-up (Fig. 1B, 1D, 1F, 1H and 1J).

The diagnosis of SNIFR was stablished after an extensive investigation and exclusion of other causes of foveomacular retinoschisis. The patient had no personal or familial risk factors for congenital juvenile X-linked retinoschisis (CXLR) and as such RS1 mutation was not investigated. Patient denied previous use of niacin or taxane derived drugs.

Three months after initial presentation the patient underwent routine phacoemulsification. Her post-operative BCVA improved to 20/25 in OD. Fundus examination and SD-OCT remained unchanged in OD (Fig. 1E) at her 6-months retina follow-up. No specific treatment for the SNIFR was recommended as visual acuity was satisfactory and patient was asymptomatic. Sixteen months after initial referral, VMA release and an important improvement of the macular schisis were observed (Fig. 1G). At 22-months follow-up, total resolution of SNIFR with the reabsorption of residual intraretinal fluid was detected in OD (Fig. 1I). BCVA remained at 20/25 in OU without metamorphopsia or other visual symptoms. Patient denied any other eye surgeries, use of carbonic anhydrase inhibitors[4] or any other plausible cause to justify the schisis resolution in OD apart from observed VMA release.

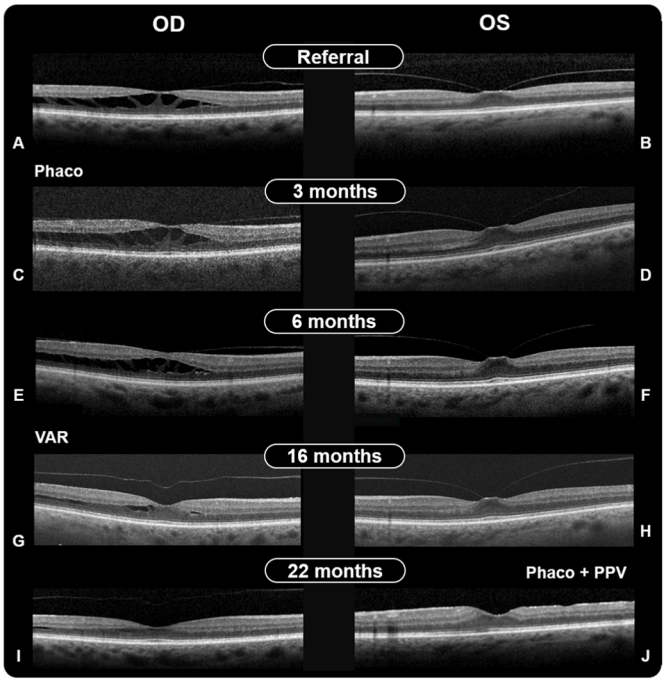

Fig. 1. Foveal scans of both eyes at different periods of the follow-up. VMA release occurred 16 months after initial referral and coincided with retinoschisis improvement – further residual fluid reabsorption and complete schisis resolution was observed in the subsequent OCT. VMT was observed in OS and was treated with Phaco + PPV.

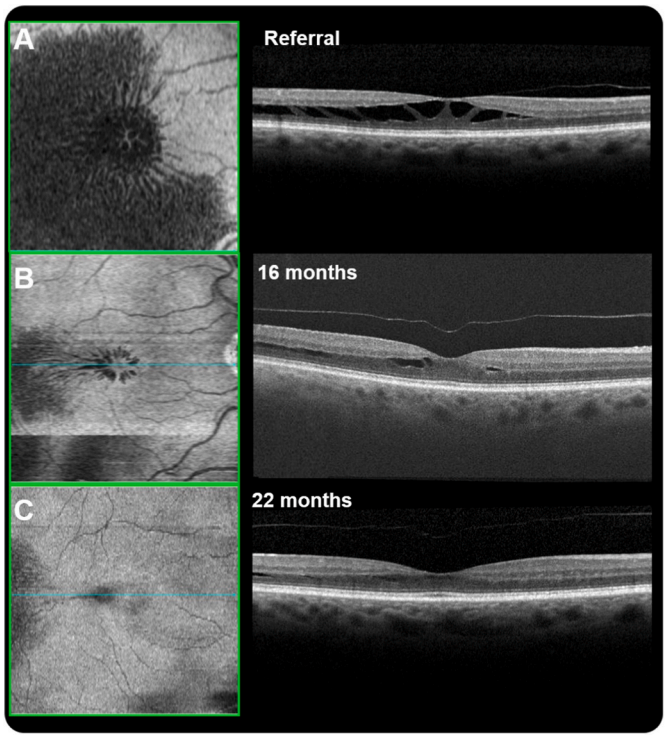

Fig. 2. En face and cross-section SD-OCTs at different periods of follow-up. Initial scans showed the classic spoke wheel aspect of the HFL splitting. An important improvement of the schisis was observed after VMA release at 16-monthfollow-up visit. Complete resolution of SNIFR was detected 22 months after referral.

Discussion

Ober et al.[1] presented a series of 22 eyes from 17 patients with foveomacular retinoschisis without any known hereditary or acquired predisposition and described it as SNIFR. This condition was observed mainly in female (94%) and myopic (72%) patients with unilateral schisis (70%). Interestingly, 19 (86%) of the eyes presented with VMA. SNIFR diagnosis is based on the presence of characteristic radial spoking around the fovea and the splitting of the retina at the level of Henle fiber layer in OCT after the exclusion of other possible causes of retinoschisis. Such conditions include CXLR,[5] myopia,[6] optic disc pit,[7] glaucoma,[8] myotonic dystrophy,[9] enhanced S-cone syndrome[10] and vitreomacular traction.[11]

Other findings in SNIFR include mild radial hypofluorescence in perifoveal region and absence of leakage/staining on fluorescein angiography. BCVA is usually 20/40 or better and metamorphopsia has also been described as a possible symptom.

In this case, other causes of macular retinoschisis were excluded. The presence of VMA associated with SNIFR in OD and VMT in OS may suggest that SNIFR could be an alternative presentation in patients with vitreomacular interface abnormalities. It is also possible that patients with SNIFR might present an underlying susceptibility for retinoschisis caused by normal vitreo-retinal interface adhesion which would not cause retinal splitting in patients without this condition. As visual acuity is usually preserved in SNIFR cases, invasive interventions to promote VMA release would rarely be warranted.

According to the International Vitreomacular Traction Study Group Classification of Vitreomacular Adhesion, Traction, and Macular Hole's Group Classification,[11] OD presented a broad VMA on SD-OCT. Interestingly, a clear spatial correlation of VMA and macular retinoschisis areas was observed (Fig. 3).

This is the first description of SNIFR spontaneous resolution after SD-OCT-documented VMA release to the best of our knowledge. Of note, the

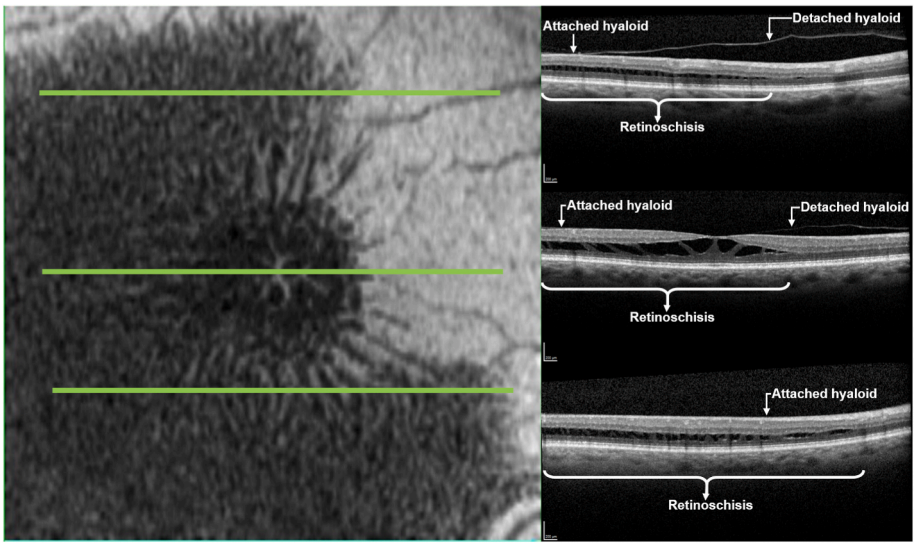

Fig. 3. Initial en face and correspondent tomographical cross sections SD-OCT showing extensive foveomacular retinoschisis. There is a clear spatial correlation of VMA areas and the presence of retinoschisis.

frequent presence of VMA in SNIFR case reports corroborates the hypothesis that VMA may contribute to the pathogenesis and persistence of this kind of foveomacular retinoschisis. A recent OCT angiography study hypothesized that the absence of local vasculature in Henle fiber layer could contribute to the occurrence of retinoschisis due to a limited mechanical resistance offered by the lack of structural support provided by retinal vessels.[3]

The tissue bridging the schisis in SNIFR cases seems to be composed by Müller cells and photoreceptor axons. In his 1999 paper, Gass proposed the hypothesis on the Müller cells function in the ultrastructure of fovea centralis and the pathogenesis of foveomacular schisis. He postulated that the Müller cell cone is the primary structural support for the fovea and that this structure provides an anatomical substrate for schisis to occur.[12] Chung et al. revisited the pathoanatomy of macular holes (MH) with new evidence provided by OCT and described an antero-posterior focal vitreous traction on Müller cell cone in the foveola as a mechanism to explain the so-called 'stage 0' MH.[13] A recent publication proposed a physical model of mechanical force transmission for the Z-shaped parafoveal Müller cells in tractional macular disorders and found an association between the grade of verticalization of the horizontal processes of this cells and the BCVA.[14] The visual acuity in SNIFR patients could be majorly preserved due to a lower mechanical stress transmitted from the Müller cells to the photoreceptors which might explain a milder verticalization of Henle fibers when compared to other conditions in which vitreo-retinal interface mechanical forces are higher such as myopic foveoschisis.

The patient presented here demonstrated a broad VMA which could explain a more diffuse mechanical vitreous force on the macula possibly leading to schisis formation. Interestingly the fellow eye displayed a focal VMA adhesion and progression to VMT. Retinal splitting might be a result of the resistance of the retina to the tractional force promoted by VMA.[15]

Similarly to MH, the cysts in SNIFR present a stellate arrangement in the HFL. This could be due to the reduction of the interstitial tissue pressure of the retina secondary to the tractional force exerted by the vitreous possibly leading to fluid influx from the retinal vasculature.[15,16] This coincidence of cystic patterns seems to be linked to Müller cells radial distribution in the parafovea where they present a Z-shaped course, spanning through the HFL in an almost horizontal orientation. In fact, a recent study by Govetto et al.[17] analyzed intraretinal cystoid spaces characteristics in tractional macular edema and described a spoke-wheel pattern of the cysts observed in the en face SD-OCT of different tractional macular disorders which is consistent with the one presented in this case (Fig. 3). Furthermore, the presence of hyporreflective cysts only at the level of HFL, as seen in this case, was more frequent in macular edemas caused by traction when compared to those caused by exudation which portrayed hyporreflective cystoid spaces in both INL and HFL-ONL.[17] In addition, it is hypothesized that a dysfunction in Müller cells hydro-ionic regulation could play a role in the genesis of such cysts.[16]

A previous report of SNIFR combined with macular telangiectasia type 2 (MacTel) could not stablish an etiologic role of the latter in SNIFR[18] even though one could speculate a possible correlation of Müller cell degeneration in MacTel type 2 and structural and hydro-ionic alterations in outer retina in retinoschisis.

Further studies are needed to investigate the possible correlation between SNIFR and VMA as well as the possibility of retinoschisis resolution after VMA release as observed in our case.

In conclusion, the here presented case of foveoschisis resolution after VMA release seems to indicate that, in fact, SNIFR could be an alternative presentation of broad VMA which may be associated with an underlying susceptibility to a structural breakdown from what would otherwise be considered normal vitreous adhesion.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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