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Spontaneous Resolution and Asynchronous Onset of Stellate Nonhereditary Idiopathic Foveomacular Retinoschisis in the Contralateral Eye

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Abstract

Purpose: To present a patient with stellate nonhereditary idiopathic foveomacular retinoschisis found with multimodal imaging. **Methods:** A single case was evaluated. **Results:** A 70-year-old woman presented with schisis cavities in the outer plexiform layer in the left eye on macular optical coherence tomography. No leakage was detected by fluorescein angiography. After ruling out other causes of nonvasogenic cystoid maculopathy, the patient was diagnosed with stellate nonhereditary idiopathic foveomacular retinoschisis. After 5 years, the best-corrected visual acuity remained stable. The retinoschisis in the left eye had resolved, and cystoid spaces appeared in the lower portion of the macula in the right eye despite the absence of vitreomacular traction. There was no history of ocular procedures or medication use that could have affected the course of the pathology during the intervening period. **Conclusions:** This case emphasizes the importance of performing multimodal imaging to monitor the characteristics of stellate nonhereditary idiopathic foveomacular retinoschisis as they evolve.

Keywords

cystoid spaces, foveoschisis, nonvasogenic macular edema, multimodal imaging, stellate nonhereditary idiopathic foveomacular retinoschisis

Introduction

Stellate nonhereditary idiopathic foveomacular retinoschisis is characterized by a star-shaped appearance of the fovea on funduscopy and schisis at the level of the outer plexiform layer (OPL) on optical coherence tomography (OCT).[1] The disease is diagnosed by the exclusion of imaging findings unexplained by other causes of foveomacular retinoschisis, such as X-linked congenital retinoschisis, myopic tractional maculopathy, optic nerve pit maculopathy, degenerative retinoschisis, or vitreomacular traction (VMT).[1–5]

Peripheral schisis is reported in up to 50% of patients with stellate nonhereditary idiopathic foveomacular retinoschisis.[6] Furthermore, the disease may be observed in the contralateral eyes without evidence of foveomacular schisis, and partial posterior vitreous detachment (PVD) is common[6,7] (Table 1).

Stellate nonhereditary idiopathic foveomacular retinoschisis is an uncommon condition, and there is limited understanding of its underlying pathophysiologic mechanisms. Previous hypotheses have focused on the role of vitreous traction.[7] Treatment options include vitrectomy or the application of topical dorzolamide in cases in which the patient is symptomatic or when the visual acuity (VA) is affected.[4]

Here, we present a patient with stellate nonhereditary idiopathic foveomacular retinoschisis in the left eye that spontaneously resolved and subsequently appeared in the contralateral eye during long-term follow-up.

Case Report

A 70-year-old woman with no significant medical history was referred to our department for cataract surgery. Preoperative macular OCT imaging of the left eye showed a partial PVD and schisis cavities in the OPL without vitreomacular adhesions (Figure 1A). At presentation, the patient's best-corrected VA (BCVA) was 20/60 OU. A slitlamp examination of the left eye showed grade 2 nuclear lens opacity, and a dilated fundus examination showed a decreased foveal reflex and bilateral peripheral

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Table 1. Main Clinical and Imaging Characteristics in Stellate Nonhereditary Idiopathic Foveomacular Retinoschisis.

Clinical/Imaging Modality	Key Findings
Symptoms	Asymptomatic or metamorphopsia
Visual acuity	20/20-20/30; usually better than 20/50
Color photograph/fundus examination	Blunted foveal reflex; apparent foveal elevation; foveal cysts in a stellate configuration
Infrared imaging/FAF	Foveal stellate configuration (more evident than clinically)
Fluorescein angiography	Absence of leakage at the macula; might have slight leakage at the optic disc
Optical coherence tomography	Increased central foveal thickness with foveomacular schisis cavities at the level of the HFL/OPL; might have SRF or peripheral schisis
Electroretinography	Normal photopic and scotopic responses
Genetic testing	Invariably negative

Abbreviations: FAF, fundus autofluorescence; HFL, Henle fiber layer; OPL, outer plexiform layer; SRF, subretinal fluid.

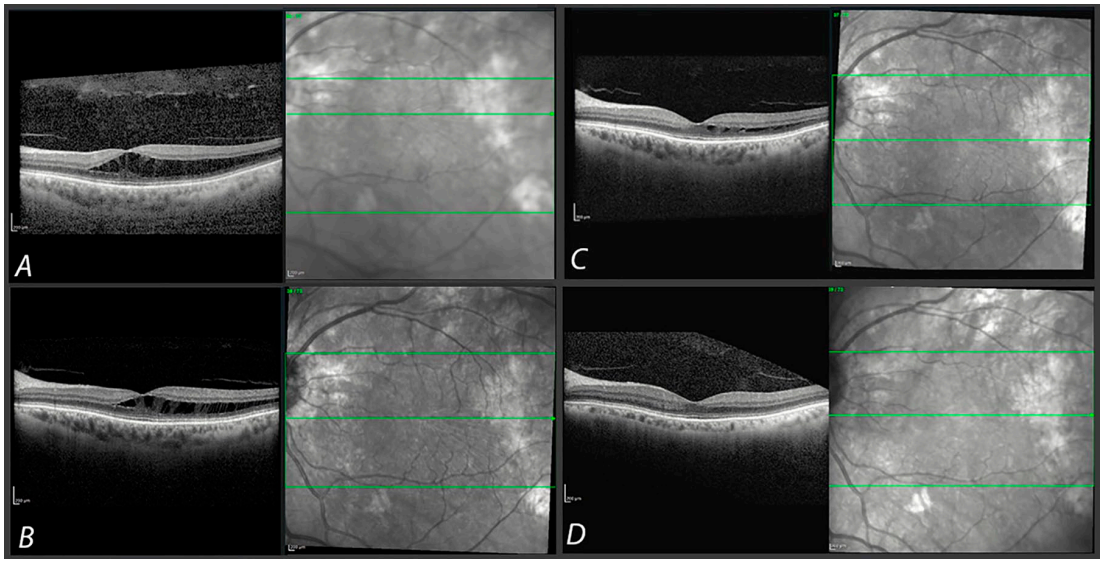

Figure 1. Macular optical coherence tomography of the left eye at presentation. (A) The presence of predominantly temporal foveoschisis without vitreomacular adhesion. (B–D) Progressive resolution of schisis over time (at 6 months, 12 months, and 18 months, respectively).

hypopigmentation with overlying peripheral schisis (Figure 2A). The right eye had a grade 2 nuclear, grade 1 cortical cataract with no other findings (Figure 3A).

Autofluorescence imaging of the left eye showed radial changes around the fovea (Figure 2B), and mild transmission hyperfluorescence was seen on fluorescein angiography (FA). However, no leakage was detected during the early or late phase (Figure 2, C and D).

Interferometry measurements found an axial length (AL) of 23.04 mm OD and 23.02 mm OS, confirming that the patient was emmetropic. Genetic causes of schisis were unlikely given the late onset of symptoms and the lack of family history. Electroretinography testing showed normal photopic and scotopic responses, eliminating X-linked congenital retinoschisis as a potential cause. A genetic panel analysis for hereditary retinal diseases was also negative.

The patient denied taking drugs that included niacin or taxanes. After other causes of nonvasogenic cystoid maculopathy were ruled out, the diagnosis of stellate nonhereditary idiopathic foveomacular retinoschisis was confirmed.

During the patient's regular follow-up, a progressive reduction in the schisis cavities in the left eye was seen (Figure 1, B–D). Notably, the resolution of schisis occurred without changes to the vitreomacular interface. Phacoemulsification with intraocular lens implantation was performed in the right eye, and the BCVA improved to 20/25 postoperatively.

After a 5-year interval, the patient returned for a follow-up examination with no history of topical or systemic carbonic anhydrase inhibitor use or ocular procedures that could have affected the course of the pathology during the intervening period. Although the BCVA remained stable and macular OCT imaging showed resolution of the retinoschisis in the left eye, cystoid spaces were observed in the lower portion of the macula in the right eye (Figure 3B). No leakage in the macula during the early phase and late phase in either eye was seen on wide-field FA (Figure 3, C and D).

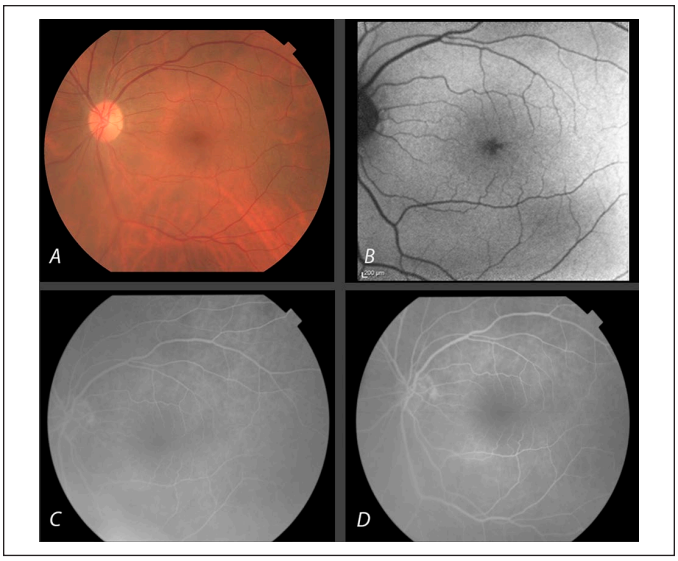

Figure 2. (A) Color photograph of the left fundus shows a decreased foveal reflex. (B) Radial pattern of foveal autofluorescence. Fluorescein angiography shows a mild transmission hyperfluorescence; however, there is no leakage during the (C) early phase and (D) late phase.

Thus, without intervention, the patient's condition in the left eye improved but subsequently developed in the right eye, with an apparently self-limiting course.

Conclusions

Stellate nonhereditary idiopathic foveomacular retinoschisis mainly occurs in women in their 70s and is often asymptomatic; however, in some cases it may cause slight vision loss or metamorphopsia.[2] Although the disease typically occurs unilaterally, it can be bilateral.[3] Its pathophysiology remains unclear; however, the absence of leakage on FA and the poor response to treatment with antivascular endothelial growth factor suggest that it is not caused by exudative maculopathy with classic cystoid spaces at the OPL level.[1]

Because partial PVD has been reported in approximately 86% of affected eyes and 42% of unaffected fellow eyes, several studies have suggested an association between stellate nonhereditary idiopathic foveomacular retinoschisis and VMT.[6] Foveomacular retinoschisis could result from vitreous traction in eyes with a fragile nerve fiber layer, and the characteristic stellate pattern could be the result of a lack of structural support from local blood vessels in the Henle layer.[1,7] These hypotheses are supported by reports of stellate nonhereditary idiopathic foveomacular retinoschisis resolving after the spontaneous resolution of VMT in the foveal region.[6,7]

Unlike other entities, the morphology of the schisis in stellate nonhereditary idiopathic foveomacular retinoschisis is dynamic, with changes in retinal thickness associated with the Valsalva maneuver.[1] The stellar appearance of the fovea is best observed using infrared imaging; OCT shows increased central thickness with cystoid spaces at the OPL, whereas FA confirms the absence of macular leakage.[5] No abnormalities in stellate nonhereditary idiopathic foveomacular retinoschisis are seen on electroretinography, helping to distinguish the disease from genetic conditions or dystrophies associated with foveomacular schisis. Furthermore, negative results of genetic studies for retinal dystrophies, most commonly for the *RS1* gene, can help with the diagnosis when it is uncertain.[1–6]

In our patient, there was no adherence of the posterior vitreous to the fovea and no changes during the follow-up. This is in contrast to the findings in other cases in which resolution of VMT occurred with the subsequent improvement of stellate nonhereditary idiopathic foveomacular retinoschisis.[6,7] Because our patient never reported decreased VA or metamorphopsia, we chose to keep her under observation.

The patient had cataract surgery with a subsequent improvement in BCVA. As previously reported, cataract surgery in patients with stellate nonhereditary idiopathic foveomacular retinoschisis does not significantly affect the macular status.[8] Although evidence is lacking to confirm the safety of cataract surgery in patients with stellate nonhereditary idiopathic foveomacular retinoschisis, to date no associated adverse events have been reported.[8]

Similar to the current case, most patients with stellate nonhereditary idiopathic foveomacular retinoschisis are asymptomatic or minimally symptomatic, have a BCVA of 20/40 or better, and are typically monitored without treatment. However, topical carbonic anhydrase inhibitors can be administered to patients with decreased vision or significant symptoms. Ajlan and Hammamji[9] reported improvement in a 27-year-old patient with stellate nonhereditary idiopathic foveomacular retinoschisis treated with dorzolamide hydrochloride eyedrops. However, evidence supporting the routine use of topical dorzolamide is insufficient.[5]

Maruko et al[2] reported using vitrectomy with internal limiting membrane peeling to successfully attain morphologic and visual improvements in patients with vision loss caused by idiopathic foveomacular retinoschisis. We believe that some patients who undergo other treatment modalities may experience spontaneous resolution of stellate nonhereditary idiopathic foveomacular retinoschisis, as in the current case.

Our patient had documented improvement in the initially affected eye, and a separate, asynchronous onset was observed in the fellow eye, which to our knowledge has not been previously reported. It is important to emphasize the absence of evolving vitreous changes. Previous studies have indicated a correlation between associated VMT and stellate nonhereditary idiopathic foveomacular retinoschisis as well as the subsequent resolution after the alleviation of vitreous traction.[2,6,7] Another previously described hypothesis regarding the development of macular schisis in this disease is related to a disruption of retinal architecture, potentially associated with areas of lesser resistance, which may indicate a dysfunction at the level of Müller cell dendrites.[4] This indicates the potential involvement of additional factors that influence the development and resolution of the pathology and have yet to be clarified.

There is significant variability among the reports of stellate nonhereditary idiopathic foveomacular retinoschisis. On the one

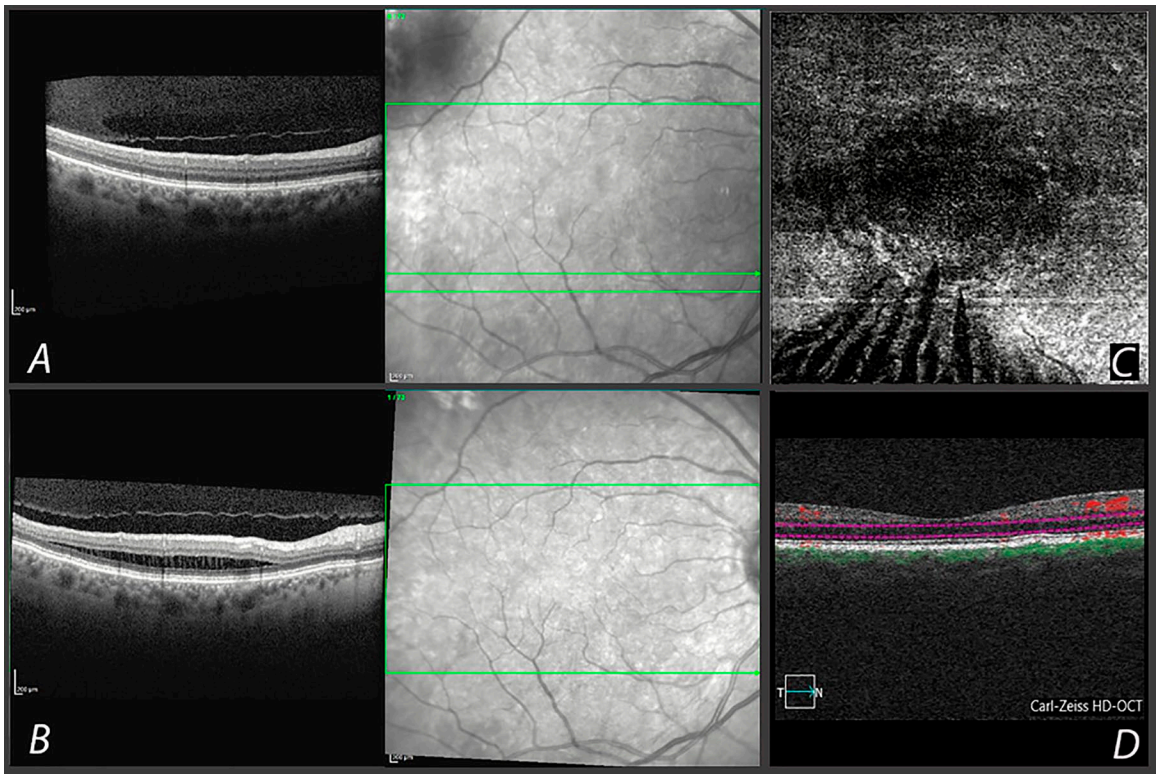

Figure 3. (A) Macular optical coherence tomography (OCT) of the right eye at presentation shows focal vitreomacular adhesion and no retinal alterations. (B) Five years later, the same section shows the onset of foveoschisis. (C and D) En face structural OCT, 3 mm × 3 mm scan with manual segmentation at the level of the outer plexiform layer highlights the extension of the foveoschisis.

hand, the classic description includes the presence of mild myopia and an attached posterior hyaloid.[1] Other studies have reported different findings, such as hyperopia with a shortened AL and the presence of PVD.[2] These discrepancies suggest that stellate nonhereditary idiopathic foveomacular retinoschisis may encompass a spectrum of pathologies rather than representing a singular clinical entity; however, the disease is likely to be underdiagnosed because it has few symptoms and recent descriptions. Therefore, using multimodal imaging during diagnosis and follow-up is essential for excluding other potential causes of foveomacular retinoschisis.

Future investigations should prioritize the acquisition of additional data regarding the pathogenesis of stellate nonhereditary idiopathic foveomacular retinoschisis. In addition, an examination of its natural course in relation to the various therapeutic strategies that have been suggested for its management would be beneficial.

Authors' Note

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Ethical Approval

This study was conducted in accordance with the principles of the Declaration of Helsinki. Ethical approval was not sought because this paper is a case report.

Statement of Informed Consent

Informed consent, including permission for publication of all photographs and images included herein, was obtained before the procedure was performed.

Declaration of Conflicting Interests

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