PROGRESSIVE DEVELOPMENT OF STELLATE NONHEREDITARY IDIOPATHIC FOVEOMACULAR RETINOSCHYSIS AFTER VITREOMACULAR TRACTION SPONTANEOUS RELEASE: MULTIMODAL IMAGES CASE REPORT

Talita Goulart Machado, João Carlos Diniz Arraes, Danielle Ramos Castro, Emanuela Pereira de Holanda Silva, Tatiana Azevedo Arraes.
Report from Hospital de Olhos do Tocantins

INTRODUCTION

Macular retinoschisis is generally associated with X-Linked Retinoschisis (XLR), however, a new form of stellate nonhereditary idiopathic foveomacular retinoschisis (SNIFR) has recently been described in women, unilaterally and without RS1 gene defect or other secondary causes (1).

METHODS

Case report with multimodal images of progressive development of SNIFR after vitreomacular traction (VMT) resolution.

CASE REPORT

In May 2015, a healthful 56-year-old white woman complained of blurred vision in OD. Ophthalmic exam revealed mild hyperopia, macular hole in OD and VMT in OS. Two months later, OCT showed partial posterior vitreous detachment (PVD) with little cystic formation in inferior fovea in OS (Figure 1).

Until May 2019, OS remained asymptomatic (20/25), but OCT showed progression of the macular retinoschisis with increased splits in outer plexiform layer (OPL) and without peripheral retinoschisis. Fluorescein angiography and fundus autofluorescence were normal (Figure 2). Red-free photography and en face OCT evidenced stellate pattern of foveomacular retinoschisis (Figure 3).

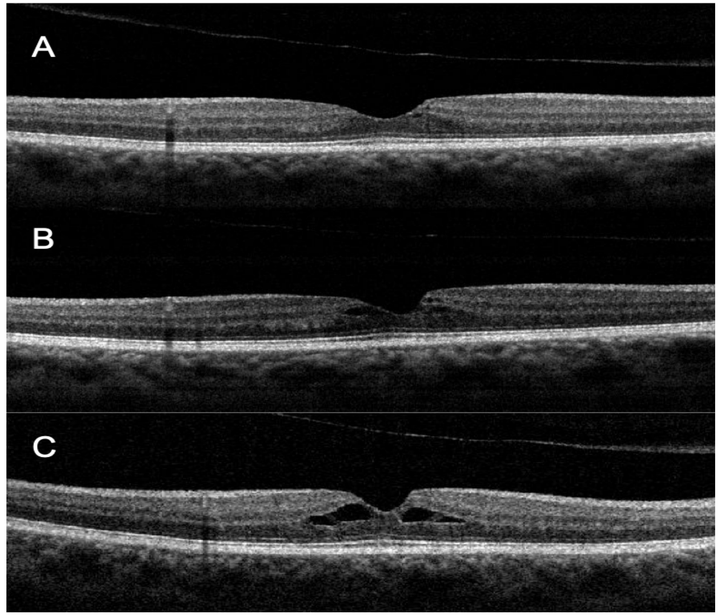

Figure 1. OCT showing progression of SNIFR in central fovea. Increase in size and number of foveomacular cysts is noted between 2015(A), 2016(B) and 2019(C).

KEYWORDS

Retinoschisis, Maculopathy, Vitreomacular traction.

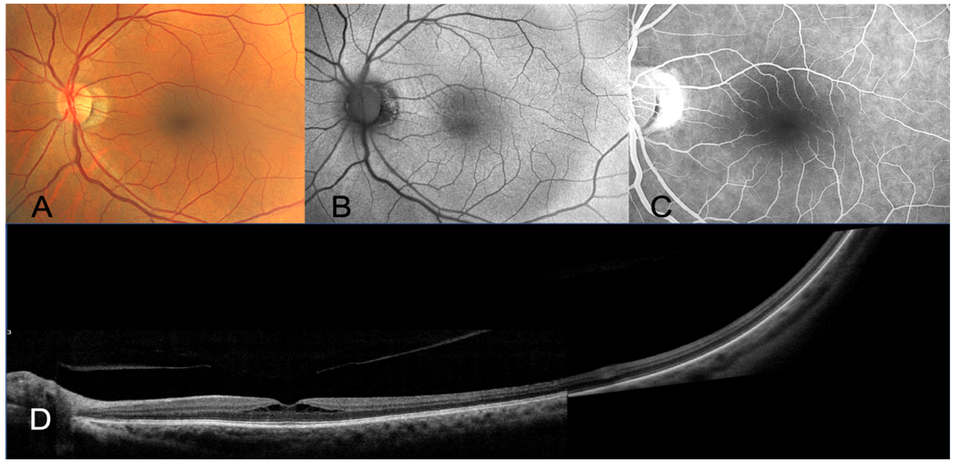

Figure 2. SNIFR without fundus autofluorescence abnormalities (B) and no signs of contrast leakage or staining in fluorescein angiography (C). Wide angle OCT showing fovemacular cysts without peripheral retinoschisis (D).

DISCUSSION

Structurally, SNIFR differs from XLR as SNIFR primarily affects the OPL (2). It has been described in association with exudative maculopathies (3), macular telangiectasias (4) and vassalva maneuvers (5). The OPL low resistance can also allow previous VMT to leave degenerative retinal splits and these may remain even after PVD (2, 6, 7).

In our case, we believe that resolution of VMT may have created microholes in internal limiting membrane and also exerted mechanical stress inducing splits in the OPL. Similar to optic disc PIT maculopathy and glaucoma associated retinoschisis(8), we believe that leakage of vitreous fluid through microholes in the ILM allowed enlargement of foveal cysts between 2015 and 2019. This is the first case reported in literature of SNIFR associated with VMT and is also the first showing progressive formation of retinal splits through a 4-year follow-up.

REFERENCES

1. Ober MD, Freund KB, Shah M, Ahmed S, Mahmoud TH, Aaberg TM, Jr., et al. Stellate nonhereditary idiopathic foveomacular retinoschisis. Ophthalmology. 2014;121(7):1406-13.
2. Fragiotta S, Leong BCS, Kaden TR, Bass SJ, Sherman J, Yannuzzi LA, et al. A proposed mechanism influencing structural patterns in X-linked retinoschisis and stellate nonhereditary idiopathic foveomacular retinoschisis. Eye (Lond). 2019;33(5):724-8.
3. Casalino G, Upendran M, Bandello F, Chakravarthy U. Stellate nonhereditary idiopathic foveomacular retinoschisis concomitant to exudative maculopathies. Eye (Lond). 2016;30(5):754-7.
4. Falb T, Malle EM, Haas A, Weger M, Wedrich A. Stellate Nonhereditary Idiopathic Foveomacular Retinoschisis in a Patient with Macular Telangiectasia Type 2. Retin Cases Brief Rep. 2018.
5. Dolz-Marco R, Kato K, Freund KB, Yannuzzi LA. Unusual Case of Stellate Nonhereditary Idiopathic Foveomacular Retinoschisis. Retin Cases Brief Rep. 2017;11 Suppl 1:S49-S53.
6. Querques G, Cristiana I, Francesco P, Noci ND. Vitreomacular traction syndrome with an outer macular hole. Retin Cases Brief Rep. 2008;2(4):266-8.
7. Hotta K, Hotta J. Retinoschisis with macular retinal detachment associated with vitreomacular traction syndrome. Retina. 2004;24(2):307-9.
8. Inoue M, Itoh Y, Rii T, Kita Y, Hirota K, Kunita D, et al. Macular retinoschisis associated with glaucomatous optic neuropathy in eyes with normal intraocular pressure. Graefes Arch Clin Exp Ophthalmol. 2015;253(9):1447-56.

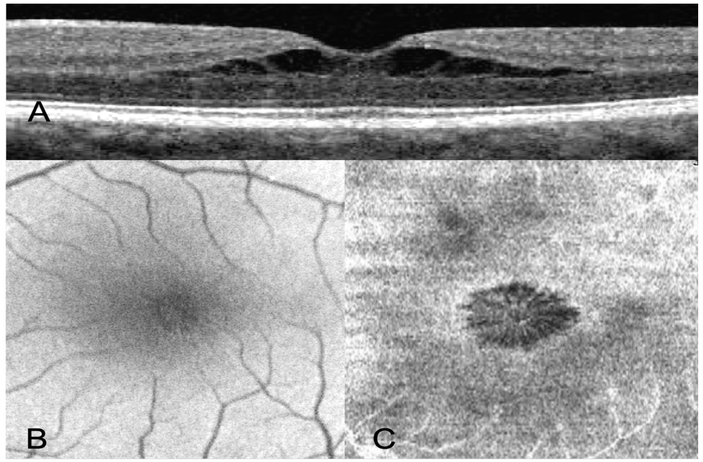

Figure 3. Spectral OCT, red-free fundus photography (A) and Macula En face OCT (B) showing stellate foveal pattern in outer plexiform layer.