

SUCCESSFUL CLOSURE OF FULL-THICKNESS MACULAR HOLES SECONDARY TO MACULAR VITELLIFORM LESIONS

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Purpose: To describe the first reported cases of full-thickness macular holes secondary to vitelliform lesions that were successfully closed with vitrectomy surgery and gas tamponade.

Methods: Two female patients developed visual loss secondary to bilateral vitelliform lesions and associated full-thickness macular holes. The patients underwent 25-gauge pars plana vitrectomy, internal limiting membrane peeling, and 26% sulfur hexafluoride gas, followed by 3 days of face-down positioning.

Results: In both patients, the macular holes remain closed 3 and 25 months post-operatively.

Conclusion: Vitrectomy surgery with gas tamponade may successfully close full-thickness macular holes secondary to macular vitelliform lesions.

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Macular vitelliform lesions (VLs) may be inherited (vitelliform macular dystrophies one to five) or acquired. The most common inherited causes include Best disease (vitelliform macular dystrophy-2, *BEST1* gene mutation, OMIM #153700), and adult-onset foveomacular vitelliform dystrophy (AFVD, vitelliform macular dystrophy-3, *RDS/PRPH2* gene mutation, OMIM #608161). Mutations in *IMPG1* and

IMPG2 have also been described. Associations of acquired VLs include age-related macular degeneration, cuticular drusen, subretinal drusenoid deposits, vitreomacular traction, central serous chorioretinopathy, and angioid streaks.¹ Rarely, macular VLs have been associated with development of full-thickness macular holes (FTMHs).^{2–5} This is believed to occur because of outer retinal thinning.^{3,5} Here, we describe the first reported cases of successful closure of FTMH secondary to VLs after vitrectomy surgery and gas tamponade.

Case Reports

Case 1

A 65-year-old woman presented with progressive visual loss in her left eye. She had two sisters with visual problems, the diagnosis of which could not be elucidated. Best-corrected visual acuities were 20/50 + 1 in the right eye and 20/85 – 1 in the left eye. Bilateral, small, yellow, hyperautofluorescent subretinal vitelliform deposits were seen at both foveae (Figure 1, A and B), and a clinical diagnosis of AFVD was made. On spectral domain optical coherence tomography (SD-OCT, Cirrus HD-OCT 5000; Carl Zeiss Meditec AG, Jena, Germany), there was a small FTMH in

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the left eye, measuring 185 μm in diameter (Figure 1C). The vitreous was detached off the fovea and an operculum present floating above it. A 25-gauge pars plana vitrectomy using ILM-Blue dye (0.025% brilliant blue G, 4% polyethylene glycol; D.O.R.C., Zuidland, Netherlands) for internal limiting membrane peeling and 26% sulfur hexafluoride gas was followed by 3 days of face-down positioning. One month later, the visual acuity had improved to 20/50 + 1, and SD-OCT confirmed closure of the hole (Figure 1D). The FTMH has remained closed, 25 months after operation.

Case 2

A 78-year-old woman was referred with a 6-week history of increasing central distortion in her left eye. She was known to have bilateral hyperautofluorescent macular VLs associated with drusen and pigmentary changes, which had been relatively stable for 7 years (Figure 2, A and B). There was no family history of eye disease, and a diagnosis of acquired VLs secondary to age-related macular degeneration was made. One year before referral, her visual acuity was 20/20 in the left eye, and SD-OCT (Spectralis HRA + OCT; Heidelberg Engineering, Heidelberg, Germany) revealed hyperreflective material between the outer retinal and retinal pigment epithelium (Figure 2C). There was no vitreomacular traction, and a posterior vitreous detachment was already present.

On examination, her visual acuities were now 20/50 – 1, pinhole 20/40 – 1 in the right eye and 20/200 – 4 pinhole 20/200 – 2 in the left eye. The vitelliform material in the left eye had erupted into the vitreous cavity, leaving a large FTMH measuring 1150 μm in diameter (Figure 2D). The vitreous was detached off the fovea. A 25-gauge pars plana vitrectomy using ILM-Blue dye for internal limiting membrane peeling and 26% sulfur

hexafluoride gas was performed, and the patient instructed to posture face down for 3 days. Closure of the hole was seen at the 1-month postoperative visit, but the left vision remained 20/200 – 1, pinhole 20/200 + 3, most likely due to the loss of photoreceptors as seen on SD-OCT (Figure 2E). The FTMH has remained closed, 3 months after operation.

Discussion

In both patients, the macular holes remain closed postoperatively. Although visual acuity improved in the first patient, it did not significantly improve in the second because of the loss of foveal photoreceptors. The preservation of photoreceptors is difficult to determine preoperatively. In Patient 2, the latest SD-OCT before FTMH development showed an intact ellipsoid line (Figure 2C). Successful repair of FTMH secondary to AFVD has been described using a heavy silicone oil tamponade, after initial attempts with gas tamponade were unsuccessful.³ However, silicone oil requires subsequent removal and may be complicated by emulsification. Our cases show that the use of gas tamponade may be sufficient in these cases.

The diagnosis of AFVD (Patient 1) and acquired VLs secondary to age-related macular degeneration (Patient 2) was based on clinical presentation.

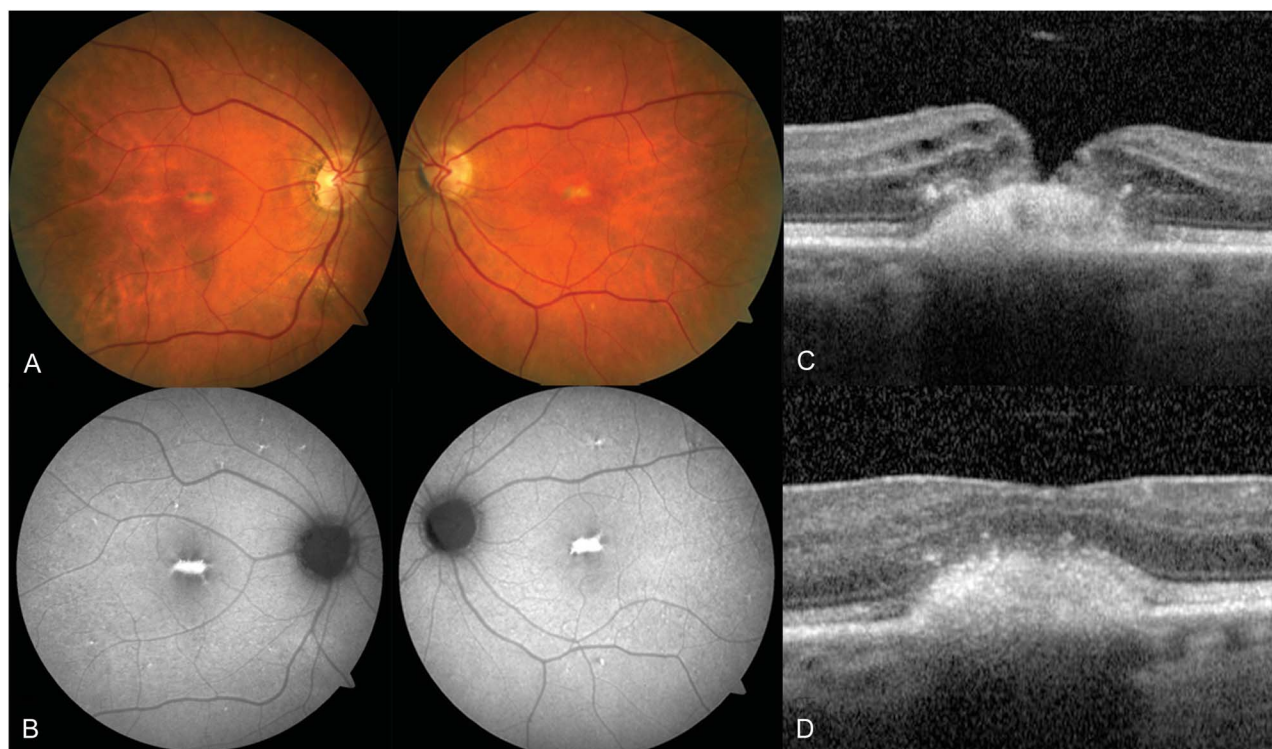


Fig. 1. A. Color fundus photographs show small, round, yellow deposits at both foveae. B. The VLs are hyperautofluorescent. C. Optical coherence tomography of the left eye shows the presence of a small FTMH above the subretinal vitelliform deposit. There is no vitreous adhesion to the fovea, and the visual acuity is 20/85 – 1. D. Two weeks after vitrectomy surgery, internal limiting membrane peeling and gas tamponade, there has been complete closure of the macular hole with an improvement in vision to 20/50 + 1.

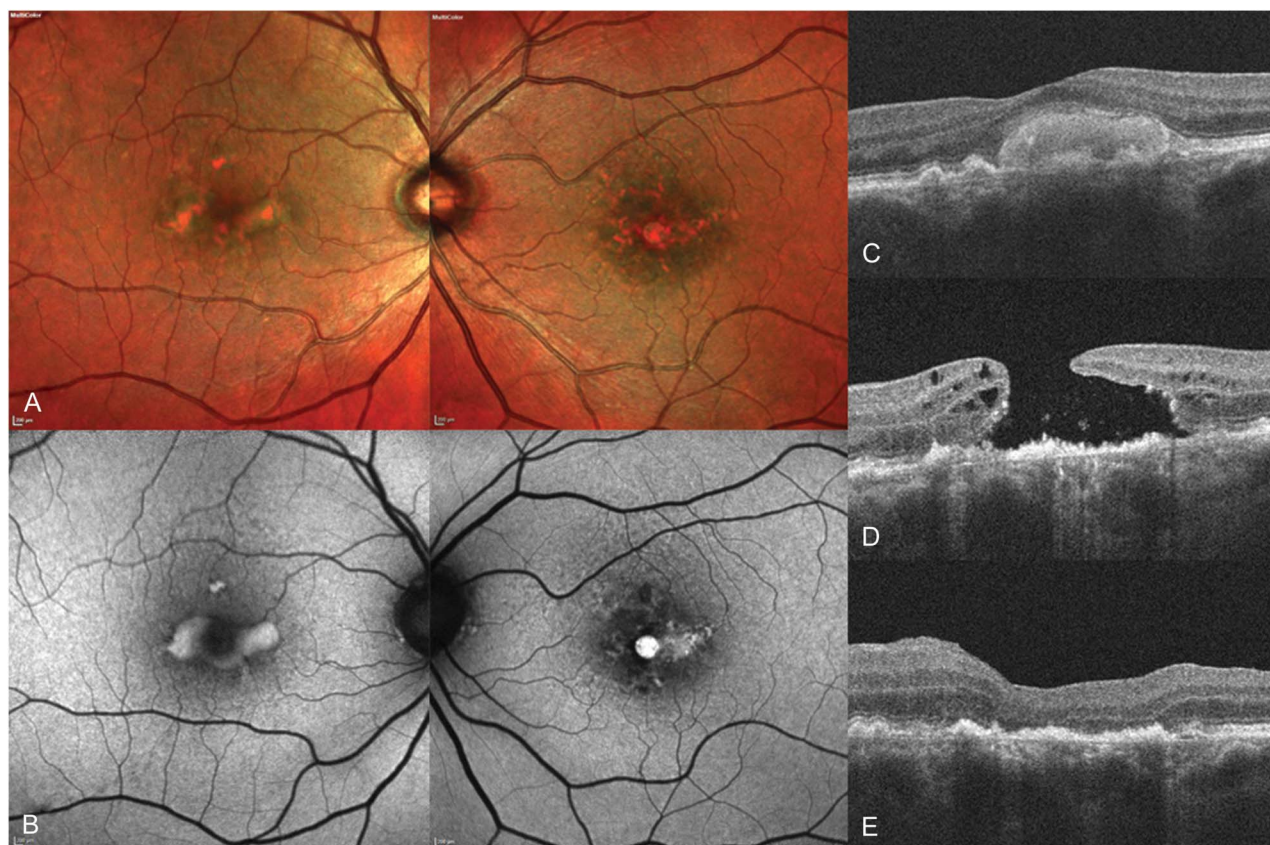


Fig. 2. **A.** Bilateral confocal-scanning laser multicolor images demonstrate yellow deposits, pigmentary changes, and drusen at both foveae. **B.** The vitelliform material is hyperautofluorescent, but there are also small patches of hypoautofluorescence due to retinal pigment epithelial atrophy. **C.** Spectral domain optical coherence tomographic images of the left eye show a subfoveal VL with drusen and an intact ellipsoid line. **D.** Progression into a large, FTMH measuring 1150 μm in diameter. **E.** Successful closure 1 month after vitrectomy, internal limiting membrane peeling, and gas tamponade.

Characteristics of AFVD include small, round, yellowish, subretinal, lipofuscin-rich, hyperautofluorescent deposits at the macula, which may be asymptomatic or present in the fourth to sixth decades of life with central scotoma and/or metamorphopsia. The diagnosis of acquired VLs secondary to age-related macular degeneration in Patient 2 was based on the older age of presentation and presence of drusen. Neither patient had a family history to suggest Best disease. Without genetic testing, confirmation of the disease cannot be absolutely certain. Nevertheless, the pathogenesis of macular VLs causing outer retinal thinning leading to FTMH is likely to be the same regardless of the underlying etiology. We conclude that vitrectomy surgery with internal limiting membrane peeling and gas tamponade may successfully close FTMH secondary to macular VLs, although the visual benefit may depend on the remaining photoreceptor integrity.

Key words: adult-onset foveomacular vitelliform dystrophy, gas tamponade, macular dystrophies, macular hole, vitelliform lesion, vitrectomy.

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