Management of a Macular Hole Following Retinal Arterial Macroaneurysm Rupture

A B C D  Xhevat Lumi
     E F  Fran Drnovšek

Corresponding Author: Xhevat Lumi, e-mail: xhlumi@hotmail.com
Conflict of interest: None declared

Patient: Female, 60-year-old
Final Diagnosis: Macular hole following retinal arterial macroaneurysm rupture
Symptoms: Sudden loss of central vision
Medication: —
Clinical Procedure: 25-gauge pars plana vitrectomy
Specialty: Ophthalmology

Objective: Unusual clinical course
Background: We report the successful treatment of a macular hole (MH) secondary to a retinal macroaneurysm (RAMA) rupture by pars plana vitrectomy (PPV) and autologous transplantation of an internal limiting membrane (ILM) during primary surgery.

Case Report: A 60-year-old woman presented with a sudden loss of central vision in her right eye. Fundus examination revealed a large sub-ILM hemorrhage involving the macula in her right eye. We performed 25-gauge PPV and ILM peel overlaying the hemorrhage. A ruptured RAMA next to the inferotemporal branch of the central retinal artery, MH, and subretinal hemorrhage were discovered. Part of the subretinal blood clot was removed through the MH. ILM was then further peeled off outside vascular arcades and transplanted as a free flap into the MH. At the end of surgery, 10% C3F8 gas was instilled as tamponade. Postoperatively, we observed anatomical closure of the MH, restoration of outer retinal layers, and improvement of visual acuity. During the follow-up period, the patient also had cataract surgery. Two years after the vitrectomy, the MH remained closed, with visual acuity improved to 10/20.

Conclusions: Autologous ILM transplantation during primary PPV can be an effective surgical approach for treatment of MH related to ruptured RAMA in selected cases.

MeSH Keywords: Retinal Artery • Retinal Perforations • Transplantation, Autologous

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/922437
Background

Sub-internal limiting membrane (ILM) hemorrhages are located between the ILM and retinal nerve fiber layer. They have been described in a large variety of clinical settings and cause severe visual impairment due to their predilection for the macular region, which is explained by the absence of firm attachments of ILM to the retina at the posterior pole peripheral to the fovea [1]. This type of intraretinal hemorrhage can be complicated further by development of a macular hole (MH). It has been reported that 5.3–12.5% of eyes with intraretinal hemorrhage following ruptured retinal arterial macroaneurysm (RAMA) developed a full-thickness MH [2,3].

Case Report

A 60-year-old woman with long history of hypertension and asthma reported to the emergency clinic 1 day after having noticed sudden, painless loss of central vision in her right eye. The vision loss developed while she was gardening in a bent-over position. The anamnesis for Valsalva maneuver and eye trauma was negative. Examination upon arrival at the clinic revealed best corrected visual acuity (BCVA) of counting fingers at 0.5 meters eccentrically in her right eye and 20/20 in her left eye. Anterior segment examination revealed normal results in both eyes. Examination of the fundus revealed a large preretinal and sub-ILM hemorrhage with a niveau formation involving the macula in her right eye. Due to a thick sub-ILM hemorrhage, it was not possible to visualize the macular area on OCT (Figures 1A, 2). The fundus of the left eye exhibited grade 2 hypertensive retinopathy with arteriolar narrowing and arteriovenous nicking. Thus, uncontrolled high blood pressure was the only known risk factor for the macular hemorrhage, and the presence of RAMA was assumed. On the third day after initial presentation of symptoms, further deterioration with vitreous hemorrhage was found. The patient underwent a 25-gauge pars plana vitrectomy (PPV). After inducing PVD, a complete vitrectomy with the ILM peel overlaying the hemorrhage was performed. A ruptured RAMA was noticed next to the inferotemporal branch of the central retinal artery (Figure 1B–1D). A thick blood clot under the ILM was removed with a vitreous cutter. After cleaning the macular area, a full-thickness MH approximately one-third the size of the optic disc was detected overlaying the submacular hemorrhage (SH) (Figure 1C, 1D). Part of the submacular clot was protruding through the hole, indicating a preexisting full-thickness MH. The protruding clot was removed through the hole using a soft-tip backflash instrument (Figure 1C).

Due to the presence of a subretinal hemorrhage, the retina at the edges of the hole was fixed and did not show any mobility. Since the MH did not exhibit a tendency to close, even after massaging it with the soft-tip backflash, and no ILM was left within the macular area, the ILM was stained with Brilliant Peel dye (Fluoron Gmbh) and then further peeled off outside the vascular arcades and transplanted as a free flap into the MH (fill technique, Figure 1E, 1F). We instilled 10% C3F8 gas as tamponade at the end of the surgery. No complications were observed during or after the surgery.

At the follow-up examinations 3 months and 1 year after the surgery, anatomical closure of the MH was observed, with completely restored outer retinal layers (Figures 3, 4). We observed a significant cataract and improvement of visual acuity to 20/200. The patient was referred for cataract surgery. Two years after surgery, the MH remained closed and the BCVA had improved to 10/20 (Figure 5).

Discussion

Hypertensive retinopathy is a common disease, present in most patients with a history of untreated hypertension, which is a predisposing factor for the development of macroaneurysms and full-thickness macular hemorrhage with intraretinal (sub-ILM) location of a hemorrhage, subretinal or even a combination of both. Although spontaneous resolution of preretinal and sub-ILM hemorrhage may occur within 2–3 months, several concerns have been raised. Persistence of blood can cause irreversible damage to the retina and cause permanent visual loss as a result of preretinal tractional membrane formation [4]. SH, on the other hand, can induce damage to the retina by 3 different mechanisms. Firstly, a blood clot acts as a diffusion barrier for nutrients and metabolites passing to and from the retina [5]. Secondly, the fibrin meshwork of the blood clot also interlinks with the photoreceptors and applies traction that can lead to sheering of the photoreceptor layer when the clot contracts [5]. Finally, there is direct toxicity to the photoreceptors caused by iron released from erythrocytes. This harmful effect to the photoreceptors is correlated with the duration of SH [5].

MHS following RAMA ruptures are also not uncommon. The primary success rate of anatomical closure following surgical intervention in this setting is lower compared to the treatment of idiopathic MH, ranging between 57.1% and 75% [2,3]. In previously published reports, different surgical approaches were used with different success rates, and it remains unclear which is most effective. A case series published by Sagara et al. described PPV with ILM peel and SF6 gas tamponade for treatment of MHS in eyes with a ruptured RAMA [2]. However, 42.9% of the patients needed more than 1 surgery with additional ILM peel in order to achieve anatomical closure of the MH. Peeling of the ILM has been well described as an important aid to achieve anatomical closure of idiopathic MHS [6–8].
Figure 1. Fundus images during the various stages of the surgery: (A) hemorrhage covering posterior pole; (B) peeling ILM over the hemorrhage; (C) removing part of the blood clot through MH; (D) image showing RAMA and MH; (E) additional ILM peel; (F) MH filled with ILM.
Figure 2. Preoperative fundus image and OCT scan showing macular hemorrhage.

Figure 3. Early postoperative OCT scan showing closed MH with disrupted outer retinal layers.

Figure 4. Postoperative fundus image and OCT scan showing macular area without signs of hemorrhage and closed macular hole 3 months after surgery.
OCT scan showing restored retinal layers 2 years after surgery.

Data on large idiopathic MHs suggests that results can be further improved by using an ILM flap [9].

Various approaches for management of concomitant SH and MH have been reported. Observation of up to 2 months, until partial resolution of the hemorrhage prior to PPV with ILM peel, and 15% C3F8 tamponade for successful treatment of MH were described by Ciardella et al. [10]. In contrast, Bakri et al. performed PPV, ILM peel, subretinal injection of t-PA, and gas tamponade 1 week after presentation of symptoms [11]. In a similar case reported by Iwakawa et al., a 2-step approach was described [12]. Due to the possibility that the enclosed residual SH and use of indocyanine green (ICG) for staining of ILM could induce impairment of the photoreceptor layer in the fovea and retinal pigment epithelium cytotoxicity, they performed ILM peeling and air tamponade first [13]. After absorbing the residual subretinal hemorrhage, the second surgery was performed. The ILM was further peeled off up to a diameter of 2 discs around the MH and transplanted into the MH, achieving closure.

In contrast to Iwakawa’s method, in our case, subretinal hemorrhage was completely displaced and resorbed, despite immediate closure of the MH during primary surgery. Brilliant Peel dye was used instead of ICG for staining the ILM prior to peeling, and was washed out before the removal of the subretinal hemorrhage, thus minimizing potential damage to the RPE [14]. At follow-up examinations, functional and anatomical results were comparable to the results of patients treated for idiopathic MHs with early restoration of the outer retinal layer. It has been shown that restoration of the external limiting membrane and ellipsoid zone after MH surgery is a positive predictive factor of visual acuity improvement [15]. Further improvement of visual function was observed following the cataract surgery. In this situation, clear lens extraction during primary PPV could be considered, as it might be more comfortable for the patient. However, due to the presence of a thick macular hemorrhage, the results of combined surgery (phaco-vitrectomy) could be compromised because of inaccurate biometry measurements in such cases. The functional outcome in this particular case with secondary cataract surgery performed was favorable and she achieved 10/20 vision.

Conclusions

To the best of our knowledge, this is the first report of successful closure of a MH following RAMA rupture by autologous ILM transplantation during primary surgery. Autologous ILM transplantation during primary PPV can be an efficient surgical approach for treatment of MH related to ruptured RAMA in selected cases. Anatomical closure of the MH in this single case was achieved, resulting in significant improvement of visual function.

Conflict of interest

None.
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