Spontaneous release of epiretinal membrane in a young weight-lifting athlete by presumed central rupture and centrifugal pull

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Abstract: This patient presented for surgery at the age of 32 years, 14 months after his initial complaint of metamorphopsia and visual loss in the right eye. Past tests demonstrated a whitish epiretinal membrane (ERM) with translucent stress lines over a thickened macula. Visual acuity was found on last presentation to be normal with minimal alteration on Amsler grid testing. A torn ERM was found in the center with left-over ERM temporally and rolled-over ERM nasally at the site of the epicenter with no posterior vitreous detachment. Visual recovery occurred gradually over several days 2 months prior to presentation apparently following heavy weight-lifting with a sensation of severe eye pressure. Sequential funduscopy and optical coherence tomography scans demonstrated the peeling of an ERM accompanied by normalization of foveal thickness. Valsalva maneuver had put excessive tension on ERM which tore in its center at the weakest line with gradual contraction of the ERM away from the fovea towards the peripapillary area. This is a new mechanism of self-separation of ERM induced by Valsalva. ERM in young subjects is subject to rupture and subsequent separation by tangential traction. There are three mechanisms for spontaneous separation of ERM: 1) posterior vitreous detachment with pulling of ERM by detaching vitreous (most common in adults); 2) the contracting forces of the immature ERM become stronger than its adhesions to the retina resulting in slow tangential traction on the edges of the ERM and gradual separation from the edges towards the center (remodeling common in youngsters); and 3) acute tearing of ERM at its weakest central point and retraction of part of the membrane towards the epicenter (current case report).

Keywords: valsalva maneuver, posterior vitreous detachment

Introduction

Epiretinal membranes (ERMs) are characterized by wrinkling of the macular surface from cell proliferation. Most commonly ERMs are noted in adults above the age of 50 years in association with posterior vitreous detachment (PVD), laser photocoagulation, or after retinal detachment repair. A majority of ERM in adults is of the cellophane macular reflex type referring to the thin transparent variety.1 ERM in children and young adults is uncommon, because a PVD is uncommon in very young subjects. Ocular trauma, pars planitis, ocular toxocariasis, ocular toxoplasmosis, combined hamartoma of the retina and retinal pigment epithelium, and Coats’ disease are leading causes of secondary ERMs in children. The prevalence of ERM in subjects less than 20 years of age is around 1 in 20,000.1 A majority of ERM in young subjects is of the thick white contractile variety having strong adherence to retinal vessels.2,3 Myofibroblasts, myoblastic differentiation of retinal pigment epithelial cells and fibrous astrocytes, as well as new collagen formation are more common in ERM of young subjects than in ERM in older subjects.4 We report a young adult with ERM that underwent spontaneous...
release in a peculiar way (ERM tearing centrally) apparently following Valsalva maneuver.

Case report
This 32 year-old healthy man complained of visual loss in the right eye 14 months prior to presentation and he was referred for surgical peeling of an ERM (Figures 1–5) by two retina experts. Two months before presentation, he indulged in heavy weight-lifting and noted repeatedly severe eye pressure during the exercise. Then he noted a gradual visual improvement in the right eye over several days. Two months later, ophthalmic exam revealed 6/6 uncorrected visual acuity with findings of a residual rolled-over ERM supranasal to the fovea (Figure 6). The patient denied ocular rubbing or previous ocular trauma. Amsler grid revealed mild distortion temporally. Past fundus photograph 1.5 years ago revealed a

Figure 1 Fundus photograph 18 months before presentation. Notes: The opaque ERM bridges between multiple fixation points on the retinal surface, obscuring the view to the macula. Note that traction lines correspond with wrinkling of the ILM. Abbreviations: ERM, epiretinal membrane; ILM, internal limiting membrane.

Figure 2 Red-free photograph of the right posterior pole. Note: An opaque epiretinal membrane covers the macula resulting in superficial radial folds.

Figure 3 Infrared photograph of the right posterior pole. Note: An opaque epiretinal membrane covers the macula with prominent traction exerted on the inner retinal surface in the form of superficial radial folds.

Figure 4 Arteriovenous phase of fluorescein angiography transit. Notes: Visible retinal vessels at the posterior pole appear tortuous. The superior and inferior vessels appear narrowed and the perimacular vessels are pulled towards the epicenter with prominent retinal striae.
dense macular gliosis with prominent radial stress line (best seen on infrared photography) throughout the posterior pole and centered around the thickest part of the membrane nasal to the disc (as noted by red-free photograph [Figure 2]). Intravenous fluorescein angiography done at the same time revealed obliteration of foveal avascular zone with tortuous macular capillaries and stretched out papillomacular vessels. Optical coherence tomography (OCT) done 1 year ago (Figure 5) revealed a diffuse ERM thin temporal to the fovea and very thick nasal to the fovea with marked thickening of the central fovea. OCT upon presentation demonstrated central peeling of ERM (Figure 7) accompanied by normalization of central foveal thickness (Figure 8). ERM rupture could be noted with one edge ruptured temporally and the nasal edge rolling over (Figure 9). The thick nasal membrane rolled over itself denoting its elastic property. There was no PVD identified by OCT or by slit lamp fundus biomicroscopy with a 90 diopter lens.

Discussion

ERM in young subjects commonly causes both metamorphopsia and reduced visual acuity from reduced axoplasmic flow, abnormal hemodynamic microcirculation, light-filtering effect of ERM, and photoreceptor distortion by tangential traction.5–7 Clinically, the superior and inferior vessels are narrowed while the perimacular vessels are pulled toward the epicenter with inner retinal striae. ERM in young people and children is rare and may be caused by congenital defects resulting from persistent adhesions of primary vitreous to the retina.7 The clinical and ultrastructural features of juvenile macular pucker reflect a more rapidly changing, contractile tissue4 compared with the usually more quiescent features in most cases involving older patients.

ERM is common in adults and rare in the young. However in adults, PVD can damage the internal limiting membrane, thereby permitting the migration of glial cells to the retinal surface.6,8 Alternate proposed hypothesis in adults is that an incomplete PVD provides the conditions suitable for membrane proliferation in the adhesion area between the vitreous and the retina. The cells involved in this process are retinal pigment epithelium metaplastic cells, glial cells (Muller cells and astrocytes), hyalocytes,9 endothelial cells, fibroblasts, myofibroblasts, monocytes, and macrophages. It appears that several growth factors10 (platelet-derived...
growth factor, tissue growth factor TGF beta1 and 2, fibroblast growth factor, vascular endothelial growth factor, nerve growth factor) could stimulate glial cells to transdifferentiate into myofibroblasts and stimulate myofibroblasts to turn on their contractile actions more so in young subjects.9–11 Moreover the role of plasminogen and metalloproteinases in contraction of ERM has been raised.12,13 Spontaneous release of ERM is a rare event but is known to occur in adults and is related to the occurrence of an acute PVD that simultaneously releases the attachment between the retina and ERM (Figures 10, 11). Separation or peeling of ERM in young subjects is quite rare.14–30 It may occur spontaneously by development of a PVD,23 or shortly after panretinal photocoagulation or Nd:YAG (neodymium-doped yttrium aluminum garnet) posterior capsulotomy27 Meyer et al15 presented six cases of spontaneous gradual “remodeling” with release of ERM in young subjects. They hypothesized that when the contracting forces of the immature ERM are stronger than its adhesions to the retina, the membrane may separate spontaneously. Only one of six cases had PVD15 and

Figure 7 Serial horizontal optical coherence tomography scans demonstrate rupture of the epiretinal membrane (ERM) temporal to fovea with retraction of remaining ERM nasally and return of the normal foveal contour.

Figure 8 Close up view of horizontal optical coherence tomography scan showing normal foveal contour.
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one case with prior PVD also had release of ERM. ERM release was more common in eyes with PVD in a study of 1,248 consecutive eyes with idiopathic ERM followed-up for around 3 years, ERM self-separation occurred in 37 eyes (3.0%), with 16 of 1,091 eyes with pre-existing PVD (1.5%) and 21 of 157 eyes without pre-existing PVD (13.4%). A higher rate of spontaneous separation was reported in Japan: Nomoto et al detected five patients with spontaneous separation of ERM among 92 patients with idiopathic ERM.

Three mechanisms (Figures 10–15) are proposed for spontaneous ERM separation including the two previously described ones: PVD (Figure 11), remodeling (contraction of myofibroblast) (Figures 12, 13), and rupture of weakest line by lifting heavy objects (Valsalva) (Figures 14, 15). Head-down position combined with weight-lifting (Table 1) and mouth closure lead to high intraocular pressure (IOP) (rise can reach up to 30 mmHg) and this places tension on tissues leading to compression of the vitreous against the retinal surface and hence retinal dehiscence in places of retinal lattice or in this case tearing off of the elastic fibrous tissue already under tangential traction at its weakest, thinnest point. Power athletes routinely utilize the Valsalva maneuver during weight-lifting. Valsalva maneuver comprises forcible exhalation against the closed glottis, thereby creating a sudden increase in the intrathoracic or intra-abdominal pressure. Ocular and systemic changes are secondary to the extreme pressure elevations that occur in the intra-abdominal, intrathoracic, intracranial, intraocular, and vascular compartments. The enormous pressures generated lead to elevations in intracranial pressure obstructing venous outflow leading to hemorrhage and elevations in IOP. In one study by Dickerman et al IOPs were significantly elevated by weight-lifting by 15 mmHg.
from a mean of 13 to a mean of 28 with one subject’s IOP reaching 46 mmHg during maximal contraction. Macdougall et al\textsuperscript{33} found severe elevations in blood pressure (BP) with mean value of 320/250 mmHg (BP exceeding 480/350 mmHg in one subject) and mouth pressures of 30–50 torr during a single maximum lift (normal mouth pressure being 15 torr). This combination of severe elevation in BP and intracranial pressure would have yielded a high incidence of ocular hemorrhage was it not for the dampening effect of high IOP. ERM sites above blood vessels appear to be under focal stress. ERM rupture during weight-lifting was not previously reported in the literature (Table 1).\textsuperscript{32–42}

The relation between weight-lifting and ERM separation could be a coincidence with the natural history of ERM taking its course. The current ERM is thought to be primary and not secondary to trauma or inflammatory chorioretinal disorders. Other factors may be involved in ERM separation like rubbing of the eye or minor trauma to the eye, neither of which could be elicited in this case. A table of ocular findings in weight-lifters is enclosed (Table 1).

In conclusion, ERMs in young subjects seems strikingly different from that of those in older individuals; they appear thicker, whiter with high prevalence of myofibroblast cells, hence the contracting forces that stretch the retina. Potential maneuvers that can put stress on ERM surface include severe acute oculopression or rubbing that can raise the IOP acutely and Valsalva maneuvers, hence triggering spontaneous resolution of ERM via rupture in its thinnest meridian with subsequent contraction of the thicker side towards the epicenter.
Table 1 Ocular and systemic changes in weight lifters

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Table 1 Ocular and systemic changes in weight lifters

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<tr>
<th>Subconjunctival hemorrhage</th>
<th>Increased intraocular pressure</th>
<th>Retinal hemorrhage</th>
<th>Premacular hemorrhage</th>
<th>Vitreous hemorrhage</th>
<th>Venous stasis retinopathy (Purtscher retinopathy)</th>
<th>Stroke</th>
<th>Cerebral hemorrhage</th>
<th>Subarachnoid hemorrhage</th>
<th>Hiatal hernia</th>
<th>Pneumothorax</th>
<th>Leg varices</th>
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Disclosure

The authors have no conflicts of interest related to this paper.

References


