Epiretinal membrane removal in patients with Stargardt disease

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Epiretinal membranes (ERMs) in Stargardt disease have been known to undergo spontaneous separation in children. Results of surgical intervention in adult patients with Stargardt disease have rarely been reported. A retrospective review of results of surgical intervention for ERM causing visual impairment in two adult patients of Stargardt disease was carried out. Both patients developed ERM in one eye during their follow-up period with the resultant drop in their preexisting visual acuity. Postsurgery, restoration of foveal contour with some improvement in visual acuity was observed in both patients. No adverse effect of surgery was noted.

Key words: Epiretinal membrane, stargardt disease, vitrectomy

Stargardt disease is an autosomal recessive condition commonly seen in early adulthood characterized by the presence of classical yellow subretinal flecks at the posterior pole. The macular changes can range from fine granularity to beaten...
Bronze metal appearance to geographic atrophy. Complications such as choroidal neovascularization and more rarely epiretinal membrane (ERM) formation are known to cause a sudden drop in vision. ERMs, rarely reported in Stargardt disease are known to undergo spontaneous separation,\(^1\) avoiding the need for surgical intervention. However, in patients where spontaneous separation does not occur, surgical intervention would eventually be needed. Literature regarding the surgical removal of an ERM in Stargardt disease and its outcome is scarce, except for a single case report.\(^2\) Herein we report our experience in two patients of Stargardt disease in whom spontaneous separation failed to occur and they underwent ERM removal, with some improvement in their visual acuity.

### Case Reports

#### Case 1
A 31-year-old male, a diagnosed case of Stargardt disease [Fig. 1a and b] on follow-up since 6 years presented with gradual diminution of vision in the right eye since 2 months. His best-corrected visual acuity (BCVA) had dropped from 20/40, N6 to 20/400, N36 in the right eye while left eye was 20/40, N6 as before. Fundus examination revealed the presence of a thick ERM with macular edema [Fig. 1c]. Spectral domain-optical coherence tomography (SD-OCT) [Fig. 2a] also documented a macular thickness of 680 µ in the right eye. 1-month later macular thickness increased to 796 µ. No obvious vascular pathology was noted that could account for increased thickness. He underwent 23 g pars plana vitrectomy with ERM removal. Intraoperatively there was dense adhesion of the posterior hyaloid face to the macula. The rapid increase in the thickness of the retina could have been due to an attempt at spontaneous posterior vitreous detachment which did not occur. Postoperative period was uneventful with release of traction due to ERM [Fig. 1d] and restoration of the foveal contour on SD-OCT [Fig. 2b] and unchanged autofluorescence pattern [Fig. 2c and d]. At 6 months follow-up, BCVA had improved to 20/120 N18.

#### Case 2
A 24-year-old myopic male, a diagnosed case of Stargardt disease [Fig. 3a] on follow-up since 3 years with a BCVA of 20/400, N36 in both eyes, presented with sudden onset metamorphopsia in the left eye of 1-week duration. Though, BCVA was unchanged, fundus showed an ERM causing traction on the macula with associated edema and subretinal hemorrhage [Fig. 3b]. Choroidal neovascular membrane was ruled out on fundus fluorescein angiography (FFA) [Fig. 4]. The ill defined leak seen on

### Figure 1
(a and b) Colour fundus picture of the postpole of right and left eyes of case 1 showing numerous flecks in the perifoveal area and posterior pole (c) colour fundus picture of the right eye 6 years later showing a thick epiretinal membrane at the macula. (d) Postoperative colour fundus picture of the right eye showing absence of epiretinal membrane with retinal pigment epithelial atrophy at the macula.

### Figure 2
(a) Preoperative Optical coherence tomography (OCT) of case 1 showing increased macular thickness secondary to thick epiretinal membrane. (b) Postoperative OCT showing restoration of foveal contour with RPE atrophy. (c) Preoperative autofluorescence image showing hypoautofluorescence corresponding to central area of retinal pigment epithelial atrophy. (d) Postoperative autofluorescence image showing no change in the hypoautofluorescent area.

### Figure 3
(a) Colour fundus picture of right eye of case 2 patient at 3 years follow-up showing beaten metal appearance at the macula. (b) Colour fundus picture of the left eye showing thick epiretinal membrane causing macular traction and associated with subretinal hemorrhage. (c) Colour fundus picture of the left eye 1-month later showing further increase in the macular traction with resolution of subretinal hemorrhage. (d) Postoperative colour fundus picture at 5 month follow-up showing traction relief at macula with retinal pigment epithelial atrophy.
FFA could be due to traction on the capillaries, which is one of the causes for leakage in ERMs on FFA. On review 1-month later [Fig. 3c], BCVA reduced to 20/600, <N36. SD-OCT revealed thick ERM with a taut posterior hyaloid and increased retinal thickness of 1198 microns [Fig. 5a]. 25 g pars plana vitrectomy with ERM removal was done. With an uneventful postoperative period, 5 months later BCVA was 20/400 N36. Restoration of foveal contour with retinal atrophy [Figs. 3d and 5b] and a slight increase in the hypoautofluorescence due to unfolding of the retina was noted [Figs. 5c and d].

**Discussion**

Vision in Stargardt disease is usually reduced due to macular changes. A thin ERM may not cause additional visual impairment. However a thick ERM may result in further visual reduction. Reports of spontaneous separation of idiopathic ERMs in the young\(^1\) as well as in a child with Stargardt disease\(^3\) suggest observation rather than surgical intervention.

Though a compromised retinal structure hampers the expected results of surgical intervention, some visual improvement can be expected if the deterioration is mainly due to the ERM. As both our patients were on regular follow-up, reduction in visual acuity corresponded to the development of a thick ERM, which further progressed during the short period of observation. Both on OCT and intraoperatively, the ERM appeared to be part of the attached, thickened posterior hyaloid. Case 1 had always had better vision than case 2, and this patient had a significant improvement following surgery.

Subretinal fibrosis and hyperlipofuscinosis have been reported previously in these patients\(^4,5\) following blunt ocular trauma with resultant visual loss suggesting the need for protection from even minor ocular trauma. In our patients, there was no apparent surgical damage to the retinal pigment epithelium, which is depicted by the preoperative and postoperative autofluorescence images. In the second patient, an apparent increase in the area of hypoautofluorescence seen temporally could have been due to unfolding of the retina following release of traction caused by the ERM.

Epiretinal membranes have been described in Stargardt disease and observation rather than surgery has been one option in the past, with isolated reports suggesting limited improvement after surgery. Our experience shows that visual improvement following surgery is possible in these eyes without compromise or worsening of the preexisting pathology.

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