Internal limiting membrane peeling in combined hamartoma of retina and retinal pigment epithelium: Does it make a difference?

Hanan Makhdoum1,2, Yahya Alzahrani3,4

Abstract:
This case study reports a pediatric case of a combined hamartoma of the retina and retinal pigment epithelium (CHR-RPE) successfully treated with pars plana vitrectomy (PPV), membrane peeling, and internal limiting membrane (ILM) peeling. In this rare tumor, adding ILM peeling to the surgical treatment of CHR-RPE with epiretinal membrane may result in a favorable outcome.

Keywords: Combined hamartoma of retina, internal limiting membrane peeling, retinal pigment epithelium

INTRODUCTION
Combined hamartoma of the retina and retinal pigment epithelium (CHR-RPE) is an uncommon, benign, hamartomatous lesion involving the fundus. First named and classified in 1973 by Gass,[1] combined hamartomas are classified based on the location: peripheral, macular, and peripapillary.

These lesions can also be classified according to the type of the predominant tissue: melanocytic, vascular, or glial.[2] Histopathologically, these lesions demonstrate a disorganized retina with dysplastic gliotic vascular tissue, which are infiltrated by cords and sheets of RPE, with duplication of the RPE layer and fibrous tissue proliferation resulting in bridging of the folds on the anterior surface.[3]

These lesions are mostly diagnosed in childhood through clinical examination. They commonly present with retinal distortion, elevation, vascular tortuosity, and gliosis.

In addition, foveal dragging, detachment, macular edema, choroidal neovascularization, vitreous hemorrhage, and retinoschisis have also been reported in these patients.[2,3]

Epiretinal membrane formation is a common finding particularly in the glial subtype, which is amenable to surgical treatment.[2,3] Previous studies have reported conflicting results regarding visual acuity following pars plana vitrectomy (PPV) with epiretinal membrane removal.[2,4‑7]

Here, we present a case with a successful anatomical outcome after pars plana vitrectomy with membrane peeling and internal limiting membrane (ILM) peeling. To the best of our knowledge, this is only the second case reported in the current literature.

CASE REPORT
A 4-year-old male patient born at 32 weeks gestational age through cesarean delivery secondary to eclampsia with a weight of 650 g was previously admitted to neonatal intensive care unit for a month. No systemic illness was previously noted. The patient received Botox injection in both eyes for strabismus 2 years before presentation of current symptoms.

The patient’s vision was noted to deteriorate over time, with the visual acuity recorded as fix and follow for both eyes. Fundus examination of the right eye revealed an elevated lesion in the macula, whitish-gray in color with...
an associated vascular distortion resulting in macular traction [Figure 1a]. Macular optical coherence tomography through the lesion showed disorganized thickened retina with traction [Figure 1b and c]. The examination of the left eye was unremarkable.

The patient underwent a pars plana vitrectomy with membrane peeling and ILM peeling.

Postoperatively, no adverse events or recurrence of the epiretinal membrane were recorded [Figure 2]. Unfortunately, the patient’s corrected vision was limited due to amblyopia and was recorded to be 20/200 at 16 months postoperatively.

**Discussion**

Combined hamartoma of the retina and RPE is commonly associated with epiretinal membrane formation. The treatment of CHR-RPE remains controversial in regard to functional results since it was first named and classified by Gass. In 1984, Schachat et al. in collaboration with the Macula Society reported three patients who underwent pars plana vitrectomy (PPV) with membrane peeling (MP), in which only one demonstrated an improvement of vision from 20/200 to 20/40. McDonald et al. reported two cases that demonstrated poor visual outcome after pars plana vitrectomy with membrane peeling. Those two patients were found to have longstanding poor vision with amblyopia which can explain the outcome.

On the other hand, several previous case reports reported improved visual outcomes. Konstantinidis et al. described two patients that showed improved visual acuity after surgical intervention. One of them had ILM peeling following membrane peeling. More recently, Shields et al. reported the visual outcomes of 79 eyes with CHR-RPE in which one patient demonstrated visual improvement from 20/100 to 20/50 after pars plana vitrectomy with membrane peeling and another had stabilized vision. In 2009, Cohn et al. reported the largest case series of 11 patients who underwent PPV with MP, eight of which demonstrated visual improvement, and three ended up with stabilized vision. The author suggested plasmin-assisted pars plana vitrectomy for cases with dense epiretinal proliferation. Among six patients who underwent plasmin-assisted PPV with MP, four were reported to have improved. Brué et al. published a more recent series of six patients in which all of them demonstrated visual improvement until 4 years postoperatively. Data regarding the recurrence of epiretinal membrane after membrane peeling are limited. In the case series reported by Cohn et al., four of 11 operated eyes developed recurrences of epiretinal membrane despite the use of plasmin. ILM peeling was used by Konstantinidis et al. in one patient, but the case was not specified, and all reported cases improved without recurrence.

Our patient demonstrated an excellent anatomical outcome, where traction was relieved without recurrence, and the retina regained a normal anatomical stratification after ILM peeling despite the limited visual outcome due to amblyopia. The surgical success of ILM peeling can be attributed to the strong adhesions of the epiretinal membrane to the basal lamina of ILM, particularly in young patients.

In summary, we report a young pediatric patient with CHR-RPE that demonstrated a successful anatomical outcome after vitrectomy with ILM peeling.

We speculate that adding ILM peeling to pars plana vitrectomy and membrane peeling in cases of CHR-RPE with traction can lead to more favorable outcomes. Further studies evaluating the outcome after pars plana vitrectomy with membrane peeling and ILM peeling are warranted.

**Literature search methodology**

A search of the MEDLINE database was carried out to identify relevant articles published in peer-reviewed journals. For this search, the term “combined hamartoma of RPE and retina” was used in combination with the terms “surgical treatment,” “LM peeling,” “membrane peeling.”

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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