Case report

Pars plana vitrectomy with intraoperative optical coherence tomography for sub-internal limiting membrane fibrosis excision in a child with Terson syndrome: Surgical and pathological correlation

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ABSTRACT

Purpose: To report the intraoperative optical coherence tomography (OCT)-guided surgery of a consolidated sub-internal limiting membrane (ILM) hemorrhage that developed into a sub-ILM fibrotic membrane in a child with a history of Terson syndrome.

Observations: A one year-old boy with a history of Terson syndrome due to a motor vehicle accident presented three months after trauma with a white feather-shaped membrane in the left macula. Preoperative OCT showed a preretinal hyperreflective tissue at the foveal center. The patient underwent pars plana vitrectomy. After separation of the posterior hyaloid, intraoperative OCT did not show any change in structural components. After peeling the ILM, the fibrotic membrane persisted. A bent 30-gauged needle was used to create a plane of dissection in the adherent sub-ILM membrane, which was then peeled with ILM forceps without complication. Postoperative OCT confirmed complete excision without evidence of macular edema. Pathology results indicated presence of fibrocellular tissue that contained hemosiderin, consistent with old organized hemorrhage as a component of the membrane.

Conclusion and importance: Sub-ILM hemorrhage may persist as a tautly adherent fibrotic membrane that can mimic the appearance of an epiretinal membrane or a chronic subhyaloidal hemorrhage during examination, especially in young children. Intraoperative OCT may aid in select complex macular surgery cases to better delineate the planes of dissection during sub-ILM fibrosis excision.

1. Introduction

Epiretinal membrane (ERM) is a common avascular fibrocellular proliferation that occurs on the surface of the retina and causes retinal thickening and wrinkling, leading to metamorphopsia and decreased visual acuity. ERMs are typically idiopathic and occur predominantly in patients over 50 years of age, in association with posterior vitreous detachment (PVD) and anomalous internal limiting membrane (ILM). PVD and ERM formation in the pediatric population is rare and often associated with an underlying etiology such as trauma, ocular inflammation, intraocular tumors, retinovascular disease or combined hamartoma of the retina and retinal pigment epithelium. These membranes are typically thicker and more adherent when found in children, with a higher incidence of recurrence. In contrast to the common ERM, sub-ILM membrane or fibrosis has only been reported in one case report to the best of the authors’ knowledge. Resolved sub-ILM hemorrhage was suggested to be the origin of the sub-ILM membrane in that report. Causes of sub-ILM hemorrhage include Terson syndrome, vasa valvina retinopathy, blood dyscrasia, shaken baby syndrome and blunt facial trauma. Prolonged contact of the retina with hemoglobin and its catabolites can possibly cause toxic retinal damage, which may be irreversible. In other case reports, sub-ILM hemorrhage from vasa valvina retinopathy resulted in a dome-shaped premacular membranes.

We report a case of a sub-ILM membrane that developed in a one year-old boy with a history of Terson syndrome that was managed surgically without complications.

2. Case report

2.1. Clinical history and examination

A one year-old male with a history of uncomplicated full term birth was involved in a motor vehicle accident (MVA) in Puerto Rico in April
2017 that resulted in cerebral hemorrhage that required a frontoparietal decompressive craniotomy, small frontal lobectomy, and temporary placement of a ventriculostomy. The patient developed bilateral vitreous hemorrhage and was diagnosed with Terson syndrome. After conclusion of his trauma hospitalization, he was transferred from Puerto Rico to our institution in order to undergo complete ocular examination under anesthesia (EUA) and appropriate management.

On examination, the patient grimaced to light in the left eye but not the right eye. Fixation was absent in both eyes. The intraocular pressures as measured with TonoPen were 10 mmHg in both eyes. The anterior segment of both eyes was unremarkable. Dilated fundus exam with 20D and 28D lenses revealed a dense dehemoglobinized vitreous hemorrhage in the right eye that precluded a view of the posterior pole. The left eye fundus had multiple peripheral retinal hemorrhages and a feather-shaped white preretinal membrane in the left eye (Fig. 1A). Fluorescein angiography (FA) revealed avascular retina in the periphery of both eyes without evidence of exudation, though the right eye examination was limited due to the dense vitreous hemorrhage. Ultrasonography confirmed that there was no retinal detachment in either eye, but showed a hyperreflective membrane in the posterior pole of the left eye (Fig. 2). Handheld optical coherence tomography (OCT) of the left eye was remarkable for a preretinal membrane (Fig. 1).

2.2. Surgical management and follow up

After completion of examination under anesthesia, risks versus benefits of surgery of both eyes were explained including extensive discussion of immediate sequential pars plana vitrectomy (PPV) surgery versus waiting for a second surgery on the left eye. The family elected to undergo immediate sequential surgery to minimize possible risks associated to anesthesia and permanent visual loss due to amblyopia.

First, the right eye underwent 25-gauge PPV to remove the vitreous hemorrhage. With an improved view, exam disclosed macular exudates, optic nerve pallor, and attenuated vessels.

The left eye then underwent 25-gauge PPV with posterior hyaloid elevation. After PVD induction, no structural change in intraoperative OCT (Fig. 3) was noted as compared to preoperative testing. Indocyanine green was then used to stain the ILM. After the ILM was completely peeled from the macula with ILM forceps, the sub-ILM fibrotic membrane was still present. A bent 30-gauge one inch long needle was then used to create a dissection plane with assisted visualization from the intraoperative OCT, and to initially separate the sub-ILM membrane from the outer retina. The bent 30-gauge needle easily reached the retina in this pediatric eye. While there is always some risk of damage to the underlying retina with the needle, the use of intraoperative OCT gives the surgeon real-time feedback to minimize this risk. Once a dissection plane was identified, ILM forceps were used to completely excise the membrane. These steps are demonstrated in Fig. 4 and can be viewed on a supplemental video file. A complete fluid-air exchange was performed with a soft-tip cannula. The retinal periphery was noted to be flat without tears. C3F8 was instilled and 0.1 mL triamcinolone acetonide (Triescence, Alcon, Ft. Worth, TX) was instilled via the cannula system prior to closing the sclerotomies.

One month after surgery, the patient reacted to light in the right eye and maintained fixation in the left eye. The right eye fundus exam revealed a clear vitreous cavity, pale optic nerve, sclerotic vessels, exudates along the inferior arcade, and scattered peripheral retinal hemorrhages. The left eye fundus exam revealed no evidence of recurrent
sub-ILM membrane (Fig. 1B). OCT confirmed the absence of the sub-ILM membrane, epiretinal membrane and macular edema 1 month after the surgery (Fig. 1B). 1 year after the initial accident, the patient maintained the visual acuity gains without any change in posterior pole findings.

2.3. Ocular pathology results

The membrane was sent for histologic evaluation, which disclosed a fibrocellular tissue that contained foci of hemosiderin (Fig. 5A) that were positive with Perl’s Prussian Blue stain (Fig. 5B). The trichrome stain was positive, consistent with collagenous composition of the tissue (Fig. 5C). CD68 immunostain was negative, indicating absence of histiocytes. CD31 immunostain was negative, indicating there was no vascular endothelium component. Glial fibrillary acidic protein (GFAP) was negative, indicating absence of glial cells.

3. Discussion

Terson syndrome is defined as vitreous or retinal hemorrhage in the presence of intracranial hemorrhage. The etiology of vitreous hemorrhage is still unclear. However, the most predominant hypothesis involves rapidly increased intracranial pressure that leads to effusion of CSF into the optic nerve sheath. The dilation of the retrobulbar optic nerve mechanically compresses the central retinal vein causing rupture of the retinal capillary endothelium.12,13

We report a case of a one-year old boy with Terson syndrome after MVA that later developed a white sub-ILM fibrotic membrane, which was histologically confirmed to have a hemosiderin component. Hemosiderin is an iron storage complex found most often in macrophages after phagocytosis of erythrocytes, and is especially abundant following hemorrhage.14 Hemosiderin is rarely observed in idiopathic ERMs; previous histological studies on surgically excised ERMs have revealed the main components to be retinal glial or myoblastic retinal pigment epithelial cells.15 The sub-ILM fibrocellular tissue in this case...
was composed of collagen and hemosiderin. The tissue did not have glial cells, endothelial cells, or histiocytes as determined by immunohistochemistry.

There has been one previously reported case of a sub-ILM membrane presenting in a four-year-old boy, which was treated similarly with PPV and membrane excision. Scanning electron microscopy revealed the membrane to be composed of collagenous fibers, fibrinoid deposits, and cell debris containing clusters of hemosiderin. The authors similarly speculated the membrane was caused by absorption and organization of a sub-ILM hemorrhage, though they were unable to determine a cause for the hemorrhage.5

In contrast to ERMs, which have a cellophane-like appearance, the sub-ILM fibrosis presented in this case had a well-demarcated dense white appearance that obscured the underlying retina. Recent reports suggest that excellent anatomic and visual outcomes may be achieved in patients with sub-ILM hemorrhage with early PPV and ILM peel, though no pediatric subjects were present. It is possible that if the child had earlier surgery, the sub-ILM hemorrhage could have been evacuated prior to membrane formation. Due to the difficulties associated with excising highly adherent sub-ILM membranes in the fovea, the availability of intraoperative OCT may give the surgeon an additional safety margin.

Conservative management may be considered in some cases given the risks of vitrectomy, including cataract progression, macular hole formation, retinal detachment, and endophthalmitis, among others. Additionally, in some cases the sub-ILM hemorrhage may resolve without intervention. In this case, surgery was indicated due to absent fixation, age, and likely permanent visual loss due to amblyopia. However, select translucent membranes may be observed.

4. Conclusions

Sub-ILM hemorrhage may lead to formation of a sub-ILM fibrotic membrane composed of collagen and hemosiderin. To our knowledge, this is the first report of sub-ILM fibrosis excision in pediatric Terson syndrome that was surgically excised with the aid of intraoperative OCT.

Patient consent

Informed legal consent was obtained from the patient’s legal guardians for publication of medical record details.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2019.100479.

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