Vitreous hemorrhage in X-linked retinoschisis

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ABSTRACT

Purpose: To report a case of preretinal hemorrhage from extraretinal neovascularization related to capillary non-perfused retina within a large schisis in a pediatric patient with X-linked retinoschisis (XLRS).

Observations: A 4-year old male with an RS1 mutation and XLRS presented with preretinal and vitreous hemorrhage in the right eye. Retinal imaging, including wide angle fluorescein angiography (FA) and optical coherence tomography (OCT), showed vitreoretinal traction on extraretinal neovascularization and capillary non-perfused retina in the schisis cavity. Laser treatment to the non-perfused retina within the schisis was successful in reducing extraretinal neovascularization.

Conclusions: Vitreous hemorrhage is a well-known occurrence in XLRS. Imaging using wide angle FA and OCT were helpful to determine the causes of hemorrhage in order to develop a management plan.

1. Introduction

X-linked retinoschisis (XLRS) is a congenital vitreoretinal dystrophy with an estimated prevalence of 1:5,000 and 1:30,000. Vitreous hemorrhage is a well-recognized occurrence and is believed to arise from vitreous traction on retinal blood vessels in the elevated schisis or on extraretinal neovascularization, and in association with full-thickness retinal breaks. We describe a patient with XLRS who presented with preretinal and vitreous hemorrhage from several causes. Imaging with wide angle fluorescein angiography (FA) and optical coherence tomography (OCT) helped in developing a management plan.

2. Case report

A 4-year old male with XLRS was referred to pediatric retina for vitreous hemorrhage in the right eye (OD). Visual acuity was 20/80 OD and 20/125 left eye (OS). There was a preretinal and vitreous hemorrhage along the inferotemporal arcade abutting an inferotemporal retinoschisis.

An examination under anesthesia with wide angle FA revealed capillary non-perfusion within the schisis cavity and fluorescein leakage from extraretinal neovascularization at the edge of the schisis. OCT demonstrated a vitreous tractional membrane on a retinal vessel superior to the schisis leaflet with underlying hyporeflective intraretinal spaces (Fig. 1). The OS had vitreous veins without traction or capillary non-perfusion (Fig. 1).

It was believed that capillary non-perfusion in the schisis cavity presented a hypoxic stimulus for the formation of extraretinal neovascularization that then bled into the vitreous. Therefore, several sessions of FA-guided laser photocoagulation were performed to the outer retina of the schisis cavity in scatter fashion to treat the non-perfused retina. The extraretinal neovascularization partly resolved. At last follow-up 7 years later, corrected visual acuity was 20/40 OD and 20/250 OS and there was no vitreous hemorrhage (Fig. 2).

3. Discussion

Vitreous hemorrhage has been reported in 3%–21% of these patients with XLRS and proposed to be secondary to retinal tears or vitreous traction on retinal blood vessels or extraretinal neovascularization. This report describes a patient with XLRS and vitreous hemorrhage due to several factors identified by FA and OCT. FA and OCT were important in determining a treatment plan.

In this case, a number of factors likely contributed to extraretinal neovascularization. On FA, there was an area of obvious nonperfused retina within the schisis cavity and leakage for extraretinal neovascularization occurred at the junction of the avascular retina and vascular retina. One cause of neovascularization may be from hypoxia that triggers the expression of multiple angiogenic factors (e.g., VEGF, angiopoietins, erythropoietin) through the stabilization of hypoxia-inducible factors. The separation of the retinal layers may have disrupted retinal perfusion within the schisis cavity and caused hypoxia to...
cells within layers of the retina that express angiogenic factors, including the RPE, glia and ganglion cells, as examples. Extraretinal neovascularization has been identified using wide angle FA in XLRS and in areas of overlying primary retinoschisis by OCT. Traction may also contribute to extraretinal neovascular growth onto a vitreous scaffold since the layers of the retina are less adherent to one another than to the vitreous in XLRS due to a defect in the expression of RS1 and retinoschisin. In our case, vitreoretinal traction was appreciated by OCT. Besides vitreous traction disrupting capillaries within the schisis and causing capillary non-perfusion, the neovascularization may then grow onto the surface of the vitreous. What is unknown is if the separation of different retinal layers is more likely to be associated with extraretinal neovascularization.

Our rationale for using laser photocoagulation to ablate the RPE was to reduce the hypoxic drive from oxygen demand of retina lacking vascular perfusion where the schisis disrupted retinal vasculature. The RPE is perfused largely by the choroid and expresses angiogenic factors in diseases known to affect the inner retina. We propose that laser ablation of the RPE reduced the angiogenic factors from the RPE and may have allowed choroidal oxygen no longer used by the RPE to diffuse to the retina and reduce hypoxic demand.

4. Conclusion

In this patient with XLRS, vitreous traction caused retinoschisis, capillary disruption and capillary non-perfusion within the schisis that contributed to hypoxia induced extraretinal neovascularization. FA and OCT helped to identify the causes of vitreous hemorrhage and develop a management plan. FA-guided laser to capillary non-perfusion in the outer retina of the schisis reduced extraretinal neovascularization and allowed spontaneous clearing of vitreous hemorrhage.

Financial

None.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Declaration of competing interest

None.

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