The Potential Role of CSF in Coloboma-Related Retinal Detachment

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1) Purpose

To describe a case of coloboma-related retinal detachment possibly caused by CSF migration.

2) Background

Choroidal colobomas may result in retinal detachment (RD), either serous or rhegmatogenous in origin. The pathogenic mechanism for serous RD is understood to be related to the presence of a scleral (or lamina cribrosa) defect enabling anomalous communications between intraocular and extraocular spaces. This permits dynamic fluctuations in the gradient between intraocular and intracranial pressures that direct the movement of fluid (vitreous humour or cerebrospinal fluid) into and under the retina. Rhegmatogenous RD occurs due to glial atrophy, schisis, and hole formation in the intercalary membrane with separation of the locus minoris resistentiae from the pigment epithelium

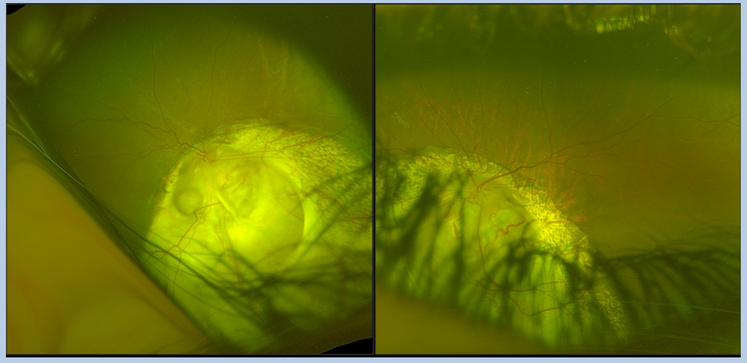


Figure 1: Optos images showing bilateral optic disc and choroidal colobomas

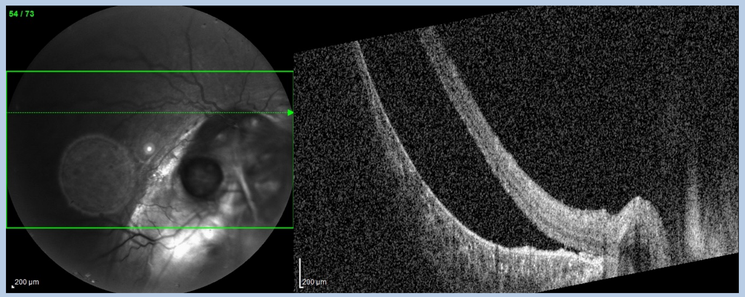
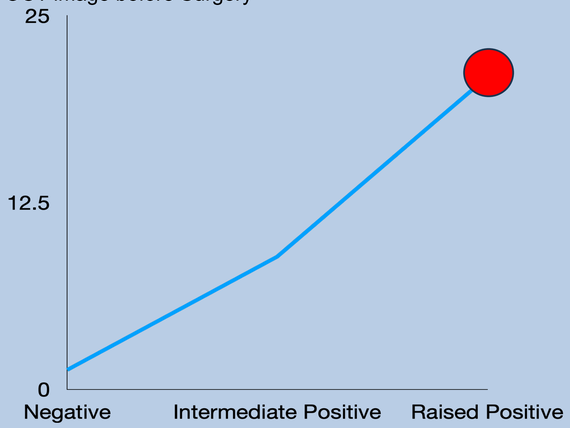


Figure 2: OCT image before Surgery



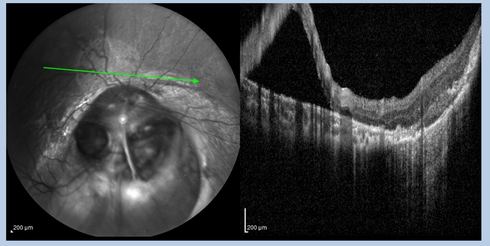


Figure 3: OCT image after surgery

No change in retinal detachment morphology after surgery

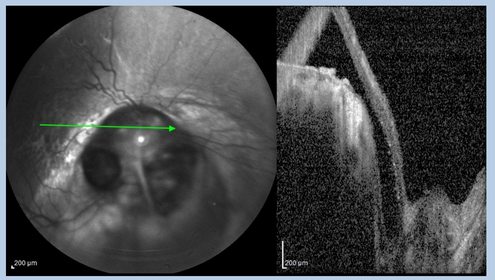


Figure 4: OCT image after surgery

Beta-trace protein levels in CSF (mg/L). <1.30 Negative; 1.31 - 8.88 Intermediate Positive; >8.89 Raised Positive. Patient result (20.9) indicated by red dot.

3) Methods

A 3-year-old boy presented with a coloboma-related retinal detachment in the setting of chronic raised intracranial pressure (ICP) controlled by a ventriculoperitoneal shunt. No retinal break was found on indirect ophthalmoscopy nor on flex-OCT survey of the detachment. Transscleral subretinal drainage and an encircling band were performed without adjunctive retinopexy.

4) Results

Analysis of subretinal aspirate confirmed a raised beta-trace protein, highly indicative of the presence of CSF. Surgical repair was unsuccessful with no change in detachment at three months. Whilst this could represent surgical failure in the presence of an undetected retinal break, the biochemistry result presented an alternative pathogenic mechanism.

5) Conclusions

CSF migration into the subretinal space could present a possible cause in the pathogenesis of coloboma-related retinal detachment in the presence of raised ICP.

References

1. Ectopic cerebrospinal-like fluid from retrobulbar cysts as a possible cause of pediatric retinal detachment associated with optic disc coloboma. Patel, C.K. et al. Arch Ophthalmol. 2012
2. Structural organization of choroidal colobomas of young and adult patients and mechanism of retinal detachment. Schubert, H.D. Transactions of the American Ophthalmological Society. 2005
3. Levels of beta-trace protein in optic disc pit with macular detachment. Makdoumi, K. et al. Acta Ophthalmologica. 2017