



## Transscleral filtration revealing a chorioretinal coloboma

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### ABSTRACT

**Purpose:** We report the case of a 19-year-old patient who presented with an ocular hypotony due to a transscleral filtration through an isolated congenital chorioretinal coloboma in his right eye.

**Observations:** The initial examination showed a decimal best corrected visual acuity (BCVA) decreased to 0.7 and a marked hypotony. A localized infero-nasal chemosis and a conjunctival hyperemia were observed. The fundus examination showed chorioretinal folds and an edematous disc. In the infero-nasal retinal periphery, a chorioretinal coloboma was seen with a full-thickness scleral defect. Ultrasound biomicroscopy showed the area of the coloboma through which the percolation of fluid occurred.

**Conclusions and importance:** A favorable outcome was observed within 6 weeks and BCVA improved to 1.0 three months later. The intraocular pressure (IOP) increased to 11 mmHg, but the bleb-like filtration could still be seen together with some persistent chorioretinal folds. Ultrasound Biomicroscopy (UBM) imaging was helpful to understand the mechanism of this uncommon complication of a coloboma.

### 1. Introduction

Colobomas have been reported in 0,5<sup>1</sup> to 2,4<sup>2</sup> per 10 000 live births. It can present in various forms, with chorioretinal colobomas accounting for 69% of cases.<sup>3</sup> We report a rare case of transscleral filtration and hypotony which led to the diagnosis of a chorioretinal coloboma.

### 2. Case report

A 19-year-old patient was referred to our department with a diagnosis of acute hypotonic uveitis of the right eye. He complained of a mild discomfort and a redness of his right eye, together with a decreased vision. His symptoms followed his fall from a ladder — on his right shoulder, without direct trauma to the head or to the eye — just before the onset of his symptoms. The patient had no significant medical history.

His best corrected decimal visual acuity (BCVA) was to 0.7 in his right eye and 1.0 in his left eye. The slit-lamp examination of his right eye showed a localized infero-nasal conjunctival hyperemia with a slight chemosis, which suggested a filtration bleb (Fig. 1). The intraocular pressure (IOP) was decreased to 4 mmHg. Descemet folds were observed as well as an anterior chamber inflammation with a 1+ flare and 0.5+ cells. The anterior chamber was relative shallow comparing to

the left eye. The examination of the fundus and OCT imaging showed choroidal folds (Fig. 2). A chorioretinal coloboma was seen in the infero-nasal periphery in the same area where the chemosis was observed. Fig. 3 shows the wide-field fundus photography and the fluorescein and indocyanine green angiographs. The examination of the left eye was unremarkable with an 18 mmHg IOP, in particular, no coloboma was observed. Ultrasound biomicroscopy of the right eye confirmed a scleral defect in the area of the coloboma (Fig. 4) and OCT imaging of the filtration bleb showed subconjunctival cysts (Fig. 5).

Topical corticosteroids were prescribed but had no effect on the anterior chamber inflammation. We hypothesized that the filtration occurred after the patient's fall and could spontaneously regress. Indeed, after a 6-week follow-up the IOP increased to 11 mmHg and decimal BCVA recovered to 1.0. The anterior chamber became completely quiescent only at the 3-month follow-up, while some residual macular folds were observed as well as a fine residual localized chemosis.

### 3. Discussion

Chorioretinal colobomas are caused by a failed closure of the embryonic fissure during the fifth and seventh weeks of fetal development.<sup>4</sup> The defect can be anywhere between the optic disc and the infero-nasal edge of the iris. It is localized in the retina and/or choroid, sometimes

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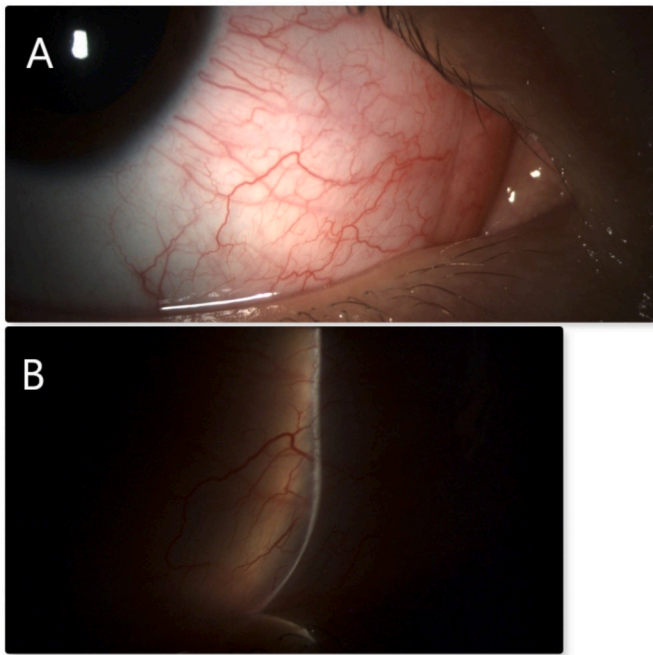
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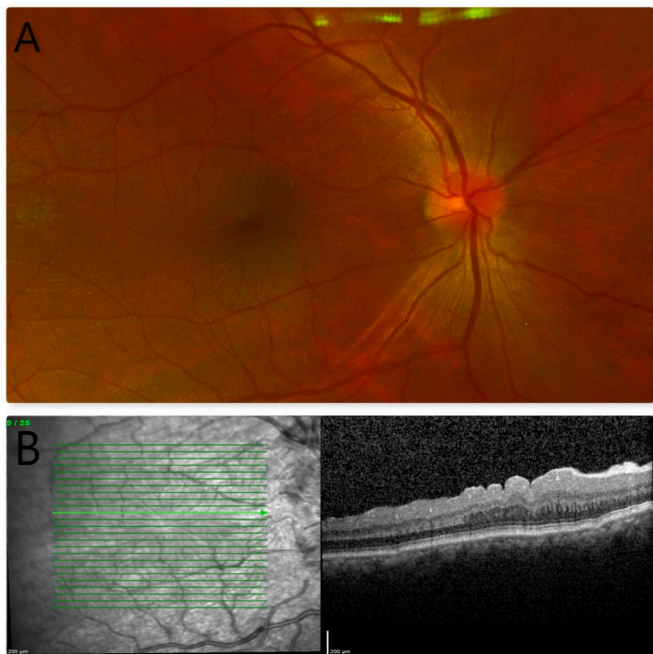
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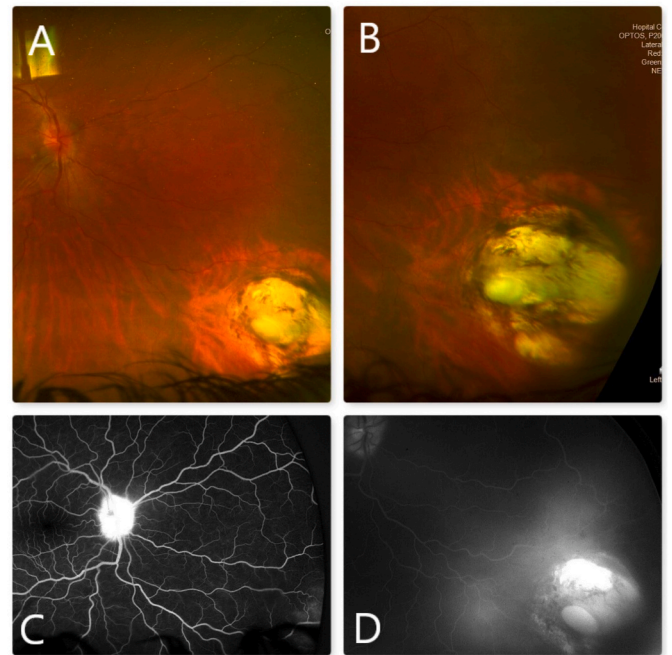
**Fig. 1.** Slip-lamp examination A: Infero-nasal conjunctival redness. B: elevated conjunctival surface due to the filtering bleb.



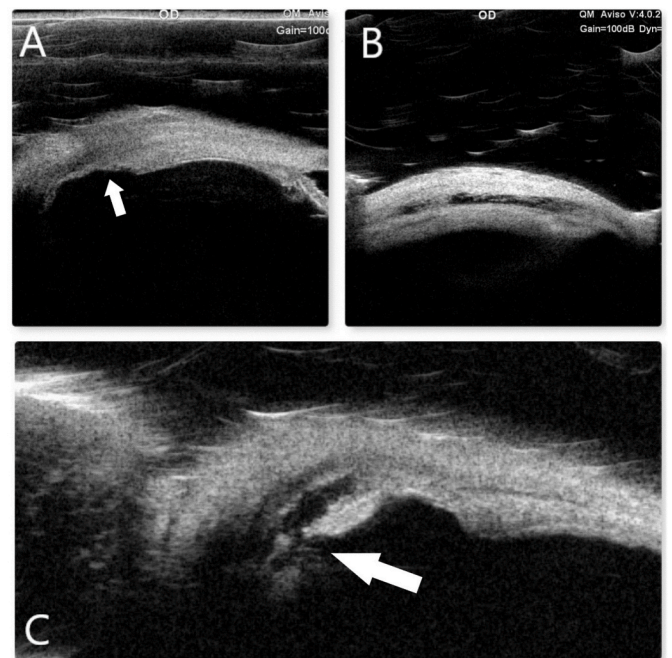
**Fig. 2.** A: Fundus photography with radial macular retinochoroidal folds and edematous disc related to the hypotony. B: OCT imaging of radial retinochoroidal folds.

with an undifferentiated retinal intercalary membrane covering the sclera. As in our case, the most common location of colobomas is in the infero-nasal quadrant.<sup>5</sup>

Pathological studies have shown that a central tear in the inner layer of the intercalary membrane, together with a ruptured outer layer at the coloboma's edge can lead to a full thickness defect.<sup>6,7</sup> In our case, we hypothesize that our patient's trauma led to the rupture of the weakened and thinned sclera and/or of the intercalary membrane. This resulted in



**Fig. 3.** Ultra-wide field imaging of the infero-nasal coloboma A&B color imaging - chorioretinal atrophic colobomatous area measuring 3 disc diameters. C&D Fluorescein angiography - C: edematous disc. D: Window defect in the area of the coloboma. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)



**Fig. 4.** UBM imagingA: Extreme thinning of the sclera at the site of the coloboma (arrow). B: Filtering bleb leading to the formation of microcysts in the subtenon space. C: Chorioretinoscleral fistula (arrow) with percolation of liquefied vitreous/aqueous humor.

the hypotonia with the liquefied vitreous and/or aqueous humor filtering through the retina and the scleral defect, with a filtering bleb as long as the percolation of the liquid lasted.

Rhegmatogenic retinal detachments are more frequent complications of chorioretinal colobomas, observed in as much as 8–43% of

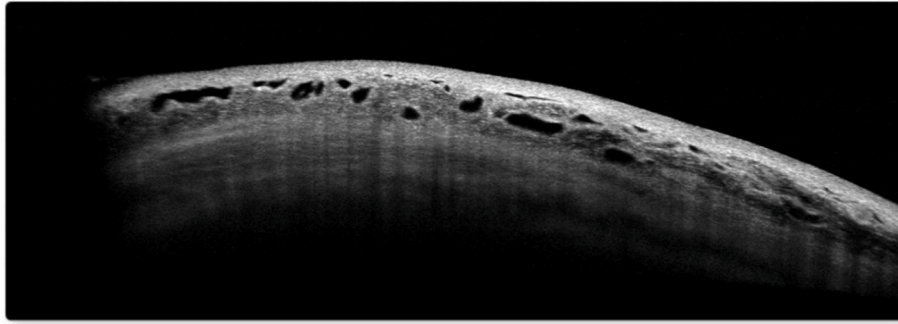


Fig. 5. Conjunctival OCT of the right eye: chemosis with a microcystic conjunctiva, resembling a post-surgical filtration bleb.

cases.<sup>4,8–10</sup> The spontaneous perforation of a chorioretinal coloboma as observed in our case is much rarer: to our knowledge, only 9 cases have been reported since 1985.<sup>11–19</sup> Most reports include various techniques of surgical interventions with the objective of closing the scleral defect. Some authors have also performed laser prophylaxis around the coloboma.<sup>20</sup> In our case, because of the mild symptoms, we chose to abstain from any therapeutic intervention. Such a spontaneous favorable outcome has already been reported only once in a 31-year-old patient who presented with mild symptoms as in our case.<sup>18</sup> The plugging of the perforation by non-liquefied vitreous and/or the intercalary membrane could explain the suspension of the filtration process.

UBM imaging of such a filtering coloboma had not been previously been shown. Our case suggests that modern imaging techniques can guide therapeutic decisions in the management of post-traumatic transscleral filtering secondary to colobomas.

#### 4. Patient consent

Consent to publish this case report has been obtained from the patient in writing. This report does not contain any personal identifying information.

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#### Authorship

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

#### Declaration of competing interest

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