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Case Report

Congenital Chorioretinal Separation in Extreme Prematurity

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Received: March 11, 2022; **Published:** May 26, 2022

Abstract

Peripheral discontinuity of the choroid, retinal pigment epithelium, and outer retinal layers is highly unusual. We report a case of unilateral curvilinear mid-peripheral chorioretinal interruption in the setting of extreme prematurity.

Keywords: *Congenital Chorioretinal Separation; Prematurity; Retinal Pigment Epithelium*

Introduction

Peripheral discontinuity of the choroid, retinal pigment epithelium and outer retinal layers occurs in unusual congenital conditions such as colobomata [1] and inherited disorders of connective tissue such as Stickler's syndrome [2] and Wagner's syndrome [3]. We report a case of unilateral curvilinear midperipheral chorioretinal interruption in a very low birth weight infant of extreme prematurity.

Case Report

A 4-week old baby girl (gestational age 26 weeks, birthweight 530 grams), and her twin sister underwent screening examinations for retinopathy of prematurity (ROP). The pregnancy had been complicated by pre-eclampsia, and the twins were delivered via emergency Cesarean section secondary to fetal distress. The course of the child in question was further complicated by grade IV perinatal intraventricular hemorrhage, neonatal seizures and chronic respiratory difficulties.

External ocular exam revealed a limited blink to light in both eyes, but was otherwise unremarkable. Further ophthalmic examination identified clear corneas and lenses bilaterally. Both optic discs were of age-appropriate size and coloration. However, we noted in the right eye mild retinal vascular tortuosity, in addition to a prominent 9-o'clock hour circumferential interruption of retinal pigment epithelium (RPE) and posterior retina, with bridging choroidal and retinal vessels (See figure 1 and 2). The margins of the break were smooth, without secondary pigmentation. Retinal examination of the left eye was unremarkable. Of note, the child's twin sister's examination revealed incomplete retinal vascularization consistent with Stage? retinopathy of prematurity, but was otherwise age-appropriate.

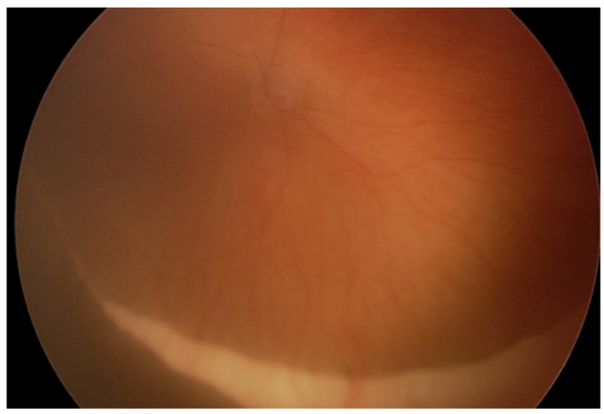

Figure 1

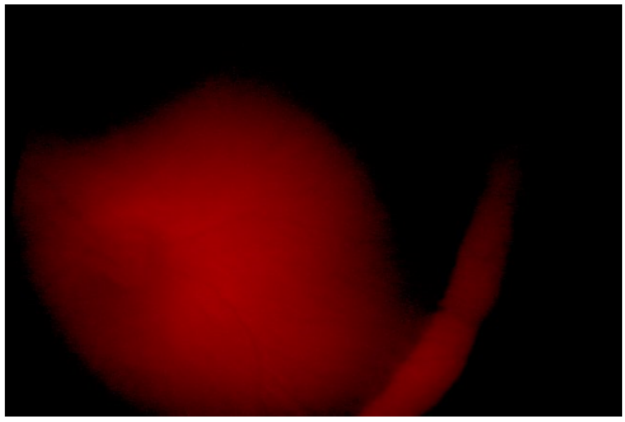

Figure 2

Figure 1 and 2: *Clinical Ret-Cam photograph of the patient's right fundus at 4 weeks of age demonstrates mid peripheral chorioretinal separation with intact retina peripheral (anterior) to the separation. Bridging retinal vessels are apparent across the separation.*

Clinical course was marked by severe developmental delay and microcephaly. Poor visual interest was apparent by 6 months of age, and the absence of any cortical responses on visual evoked testing supported a diagnosis of cortical visual loss.

Examination under anesthesia at 12 months of age revealed a mildly shallow right anterior chamber, widening of the chorioretinal separation with secondary pigment deposition, and scalloping of the anterior and posterior edges (See figure 3 and 4). Bilateral optic pallor was evident. Retinoscopy revealed approximately 10.50 to 11.50 diopters of myopia, and an axial length of 21.43 mm in the eye with the retinal anomaly. In comparison, the average axial length for infants aged 10 - 18 months is 20.14 mm [4].

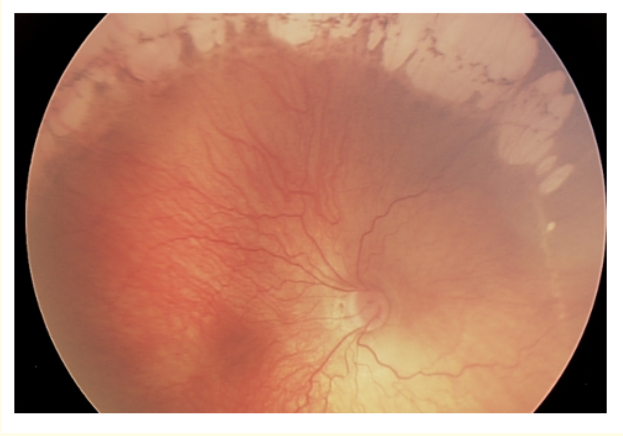

Figure 3

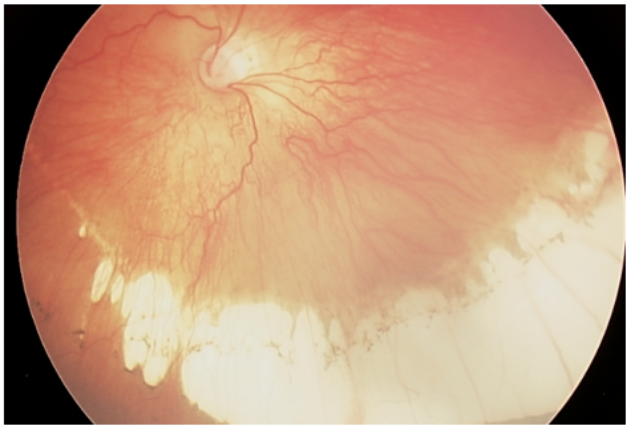

Figure 4

Figure 3 and 4: *Fundus photographs at 12 months of age, demonstrating widening of the chorioretinal separation with scalloping of the borders and pigment deposition.*

Discussion

To our knowledge, this is the first reported case of non-colobomatous congenital chorioretinal separation. Giant tears of the retinal pigment epithelium have been rarely described, primarily in relation to age-related macular degeneration [5], but also following glaucoma surgery [6], in association with choroidal effusion [7], blunt trauma [8], acute retinal necrosis [9] and panuveitis [10]. There have been numerous reports establishing the relationship between ocular pathology and risk factors such as very low birth weight, extreme prematurity, and intraventricular hemorrhage, all of which were present in our patient. However, none list chorioretinal separation as an observed complication [11-16]. Thus, given the patient's clinical history, the etiology of such a break is unclear. Although RPE and outer retinal atrophy may occur secondarily due to failure of choroidal vascular development, there was no apparent reason for the choroid to be selectively absent in the areas corresponding to the observed separation. Similarly, eclampsia has been proposed before as a cause of retinal and choroidal vessel ischemia, leading to an ischemic pigment epithelial detachment [17], but it has never before been documented. Blunt trauma during the Cesarean section might also have produced such a break, but there was no external or internal evidence of such perinatal trauma. We hypothesize that horizontal traction caused a break with continued traction producing an "unzippering" effect.

The patient's unilateral high myopia provides interesting comparisons to more common instances of myopia of prematurity. Extreme myopia occurs with relatively high frequency in the setting of prematurity, particularly following cryotherapy or laser treatment of retinopathy of prematurity. Quinn and others have suggested that ROP treatment produces a mechanical stunting of development of anterior segment structures, occurring at a time and anatomic location normally characterized by significant growth. Evidence for this theory includes signs of anterior segment maldevelopment (microcornea, steeper corneas, shallow anterior chamber, and increased lenticular curvature), as well as the relative absence of axial elongation in the face of extreme myopia [17]. The preservation of bridging chorioretinal vasculature suggests that these elements are not primarily involved in the anterior segment dysgenesis.

Conclusion

This case report of congenital chorioretinal separation in extreme prematurity adds to the literature of this unusual condition. Examiners of retinopathy of prematurity should be aware of this clinical presentation.

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Volume 13 Issue 6 June 2022