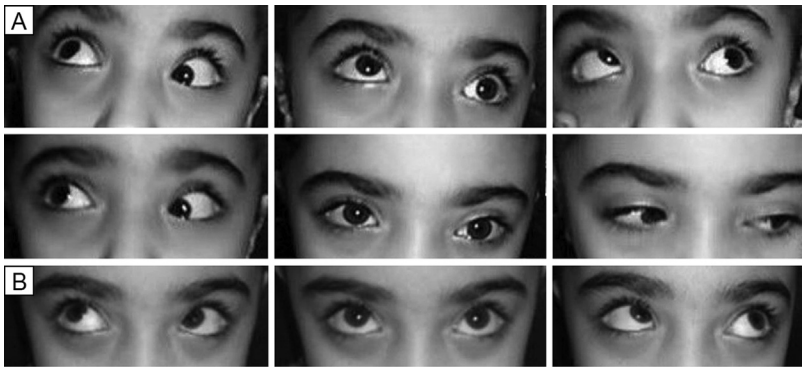

FIG 1. A, Ocular motility examination at presentation. There was marked limitation of elevation of the left eye in adduction, moderate limitation of elevation in the primary position, and mild limitation of elevation in abduction. There was also downshoot in adduction, mild hypotropia in the primary position, with normal abduction. B, Ocular motility 1 week following treatment. There was marked improvement of elevation of the left eye in adduction, with near normal elevation in the primary position, and in elevation in abduction.

repeated orbital methylprednisolone acetate injections have been reported in children.[9]

While spontaneous improvement of acquired Brown syndrome has been previously reported,[2] the rapid and dramatic improvement of the condition following treatment in this case suggests that improvement was due to treatment with a single intramuscular systemic depot steroid injection.

Literature Search

PubMed was searched on January 5, 2019, without date restriction, for English-language results, using the following terms: *acquired Brown syndrome*, *treatment*, *systemic steroids*, *intramuscular systemic steroid*, and *depot steroid injection*.

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Circumferential retinal hemorrhages after ophthalmic examination with scleral depression in an infant with anti-VEGF treated retinopathy of prematurity

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We report a case of retinal hemorrhages in a baby with retinopathy of prematurity (ROP) following examination with indirect ophthalmoscopy and scleral depression. There have been rare reports of examination-induced retinal hemorrhages during ROP screening, although those hemorrhages were diffusely scattered in the posterior pole. In this report the hemorrhages were found on the surface of the neovascular ridge. Changes in intraocular pressure caused by scleral depression may result in rupture of the fragile and immature retinal vessels, which have poor autoregulation in these premature babies. Ophthalmologists performing ROP screening examinations should be

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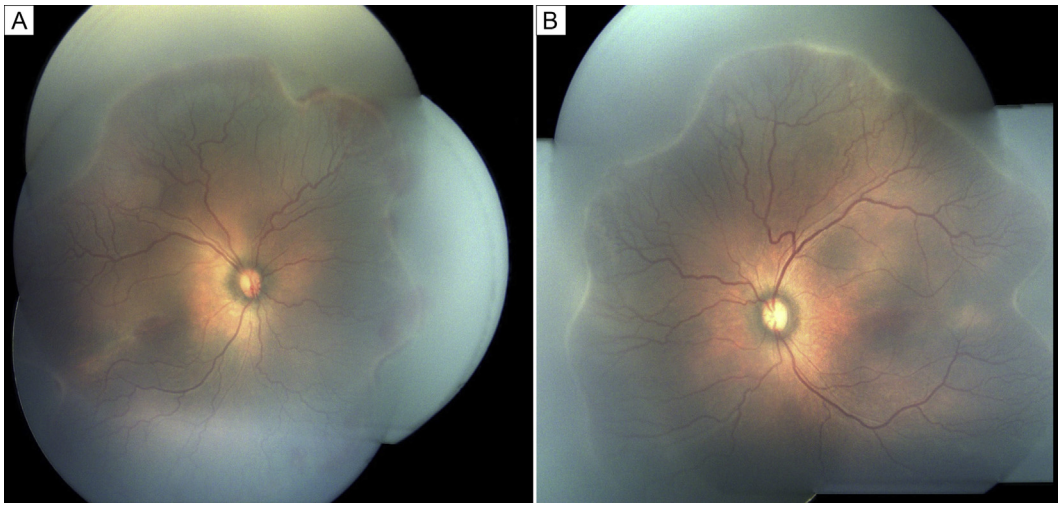

FIG 1. Fundus photographs of the right eye (A) and left eye (B) showing zone I, stage 3 retinopathy of prematurity (ROP) for 12 clock hours with plus disease (aggressive posterior ROP).

aware of the possibility of causing retinal hemorrhages with scleral depression, although the hemorrhages will resolve spontaneously.

Case Report

A 10-week-old girl, born at a gestational age (GA) of 25 weeks and a birth weight of 718 g, underwent a follow-up retinopathy of prematurity (ROP) examination with scleral depression at Jackson Memorial Hospital/Bascom Palmer Eye Institute. The newborn was delivered by cesarean section due to placental abruption. There was no history of trauma, sepsis, or other bleeding. Perinatal medical history included patent ductus arteriosus (treated with ligation), apnea of prematurity, anemia of prematurity, seizure-like activity, and mild metabolic bone disease. She received supplemental oxygen during her neonatal intensive care unit stay.

Both eyes had stage 3, zone I ROP for 12 clock hours with plus disease (aggressive posterior ROP. See Figure 1). Anterior segment examination was unremarkable. The baby was first screened at 32 weeks' GA and then treated with intravitreal bevacizumab 0.625 mg/0.05 ml in both eyes at 33 weeks, with resolution of plus disease. During a follow-up fundus examination at 35 weeks, 2 ophthalmologists performed scleral depression prior to photographic documentation with the RetCam (Clarity Medical Systems, Pleasanton, CA). Neither ophthalmologist noted retinal hemorrhages during the time of examination. At the time of RetCam photography, just minutes after scleral depression, circumferential retinal hemorrhages were noted at the surface of the neovascular ridge in the left eye (Figure 2).

Complete blood count did not show evidence of thrombocytopenia. During the admission, the baby remained afebrile, and a full sepsis workup (performed due to perinatal hypotension) was negative. No additional workup or neuroimaging was deemed necessary, given the immediate appearance of the retinal hemorrhages after scleral depression. The hemorrhages resolved approximately 2 weeks later.

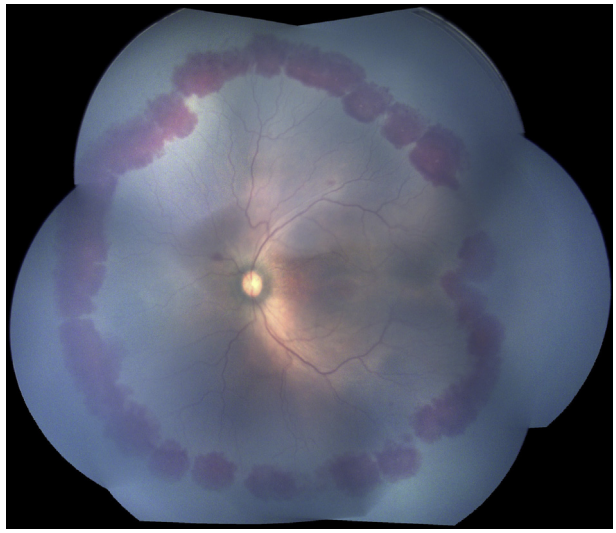

FIG 2. Fundus photograph showing a circular pattern of retinal hemorrhages arising from the neovascular ridge in the left eye.

Discussion

There are rare case reports of widespread posterior pole retinal hemorrhages following ophthalmic examination with scleral depression in babies with active ROP.[1,2] Similarly, retinal hemorrhages have been reported in association with laser photocoagulation in a case of threshold ROP, in which the hemorrhages were postulated to be secondary to scleral depression.[3] Retinal hemorrhages have been reported after RetCam screening as well.[4,5] Our

case differs from previous reports in that the hemorrhages appeared in a circumferential pattern along the neovascular ridge, similar in appearance to a beaded necklace.

The exact mechanism of the retinal hemorrhages is not fully understood; however, it may be related to changes in intraocular pressure (IOP) during scleral depression, which may damage the immature pathologic vessels seen in babies with ROP. Immature retinal vasculature is relatively fragile because of a lack of structural support from smooth muscle, collagen, pericytes, and elastin, making it more susceptible to rupture.[6] An immature autoregulatory system in preterm babies may further predispose to ischemia and hemorrhage.[7]

The baby in this report was treated with intravitreal bevacizumab 2 weeks prior to the incident, further differentiating this case from previous reports. One might have expected the vascular endothelial growth factor inhibitor to lower the risk of bleeding in this baby, because it causes regression of neovascularization. It is possible, though, that the bevacizumab increased the fragility of the vessels. We speculate that this bleeding was caused by the trauma of the examination, which also may have been exacerbated by pressure from the lid speculum and RetCam. A very high IOP during repeat scleral depression, followed by release of that tension could have caused the hemorrhages.

Physicians performing ROP examinations should be aware of the possibility of causing retinal hemorrhages with scleral depression and proceed with appropriate caution. The retinal hemorrhages resolved spontaneously in all reported cases, including the current case. To the best of our knowledge, examination-induced retinal hemorrhages in children have never been reported in the absence of active ROP.

Literature Search

PubMed was searched on November 5, 2018, without date restriction, for English-language records using the following terms: *retinal hemorrhages* AND *scleral depression*.

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Exophthalmos in Kearns-Sayre syndrome

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Kearns-Sayre syndrome (KSS) is a rare mitochondrial DNA (mtDNA) deletion syndrome that typically presents before 20 years of age and is characterized by chronic progressive external ophthalmoplegia, pigmentary retinopathy, and a combination of cardiac conduction defects, cerebellar ataxia, and elevated cerebrospinal fluid protein levels. The mtDNA defects interfere with oxidative phosphorylation and can affect a number of cellular energy processes in various organs. We report the case of a 15-year-old girl with KSS that was uniquely associated with bilateral, symmetrical exophthalmos.

Case Report

A 15-year-old girl presented emergently at Montefiore Medical Center with bilateral eye pain. She was in the United States from the Caribbean visiting relatives, who noted more pronounced proptosis and ptosis than they recalled seeing several years before. The patient reported that these features had progressed gradually over 4 years' time, but pain began only 1 month prior to presentation. She had no known relevant medical or developmental history, and there was no relevant family history.

On examination, visual acuity was 20/30 in the right eye and 20/25 in the left eye. The examination was significant for bilateral ophthalmoplegia (Figure 1), exophthalmos (27 mm in each eye by Hertel exophthalmometer), ptosis (right eye, MRD1 −3; left eye, MRD1 −2), and a lack of diplopia. Alternate cover testing did not reveal any movement because of the ophthalmoplegia; however, exotropia of approximately 30^{Δ} in primary gaze was measured by Hirschberg testing. On slit-lamp examination, there was no conjunctival injection or chemosis. No corneal microcysts were present; however, there were 1+ punctate epithelial erosions over the inferior cornea bilaterally. Dilated fundus examination disclosed bilateral pigmentary retinopathy. Magnetic resonance imaging of the brain and orbits revealed global cerebral and cerebellar volume loss,

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