Retinal Pigment Epithelial Tear in Shaken Baby Syndrome

Retinal hemorrhages are the most commonly reported ocular findings in shaken baby syndrome (SBS) in children. The intraocular hemorrhages can vary considerably not only in size and severity but also in location. Shaken baby syndrome can also cause several structural retinal abnormalities including perimacular folds, traumatic retinoschisis, vitreomacular traction, retinal pigmentary changes, and macular holes. Although these observations have been well documented in the literature, retinal pigment epithelial (RPE) tears have not been described in the setting of SBS.

Report of a Case. A full-term, 6.5-month-old male infant was found unresponsive at home under the care of his mother and grandparents. On his arrival at the hospital, he was found to have bilateral intracranial hemorrhages and obstructive hydrocephalus. Ophthalmologic examination revealed a dense vitreous hemorrhage overlying the macula in the right eye, subhyaloid and subinternal limiting membrane hemorrhage in the left eye, and 4 quadrants of intraretinal hemorrhages in both eyes. A diagnosis of SBS was made based on the history from witnesses and clinical examination.
Six weeks after the initial hospital admission, examination under anesthesia confirmed that the vitreous hemorrhage in the right eye and the subhyaloid and subinternal limiting membrane hemorrhage in the left eye failed to resolve (Figure 1). Owing to the amblyogenic potential of the nonclearing intraocular hemorrhages, the patient underwent lens-sparing vitrectomy in both eyes. The fundus and fluorescein angiographic findings are shown in Figure 2. Hyperfluorescence is apparent in the base of the RPE window defect in the arteriovenous phase (Figure 2B). Blockage of choroidal fluorescence is apparent nasal to the crescent-shaped window defect, consistent with retraction of the RPE. By way of comparison, choroidal ruptures are typically curvilinear and concentric with the optic nerve, with hypofluorescence during the early phase of the fluorescein angiogram due to disruption of the choriocapillaris.

Comment. Retinal pigment epithelial tears are a well-known complication of neovascular macular degeneration, occurring both spontaneously as well as following treatment. They may occur as a consequence of several different mechanisms, including blunt trauma in adults, but have never been described in SBS. In neovascular macular degeneration, new blood vessels invade Bruch’s membrane and may lead to a serious RPE detachment, at which point a tear occurs at the junction of attached and detached pigment epithelium. In the era of anti-vascular endothelial growth factor treatments, sudden contraction of the fibrovascular complex may accelerate the tractional forces contributing to an RPE tear. Vitreomacular traction may also induce direct mechanical forces at the edge of a pigment epithelial detachment, leading to an RPE tear.

Owing to the strong adherence of the vitreous to the retina in infants, the hemorrhages that occur in SBS are thought to be caused by the mechanical effect from shaking through repeated vitreoretinal traction forces. We hypothesize that a possible mechanism for the development of an RPE tear in SBS is clot retraction contraction of the subretinal hemorrhage, producing horizontal tractional forces leading to an RPE tear. We are unable to definitively determine whether the RPE tear occurred at the time of trauma or some time thereafter due to the vitreous hemorrhage until vitrectomy was performed.

Our case illustrates a previously unreported but clinically significant finding related to SBS that may have prognostic relevance for visual outcomes. Unfortunately, infants who are victims of SBS often have structural damage to the retina, optic nerve, or posterior visual pathways that may further contribute to limitations in visual and functional potential.

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We report a case that demonstrates the appearance of perimacular folds associated with extensive retinal hemorrhages occurring in the absence of trauma in a patient with acute myeloid leukemia. We discuss the clinical features, relevant literature, and suggested mechanisms for perimacular folds in such a situation.

Report of a Case. A 14-year-old boy with acute myeloid leukemia visited the eye department with blurred vision of recent onset. Visual acuity was 6/48 OD and 6/18 OS. Examination revealed bilateral extensive retinal hemorrhages over the posterior pole involving all retinal layers (Figure 1). The patient’s right eye had a dome-shaped cavity centered over the macula. This cavity was filled with fluid blood and surrounded by a raised ridge outlined by a shiny retinal reflex resembling a perimacular fold (Figure 1A). The patient had no history of head injury and no signs of intracranial hemorrhage. Hematological investigations showed a markedly reduced platelet count (18 × 10^3/μL; to convert to × 10^6 per liter, multiply by 1.0) and hemoglobin level (6.7 g/dL; to convert to grams per liter, multiply by 10.0); the prothrombin time was 32 seconds (reference range, 26-35 seconds).

Six weeks later, the premacular hemorrhage in the right eye persisted (Figure 2A). Nd:YAG laser membranotomy drained the blood into the vitreous cavity and his visual acuity improved to 6/6 (Figure 2B).

Comment. Traumatic retinoschisis with perimacular fold has been described following abusive head injury, particularly when characterized by the repetitive acceleration and deceleration forces seen in shaken baby syndrome. A similar ophthalmoscopic appearance was reported twice in fatal crush head injury of childhood and in adult Terson syndrome. In shaken baby syndrome, violent shaking is believed to generate shearing forces at the vitreoretinal interface, resulting in traction on the retina that causes traumatic retinoschisis and creates perimacular retinal folds. Retinoschisis may involve only separation of the internal limiting membrane.

Subinternal Limiting Membrane Hemorrhage With Perimacular Fold in Leukemia