Six weeks after the initial hospital admission, examination under anesthesia confirmed that the vitreous hemorrhage in the right eye and the subhyaloid and subtemporal 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 serous RPE detachment, 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.

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 × 10^9/L; to convert to × 10^11/L, 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 13 seconds (reference range for this age group, 9-13 seconds) and the activated partial thromboplastin 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 membraneotomy 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.
(ILM) or a more sight-threatening split in deeper retinal layers. Elevated retinal blood vessels over the cavity aid in differentiation.

We believe the reduced platelet count in our patient caused extravasation of blood from retinal vessels, resulting in rapid accumulation of blood in the sub-ILM space in the right eye. This raised the ILM from the inner retina and created a perimacular fold at the edge where the ILM remained attached.

Retinal hemorrhages at any level are often seen in leukemia with thrombocytopenia. It is unlikely that the appearance of a perimacular fold is unique to this setting. It may be seen in eyes with rapid accumulation of large premacular sub-ILM hemorrhage due to any cause and may be more common than reported in the literature. Published photographs of a large premacular hemorrhage seen in a patient with lymphoma and in Valsalva retinopathy show what appears to be a perimacular retinal fold. The fact that this has not been commented on in these reports perhaps reflects the lack of familiarity with this sign among ophthalmologists not used to seeing patients with shaken baby syndrome.

Figure 1. Fundus photographs showing extensive retinal hemorrhages over the posterior poles of both eyes. A, The right eye has a dome-shaped cavity centered over the macula. The cavity is filled with fluid blood and surrounded by a raised ridge outlined by a shiny retinal reflex resembling a perimacular fold (arrow). B, The left eye has superficial flame-shaped hemorrhages and a premacular hemorrhage. No perimacular fold is present.

Figure 2. Fundus photographs showing persistent premacular blood 6 weeks later (A) and showing the eye after Nd:YAG laser membranotomy to drain blood into the vitreous cavity (B). Note that the appearance of the perimacular fold (arrows) remained unchanged.
This case highlights the facts that perimacular folds may be seen with large sub-ILM hemorrhage in a non-abusive setting and that hematological diseases should be excluded in all cases. However, the term retinoschisis resulting in perimacular fold may be reserved for traumatic settings where shearing forces at the vitreoretinal interface cause perimacular folds.

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Intracisternal Irrigation of Papaverine Leading to Choroidal Infarction

Papaverine hydrochloride, a potent vasodilator whose mechanism of action is thought to arise from inhibition of cyclic adenosine monophosphate and cyclic guanosine monophosphate phosphodiesterases in smooth muscle, is the most researched pharmacologic agent for the treatment of cerebral vasospasm. The successful use of intracranial irrigation of papaverine by neurosurgeons to prevent vasospasm after clipping of a cerebral aneurysm has been demonstrated, leading to its widespread use. Although it is gaining popularity, its ocular complications still remain unclear. Transient cranial nerve palsies have been documented after papaverine irrigation. These cranial nerve abnormalities include facial nerve palsy and ipsilateral, bilateral, and contralateral pupillary dilation. One case of unexplained transient monocular blindness has been reported. We report a new concern of choroidal infarction causing permanent vision loss after intracranial irrigation with papaverine.

Report of a Case. A 55-year-old woman with no visual symptoms visited our medical center with an 8-mm left middle cerebral artery aneurysm. The neurosurgical team performed surgical clipping of the aneurysm and intracranial irrigation with 3 mL of papaverine hydrochloride (30 mg/mL) to prevent postoperative vasospasm. The middle cerebral artery was accessed via a transfrontal approach whereby the neurosurgical team created a small keyhole in the sphenoid bone. The following day, the patient’s visual acuity was no light perception, she had proptosis, and she had a frozen globe. Fundus examination results were initially unremarkable. Orbital computed tomography revealed left orbital swelling. Two days postoperatively her proptosis resolved spontaneously, although her visual acuity remained no light perception. When stable 1 week postoperatively, fundus photography of her left eye revealed arteriolar narrowing and diffuse areas of retinal pigment epithelial hyperpigmentation and atrophy, which was more pronounced in the macula (Figure 1). Fluorescein angiography revealed delayed retinal filling and areas of hyperfluorescence and hypofluorescence correlating to the changes seen on color photography.