Retinal Folds in the Shaken Baby Syndrome

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We examined two children with presumed shaken baby syndrome. Both children suffered severe, indirect closed head trauma with intracranial hemorrhage, sharply increased intracranial pressure, and extensive neurologic damage. In addition to extensive retinal and preretinal hemorrhages, bilateral symmetric white ring-shaped retinal folds were seen encircling the macula outside the vascular arcades. These retinal folds may be a hallmark of shaking injuries in child abuse victims.

OCULAR MANIFESTATIONS of the shaken baby syndrome consist primarily of diffuse bilateral subretinal, intraretinal, preretinal, and vitreous hemorrhages.[1,2] Intraretinal blood-filled schisis cavities have also been described.[3] Typically, signs of external or anterior segment ocular trauma are absent. We studied two cases of bilateral symmetric ring-shaped retinal folds in presumed child abuse victims.

Case Reports

Case 1

A 4-month-old girl was hospitalized for respiratory arrest. According to the parents, the child's babysitter noted that the baby suddenly became cyanotic while crying. She immediately turned the child upside down and performed the Heimlich maneuver. The child made no ventilatory effort and paramedics were called. In the emergency room, the baby was comatose and was undergoing grand mal seizures. There were no external signs of contusion or other trauma. Computed tomography demonstrated a right frontal subarachnoid and tentorial hemorrhage. The child was intubated and monitored for increased intracranial pressure. Subsequently, she underwent placement of a ventriculoperitoneal shunt.

Ophthalmologic examination on the second hospital day showed no response to bright light. The pupils were fixed at 5 mm. There were no external signs of ocular trauma and results of the anterior segment examination were unremarkable. Ophthalmoscopic examination showed bilateral diffuse subretinal, intraretinal, and preretinal hemorrhages. Elevated ring-shaped white retinal or subretinal ridges were also noted surrounding both maculae. It appeared as though the retinas had folded over themselves, forming a circular ridge around each posterior pole. The vitreous was clear.

Follow-up examination three weeks later showed dense vitreous hemorrhages bilaterally, which prevented visualization of the fundus. B-scan ultrasonography showed vitreous hemorrhage and a slight elevation of the retina in both posterior poles corresponding to the areas where retinal folds were previously seen. There was no retinal or choroidal detachment. The vitreous hemorrhages failed to clear. The child never regained consciousness and remains quadriplegic. She was placed in the custody of the court because of the suspicion of child abuse.

Case 2

A 10-month-old girl was admitted to the intensive care unit in a coma, with decerebrate posturing and grand mal seizures. She was previously in good health when she reportedly fell 5 feet to the floor from her stepfather's arms while being carried down a stairway. No history of physical abuse could be elicited. Except for two small erythematous bruises on the left lower abdomen and left thigh, there were no other external signs of trauma. Computed tomography demonstrated a left frontal subarachnoid hemorrhage and a right occipital subarachnoid hemorrhage. She was intubated

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and treated with pancuronium bromide and phenobarbital.

Ophthalmologic examination on the second hospital day showed no objective response to bright light. The pupils were fixed at 2 mm. Results of external and anterior segment examination were normal. Ophthalmoscopy showed extensive subretinal, intraretinal, and preretinal hemorrhages. The optic disks were edematous, with marked nerve fiber layer hemorrhages. Scattered areas of retinal edema were also noted. White elevated retinal or subretinal ridges were present surrounding both maculae outside the vascular arcades (Figure). They were especially prominent temporally.

On reexamination one week later there was a slight avoidance response to bright light in the right eye only. The pupils remained fixed. Ophthalmoscopy showed substantial resolution of the retinal hemorrhages, but the retinal ridges persisted.

The child recovered only partial consciousness, responding only to noxious stimuli. Magnetic resonance imaging demonstrated prominent frontal sulci and large hemorrhages in both occipital lobes. Despite this, visual-evoked potentials were normal. At the time of discharge, the child was severely impaired neurologically, with poor head control and quadriparesis. The child was made a ward of the court because of the strong suspicion of physical abuse.

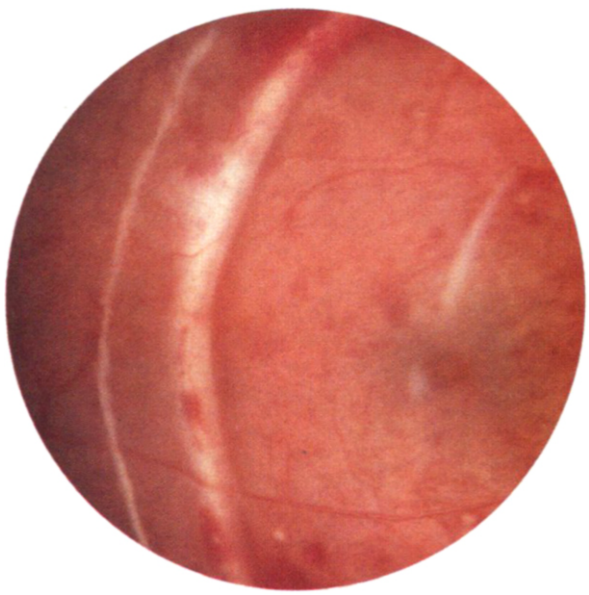

Figure (Gaynon and associates). Arcuate retinal fold along the temporal aspect of the right macula.

Discussion

These two children with nearly identical ophthalmoscopic changes were probable victims of child abuse. Both had severe, indirect head trauma leading to subarachnoid or subdural hemorrhages and resulting in increased intracranial pressures with devastating neurologic sequelae. Diffuse retinal hemorrhages were present and have been described previously in the shaken baby syndrome.[1-3] It is uncertain whether these hemorrhages result from vitreous traction or increased intracranial pressure, such as that seen in Terson's syndrome.[4]

The pupils were fixed in both infants, and a weakly suggestive light response was seen in only one eye of one child. The absent pupillary response and the weak to absent light response were probably the result of central nervous system damage as opposed to retinal or optic nerve injury in light of the intact visual-evoked potentials in Patient 2.

Retinal folds of varying orientation are found in a variety of conditions. Spontaneous folds occur in posterior persistent hyperplastic primary vitreous. Folds may develop as a result of nematode infections, retinopathy of prematurity, and familial exudative vitreoretinopathy. Vitreoretinal traction may induce folds in proliferative diabetic retinopathy, proliferative vitreoretinopathy, or incomplete posterior vitreous separation. Chorioretinal folds are seen in hypotony and orbital tumors. Iatrogenic folds may result from vitreous or retinal incarceration in a scleral wound, overly tight or incorrectly oriented scleral buckles, scleral resection, overly rapid drainage of subretinal fluid, slippage of giant tears, and occasionally from scleral buckling with an intraocular gas injection.[5] Neither infant in our study showed peripheral giant tears or dialyses.

In both infants, the ridges were depigmented, suggesting either a rupture of the retinal pigment epithelium or rarified pigmentation caused by mechanical deformation. We believe rupture of the retinal pigment epithelium is the more likely explanation because normal pigmentation remained in irregular areas on the crest of the folds. The folds appeared elevated relative to the surrounding retina based upon

their stereoscopic appearance, the suggestive distortion in the course of overlying vessels, and, in Case 1, the ultrasound findings.

Greenwald described postmortem disease in five abused children with retinal folds or ridges in a crater-like formation in the macular region (unpublished data). He also found damage in Bruch's membrane, the retinal pigment epithelium, and the retinal internal limiting membrane. He postulated that severe shaking resulted in abrupt vitreoretinal traction, leading to formation of the retinal folds.

Perhaps the neurosensory retina is displaced laterally by vitreous traction during the shaking maneuver, and comes to rest in a redundant arcuate fold. The predilection for the perimacular region and the relative sparing of the nasal retina in our patients may reflect the peculiarities of sudden, oscillating vitreous traction in children with formed vitreous gel.

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OPHTHALMIC MINIATURE

Why has not man a microscopic eye?
For this plain reason, man is not a fly.

Alexander Pope, *An Essay on Man*, line 193