0749-5161/97/1303-0183/$03.00/0
PEDIATRIC EMERGENCY CARE

Vol. 13, No. 3
Printed in U.S.A.

The significance of white-centered retinal hemorrhages in the shaken baby syndrome

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Retinal hemorrhages in healthy children with or without a history of associated trauma are a strong indicator of child abuse. This report describes six cases of battered infants who presented with white-centered retinal hemorrhages. We discuss potential mechanisms for the presence of white-centered retinal hemorrhages in battered children.

INTRODUCTION

Over the past three decades, the identification of victims of child abuse has increasingly become a major health concern. Recently reported incidence studies have revealed that more than 1.5 million children are abused each year,[1] and 25% of these children sustain serious physical injury. Emergency physicians are usually the first authorities to evaluate the abused child. Since abused children may not show obvious evidence of abuse, it is important that emergency physicians become aware of retinal hemorrhages, the hidden stigmata of child abuse, in order that abuse may be properly diagnosed.

Retinal hemorrhages are a frequent finding in the "whiplash shaken infant," a term first used by Caffey[2,3] to describe the relationship between retinal hemorrhages, brain damage, mental retardation, and the whiplash injury produced by casually or vigorously shaking children. The term "shaken baby syndrome" is now widely used to describe these abused infants. The syndrome is defined in broad terms to include battered infants who have sustained violent shaking, choking, or direct eye, head, or chest trauma.[4–6]

This report emphasizes the importance of recognizing and thoroughly investigating white-centered retinal hemorrhages (WCRH) as well as the more common dot and blot hemorrhages, in otherwise healthy children, who may or may not have a history of head injury or abuse. The presence of such hemorrhages is a strong indicator of abuse.

CASE REPORTS

Case 1. A six-month-old black male infant had apparently fallen 2 1/2 feet from a bed onto a hardwood floor. On the scene, the child was comatose (Glascow coma scale 6–7) and required assisted ventilation. In the emergency department (ED), a computed tomographic (CT) scan of his brain revealed a left-sided epidural and parietal hematoma that required surgical evacuation. Ophthalmologic evaluation showed multiple intraretinal hemorrhages (both with and without white centers) in the posterior pole of both eyes. The child improved but retained severe visual and neurologic deficits.

Case 2. A two-month-old white female infant had presumably fallen 3 feet from a bed onto a padded carpet. On the scene, transient seizure-like activity was noted. The patient was taken to the ED, where she required assisted ventilation. A CT scan of her brain revealed a left subdural hematoma. Chest x-rays showed an old left anterior rib fracture. On ophthalmologic evaluation, multiple WCRH and dot/blot intraretinal hemorrhages were noted in the posterior pole of the left eye. The hemorrhages resolved, and the child recovered.

Case 3. A three-month-old white male infant was seen in the ED in status epilepticus with apnea. A CT scan of the brain showed a small subdural hematoma and fluid collection in the right frontoparietal convexity. Chest x-rays revealed a fracture in the right 8th thoracic rib. A skull x-ray showed a spread of cranial sutures. An ophthalmologic examination showed WCRH and dot/blot hemorrhages scattered in the posterior pole of both eyes and preretinal hemorrhages in the macular area of the right eye. The child improved, but was left with impaired vision in his right eye.

Case 4. An 11-week-old black female infant was seen in the ED status postcardiac arrest with a history of having been found at home by the family pale and unresponsive when they attempted to feed her. A CT scan of the brain showed a diffuse subarachnoid hemorrhage, and an electroencephalogram showed no brain activity. On ophthalmologic evaluation, multiple dot and blot retinal hemorrhages, as well as many white-centered type, were noted in the posterior pole of both eyes. The patient died within 24 hours of initial evaluation in the ED. Autopsy revealed bilateral subdural hematomas as well as retinal and optic nerve hemorrhages.

Case 5. A four-month-old black female infant was brought lethargic to the ED with a history of having become unresponsive at home. On ophthalmologic evaluation, multiple WCRH and few dot and blot ones, were noted in the posterior pole of both eyes. A CT scan of the brain showed diffuse cerebral atrophy with intraventricular hemorrhage, and chest x-rays showed multiple old fractures of ribs. The child died shortly after admission.

Case 6. A six-week-old white male infant was seen in the ED with a history of recent onset of seizures without a history of trauma or fever. On ophthalmologic evaluation, bilateral white-centered as well as flame-shaped retinal hemorrhages were noted (see Fig. 1). CT scan and magnetic resonance imaging of the brain showed a left subdural hematoma. A bone survey revealed multiple

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Supported in part by a Research to Prevent Blindness Development Grant.
Key Words: Retina, retinal hemorrhage, shaken baby syndrome, trauma, whiplash

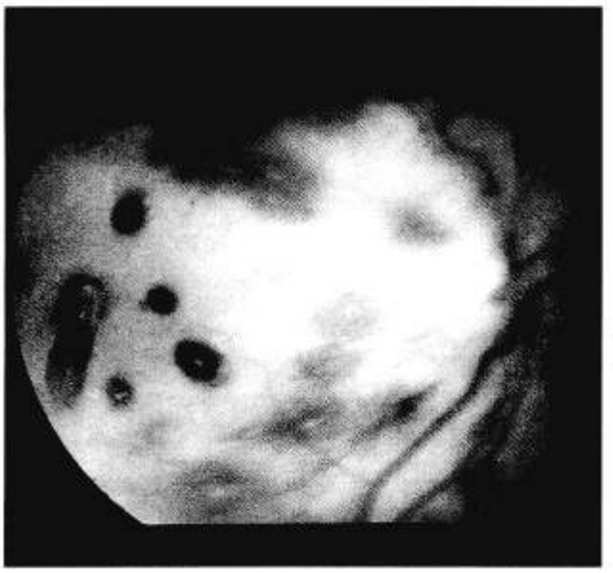

FIG 1. Fundus photograph of intraretinal hemorrhages: Left eye of a six-week-old white male infant. Note the four round WCRH inferior and temporal to the macular area in the posterior pole. Multiple other less distinct flamelike WCRH are seen in the temporal border of the left disc. Large blotchy hemorrhages are also seen in the superior arcade.

old fractures of ribs and long bones. The child recovered with residual neurologic deficits.

DISCUSSION

Several studies[3,7–9] have implicated retinal hemorrhages as indicators of child abuse. It is therefore important that all retinal hemorrhages in otherwise healthy children be thoroughly investigated. The differential diagnoses include birth trauma (flame hemorrhages resolve within one week after birth, dot/blot hemorrhages within six weeks), hematologic conditions, such as severe anemia, thrombocytopenia, acute leukemia and coagulopathies, and the shaken baby syndrome, among other disorders.[10,11]

Eisenbrey[8] reported that, in children who are less than three years old and who have no significant head injury or medical risk factors, the presence of retinal hemorrhages should be considered pathognomonic of child abuse. This diagnosis has been confirmed in several other clinical studies of trauma in children.[9,12–14]

Several epidemiologic studies[9,13,15] have shown that in children who have accidentally sustained a mild to moderate head injury, eg, a fall from bed), retinal hemorrhages are usually absent. In cases of severe accidental head trauma (eg, a child thrown from a car), retinal hemorrhages are noted in fewer than 3% of the studied children.[9] Therefore, the presence of retinal hemorrhages in a child with a head injury should alert physicians to the likelihood of abuse (nonaccidental head trauma). Goldstein et al.[14] reported in prospective clinical studies that child abuse was always predicted correctly when retinal hemorrhages were present, there were inconsistencies in the explanation for the accident, and parental risk factors were present. Risk factors include previous social service involvement in the family, a history of spousal abuse, previous referral to Child Protective Services, a single-parent family, welfare recipients, parental age less than 18 years, drug or alcohol abuse, parent who did not finish high school, or a past history of child abuse or neglect.

The reported incidence of retinal hemorrhages in the shaken baby syndrome can vary from 50 to 100%.[6,16–18] A retinal hemorrhage may be the first sign of a head injury, and it may precede recognizable clinical and radiologic evidence of a subdural hematoma.[3,19] Retinal hemorrhages can occur in any layer of the retina,[20] but they occur most often in the intraretinal region. Dot/blot hemorrhages, for example, occur in the plexiform retinal layer. Flame hemorrhages, which occur in the more superficial nerve-fiber layer of the retina, are also frequent. Preretinal and subretinal hemorrhages can also occur.

We noted WCRH as the predominant finding in the fundus of our six reported cases of battered children. After the emergency physician alerted social services and the pediatric team, a thorough investigation confirmed child abuse. All infectious etiologies and blood disorders were excluded as possible causes in these children. Our review of the literature did not reveal previous reports that correlated WCRH and the shaken baby syndrome.

The WCRH was first reported by Roth[21] in 1872 in two clinical papers describing these hemorrhages in patients with sepsis from various causes, including subacute bacterial endocarditis. He postulated that the white centers represented embolic bacterial infiltrates that produced localized retinal abscesses. His hypothesis was confirmed by light microscopy studies on postmortem specimens.

Litten,[22] in 1878, corroborated this finding and was actually the first to fully describe WCRH. He too associated it with subacute bacterial endocarditis. He gave the name "Roth spots" to such hemorrhages.

Over the past decades, WCRH has been observed and documented in a variety of noninfectious diseases.[23,24] Because there are many causes for WCRH, it is no longer considered to be pathognomonic for any single disease. WCRH has been reported in subacute bacterial endocarditis, anemia, leukemia, poorly controlled arterial hypertensive states, collagen vascular diseases, cerebral arteriovenous malformations, complicated labor and delivery, carbon monoxide poisoning, and in rare instances, diabetes and AIDS.

Our new hypothesis for the presence of WCRH in battered children includes the possibility of posttraumatic capillary rupture with subsequent extravasation and formation of a fibrin platelet plug[24] or the presence of retinal ischemia with resulting infarct of the nerve-fiber layer.[25] Because WCRH was present in each of our six cases, we recommend further investigation to determine the incidence, prevalence, and significance of WCRH in battered children. These data should be correlated with the children's visual and neurologic outcomes.

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