Purtscher Retinopathy in the Battered Child Syndrome

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• **Purtscher retinopathy is a hemorrhagic angiopathy that occurs after sudden compression of the thorax. Virtually all reported cases have been in adults who have decreased visual acuity, retinal hemorrhages and exudates, and no other neurological signs. By contrast, in infants, hemorrhagic retinopathy is rarely benign, and generally is considered to indicate intracranial hemorrhage, usually an acute subdural hematoma.**

Two battered infants had seizures and associated chest injury. There were retinal hemorrhages and exudates, unaccompanied by clinically important intracranial hemorrhage. At follow-up, the hemorrhagic retinopathy had resolved without sequelae; development was normal, and seizures had not recurred.

Purtscher retinopathy thus should be added both to the differential diagnosis of hemorrhagic retinopathy in infancy and to the list of physical signs suggesting child abuse.

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Purtscher retinopathy[1-3] is a hemorrhagic retinal angiopathy characterized by preretinal and retinal hemorrhages, retinal exudates, and decreased visual acuity. It follows a sudden compression of the thoracic cage, which results in the transmission of an acute increase in intravascular pressure to the head and eyes. This gives rise to the retinal hemorrhages.[2,3] Almost all reported cases have been in adults who experienced decreased visual acuity after sudden chest compression or a prolonged Valsalva maneuver.[4] Recently, several cases have been reported in patients who were wearing shoulder belts during minor traffic accidents.[5] Findings of the remainder of the neurological examination in such patients have been normal, and visual acuity improves with resolution of the retinal hemorrhages.

See also pp 1265 and 1338.

In childhood, a benign hemorrhagic retinopathy of this type has been reported only in the neonatal period.[6,7] The appearance of retinal hemorrhages in newborns has been related to the birth process, with an average frequency of 20% in vaginal deliveries, as contrasted to 3% in children born by cesarean section. In older infants, on the other hand, the presence of preretinal and retinal hemorrhages is considered to be a reliable indicator of intracranial hemorrhage. Preretinal and retinal hemorrhages are present in 50% to 70% of children with subdural hematomas,[8,9] the most common form of intracranial hemorrhage in infants beyond the newborn period. The presence of a subdural hematoma in a battered infant is a serious threat to the child's survival[10] and to his prognosis for normal development.[11]

Recently, there have been a number of publications concerning the ophthalmological manifestations of the battered child syndrome, relating preretinal and retinal hemorrhages to severe intracranial injury.[12,13] In this article, two battered infants with preretinal and retinal hemorrhages will be discussed. The neurological evaluation of these infants and their clinical course demonstrated that noteworthy intracranial hemorrhage did not occur, and suggested that the preretinal and retinal hemorrhages were caused by thoracic compression. This led to the diagnosis of Purtscher retinopathy, which should be added to the list of signs of physical abuse in infancy.

REPORT OF CASES

CASE 1.—An 18-week-old female infant was admitted to the Boston City Hospital, having had two tonic seizures during the

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previous hour while sleeping. Each had lasted approximately three minutes. The child had not been ill, and there had been no previous seizures. The infant weighed 3.1 kg (6.8 lb) at birth and had been the product of an uncomplicated full-term pregnancy in a 16-year-old, unmarried girl. The infant lived with both parents, maternal grandparents, and four siblings of the mother. One of the family members is said to "look after the infant" while the mother attends school.

On the patient's admission to the hospital, examination disclosed an alert child without dysplastic features. Vital signs were normal, and the height and weight were at the 50th percentile. Ecchymoses were noted on the right cheek, buttock, thigh, and left foot. A large ecchymotic area, seen over the right hemithorax, outlined an adult hand, including prints of the five digits and nails. Neurological examination showed a smiling infant who functioned at the 3- to 4-month level on the Denver Developmental Screening Test. The scalp was free of contusions. The head circumference was 40.5 cm (15.9 in) (25th percentile). The anterior fontanelle measured 3×2 cm (1.2×0.8 in) and was pulsatile. Transillumination was questionably increased over the right hemicranium. No focal neurological deficits were noted. On ophthalmological examination, the infant would fix and follow light. Retinal hemorrhages were present bilaterally, and retinal exudates were noted in the left fundus. No papilledema or optic atrophy was seen. Findings of the remainder of the ophthalmological examination were normal. Because of the striking retinal hemorrhages, bilateral subdural taps were performed, 0.25 cm (0.1 in) and 1.25 cm (0.5 in) lateral to the anterior fontanelle, with negative results. A lumbar puncture done at that time was traumatic. The third tube of cerebrospinal fluid (CSF) contained colorless fluid that had a cell count of 923 red blood cells (RBC) per cubic millimeter, two white blood cells (WBC) per cubic millimeter, a total protein level of 48 mg/100 ml, and a glucose level of 116 mg/100 ml. An electroencephalogram done 18 hours after the patient's admission was normal. Other laboratory studies included a hematocrit reading of 25%, with 8% to 10% reticulocytes and a normal platelet count. Hemoglobin electrophoresis, glucose-6 phosphate dehydrogenase level, serum electrolytes levels, calcium and blood glucose levels, blood urea nitrogen (BUN) level, prothrombin time, and partial thromboplastin time were normal. Roentgenograms of the skull and long bones were normal.

During the ten-day hospitalization, while awaiting disposition, findings of the patient's physical and neurological examination remained normal. No seizures were noted. On the fifth hospital day, a brain scan with Technetium Sulfide Tc 99m showed no evidence of an arterial-venous malformation, but suggested an increased uptake of isotope on the left side near the convexity. A second set of subdural taps were done: the left side was negative, but the right side yielded 3 ml of slightly xanthochromatic fluid with a protein content of 61 mg/100 ml. Cerebrospinal fluid obtained from a lumbar puncture at that time was clear and had a protein content of 51 mg/100 ml.

The child was seen for follow-up examination two months after her hospitalization. There had been no episodes suggesting further seizures. She was playful, and her performance on the Denver Developmental Screening Test remained normal for her age. Her head circumference continued to be in the 25th percentile. Results of transillumination of the skull were normal. Funduscopic examination disclosed almost complete resolution of the hemorrhages. A mild tightness of the hip adductors was noted.

Case 2.—A 23-month-old female infant was brought to the emergency room of the Boston City Hospital by her babysitter who said that the child had "swallowed a spoon," and then had a seizure. On initial examination, tonic clonic activity was noted in both upper extremities; this subsided in two minutes without medication. There was no history of previous seizures, and the child had not been ill. The infant had weighed 3.3 kg (7.3 lb) at birth, and growth and development had been normal. The mother was 21 years old, unmarried, and had accused a house guest of frequently beating her child. A 13-month-old sibling had been hospitalized with third-degree burns on the right hand.

On admission to the hospital, examination of the patient showed a well-developed child with normal vital signs, and a height and weight in the tenth percentile. Ecchymoses were noted on the face, circumferentially around the neck and chest, and on the buttocks and legs. On neurological examination, the child was lethargic and withdrawn, but would respond to her name. Head circumference was 47 cm (18.5 in) (25th percentile). No focal neurological deficits were noted other than bilateral extensor plantar responses. The child would fix and follow light. Preretinal hemorrhages were present bilaterally. A subhyaloid fluid level and exudates were seen in the right fundus. No papilledema or optic atrophy was noted. Because of concern about the probable presence of increased intracranial pressure, the child was given intravenous fluids and dexamethasone. A lumbar puncture was deferred. Laboratory studies included a normal skull roentgenogram and a normal EEG. Within 24 hours, the child was more alert, and the plantar responses became flexor. Lumbar puncture was nontraumatic, with an opening pressure of 150 mm H_2O. The clear, colorless fluid contained 456 RBC per cubic millimeter and 11 lymphocytes per cubic millimeter. Total protein content of the CSF was 20 mg/100 ml. Dexamethasone therapy was discontinued. During this first hospital day, cerebral angiography was considered, but not performed because of the child's rapid clinical improvement. The results of a complete blood cell count, platelet count, and urinalysis were normal. Normal results of other studies included serum electrolytes and calcium levels, blood glucose level, BUN level, and prothrombin time. Skeletal roentgenograms were normal.

The child remained in the hospital for nine days, awaiting foster home placement. During this time, findings from her neurological examination remained normal, and the apathy that had characterized the early days of her hospital stay disappeared. No further seizures were noted, and the child was discharged receiving no medication.

The preretinal hemorrhages resolved completely, without sequelae. When the patient was last seen one year after the hospitalization, there had been no further seizures, findings from the neurological examination were normal, and the child's development was normal for her age.

COMMENT

The infants described were of great interest since both had preretinal and retinal hemorrhages in the absence of intracranial hemorrhage, particularly, subdural hematoma. The unusual histories, coupled with the multiple ecchymoses seen on both infants, led to a consideration of the battered child syndrome. This diagnosis was substantiated in both children after further investigation.

Despite the relatively normal findings from neurological examinations, the presence of the bilateral retinal hemorrhages (case 1) and preretinal hemorrhages (case 2) in these battered infants[12,13] suggested traumatic intracranial hemorrhage.[8,9] Pathogenetically, this hemorrhagic retinopathy is thought to arise from an abrupt increase in intracranial pres-

sure caused by the intracranial hemorrhage, which is then transmitted to the retina.[8] The skull roentgenograms and EEGs were normal in both children. In the first case, bilateral subdural taps done on the first hospital day were negative. On the fifth hospital day another set of subdural taps in this patient yielded 3 ml of fluid, which was considered to be from the cerebral subarachnoid space since the protein content of this fluid was only slightly greater than that of the lumbar CSF obtained at the same time.[9] Follow-up examination in both children showed them to be developing normally and to have remained seizure-free, without anticonvulsant medication. These data are consistent with the conclusion that no clinically important intracranial hemorrhage, particularly in the subdural space, had occurred in either case, and that the hemorrhagic retinopathy in these two infants had not been secondary to increased intracranial pressure. Other diagnostic possibilities that were considered, but excluded, were subacute bacterial endocarditis, idiopathic thrombocytopenia, and acute leukemia. The bilateral symmetry of the retinal hemorrhages made local trauma unlikely. These two cases, thus, fulfill the diagnostic criteria for Purtscher retinopathy.[5]

A review of the literature revealed only one report of Purtscher retinopathy in the pediatric age group.[14] The patient was a young boy who was run over by a small cart. On his initial visit to the emergency room, examination showed a contusion of the chest wall. The remainder of the examination results, including a chest x-ray film, were normal. The child later returned to the hospital, complaining of decreased visual acuity, and funduscopic examination disclosed retinal hemorrhages that resolved without sequelae.

Purtscher retinopathy, thought to be caused by a sudden rise in intracranial and intraocular venous pressure following thoracic compression, has been suggested as a clinical sign of physical abuse in childhood.[15] The two battered infants described in this article provide support for this hypothesis. Perhaps the history of seizures, which resulted in a detailed neurological evaluation of these patients, was instrumental in establishing the diagnosis. The addition of Purtscher retinopathy to the list of findings seen in the battered child syndrome stresses the importance of a complete ophthalmological evaluation in any infant in whom a diagnosis of battering is considered. Purtscher retinopathy is likely a mild form of the more severe ocular hemorrhages attributed to chest compression by Caffey.[16]

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