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Sir,

Vertical sensory nystagmus associated with intraocular haemorrhages in the shaken baby syndrome

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The shaken baby syndrome is a serious and clearly definable form of child abuse caused by the violent shaking of young children with or without impact injuries.[1] These children typically have intracranial and intraocular haemorrhages.[2] Survivors may demonstrate a variety of neurological, intellectual, and ocular motility sequelae.[2] We present a case of vertical nystagmus following a shaking injury in an infant.

Case report

An 8-week-old male infant was referred to The Hospital for Sick Children for management of uncontrolled seizures following diffuse brain injury resulting from a shaking injury. The diagnosis of shaken baby syndrome was made at the referring hospital on the basis of the clinical findings: subdural intracranial haemorrhage and retinal haemorrhages occurring in a characteristic pattern[2] in the absence of any other identifiable medical cause. A second ophthalmologic consult was requested as the child had been noticed to have abnormal eye movements.

On examination, there was a blink reflex to bright light, but the child was unable to fix and follow, and there was no optokinetic response. The pupillary light reactions were sluggish bilaterally; there was no relative afferent pupillary defect. There was a constant, large-amplitude vertical pendular nystagmus of moderate frequency. The anterior segment examination was normal. Dilated fundus examination revealed vitreous and posterior pole haemorrhages in the right eye, involving the macula. In the left eye, there was a dense vitreous haemorrhage with no view of the retina. B-scan ultrasound demonstrated no retinal detachment in either eye. Magnetic resonance imaging (MRI) of the brain revealed bilateral subdural haemorrhages and diffuse infarction of cerebral cortex, with extensive loss of cortex consistent with a profound ischaemic event, but relative preservation of the brainstem.

At follow-up examinations 4 and 7 months later, the vertical nystagmus had resolved, though the eyes showed conjugate, random movements. Visual function was unchanged. Fundus examination showed resolution of all intraocular haemorrhages, bilateral optic atrophy, macular scarring, and vitreous veil formation in the left eye. There was a preretinal gliotic membrane bilaterally (Figure 1). Neuroimaging demonstrated severe atrophic changes throughout the cerebral cortex.

Comment

To our knowledge, there are no reports in the literature of nystagmus following shaking injuries in children. The shaken baby syndrome is a serious and clearly definable form of child abuse caused by the violent shaking of young children with or without impact injuries.[1] Our case was typical of shaking injuries in most respects. These children typically have subdural or subarachnoid intracranial haemorrhage visible on neuroimaging.[2] Intraocular haemorrhages are common following injuries occurring in at least 80% of cases, usually bilaterally.[2] The injuries are fatal in nearly one-third of the children.[2] Approximately 20% of survivors have reduced vision that is usually cortical in origin and highly correlated with severe neurologic outcome. Poor vision may sometimes, however, result from anterior pathway injuries such as optic atrophy or vitreous haemorrhage.[2]

Sensory nystagmus results from bilateral anterior visual pathway disease, in this case intraocular haemorrhages, whereas cortical visual loss typically does not cause sensory nystagmus. Sensory nystagmus is

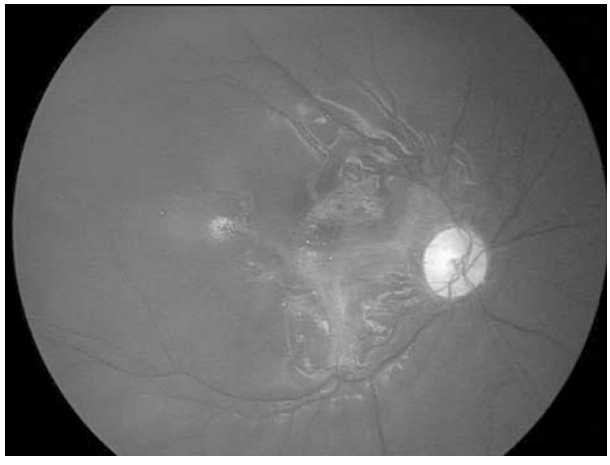

Figure 1 Right fundus photograph 4 months following shaking injury. The intraocular haemorrhages have cleared, revealing optic disc pallor, atrophic, and pigmentary change at the macula and preretinal gliosis with retinal wrinkling.

usually horizontal or rotary, but may sometimes be vertical. Vertical nystagmus in adults is most commonly due to intracranial pathology. Children however may demonstrate vertical nystagmus in the presence of normal neuroimaging, this nystagmus is invariably associated with poor visual acuity.[3-5] In most children where the sensory nystagmus persists, the nystagmus eventually becomes horizontal in direction.[4]

The child described above developed vertical, pendular sensory-type nystagmus following a shaking injury with documented intracranial and intraocular haemorrhages. Resolution of the nystagmus coincided with clearing of the intraocular haemorrhages. The ocular movements then reverted to conjugate, random movements characteristic of cortical visual impairment. We postulate that this child developed reversible sensory nystagmus as a result of the occlusive effect of the intraocular haemorrhages. Future reporting of similar cases may clarify whether sensory nystagmus is more likely to be vertical in the presence of diffuse brain injury from shaking or other causes.

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Sir,

Leukaemic infiltration of the optic nerve as the initial manifestation of leukaemic relapse

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Ocular problems in patients who have leukaemia are commonly observed by the ophthamologists.[1] Cases of leukaemic involvement of the central nervous system (CNS) are becoming more frequent because of the increased survival rate associated with more effective treatment including combination therapy of systemic chemotherapy, prophylactic irradiation, and intrathecal injection of cytotoxic drugs.[1–5] Direct invasion of the orbit with the neoplastic cells is common, but the involvement of the optic nerve, uveal tract, and retina is relatively rare.[2,3] The optic nerve is known to be one of the disease-relapse sites in a patient with systemic or meningeal leukaemia,[2,4] but it was rarely reported as the initial isolated presentation for the relapse in a patient with complete remission. Moreover, the optic nerve had been characterized to be a pharmacologic sanctuary, relatively unaffected by systemic chemotherapy.[3,5] Thus, a separate treatment modality, often radiotherapy, is required for the optic nerve involvement of leukaemia.[2,5,6] Here, we report three cases of leukaemia with the leukaemic infiltration of the optic nerve as the initial isolated presentation of disease relapse. Systemic remission was proved by bone marrow aspiration and peripheral blood studies at the time of presentation.

Case report

Patient 1

Acute myeloid leukaemia was diagnosed in the 36-year-old male on November 1987. He was treated with cytotoxic drugs including cytarabine, 6-mercaptopurine, and daunomycin. Systemic remission was achieved after three complete courses of chemotherapy.

He complained of pain and mild visual distortion in his left eye on February 1989. His best-corrected visual acuity was 6/6 in the right eye and 6/6.7 in the left eye. Ophthalmic examination of the anterior segment was unremarkable in both eyes. Ophthalmoscopy showed normal optic disc and retina in the right eye. In the left eye, the optic disc was markedly swelling. The retinal vessels were engorged with dot-shape haemorrhage. The vitreous was clear without cell.

After 2 weeks, visual acuity in the left eye decreased to 6/30. Computed tomography showed an enlargement of the left optic nerve (Figure 1). Then, he was admitted for evaluation and for another course of maintenance chemotherapy. Bone marrow aspiration revealed a state