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Saccadic intrusions in pediatric non-accidental trauma

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ABSTRACT

Purpose: To report a case of saccadic intrusions in a 9-month-old under the context of non-accidental trauma.
Observations: A 9-month-old female presented with the eye finding of intermittent ocular flutter. Upon imaging for neuroblastoma, she was discovered to have bilateral supratentorial subdural hematomas with internal septations, moderate mass effect on adjacent cerebral parenchyma, pachymeningeal and tentorial enhancement, and cortical vein thrombosis. This constellation of findings was concerning for non-accidental trauma.
Conclusions and importance: Although an extremely rare occurrence, ocular flutter may be a presenting sign in non-accidental trauma, and imaging should absolutely be considered under such circumstances.

1. Introduction

Non-accidental trauma (NAT) is defined by the intentional abuse of a child. The incidence is estimated to be about 550,000 cases per year in the United States alone.[1] Abusive head trauma tends to occur with a complex presentation of ocular, intracranial, and other injuries.[2,3] This most commonly occurs in children under age 5, and even more so in children under 12 months.[2,4]

Abusive head trauma is thought to result from a series of repetitive, and sudden, accelerations and decelerations of the child's head. This can result in subdural hematomas, subarachnoid hemorrhage, intracranial edema, ischemia, contusion, and chronic cerebral atrophy.[2]

Ocular involvement is present in about 80% of cases, with retinal hemorrhage being the most common finding, seen in all layers of the retina.[3] This usually involves the posterior pole more than the periphery, may be bilateral, and may present with vitreous hemorrhage.[3]

To the best of the authors' knowledge, saccadic intrusions and/or nystagmus have only been reported once in literature in association with NAT.[5] This was a case of an 8-week-old with shaken baby syndrome associated with the finding of a constant, large-amplitude vertical pendular nystagmus of moderate frequency.

2. Case report

A 9-month-old infant presented to the eye clinic with her grandmother for the concern of new onset "rhythmic eye movements" over the past 4–5 months, difficulty making eye contact, along with not meeting her developmental milestones appropriately. She was born at 31 weeks to a 20-year-old female via cesarean delivery for pre-eclampsia with severe features. The mother was insulin-dependent and had a history of substance abuse, positive for tetrahydrocannabinol (THC). The child's perinatal Apgar scores were 2 and 8, and she had an extended stay in the neonatal intensive care unit (NICU) for intrauterine growth retardation (IUGR) and prematurity requiring respiratory support. She was found to have a small ventricular septal defect. Genetic workup was unremarkable. Screening head ultrasound was normal at the time, and she required no further imaging. Her initial eye screen at 4 weeks showed zone III retinopathy of prematurity (ROP) with no plus disease in both eyes. Her mother had a significant past medical and ocular history of psychiatric illness, refractive error, and ocular hypertension. Her father had a questionable history of amblyopia.

Clinic exam findings showed a macrocephalic child with central, intermittently unsteady, and intermittently maintained acuity. She had equally reactive pupils with no relative afferent pupillary defect. Indirect ophthalmoscopy with a 20 diopter lens showed a normal anterior segment and posterior fundus exam. There were no hemorrhages or abnormalities in any ocular structure. Her cycloplegic refraction was +1.50 D in the right eye and +1.00 D in the left eye.

During the sensorimotor exam, there was no strabismus, but an intermittent jerk nystagmus superimposed with intermittent saccadic intrusions most similar to ocular flutter. No normal intersaccadic intervals were noted. There was no null point identified. The primary differential diagnosis was concern for neuroblastoma, upon which an urgent MRI of the brain, sympathetic chain, and adrenal glands was ordered.

Pediatric neuroradiology read the MRI report as bilateral

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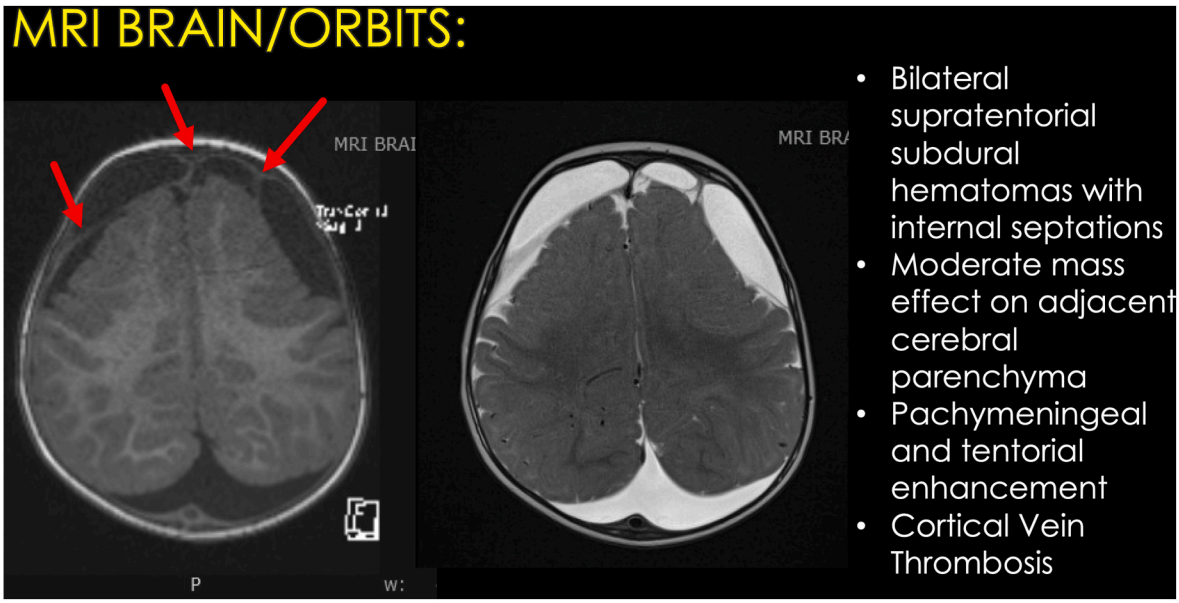

Fig. 1. Axial MRI of the brain of the 9-month-old child showing bilateral supratentorial subdural hematomas with internal septations.

supratentorial subdural hematomas with internal septations, moderate mass effect on adjacent cerebral parenchyma, pachymeningeal and tentorial enhancement, and cortical vein thrombosis (Fig. 1). This constellation of findings was concerning for NAT. The child was admitted to the pediatric service for further workup. During her hospital stay, she was started on hyperhydration for cortical vein thrombosis but required no acute interventions otherwise. A repeat MRI five days later demonstrated stable findings and patient was discharged to foster care. Of note, the patient had been under the custody of 4 different family members as her mother was incarcerated for abandoning the child in a motel 2 months prior. Over the course of the next year, her brain MRI continued to show stable findings while the nystagmus and saccadic intrusions gradually improved without requiring further interventions.

3. Discussion

Ocular flutter and opsoclonus are saccadic intrusions characterized by bursts of back-to-back saccades that oscillate about the midline without an intersaccadic interval. It is termed ocular flutter when the saccades occur only in the horizontal plane and opsoclonus when saccades also have vertical and/or torsional components.[6] They are typically seen in association with diseases involving the brainstem or cerebellum, in particular viral encephalitis, paraneoplastic syndromes, drug intoxications, or abnormal metabolic states.[6,7] In children, neural crest tumors (such as neurobloastoma) are the most commonly implicated underlying malignancy, and opsoclonus may be the initial presenting sign in some cases.[8,9] Thus, appropriate diagnostic studies should be pursued whenever flutter or opsoclonus is observed.

In rare instances, ocular flutter has been reported as a consequence of both severe and mild traumatic brain injury.[10–12] Despite some theoretical similarity in the mechanism of such injuries, we report the first case of ocular flutter in association with shaking impact injury in an infant. Due to the nonspecific symptoms and frequently unreliable history, NAT cases are often challenging to diagnose in young children and require high clinical suspicion.[13] We hope that increasing awareness of unusual presenting signs of NAT such as this case will increase the likelihood of more cases of NAT being detected.

Shaken baby syndrome is a serious and clearly definable form of NAT resulting from extreme rotational cranial accelerations and decelerations induced by violent shaking and/or impact.[2] Risk factors include young parents, unstable family situations, low socioeconomic status, and disability or prematurity of the child.[14] The classic constellation of clinical findings in infants include subdural and/or subarachnoid hemorrhage, traction-type metaphyseal fractures, and retinal hemorrhages.[15] Computed tomographic (CT) scanning is generally the method of choice for demonstrating subarachnoid hemorrhage, mass effect, and large extra-axial hemorrhages, but may miss more subtle findings early in the evolution of cerebral edema.[2] Magnetic resonance imaging (MRI) is more sensitive in detecting and characterizing small extra-axial hemorrhages and identifying parenchymal contusions.[16] The intracranial findings in our case were typical for NAT but would not have been discovered were imaging not pursued for a neuroblastoma workup. This highlights the importance of maintaining a low threshold for imaging whenever NAT is suspected.

To our knowledge, only one case of abnormal eye oscillation in association with NAT has been reported previously. This was an 8-week-old with shaken baby syndrome associated with a constant, large-amplitude vertical pendular nystagmus of moderate frequency.[5] Nystagmus and saccadic intrusions are differentiated by the type of eye movement that initiates the shift in eye position from fixation. Unlike nystagmus which begins with a slow drift of the eyes called the slow phase, saccadic intrusions are initiated by a fast saccadic eye movement away from fixation. Nystagmus also tends to be rhythmic and regular and, if present in central gaze, continuous and sustained while saccadic intrusions are more often nonrhythmic, intermittent, and unsustained.[17] It can be challenging to distinguish saccadic intrusions from nystagmus clinically and eye movement recordings may be required.[6] However, distinguishing them from each other is clinically important as each condition is associated with a different set of differential diagnosis and may influence the decision of pursuing additional imaging.[6]

It is also worth noting that the patient in our case was born prematurely at 31 weeks of gestation. Although premature children have been reported as having higher rates of infantile nystagmus, most cases have an identifiable organic cause such as ocular diseases or other genetic or neurological disorders.[18] Studies have also reported that children born prematurely had deficits in the voluntary control of saccades compared to full term controls, especially the inhibitory control of antisaccades.[19] There was also a high occurrence of intrusive saccades during pursuit movement.[20] This is likely explained by immature brain structures, particularly in the frontal and parietal region. In the case of our patient, with no abnormal structural findings on her initial screening head ultrasound or ocular exam, the saccadic intrusions observed were most

likely attributable to the intracranial findings of NAT.

4. Conclusions

Clinical suspicion of NAT should be considered with new onset nystagmus or saccadic intrusions. Opsoclonus or ocular flutter in a child should receive imaging to rule out a malignancy or paraneoplastic phenomenon as standard of care.

Patient consent

The patient's legal guardian consented to publication of the case in writing.

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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