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Brief Communication

Neonatal Neurocritical Care: Overlooked Neurologic Syndromes

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ABSTRACT

Although encephalopathy, seizures, and changes in tone comprise the most common signs of neurologic disruption in neonates (and in older children and adults), careful neurologic examination can yield additional clues to assist with the management of underlying neurologic conditions.

Introduction

Neonatal neurocritical care is an emerging subspecialty that combines expertise in neurology, neurosurgery, and critical care medicine. Neonates often demonstrate a limited repertoire of signs and features in the face of diverse illnesses. Although encephalopathy, seizures, and changes in tone may indicate one of several neonatal conditions from infection to global hypoxia to focal hemorrhages, careful neurologic examination reveals clues to localization and differential diagnoses that can help with the management of underlying neurologic conditions [1].

Case Reports

Patient 1: Cerebellar tonsillar herniation

A 1-day-old neonate was examined for "jitteriness" after emergent cesarean section. Bedside examination by the child neurologist revealed a full, tense fontanel, weakly reactive midsized pupils, symmetric hyperreflexia, jittery movements, and extensor posturing with stimulation. Urgent magnetic resonance imaging demonstrated bilateral anterior circulation strokes, with severe cytotoxic edema causing a downward brain herniation of the cerebellar tonsils into the foramen magnum. After discussions with the family, the patient was transitioned to palliative care and died on day 2 of age.

In neonates, brain herniation is extremely rare because of the high compliance of the cranial vault due to open sutures, which allows the skull to expand with rising intracranial pressure. Nevertheless, large and rapidly growing lesions can, on rare occasions, cause herniation in this population [2].

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Patient 2: Cushing response

A 10-day-old neonate presented with 24 hours of lethargy and poor feeding. The pregnancy and delivery were unremarkable. A bedside examination revealed hypertension with bradycardia in response to stimulation. Brainstem reflexes were absent. The motor response involved stereotyped extensor posturing to central pain. Urgent magnetic resonance imaging detected symmetric reduced diffusion and T_2 hyperintensity involving the deep white matter, ventromedial thalami, lentiform nuclei, cerebral peduncles, and dorsal medulla. Magnetic resonance spectroscopy demonstrated a markedly elevated lactate peak. The clinical and radiographic findings were consistent with Leigh syndrome. The patient manifested rapid multiorgan deterioration, and died after a transition to palliative care.

The Cushing response (i.e., progressive hypertension associated with bradycardia and diminished respiratory effort) involves a central nervous system response to raised intracranial pressure. In adults and older children, the Cushing response comprises an ominous finding that may indicate imminent brain herniation. Localization is thought to occur in the medulla and spinal cord. The pathogenesis of findings in this patient was uncertain.

Patient 3: Parinaud syndrome

A 3-week-old neonate with a history of a repaired congenital diaphragmatic hernia was evaluated after episodic nystagmus had worsened during a 2-day period. Bedside neurologic and ophthalmologic examinations indicated a preference for downward gaze, with intermittent, conjugate upward beating nystagmus provoked by upward gaze, as well as intermittent bilateral upper eyelid retraction. The neurologic examination produced otherwise normal results. Cranial ultrasound and magnetic resonance imaging indicated communicating hydrocephalus, presumably because of high venous pressures.

Parinaud (or dorsal midbrain) syndrome is caused by lesions of the upper brainstem, and consists of a classic constellation of signs including: (1) paralysis of upward gaze, (2) convergence-retraction nystagmus, (3) light to near dissociation of the pupillary response, (4)

eyelid retraction (Collier's sign), and (5) conjugate downward gaze in the primary position ("sunsetting"). The syndrome is most commonly observed in children with tectal tumors. However, hydrocephalus is also a known cause [3].

Discussion

These three neonates presented with classic neurologic syndromes that initially went unrecognized. Careful bedside neurologic examinations permitted the localization of findings, and facilitated appropriate diagnoses and management.

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