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Facial Weakness and Ophthalmoplegia in a 4-Day-Old Infant

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We present a neonate with neurologic deficits recognized at 4 days of age. A male infant was born at term via emergency cesarian section due to failure to progress and fetal decelerations. He underwent therapeutic hypothermia for hypoxic ischemic encephalopathy. Upon completion of rewarming, he was noted to have left facial palsy, abduction deficit on the left eye past the midline, and nystagmus involving the right eye. Brain magnetic resonance imaging showed a pontine stroke, and computed tomography angiogram revealed basilar artery thrombosis. He was treated with enoxaparin for 3 months, followed by low-dose aspirin. The mechanism of the stroke remains unclear, and there is limited evidence to guide management.
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Introduction

Perinatal ischemic stroke is recognized with increasing frequency with the use of modern neuroimaging. The incidence is reported to be 1 of 2300-5000 live births and is a significant cause of long-term disability and mortality.[1] Basilar artery occlusion is an uncommon cause of pediatric stroke, accounting for 6% of pediatric strokes excluding neonates in a recent analysis of data from a large pediatric stroke registry. It is even less common in the neonatal period (1 basilar artery stroke out of 163 neonatal strokes in the stroke registry). Infarcts most commonly involve the pons followed by the midbrain and the medulla, and nearly half of the aforementioned cases had basilar artery occlusion.[2] We describe a neonate with long-segment basilar artery occlusion detected after therapeutic hypothermia.

Case Report

A male infant was the 2.806 kg (~10th percentile) product of a full-term pregnancy complicated by maternal multiple sclerosis (mother discontinued natalizumab before pregnancy), severe oligohydramnios in previous pregnancy, and history of C-section. After 72 hours of labor, complicated by shoulder dystocia, fetal decelerations, and meconium-stained amniotic fluid, an emergent C-section was performed. Cephalic delivery was attempted, but owing to the engaged fetal vertex, the baby was delivered via breech maneuvers. Apgar scores were 2 at 1 minute, 5 at 5 minutes, and 7 at 10 minutes. Initial blood gas had a pH of 7.01 and base deficit of −16. The baby had moderate hypoxic ischemic encephalopathy (HIE) on examination (decreased activity, decerebrate posture, constricted pupils, bradycardia, periodic breathing, flaccid tone, absent suck, incomplete Moro, and lethargy). Fracture of left distal humerus was noticed right after birth. Therapeutic hypothermia was initiated at approximately 2 hours of life. Rewarming was started on day of life 3. He had abnormal eye movements on day of life 3; otherwise, he had no spells concerning for seizures. Continuous electroencephalogram (performed per protocol during rewarming) showed multifocal right and left hemispheric sharps, but no clinical or subclinical seizures. Approximately 29 hours after rewarming was started, he developed repeated episodes of apnea and bradycardia. He was evaluated and treated for possible sepsis, given caffeine, and supported with neurally adjusted ventilatory assist. After rewarming period was completed on day of life (DOL) 4, facial asymmetry was observed; the left side of the face did not move as well as the right. He underwent cranial magnetic resonance imaging (MRI) 5 hours after the asymmetry was recognized. The brain MRI showed diffusion restriction in the pons with corresponding decreased apparent diffusion coefficient (ADC) signal, left more than

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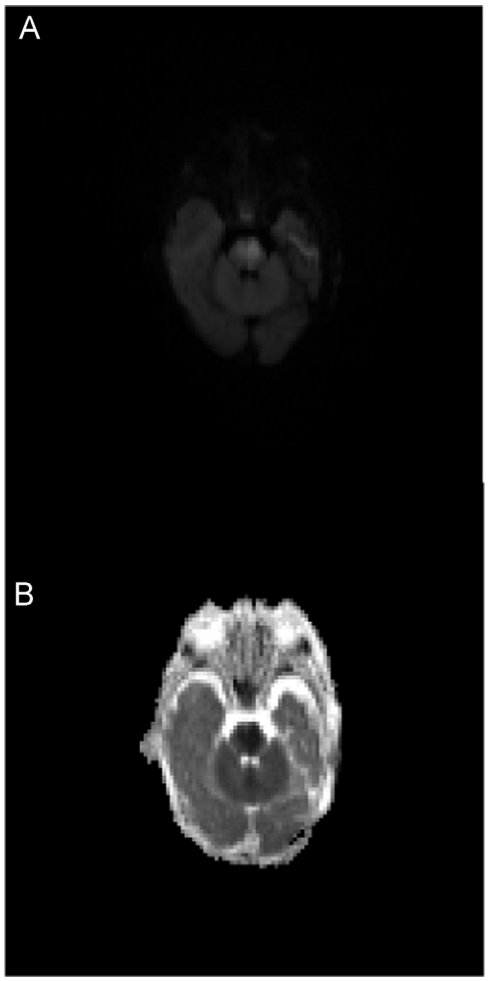

Figure 1 (A and B) Brain MRI, DOL 7, axial view DWI, and ADC sequences: DWI sequence shows hyperintensity in pons with corresponding decreased ADC signal.

right (Fig. 1). Computed tomography angiogram (CTA) revealed long-segment basilar artery thrombosis (Fig. 2).

On DOL 6, the baby was alert, did not fully close his left eye, had flattening of left nasolabial fold; on left gaze, he was unable to abduct the left eye past midline with Doll eye maneuver and had nystagmus of right eye.

Hematology was consulted, and testing for factor V Leiden mutation (c.1601G > A) and c.*97G > A mutation of factor II mutation was negative. Because of the location of the clot, uncertainty about timing and risk associated with progression, he was started on enoxaparin 3 mg/kg/d.

The baby was discharged on DOL 15, feeding well. At 1 month, his left eye still did not fully close while sleeping or while crying. The left eye also continued to have limited abduction and did not pass the midline. At 3 months of age, he had bilateral lower extremity hyperreflexia, with left facial weakness and restricted left eye movement. MRI brain showed chronic ischemic changes (Fig. 3), and MR angiogram revealed patent basilar artery (Fig. 4), so enoxaparin was discontinued and aspirin was started at 3 mg/kg/d for a total of 12 months. At 12 months of age, his ocular findings are stable. He has increased tone involving the right upper and lower extremities. He holds his head; he has been sitting well and is able to roll

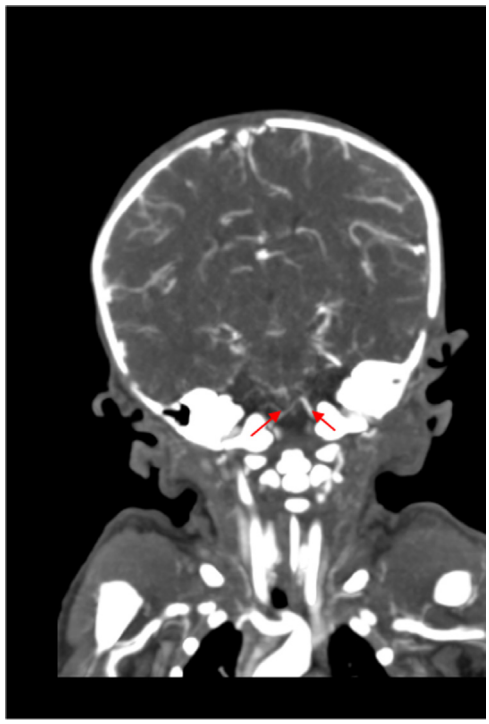

Figure 2 CTA head and neck, coronal view: no basilar artery is visualized in CT angiogram of head and neck suggesting long-segment basilar artery thrombosis (arrows point to V4 segments of vertebral arteries).

over both sides, not able to crawl yet. He has mature pincer grasp, tends to use his left hand more than his right, drinks from a cup, has 3-4 words, and makes good eye contact.

Discussion

We describe a rare case of neonatal long-segment basilar artery thrombosis leading to pontine infarction. Our patient presented with brainstem dysfunction not recognized until day of life 4 after rewarming and basilar artery thrombosis was confirmed by CTA.

The cause of the thrombosis in our case remains unclear, and assigning causation is complicated by the fact that physical examination abnormalities were first recognized after rewarming. It is possible that the stroke occurred in the peripartum period and was not recognized immediately after birth. The difficult extraction and humerus fracture suggest the possibility of vertebral artery dissection secondary to head and neck positioning during labor and delivery, although residual vascular stenosis was not noted in repeat neuroimaging studies. Less likely causes were also considered. The fractured humerus raised the possibility of cerebral fat embolism; however, this is typically associated with small, scattered, nonconfluent hyperintense intracerebral lesions on T2-weighted scans ("starfield pattern"), not with a basilar artery thrombosis.[3] Placental or systemic thromboembolism is presumed to be among the more common causes of neonatal stroke. In a cohort of 121 neonates, 34% of neonatal strokes were grouped as unclassifiable (largest subgroup), followed by