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Case report

Perinatal stroke involving the anterior cerebral artery distribution

Mahmoud Alhosiny Fayed[1,2], Usha Rajram[2], Rabab Yasin[3], Eissa Sayed Ahmed[2]

[1]Faculty of Medicine, Helwan University, Egypt
[2]Neonatal Intensive Care Unit, Jahra Hospital, Kuwait
[3]Radiology Department, Jahra Hospital, Kuwait

Abstract

Perinatal stroke is not rare; the estimated incidence of ischemic perinatal stroke ranks second to the incidence of ischemic stroke in older adults, and exceeds the incidence in childhood by approximately 10-fold. It is defined as cerebro-vascular events that occur between 20 weeks of fetal life and 28 postnatal days. Herein, we report a full-term female infant with multiple recent infarcts involving the anterior cerebral artery distribution. The diagnosis was made through diffusion-weighted magnetic resonance imaging.

Keywords

Perinatal arterial ischemic stroke, anterior cerebral artery syndrome.

Corresponding author

Dr Mahmoud Alhosiny Fayed, MD, Lecturer of Pediatrics and Neonatology, Faculty of Medicine, Helwan University, Egypt; Senior Registrar, Neonatal Intensive Care Unit, Jahra Hospital, Kuwait; phone: 0096560979369; email: drmahmoud38@hotmail.com.

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Introduction

Perinatal stroke is not rare; the estimated incidence of ischemic perinatal stroke ranks second to the incidence of ischemic stroke in older adults, and exceeds the incidence in childhood by approximately 10-fold. The estimated incidence of perinatal stroke is between one in 1,600 and one in 3,000 live births and the estimated mortality rate is 3.49/100,000 annually. It is defined as cerebrovascular events that occur between 20 weeks of fetal life and 28 postnatal days [1]. The mode of presentation distinguishes 2 varieties; acute perinatal stroke, which occurs in newborn infants at or near birth, and presumed perinatal stroke, which refers to chronic infarcts, diagnosed in a delayed manner [2]. Herein, we report a full-term female infant with multiple recent infarcts involving the anterior cerebral artery (ACA) distribution. The diagnosis was made through diffusion-weighted magnetic resonance imaging (DWMRI).

Case report

A female, full term infant was delivered by spontaneous vaginal delivery. Apgar score was 7 and 9 at 1 and 5 minutes, respectively. She was appropriate for gestational age, with birth weight 2.930 kg. Mother was Para 2 with no known risk factor. The baby did not need active resuscitation and was shifted beside her mother after routine care. At the age of 25 hours, the baby was noted to have right-sided focal colonic convulsion that was aborted by phenytoin. The baby was admitted to Neonatal Intensive Care Unit (NICU) and, immediately, random blood sugar, calcium, magnesium and sodium were checked and all were normal. On examination, there were no dysmorphic features. No skin rash, no hypo- or hyperpigmentation. The vital signs were normal; temperature = 37°C, heart rate = 140/min, respiratory rate = 40, saturation = 99% in room air, normal blood pressure for age and sex. Normal bilateral breath sound. Cardiovascular examination was normal. The abdomen was soft, with no hepato-splenomegaly. Normal neurological examination. Blood gases were normal in room air, CBC within normal limit, septic workup including CSF culture was negative. Chemistry within normal limits. Cranial ultrasound (US) was normal. Magnetic resonance imaging (MRI) brain was done at day 4 of life and revealed multiple recent ischemic insults (infarction) involving left frontal parasagittal cortical and subcortical area of ACA territory, bilateral corona radiata, genu of the corpus callosum, left external capsule. The left frontal area exerted a mild mass effect in the form of effacement of the related sulci and subtle frontal midline shift (**Fig. 1A-E**). In view of these findings, magnetic resonance angiography (MRA) and venography (MRV) were requested and showed absent A1 segment of right ACA (normal variant), otherwise normal MRA (**Fig. 1F**) with normal MRV. Further workup was done in the form of sickling test, coagulation profile, protein S, protein C, antithrombin III, factor V Leiden, lupus anticoagulant, amino acid chromatography, free and the total homocysteine level, official ECG, ECHO, and all came to be normal. Follow-up MRI was repeated at the age of 2 weeks and revealed chronicity of the left frontal parasagittal ischemia developing encephalomalacia with surrounding gliosis. We observed reversible changes (total recovery) of the previously mentioned diffusion restriction within the genu of corpus, corona radiata, internal and left external capsules. The patient was kept under maintenance leviteracitam and discharged home under a multidisciplinary team follow-up.

Discussion

Perinatal stroke includes both ischemic and hemorrhagic events resulting from the disruption of either arteries or veins from early gestation through the first month of life [2]. The clinical scenario and the MRI findings of this neonate were consistent with perinatal stroke of arterial ischemic type (PAIS). PAIS is a common cause of acute neonatal encephalopathy, and may manifest as altered mental status, seizures, sensorimotor deficits, hypotonia, and/or with subtle features like lethargy, feeding difficulties, temperature or hemodynamic instability. Our neonate presented with focal colonic convulsion involving the contralateral limb (right side), consistent with early symptomatic PAIS (60%), as the most common presenting symptom is neonatal seizures, of which 70-83% are focal seizures in the first 3 days of life [3]. Most cases of PAIS result from arterial infarction in the distribution of the middle cerebral artery; the main branch, a distal cortical branch, or smaller lenticulostriate branches can be involved. In a consecutive cohort study of 94 infants with 166 PAIS confirmed by MRI, the proportion of infarcts in the distribution of the middle cerebral,

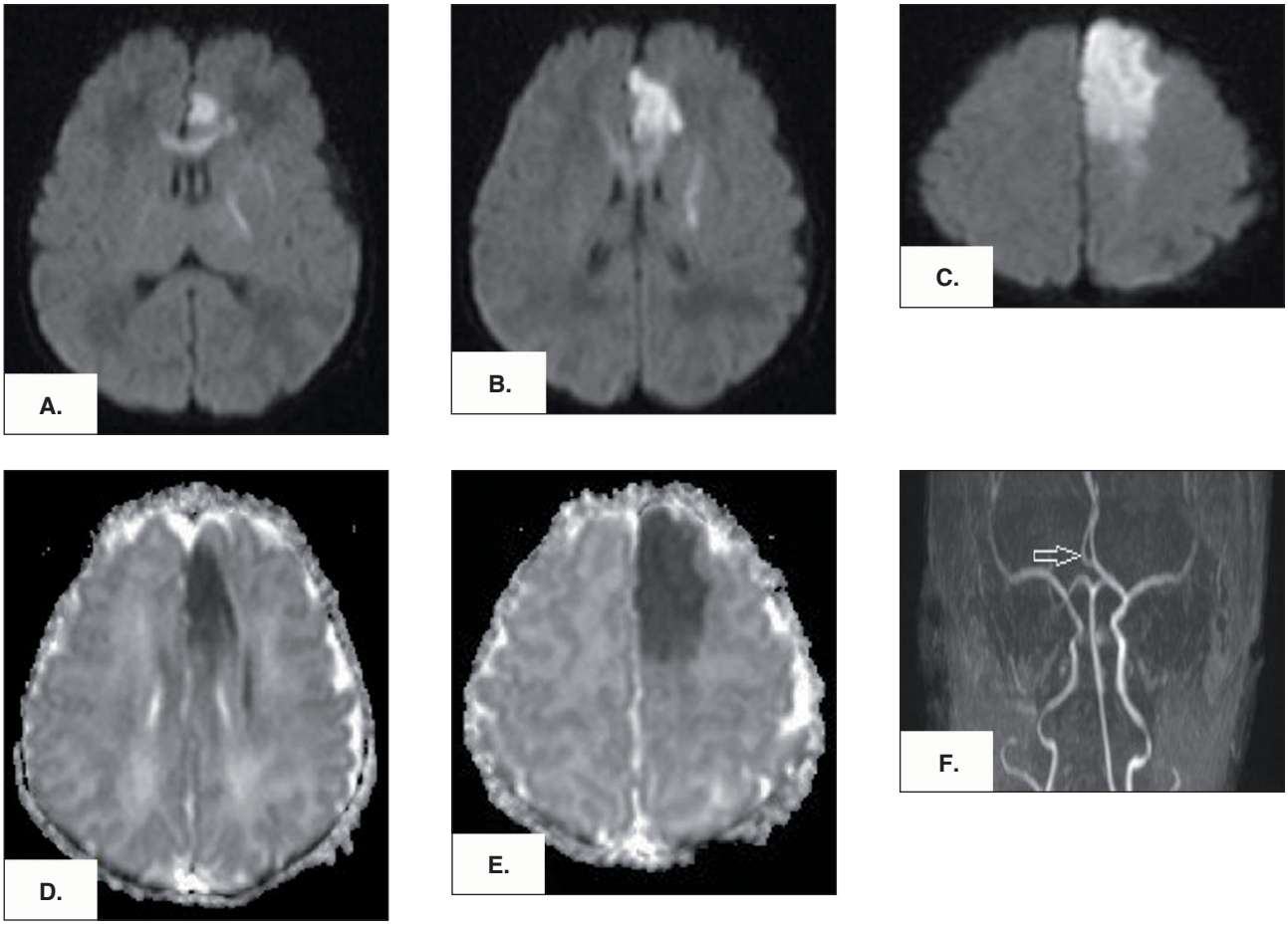

Figure 1. DWMRI (b value 1,000) revealed abnormal high DWMRI in genu of corpus callosum, both internal capsules more evident on the left side. Left external capsule and left medial frontal lobe (**A**, **B** and **C**) with corresponding low ADC value (**D** and **E**) indicating true diffusion restriction. On MRA (**F**), there was absent A1 segment of right ACA with A2 segment on right side supplied from left side.

DWMRI: diffusion-weighted magnetic resonance imaging; ADC: apparent diffusion coefficient; MRA: magnetic resonance angiography; ACA: anterior cerebral artery.

anterior cerebral and posterior cerebral arteries was 51%, 19%, and 18%, respectively. Cerebellar infarcts made up another 9% [4].

PAIS is thought to have a diverse underlying etiology and several potential risk factors in term infants are frequently cited. These include hereditary or acquired thrombophilias and environmental factors that occur before, during, and after delivery. However, establishing a causal role for many risk factors awaits larger, more definitive, prospective or case-controlled studies [5]. Thrombophilia evaluation in the neonate has limited clinical utility because levels of protein C, protein S, antithrombin, and factor XI are normally decreased to 30% of adults levels, and these levels only approach adult levels at various time points during childhood. Moreover, thrombophilia testing for the mentioned proteins in the neonatal period may be misleading and requires repeat testing for a confirmatory diagnosis [2].

On the other hand, thrombosis of placental vessels normally occurs as pregnancy ends, and emboli may be released into the fetal circulation as the placenta separates at birth. Moreover, placental pathology may lead to direct embolization into the fetal circulation or cause an inflammatory and prothrombotic state that promotes thrombus formation in the placenta and fetus [6]. This could explain the etiology in our case, as there was no determined risk factor. As a result, we recommend routine histopathological examination of the placenta.

Cranial MRI, especially DWMRI, is the most sensitive imaging modality for detecting PAIS, dating the injury, predicting the motor outcome of the child. Although ultrasonography with Doppler imaging of cerebral blood flow is useful in the neonate who is too ill to transport [7], our neonate trans-fontanel US was normal, consistent with previously reported low sensitivity of US

in PAIS, especially in small and deep-seated infarction [8].

Supportive care measures for PAIS in neonates include the control of seizures, the optimization of oxygenation, and the correction of dehydration and anemia. Antiplatelet therapy such as aspirin and anticoagulation with low-molecular-weight heparin or un-fractionated heparin are rarely indicated because of the low risk of recurrent stroke after neonatal PAIS [9].

The majority of neonates with PAIS experience residual neurological deficits. Golomb et al. in 2007 [10] summarized 111 children with perinatal stroke, including 67 who presented as neonates and 44 whose strokes were discovered later. Seventy-six children (68%) exhibited cerebral palsy, and 55 of these individuals had at least 1 additional disability; 45 (59%) experienced cognitive or speech impairment, and 36 (47%) had epilepsy. An early intervention program based on the best available evidence of interventions that work in older children (Goals – Activity – Motor Enrichment [GAME] protocol) was evaluated in infants in a single randomized trial with promising results, showing improved motor outcomes of participants compared with standard care [11].

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Declaration of interest

The Authors have no conflicts of interest to disclose.

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