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MRI of bilirubin encephalopathy (kernicterus): A case series of 4 patients from Sub-Saharan Africa, May 2017

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ABSTRACT

Characteristic magnetic resonance imaging (MRI) findings in patients with chronic kernicterus are bilateral and symmetric T2-weighted hyperintensities in the globus pallidus. We report 4 cases of infants with clinical, laboratory, and MRI findings of kernicterus in this case series. This is the first MRI report of kernicterus in Ethiopia. Awareness of the disease is raised in this report, and the role of magnetic resonance in detecting signal abnormalities associated with kernicterus in the globus pallidi is underscored. We recommend MRI to be part of the investigation in neonates with jaundice.

Introduction

Kernicterus, also known as chronic bilirubin encephalopathy, describes the chronic, toxic, and permanent sequelae of high levels of unconjugated bilirubin on the central nervous system of infants. It is part of the spectrum of bilirubin-induced neurologic dysfunction, which also includes acute bilirubin encephalopathy. Kernicterus is thought to be very rare and decreasing in incidence, although the exact number of incidence is unknown [1,2].

Schmorl first used the term kernicterus in1904 to describe the yellowish staining in the areas of the brain stem and basal ganglia at autopsy in babies who had marked hyperbilirubinemia before they died [1]. Kernicterus literally means" yellow kern", in which kern indicates the commonly affected region of the brain, namely the nuclear regions. The basal ganglia, hippocampus, geniculate bodies, and cranial nerve nuclei including the oculomotor, vestibular, cochlea, and dentate nuclei are most commonly involved [2].

There is no disease-modifying treatment available for this condition, and prognosis is poor. Early management of

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neonatal hyperbilirubinemia, with therapies such as phototherapy and exchange transfusion, should be employed to prevent kernicterus.

Magnetic resonance imaging (MRI) has demonstrated high sensitivity in kernicterus and is the imaging modality of choice [3–5]. The posteromedial borders of the globus pallidi seem to be the most sensitive regions of the brain in detecting signal anomalies associated with kernicterus. These signal anomalies typically and initially show on T1-weighted sequences hyperintense signals in the acute phase but eventually become hyperintense on T2-weighted sequences as the disease progresses. Diffusion-weighted images show normal appearance. Magnetic resonance spectroscopy has been scantly mentioned in the literature in the diagnosis of kernicterus, and studies have reported increased levels of glutamate and decreased levels of choline and N-acetyl-aspartate [3–6].

Here we report 4 cases of patients with kernicterus who had perinatal hyperbilirubinemia and who subsequently had choreoathetoid and dystonic motor disorders. Brain MRI showedT2-weighted hyperintensities in the globus pallidi of the 4 cases.

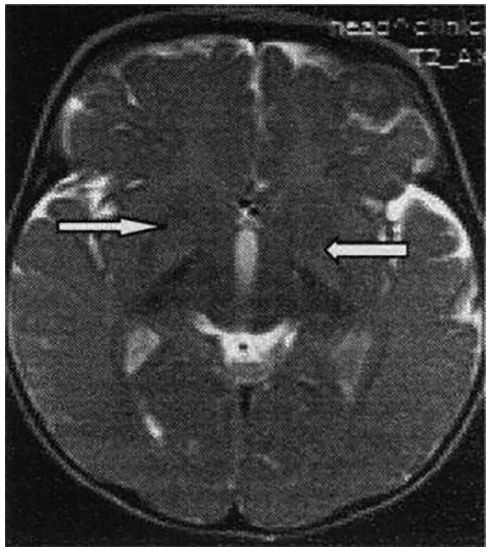

Fig. 1 – Case 1. Axial T2W magnetic resonance image showing bilateral T2 hyperintensities of the globus pallidus (arrows).

Case I

A full-term male infant was born by spontaneous vaginal delivery after normal pregnancy. He was well till the fourth day of life when his mother noticed a yellowish discoloration in the eyes and skin, as well as failure to suck. Hyperbilirubinemia was the cause of A, B, O blood group incompatibility and the serum bilirubin level at that time was 40 mg/dL (normal bilirubin level is <12 mg/dL) with direct bilirubin level at 1.4 mg/dL. He needed an exchange transfusion and was discharged after a 5-day hospital course.

At 6 months of age, the patient presented with abnormal, contorting limb and torso movements, and delayed motor milestones—he was not able to roll over. It was found on physical examination that head circumference (HC) was 43.5 cm between the mean and two standard deviation (+2SD) for age and gender. His eyes were crossed, he had dystonic extremities, and subjective hearing loss. MRI of the brain taken at presentation (age of 6 months) to explain the dystonic motor disorders that the patient had showed T2-weighted hyperintensities in the globus pallidus bilaterally (Fig. 1), and diagnosis was confirmed on MRI.

Case II

A full-term female infant, who was born by cesarean section because of previous cesarean delivery of the mother, weighed 2 kg on the third day of life. Her mother noticed yellowish discoloration of the eyes and the skin. Serum bilirubin level at that time was reportedly greater than 50 mg/dL. On physical examination, her HC was 45.5 cm between the mean and +2SD for age and gender, and she had generalized dystonia with some oral movements. At 9 months of age, she presented with generalized dystonia with some oral movements. MRI, taken at the time of presentation, showed T2-weighted hyperintensities in the globus pallidus (Fig. 2).

The hyperbilirubinemia that the baby had after delivery was due to hemolysis caused by Rhesus factor (RH) incompatibility. Her mother's blood type was O and RH-negative but she was RH-positive.

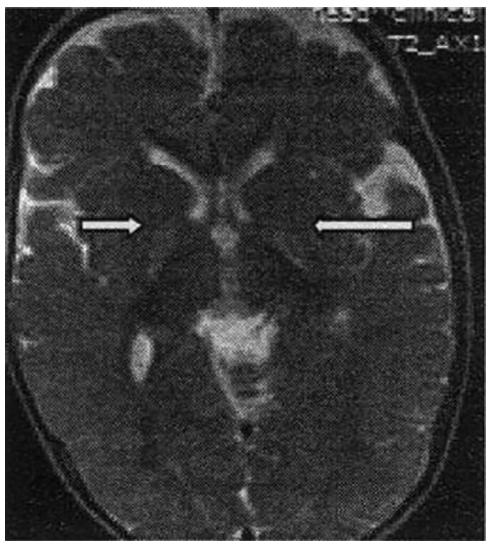

Fig. 2 – Case 2. Axial T2W magnetic resonance image showing symmetric hyperintensities in the globus Pallidus (arrows).