Letters to the Editor

Neonatal Hyperbilirubinemia due to Hereditary Spherocytosis

Hereditary spherocytosis is a rare cause of neonatal hyperbilirubinemia and Medline search from 1966 onwards revealed only one case from India(1). Fifty percent of patients with hereditary spherocytosis give a history of jaundice in the neonatal period but it is often passed over as physiological jaundice(2). We present a neonate who was diagnosed after seeing spherocytes on the peripheral smear.

A term, 2.9 Kg baby born to a B+ve primi gravida mother developed jaundice at 3 days of age. His serum bilirubin was 18mg/L (direct component 1 mg/L). His PCV was 40, reticulocyte count was 12%, G6PD levels were normal, blood group was B+ and peripheral smear examination revealed spherocytes. Mother gave a history of splenectomy at 10 years of age and she had been diagnosed to have spherocytosis. She has been asymptomatic with a hemoglobin of 11 g/dL.

Baby's DCT was negative, MCV was 84.2 fl and MCHC was 36.2%. Osmotic fragility was significantly increased. Hemolysis started at 0.72% NaCI and completed at 0.56% NaCl as against the control sample in which hemolysis started at 0.48% NaCl and completed at 0.4% NaCl.

Phototherapy was given for 72 hours during which serum bilirubin initially rose to 21 mg/dL and subsequently decreased to 10mg/dL. Patient was started on oral folic acid and parents were counseled as to long term outcome.

Diagnosis of hereditary spherocytosis is suspected when spherocytes are seen on the peripheral smear. Spherocytes can occasionally be seen in normal newborns, in ABO incompatibility and autoimmune hemolytic anemia. Blood grouping and DCT help to rule out the latter two. The mean corpuscular hemoglobin concentrations (MCHC) are elevated (35-36%) and MCV is in the normal range as seen in our patient(3). Osmotic fragility may be done using neonatal controls as the newborn RBC is relatively more resistant to hemolysis(4). Treatment in the neonatal period is directed towards treating hyperbilirubinemia. Rarely, packed cell transfusion for symptomatic anemia may be required. The definitive treatment is splenectomy which is best deferred till 5 years of age.

Gouri Rao Passi,
Shashwat Saran,
Department of Pediatrics,
Choithram Hospital & Research Center,
Indore 452 001, India.
E-mail: gouripassi@hotmail.com

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