Case Reports

Neonatal Lupus Mimicking Extra Hepatic Biliary Atresia

Madhuri Kanitkar
K.P.V. Rohini
Bipin Puri*
M.N.G. Nair

Neonatal Lupus Erythematosus (NLE) is an uncommon condition manifesting with congenital complete heart block and occasionally other manifestations like hepatitis. Neonatal Lupus with severe cholestasis with positive anti SS-A/Ro and anti SS-B/La antinuclear antibodies in the mother and child is being reported.

Key words: *Neonatal hepatitis, Congenital heart block, Cholestatic jaundice.*

Neonatal Lupus Erythematosus (NLE) results from maternal transfer of IgG autoantibodies, between 12th and 16th week of gestation. The principal serological markers are anti SS-A/Ro and anti SS-B/La maternal antinuclear antibodies that are transferred across the placenta and can be detected in the affected child for the first few months of life. Manifestations include congenital heart block, cutaneous lesions, thrombocytopenia, neutropenia, pulmonary or neurological disease and rarely hepatitis(1). Of these congenital heart block and cutaneous lesions are the most common occurring in 54% and 37% of cases respectively(2). During the past decade it has become clear that hepatobiliary disease may also occur as a manifestation of lupus. Although several hundred cases of neonatal lupus have been reported only a handful have been noted to have hepatobiliary disease. Hepatobiliary disease is a rare manifestation of neonatal lupus and may pose a diagnostic challenge. We report a case of neonatal lupus presenting with cholestatic jaundice mimicking extra hepatic biliary atresia (EHBA) and a complete heart block, born to an asymptomatic mother.

From the Departments of Pediatrics and Pediatric Surgery, Command Hospital & Armed Forces Medical College, Pune 411 040, India.

Correspondence to Lt. Col. Madhuri Kanitkar, Associate Professor, Department of Pediatrics, Command Hospital, Pune 411040, India. E-mail: madhurikanitkar@rediffmail.com

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Case Report

A 45-day-old baby presented with progressively increasing jaundice noted since second week of life accompanied by pale stools and high colored urine. Baby was born at term to a healthy 28 year old mother with no history of previous abortions or stillbirths, an uneventful antenatal period and with one previous three year old healthy female child. At presentation the baby appeared to be thriving and feeding well with a weight of 3.6 kg (birth weight 2.7 kg) but having bradycardia (heart rate 66/min) and hepatosplenomegaly (liver - 5 cm, spleen -2 cm) with an ECG showing complete heart block. There were no skin rashes or murmurs. Hematological investigations were normal except for a conjugated hyperbilirubinemia and raised liver enzymes (Serum bilirubin 17.2 mg/dL, SGOT 632 IU/L, SGPT 236 IU/L, Alkaline phosphatase 2512 IU/L). TORCH titers, viral markers and thyroid function tests

were normal for both mother and baby. Echocardiographic evaluation revealed a small persistent foramen ovale defect with no other abnormality. The technetium-99m diisopropyl iminodiacetic acid hepatobiliary scintigraphy and Magnetic Resonanance Imaging were consistent with the diagnosis of EHBA. In view of the urgency for surgery in EHBA, while awaiting serological evaluation for neonatal lupus, a diagnostic laparotomy with peroperative cholangiogram was performed with a possible Kasai procedure planned. This revealed a healthy gall bladder with free flow of dye into the duodenum ruling out an EHBA. Liver biopsy showed fibrosis with evidence of cholestasis. In view of the persistence of hepatitis as evidenced by raised enzymes, a short course of steroids was given with a decrease in serum bilirubin and hepatic enzymes. Serological reports available subsequently revealed significantly high anti SS-A/Ro and anti-S-B/La antinuclear antibodies but negative Anti-ds DNA titers in mother and baby.

Discussion

Neonatal lupus erythematosus is a rare disorder noted in only 1% of infants with positive maternal auto antibodies and the principal clinical characteristics are cardiac disease, notably congenital heart block, and lesions of cutaneous lupus(3). Mothers may be asymptomatic at the time of childbirth without signs or symptoms of lupus erythematosus or other collagen vascular disease. Heart block occurs in approximately 1-5% of pregnancies in mothers with anti SS-A/Ro and anti-SS-B/La antinuclear antibodies, independent of mothers disease status and in 15-20%of of pregnancies following the birth of a child with neonatal lupus(4). Hepatobiliary disease in NLE may be missed in case of asymptomatic mothers because of its often mild nature, however around 10-26% may have significant hepatobiliary involvement(5) However, idiopathic neonatal cholestasis without cardiac or cutaneous findings may not represent NLE(6).

Three clinical variants have been described namely severe liver failure, conjugated hyperbilirubinemia with mildly raised enzymes, or a mild elevation of enzymes alone(7). Our patient had conjugated hyperbilirubinemia with markedly raised enzymes, but the two commonly used modalities for imaging the biliary tract namely, the technetium-99m diisopropyl iminodiacetic acid hepatobiliary scintigraphy and Magnetic Resonanance Imaging were suggestive of EHBA and the baby underwent a per operative cholangiogram to avoid delay in the surgical management of biliary atresia. A serum gamma glutamyl transpeptidase level (>150 u/L) if used in conjunction with the hepatic scintigraphy has been reported to increase the sensitivity of the scintigraphy to 100% for EHBA and may result in a reduction of the false positivity observed with these tests individually(8) as happened in our patient.

Even though the prognosis is excellent in hepatobiliary disease, considering the fibrotic changes noted on liver biopsy in our case, a long term follow up is necessary to look for progress to a chronic liver disease.

Contributors: MK treated the case, revised the manuscript and will act as guarantor for the paper. KPVR did the literature search and prepared the manuscript. BP was involved in the operative management of the patient and MNGN critically reviewed the document.

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Scrub Typhus

Sreeja Pavithran
Elizabeth Mathai*
Prabhakar D. Moses

Scrub typhus is being increasingly reported in adults in India.It should be considered a strong possibility in all undifferentiated fevers. Two children with this infection are being reported highlighting the wide variation in clinical presentation. Specific tests should be preferred over Weil Felix test wherever possible especially in areas reporting a high incidence of the infection.

Key words: *India, Scrub typhus.*

Scrub typhus is a zoonosis, widely prevalent in many parts of Asia including India(1). Though there are reports of this infection occurring not uncommonly in South India(2,3), in clinical settings, the index of suspicion is still low. The typical rash and eschar may not be always present(3), leading to missed diagnosis. Scrub typhus is associated with about 10% mortality in our area in adults(3). There are no reports, to the best of our knowledge, on manifestations of scrub typhus in children in India. We report scrub typhus in 2 girls aged 10 and 12 years, respectively.

Case Reports

Two girls, both from areas around Vellore were admitted in November 2003 with prolonged fever. Their specific features are presented below.

From the Department of Child Health and Department of Clinical Microbiology, Christian Medical College, Vellore 632 004, India.

Correspondence to Dr. Prabhakar D. Moses, Professor and Head Department of Child Health, Christian Medical College, Vellore 632 004, India. E-mail: child3@cmcvellore.ac.in

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