ACTA SCIENTIFIC MEDICAL SCIENCES (ISSN: 2582-0931)

Volume 3 Issue 10 October 2019

Case Report

A Case of Biliary Atresia and Intracranial Bleeding Associated with Cytomegalovirus Infection

Rabia Noreen[1], Rafaqat Bota[2], FNU Farukhuddin[3]*, Mushtaq Ahmed[4] and Neelam Bashir[5]

[1]*Resident Paediatrics, civil hospital Karachi, Pakistan*

[2]*Dow Medical College, Karachi, Pakistan*

[3]*University Hospital, Cleavland Medical Center, Cleavland, USA*

[4]*FCPS-II, Senior resident Paediatrics-III, Civil Hospital Karachi, Pakistan*

[5]*Intern, Lyari General Hospital, Karachi, Pakistan*

***Corresponding Author:** Farukhuddin, Neurology, University Hospital, Cleavland Medical Center, Cleavland, USA.

Received: August 23, 2013; **Published:** September 12, 2019

DOI: 10.31080/ASMS.2019.03.0407

Abstract

In the literature, the association of biliary atresia with cytomegalovirus infection and presentation of infants with intracranial bleeding has been reported. We present a case of 02 months old female child who presented to Paediatrics ER with deep jaundice and intracranial bleeding. CT scan findings were suggestive of massive frontoparietal bleeding and patient's torch profile (cytomegalovirus Ig M) was also positive. After stabilization, the patient was referred to pediatrics surgery after a HIDA scan confirmed the findings of biliary atresia. In conclusion, biliary atresia should be considered in all patients with cholestasis and intracranial bleed and should also be searched for cytomegalovirus infection.

Keywords: Biliary Atresia; Intracranial Bleeding; Cytomegalovirus Infection

Abbreviations

CT scan: Computed Tomography; ER: Emergency; HIDA: Hydroxy-Imino-Diacetic Acid; BA: Biliary Atresia; CMV: Cytomegalovirus; USG: Ultrasonography; PCR: Polymerase Chain Reaction.

Introduction

Biliary atresia (BA) is a serious cause of infantile cholestasis leading to liver transplantation in children. BA is a rare disease characterized by the progressive fibrosis and obliteration of the extrahepatic biliary system with the incidence of 1 in 10,000 live births, more commonly in Asians [1]. The etiopathogenesis of biliary atresia is poorly understood yet but cytomegalovirus has been associated in some cases and Fischler., *et al*. found cytomegalovirus infection in 39% of patients with biliary atresia [2]. We present a case of cholestatic jaundice who presented with massive intracranial bleed and also tested positive for CMV, which has never been published before.

Case Report

We present a case of 2 months old girl born of consanguineous marriage at full term with a birth weight of 2.5 kg presented with sudden unresponsiveness, seizures (in the form of uprolling of eyes) and deep jaundice with yellow-colored stools, history of voiding high colored urine since birth, increasing pallor and right focal convulsion. There was no fever or abdominal distension. She was on exclusive breastfeeds. On examination, weight was 3.8 kg, the length was 54 cms, she had pallor, icterus, tense bulging anterior fontanelle and hepatosplenomegaly. There were abnormal posturing and right side focal neurological deficit. Investigations showed hemoglobin of 3.7 g/dL, white cell count of 18,500/cumm (52% neutrophils, 43% lymphocytes), platelets of 5,81000/cumm. Her bilirubin was 9.06 mg/dL (direct bilirubin= 8.39 mg/dL), SGPT = 94 IU/L, total proteins = 3.8 g/dL, albumin=2.9 g/dL. Prothrombin time was 25.2 seconds and partial thromboplastin time was

49.7 seconds. Serum electrolytes, blood gases, and creatinine were normal. CT brain showed acute hemorrhage in Left frontoparietal region with midline shift towards the right side and edema all-over the head and effacement of sulci and gyri along the left cerebrum and mass effect with compression of the left lateral ventricle. USG abdomen showed hepatomegaly with normal texture, regular margins, no duct dilations and no triangular cord sign at porta hepatis was seen. There was also splenomegaly and contracted gall bladder. She immediately was treated with blood transfusion (Packed Red Cells) and anticonvulsants. Her TORCH profile (sent before blood transfusion) showed positive CMV IgM after that CMV antigen and PCR were sent which came out to be positive. Hepatobiliary scintigraphy (HIDA) done with TC-99m mebrofenin after 5 days priming with phenobarbitone showed good tracer uptake but no excretion at 4 hours and even after 24 hours in the intestine. Findings of the HIDA scan confirmed the diagnosis of biliary atresia after which the patient was referred to pediatrics surgery for Kasai procedure.

Discussion

In the literature, the relationship between cytomegalovirus infection and neonatal cholestasis have been reported but CMV causes mechanism that lead to pathogenesis of biliary atresia is not well established [2]. A study conducted by Fischler., et al. in Sweden reported that cytomegalovirus DNA was found positive in 50% of their patient with biliary atresia [3]. Another study conducted by Tarr., *et al*. reported that CMV infection was positive in 24% of their patients with biliary atresia and that study suggested that infants with cholestasis and cytomegalovirus infection should be searched for biliary atresia [4].

CMV replicates in both hepatocytes and cholangiocytes during infection and liver damage may be related to direct cytopathic effect or the immune response of the host [5]. Chang., *et al*. have reported that CMV DNA was detected in 46% of patients with neonatal hepatitis and suggested that CMV could play a major role in the pathogenesis of neonatal hepatitis [6]. Fischler., *et al*. [7] found that in patients with biliary atresia and CMV, IgM deposits were significantly higher in liver biopsies than in biliary atresia patients without CMV, suggesting that CMV may be triggering immunologic processes causing biliary atresia. Our case is unique for quite a few reasons. Firstly, intracranial hemorrhage as a presenting sign of biliary atresia is extremely rare and potentially incurable. Secondly, we have not identified any cases published to date in which infants who presented with this picture and tested also positive for cytomegalovirus infection.

Houwen., *et al*. reported a case series of 4 patients who presented with bleeding as the first sign of biliary atresia [8]. Of these 4 patients, 1 presented with intracranial bleeding. This case is important because it is typically thought that biliary atresia is a disease of the newborn and delay in diagnosis can be worrying especially where early diagnosis and management leads to improved outcomes. The third reason that our case is unique is that this is the first time that we are aware of in which a patient with biliary atresia complicated by intracranial bleeding with positive cytomegalovirus infection. Therefore any infant presenting with unexplained intracranial bleeding should be evaluated for biliary atresia and should be searched for cytomegalovirus infection.

In conclusion, we may hypothesize by our case that cytomegalovirus may play a pivotal role in the etiopathogenesis of biliary atresia and it is also suggested that infants presented with intracranial bleeding and cholestasis should be investigated for biliary atresia and cytomegalovirus infection.

Bibliography

1. Dellert S and Balistreri W. "Chapter 52 - Neonatal Cholestasis". In: Walker W, Durie P, Hamilton J, *et al* (eds). Pediatric Gastrointestinal Disease, third edition. Lewiston, NY: B.C. Decker Inc (2000): 880-891.

2. Fischler B., *et al*. "Early cytomegalovirus infection and the long-term outcome of biliary atresia". *Acta Paediatrics* 98 (2009): 1600-1602.

3. Fischler B., *et al*. "The viral association of neonatal cholestasis in Sweden: a possible link between cytomegalovirus infection and extrahepatic biliary atresia". *Journal of Pediatric Gastroenterology and Nutrition* 27 (1998): 57-64.

4. Tarr PI., *et al*. "Biliary atresia, cytomegalovirus, and age at referral". *Pediatrics* 97 (1996): 828-831.

5. Ozkan TB., *et al*. "Antiviral therapy in neonatal cholestatic cytomegalovirus hepatitis". *BMC Gastroenterology* 7 (2007): 9.

6. Chang MH., *et al*. "Polymerase chain reaction to detect human cytomegalovirus in livers of infants with neonatal hepatitis". *Gastroenterology* 103 (1992): 1022-1025.

7. Fischler B., *et al*. "Immunoglobulin deposits in liver tissue from infants with biliary atresia and the correlation to cytomegalovirus infection". *Journal of Pediatric Surgery* 40 (2005): 541-6.

8. Houwen RH., *et al*. "Bleeding as the first symptom of extrahepatic biliary atresia". *European Journal of Pediatrics* 146.4 (1987): 425-426.

Volume 3 Issue 10 October 2019