Case Report

Biliary Atresia with Situs Inversus: An experience shared

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Abstract

Biliary Atresia (BA) is a well-known entity and can present with multiple congenital anomalies. BA is one of the most common conditions in which pediatric liver transplant is performed. Identification of Biliary atresia with situs inversus (SI) has not been documented in Pakistan. We report two such cases. First was an eighty-day-old baby boy, icteric from day of birth. On further evaluation had dextrocardia, SI, gross hydronephrosis (HN) of left kidney and stasis at pelvi ureteric junction (PUJ). Liver biopsy showed biliary cirrhosis secondary to extra hepatic biliary atresia (EHBA). The second baby presented at two months of age. Ultrasound abdomen and hepatobiliary scintigraphy confirmed liver in left hypochondrium (SI) and findings suggestive of BA. Echocardiography confirmed SI with mesocardia. In this paper we have described the association of BA with SI in two patients presenting at the pediatric Gastroenterology, hepatology and nutrition unit.

Introduction

Among the congenital cholangiopathies, BA is the most common cause of end stage cholestatic liver disease leading to liver transplantation in pediatric population.[1] The disease is said to be associated with congenital anomalies that include polysplenia, preduodenal portal vein, malrotation, absence of Inferior Vena Cava, cardiac anomalies, intrapulmonary shunting, asplenia, pancreatic anomalies and Situs Inversus.[2,3] SI is a condition of unknown etiology[1], in which abdominal organs and lungs are reversed, left atrium is to the right and right atrium to the left. Although the two anomalies have been linked previously but no documentation is available in this respect in published literature of Pakistan. The objective of this study is to highlight the association of BA and SI in Pakistani population.

Case No. 1

Eighty days old male baby, the only child of his parents was reported to be icteric from first day of his life. He was delivered via caesarian section due to premature rupture of membranes with an unremarkable antenatal history. At birth he had normal height, weight and a good Apgar score. The child remained in nursery and received phototherapy, which mildly reduced his jaundice. On further evaluation, he was found to have dextrocardia and SI with gross hydronephrosis of left kidney and mild hydronephrosis of right kidney. His Urea, Creatinine and electrolytes (UCE) were within normal limits (Urea 12 mg/dl, Creatinine 0.19 mg/dl, Sodium 138 mEq/L, Potasium 4.7 mEq/L, Chloride 109 mEq/L, and Bicarbonate 22 mEq/L). X-Ray chest and CT scan showed cardiac apex directed towards right, stomach bubble in right hypochondrium and liver shadow in left hypochondrium (Figure 1a and 1b). Renal scintigraphy concluded a good functioning right kidney 60% relative function and no outflow stasis. Reduced functioning left kidney 40%, with complete outflow stasis at pelvi-ureteral junction. No intervention was done.

For his non-resolving jaundice hepatology review

Figure 1a.

was taken. His liver function tests (LFT) were, total bilirubin 8.8 mg/dl, direct bilirubin 6 mg/dl, Alanine transferase (ALT) 44 U/L, Aspartate transferase (AST) 106 U/L, Gamma glutamyl transferase (GGT) 1182 U/L, Alkaline phosphate 943 U/L, Prothrombin time (PT) 14.8/10.7 sec, Activated partial thromboplastin time (APTT) 35.7/28.7 sec and International normalization ratio (INR) 1.24. Ultrasound abdomen showed coarse echotexture of liver. Gall bladder and common bile duct were not visualized. Other investigations performed included Hepatitis B surface antigen (HbsAg), Hepatitis C antibody (Anti HCV) and Cytomegalovirus (CMV) PCR were negative. Toxoplasmosis, rubella, CMV and Herpes (TORCH) screen was non-conclusive and alpha-1 Anti trypsin was within normal limits.

Figure 1b.

Echocardiography showed dextrocardia with SI, hypertrophied Ventricles with ejection fraction (EF) 80%. Fine ventricular septal defect (VSD), moderate to severe tricuspid regurgitation (TR) and dilated left atrium (LA). Hepatic imino di acetic acid (HIDA) scan reported reduced hepatocyte uptake and impaired clearance, non-visualization of biliary tree and gut motility, findings suggestive of BA. Perhepatic cholangiography done, showed failure of visualization of biliary channels. Liver biopsy revealed hepatocyte arranged in cords with mild degeneration and cholestasis and sinusoids dilatation. Portal areas showed marked fibrosis with thick periportal band formation, many normal looking bile ducts seen surrounded by mononuclear and polymorph infiltrate. Impression was of biliary cirrhosis, secondary to extra-hepatic biliary atresia or mild form of intrahepatic biliary atresia.

Case No. 2

The patient was the third child of consanguineous parents with two healthy daughters. He was born at term via normal vaginal delivery following an uneventful pregnancy. His birth weight and height were less than fifth centile by National center of health statistics (NCHS) standard[4], occipitofrontal circumference (OFC) was normal, with no significant history of birth asphyxia or cyanosis reported. On third day of life on routine examination, he was suspected to have congenital heart disease. At 10th day of life due to umbilical bleeding and low hemoglobin, the child was transfused. He also received stitches at the site of umbilicus. This was followed by gradually deepening jaundice and clay colored stools. When he was two months old he was admitted in hospital for icterus and low-grade fever. His liver and spleen were moderately enlarged.

His LFT's at that time were, total bilirubin of 32.8 mg/dl, direct 22.1 mg/dl, Alkaline phosphatase 465 U/L, ALT 183 U/L, AST 271 U/L, GGT 474 U/L, PT 22.3/11.4 sec and INR 1.33. Microcytic hypochromic, iron deficiency anaemia was reported on peripheral blood film. HbsAg was negative and CMV IgM was positive. On echocardiography the child had situs inversus with mesocardia and complete atrioventricular canal defect committed to right ventricle (RV). Transposed great arteries with valvular and infundibular pulmonary stenosis were observed. This gave a conclusive diagnosis of complete cyanotic congenital heart disease. Ultrasound abdomen showed left sided, normal sized liver whereas gall bladder and common bile duct were not visualized. Hepatobiliary scintigraphy showed that the liver was located in the left hypochondrium (SI), with fair hepatocyte uptake and impaired clearance, non-visualized gall bladder and common bile duct. Findings were suggestive of BA.

Discussion

BA, also called progressive obliterative cholangiopathy in affected infants is present since birth. Genetic factors are suggested and inference of analysis of segregation patterns rationalizes the existence of two groups, one with various anomalies within the laterality

sequence and the other involving the cardiac, gastrointestinal and urinary systems.[5] These two patients identify with the first group mentioned above.

Also, to ones interest the first child had unilateral complete outflow stasis at pelviureteral junction leading to reduced functioning of kidney, which in our knowledge has not, been documented before with biliary atresia. This is an important finding as we can easily miss the co existence of BA in a patient suffering from pelviureteric junction obstruction and vice versa unless one has in mind the coalition between the two.

BA is said to occur in 28% of infants born with SI as compared to the 0.01% of the general population.[6] These figures suggest a significant incidence of this entity, yet not reported in the past in Pakistan. As the presence of two conditions simultaneously modifies the pre operative evaluation[7], standard operative techniques of liver transplantation[7,8] and post operative complications specific for Situs Inversus[6], we have in particular shared this experience here.

References

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