CASE REPORT

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Cystic Biliary Atresia: A Delayed Diagnosis of Obstructive Jaundice

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ABSTRACT Cystic biliary atresia is an uncommon variant of biliary atresia. It is the most usual cause of neonatal jaundice which requires surgical intervention and liver transplantation in children. Herein, we present a case of a 2-month-old baby with persistent obstructive jaundice, who was not properly investigated prior to this. After the diagnosis has been established, we proceeded with the Kasai-type portoenterostomy. Even slightly delayed, the patient recovered well postoperatively and subsequently jaundice resolved.

Keywords: Biliary Atresia; hyperbilirubinemia, neonatal; portoenterostomy, hepatic

Biliary atresia (BA) is a progressive inflammatory process of intra- and extrahepatic ducts resulting in fibrosis and subsequent obliteration of biliary tract in neonates.[1] It is a rare disease affecting from 5/100,000 to 32/100,000 of live births with highest incidence in Asia-Pacific region.[2] BA is unknown in aetiology; but postulated theories include failure of recanalization of the bile ducts, vascular insufficiency, genetic influences, environmental teratogenesis and perinatal viral infections namely cytomegalovirus, Epstein-Barr and human papilloma virus.[2] Without surgery, the patient will suffer from liver failure, cirrhosis and mortality within the first year of life.[2] We present a 2-month-old baby with delayed presentation of biliary atresia after having persistent episodes of obstructive jaundice.

CASE REPORT

A male infant, born to term was referred to us at 2 months of age with history of prolonged obstructive jaundice. The mother denied any eventful circumstances during perinatal periods. His serum total bilirubin initially was reducing in trend from 199.5 umol/L at day 1 to 112.7 umol/L at day 15 of life. However, at 2 months of age, the repeated serum total bilirubin was 155 umol/L. He had no prior workup done for the prolonged jaundice.

On examination, he was active with yellowish discolouration of the body extending up to the thighs. His abdomen was soft and the liver was palpable 3 cm below and upper border at 6th intercostal space. Spleen was

not palpable but he had a small umbilical hernia. The stool was pale and clay-like with dark yellowish urine. Liver function test showed a further rise in serum total bilirubin to 185 umol/L of which direct was 133.9 umol/L. Alanine transferase (ALT) was 369 U/L with alkaline phosphatase (ALP) of 797 U/L.

Ultrasonography (USG) of the abdomen revealed a well-defined cystic lesion at porta hepatis measuring 0.9x1.2 cm. Patient was subjected for a surgery. On-table cholangiogram was performed to supplement the diagnosis. Intraoperative findings revealed a cirrhotic liver with neovascularization and cystic biliary atresia (CBA) (Figures 1, 2). A Kasai-type portoenterostomy was performed following which bile flow was restored and his jaundice cleared (Figure 3). He recovered well postoperatively.

DISCUSSION

Cystic BA characteristically presents with dilatation of obliterated extrahepatic biliary tree. It accounts for 5% of large series and can be detected during maternal USG.[3,4] Conjugated jaundice and pale stools are consistent findings in neonatal period. The CBA is classified according to Japanese Association of Paediatric Surgeon (JAPS) which based on the level of the most proximal obstruction; type I is at the level of the common bile duct, type II at the level of the common hepatic duct and type III at the level of the porta hepatis.[5]

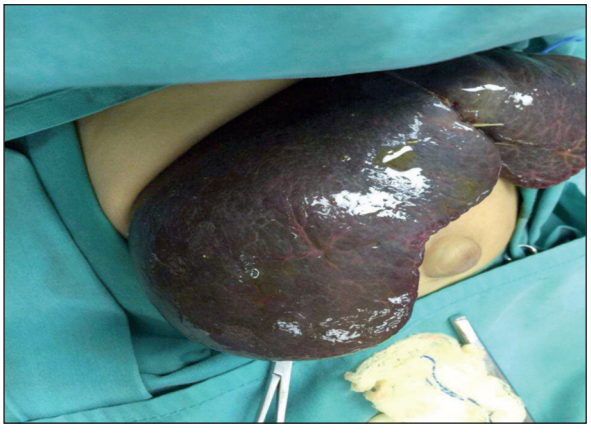

FIGURE 1: Intraoperative finding revealed hepatomegaly with near cirrhotic liver.

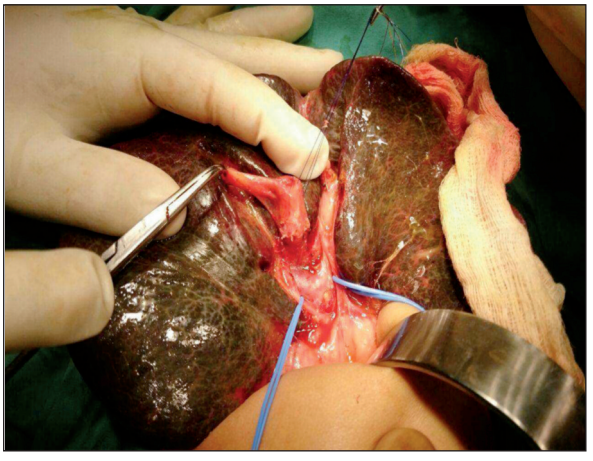

FIGURE 2: Cystic biliary atresia with evidence of obliteration of distal common bile duct and dilated proximal biliary system.

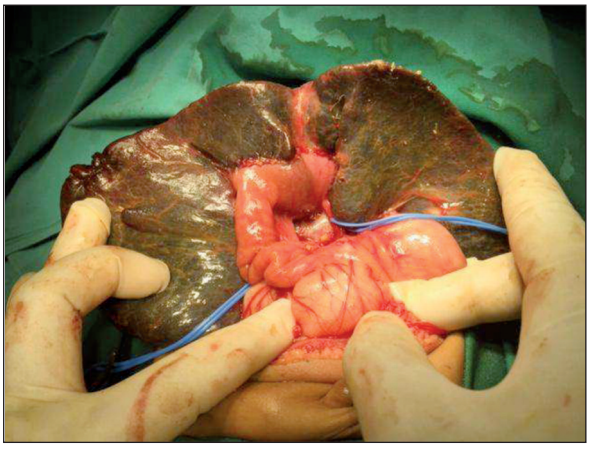

FIGURE 3: Kasai-type portoenterostomy.

USG plays a crucial role for investigating infant with persistent jaundice; to establish the choledochal structural anomalies, inspissated bile syndrome, perforated biliary duct, and to confirm the diagnosis of BA.[5] For non-cystic BA, the 'triangular cord' is the pathognomonic sign for establishing the diagnosis.[6] Triangular cord is characterized by echogenic appearance anterior to the wall of the right portal vein of >4 mm on longitudinal scan and corresponds to the obliterated proximal remnant in the porta hepatis.[7] A study has shown that this sign has a sensitivity over 90% and 98% specificity and particularly useful to differentiate between CBA and choledochal cyst.[7,8]

It is recommended that an urgent laparotomy and reconstructive surgery are indicated upon confirmation of presence of cyst, non-dilated intrahep-

atic biliary tree and abnormal liver profiles.[3] The main surgical procedure is to create hepatic portoenterostomy, first devised by Japanese surgeon, Morio Kasai in 1955.[9] Complete clearance of jaundice through promotion of bile flow into intestine, restoration of liver function and healthy hepatic growth are the purposes of this surgery. It is usually performed after delineation of the anatomy via diagnostic cholangiogram. In this procedure, the fibrous tissues that obstruct the communicating intrahepatic bile system will be transected at portal plate. Once the channel is opened, the bile flows will be established through an anastomosis of jejunal Roux-en-Y.[9] The extent of liver fibrosis will be determined by the liver biopsy, obtained during the surgery.

One of the factors that may determine outcome of surgery is age. In a series of 29 infants with CBA, it has been demonstrated that the CBA variant showed a clear relationship between age at portoenterostomy and clearance of jaundice. Those who are operated at <40 days had successfully cleared their jaundice compared to none at >70 days.[4] Better bile flow is established in patients undergoing surgery at younger age (less than 6-8 weeks of age) as the disease has a better prognosis in congenital forms of biliary atresia. Success is defined when the infant becomes non-icteric (no jaundice) by 3 months postoperatively and 50% have normal or near normal liver function. Besides, other prognostic factors include the accessibility to liver transplantation and experience of the centre managing BA patients.[2]

Ascending cholangitis, portal hypertension, hepatopulmonary syndrome and malignancies are among the serious and dreaded complications; therefore, patient should be closely follow-up after a successful surgery.[2] A combination with liver transplant, children will obtain a 90% of 4-year survival rates.[10]

In conclusion it cannot be stressed enough on how important is early workup to establish proper diagnosis of CBA. Even slightly delayed at 2 months of age, prognosis is excellent provided that the patient is managed in specialized center with an excellent surgical technique and perioperative care.

Informed Consent

Informed consent was obtained from the parents of the patient.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Aishath Azna Ali, Marjmin Osman;* ***Design:*** *Nornazirah Azizan;* ***Control/Supervision:*** *Che Ismail Che Noh, Marjmin Osman;* ***Literature Review:*** *Aishath Azna Ali,* ***Writing the Article:*** *Aishath Azna Ali, Firdaus Hayati,* ***Critical Review:*** *Che Ismail Che Noh.*

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