CASE REPORT

A Rare Case of Association of Jejunal Atresia with Biliary Atresia

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ABSTRACT

The incidence of biliary atresia (BA) is about 1 in 10000 births. It is fatal if left untreated. BA is known to be associated with various congenital anomalies however, the association of BA with jejunoileal atresia is extremely rare (0-0.5%). We encountered a case of a term baby who was born term with no significant antenatal history. She presented with intestinal obstruction concluded as jejunal atresia which was later associated with BA after extensive investigations. It caused challenging management and dilemmas in diagnostic due to multifactorial. Hence, keeping a high index of suspensions is important as the association causes detrimental effects to the patient. Further advanced radiological investigations may help in this challenging scenario.

Keywords: Biliary atresia, Jejuno-ileal atresia, Cholestatic jaundice, KASAI procedure, Total parenteral nutrition.

INTRODUCTION

Biliary atresia (BA) is an inflammatory and fibrotic obliteration of the biliary tree causing cholestasis and progressive liver failure, which account for approximately 1 in 10000 births. It is one of the most common causes of surgically treated neonatal cholestasis. However, if left untreated, it leads to mortality due to hepatic failure in the first two years of life birth (1).

Most patients diagnosed with BA are classified as classical BA with no associated congenital anomaly. Nevertheless, some patients do have associations with other congenital anomalies such as vascular anomalies, cardiac lesions, gastrointestinal anomalies, and others (2). We reported a rare case of biliary atresia in a patient with jejunal atresia which operated earlier. The association of BA with intestinal atresia is very rare with incidence reported worldwide widely range 0-0.5%. The percentage of BA with jejunal atresia was documented at around 0-5% (2). Having both biliary and jejunoileal atresia put us a challenge in terms of early diagnosis and management of the patient as post-operatively they will be commenced on Total Parenteral Nutrition (TPN) which might cause the patient to have TPN cholestasis if this patient developed jaundice post operatively. This contributes to the delay in diagnosing BA and causes detrimental results later.

CASE REPORT

SNA’s baby is 3 months old Malay girl, born via spontaneous vaginal delivery at 40 weeks and 2 days with a birth weight of 3.8 kg at the district hospital. The mother’s antenatal and delivery history was uneventful. At 17 hours of life, she developed episodes of vomiting (yellowish to greenish vomitus). She was admitted for clinical sepsis and was kept nil by mouth. Her first bowel movement was at 45 hours of life. Subsequently, noted to have fecal content aspirated from the Ryle tube at 51 hours of life. Abdominal x-ray showed an absence of bowel gas on the right side. On day 3 of life, she was transferred to the tertiary center for suspected intestinal obstruction. Urgent ultrasound abdomen was normal. However, a lower gastrointestinal study revealed a micro colon with colonic atresia, thus proceeding with laparotomy, small bowel resection, and primary anastomosis at day 4 of life. Intraoperatively, there was jejunal atresia type 3a with a proximal blind end 56 cm from the duodenojejunal junction, a mesenteric defect, small bowel kinking distally, large bowel collapse, and a micro colon beginning from the ascending colon.

However, she manifested obstructive jaundice features: pale-colored stool and tea-colored urine at 10 days post-operation. Simultaneously, her liver function deteriorated and showed persistent conjugated hyperbilirubinemia with the highest direct bilirubin 278 micromole/L (72%) with transaminitis. Full blood counts showed thrombocytosis (highest platelet of 672x10⁹/L). Blood culture and sensitivity yield MRCONS which are sensitive to vancomycin. Other infective screening...
was normal. To note that, she was commenced on total parental nutrition for 2 weeks duration post operatively. She did not have any other associated congenital anomalies and no genetic study was done.

Clinically, she was not dysmorphic, jaundiced, hepatomegaly 2 cm below the right subcostal margin. Otherwise, there was no splenomegaly, ascites, or other signs of chronic liver disease, and no other congenital anomalies were detected.

The first hepatobiliary ultrasound performed on day 14 of life revealed evidence of portal venous gas, most likely following corrective surgery, but no evidence of BA. Repeated ultrasounds of the hepatobiliary system done on day 24 of life also revealed normal findings. However, given persistent conjugated hyperbilirubinemia, magnetic resonance cholangiopancreatography (MRCP) and hepatobiliary iminodiacetic acid (HIDA) were done. MRCP on day 29 of life confirmed biliary atresia, most likely cystic type. However, surgical intervention was not immediately done as previous intraoperative findings revealed a normal gall bladder and the hepatobiliary system. Thus, the HIDA scan done on day 49 of life showed evidence of biliary obstruction and was unable to exclude the biliary atresia. The Kasai procedure was performed on day 57 of life. Intraoperatively, small bowel adhesion and dense adhesion between the liver and small bowel were noted. Histopathological examination (HPE) revealed features of cholestasis which consistent with BA.

DISCUSSION

The commonest cause of small bowel atresia is jejuno-ileal atresia (2). The cause of jejunal atresia in our patient was not ascertained as she had no meconium-stained upon review, neither significant antenatal history nor positive family history. It could indicate of congenital anomaly.

Unfortunately for our patient, she developed conjugated hyperbilirubinemia with transaminitis, and later turned out to be biliary atresia. It was difficult to differentiate between BA and TPN cholestasis as she was maintained with TPN post-operation, and the operation was documented as normal gallbladder during the first operation. Cholestatic jaundice is one of the known side effects of total parenteral nutrition. This TPN-induced jaundice will usually occur in a patient who is on prolonged TPN for more than 2 weeks (3).

The patient underwent serial ultrasound Hepatobiliary which revealed normal results. Ultrasound HBS is the first-line modality for cholestatic jaundice with a positive prediction value of 95% if the operator can delineate the presence of a triangular cord sign (the visualization of the fibrotic cord in the portal vein). Nevertheless, due to persistent obstructive jaundice features, BA could not be ruled out. There were multiple modalities of imaging in diagnosing BA including ultrasound HBS, MRCP, and HIDA scan. Based on a few studies, all concluded that the HIDA scan had the most sensitive percentage in diagnosing BA and a very small margin of false negatives. However, MRCP also yielded positive findings as they also had a high percentage of sensitivity.(4). HIDA scan showed a sensitivity of 98.7% and specificity of 70% in diagnosing BA. Other modalities such as MRCP showed a sensitivity of 99%, and a specificity of 36% and USG HBS had a specificity of 68.5%. (5). Thus, our patient did undergo MRCP and was consistent with the findings of BA, however, the surgical team still waited for the HIDA scan for confirmation of diagnosis. In the future, probably a case like this may need to be considered to proceed with on-table Cholangiogram (OTC) rather than wait for HIDA as delay in intervention may give rise to further complications and affect the survival rate for the patient.

She was discharged well at 2 months old and still jaundiced. Throughout the hospital stay, she completed multiple courses of antibiotics and was discharged with oral ursodeoxycholic acid. She was readmitted to the general hospital at 3 months old due to septicemia and subsequently succumbed to her illness.

![Figure 1: MRCP T1 sequence, showing tubular cyst structure with hypointense signal. The feature is in keeping with biliary atresia (cystic type).](image1)

![Figure 2: MRCP T2 sequence, showing tubular cyst structure with hyperintense signal. The feature is in keeping with biliary atresia (cystic type).](image2)

![Figure 3: HIDA scan: Good and homogenous tracer uptake in the liver. No significant tracer activity in biliary system, gallbladder, and upper small bowel at the end of 60-minutes. HIDA scan is suggestive of biliary obstruction.](image3)
Definite treatment for BA is the KASAI procedure (Portoenterostomy) and the time of intervention is crucial as it determines the prognosis of the patient. Around 70% of patient will have good bile flow if the operation done in the first 60 days of life.(5) Therefore, early recognition and ideal timing of intervention may yield a better outcome.

CONCLUSIONS

The association of JIA and BA is extremely rare. Keeping a high index of suspicion is important to monitor the association of mentioned anomalies. Other imaging modalities may help support the diagnosis of BA however, clinical evaluation and a high index of suspicion remained the pillar of diagnosis.

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REFERENCES