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Case Report

Extrahepatic cholestasis arising from congenital biliary cistern adhesion: a case report

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ABSTRACT

Biliary atresia (BA) is a progressive fibrotic obstructive condition that affects the biliary system, leads bile flow obstruction and the development of newborn jaundice. It is lethal if not treated, with a survival rate of less than 10% by the age of three. Bowel adhesions are anomalous bands of scar tissue that develop between bowel loops. The incidence rate for these bands remains uncertain. This study presents a 2-month-old infant with a progressive yellow discoloration dan diagnosed as BA. Intra operative colangiography was performed and congenital adhesions in the form of bands were discovered.

Keywords: BA, Congenital, Adhesion

INTRODUCTION

Biliary atresia (BA) is a progressive fibrotic obstructive condition that affects the biliary system both intrahepatic and extrahepatic. This leads bile flow obstruction and the development of newborn jaundice. Histopathological examination of liver biopsies reveals portal tract enlargement, edematous fibroplasia, bile ductule proliferation, and the presence of bile plugs in the duct lumen.[1] It manifests chronic jaundice, pale stools, and hepatomegaly in the newborn era. It is lethal if not treated, with a survival rate of less than 10% by age of 3.[2]

Bowel adhesions are anomalous bands of scar tissue that develop between bowel loops, typically not naturally connected to each other.[3] Congenital adhesion bands are commonly detected in pediatric patients, although the precise incidence rate for these bands remains uncertain.[4]

The only way to effectively diagnose and treat BA is through surgical investigation.[2] As long as the etiology of BA is unknown, surgery is the only treatment option for those suffering from this serious illness.[5]

CASE REPORT

A 2-month-old infant was brought to our center with a chief complaint of progressive yellow discoloration observed on the head, chest, and abdomen over the past two months. The patient's did not have any associated symptoms such as fever, cough, nasal congestion, vomiting, or respiratory distress. However, the patient had abdominal distension, pale stool, and yellow urine. The patient continued to feed adequately on both breast milk and formula. The patient was taken to the pediatrician's clinic and prescribed powdered medication, ursofalk, and apyalis. One week before, the jaundice and the unusual stool color once a day still remain. The birth history shows that the patient was born full-term, weight 2400 grams, length 46 cm, and did not cry immediately after birth. COVID Screening shows no suspicious to COVID and no history of allergies.

The pediatric assessment triangle (PAT) shows stable condition, weight 6700 grams, icteric sclera in both eyes, and Kramer's criteria grade III (Figure 1). On the first blood test obtained Hemoglobin 11.1, hematocrit 33,

leukocytes 26.0, platelets 245.000, erythrocytes 4.34, neutrophils 31.00, lymphocytes 57.70, SGOT 1.153, SGPT 618, total bilirubin 12.19, direct bilirubin 9.44, indirect bilirubin 1.11, PT 15.6, APTT 41.1, INR 1.210, Gamma GT 76, albumin 3.9, IgM anti CMV negative, and IgG anti CMV positive. Urinalysis shows 2+ bilirubin, and 2-phase abdominal ultrasound shows cholestasis with enlarged postprandial GB (Contractility index-7%) (Figure 2).

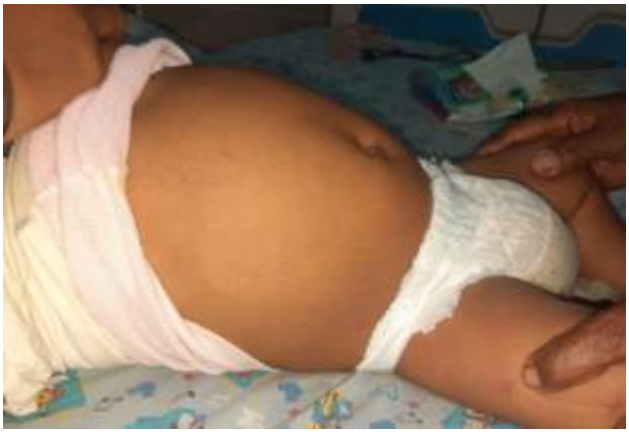

Figure 1: The patient had abdominal distension and icteric on the head, chest, and abdomen (Krammer III) over the past two months.

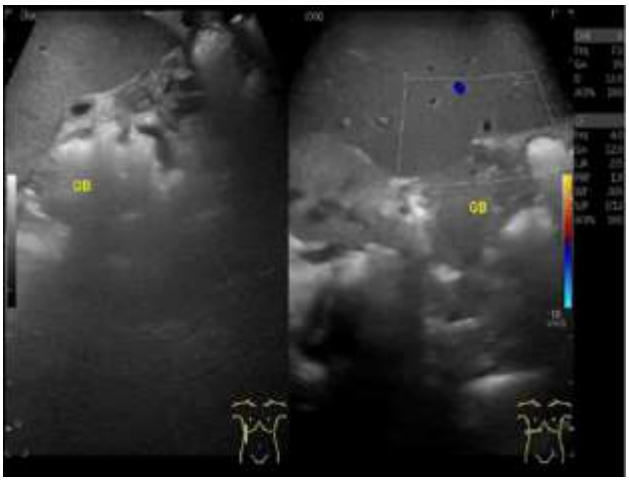

Figure 2: Two-phase abdominal ultrasound shows cholestasis with enlarged postprandial GB (Contractility index-7%),

The assessment of the patient's condition for the case revealed several conditions. Firstly, there was an elevation in the levels of transaminase enzymes. This raised suspicions of extrahepatic cholestasis, with a possible differential diagnosis including biliary stenosis or even BA. Additionally, intra-hepatal cholestasis was considered, with the potential involvement of congenital CMV infection as a differential diagnosis. Lastly, the patient exhibited microcytic hypochromic anemia, which could be attributed to iron deficiency or underlying chronic illness.

The treatment from pediatric department included oxygen therapy within the hospital room to ensure proper oxygenation. Additionally, the patient was provided with breast milk on demand as part of their dietary plan. Intravenous fluids consisting of D5 ¼ NS at a rate of 28 ml per hour were administered during fasting periods, and when intake was resumed, the rate was adjusted to D5 ¼ NS at 4 ml per hour. Furthermore, a combination of medications was prescribed, which consisted of N-acetylcysteine (NAC) at a dosage of 10 mg/kg every 8 hours, totaling 70 mg every 8 hours orally, Aaialys at a dose of 0.6 ml per day orally, vitamin E at 100 IU per day orally, vitamin K at 2.5 mg per day orally, Urdafalk at a dose of 10 mg/kg every 8 hours, equating to 70 mg every 8 hours orally, and Curcuma at a dosage of 3 mg/kg every 8 hours, summing up to 15 mg every 8 hours orally.

After 2 days of treatment, the feces color changed to yellow gradually but the jaundice still remains. Peripheral blood morphology shoes erythrocytes displayed characteristics of hypochromia and microcytosis, along with the presence of teardrop cells, cigar-shaped cells, target cells, and the absence of erythroblasts. In the leukocyte count, there was an elevation in the total count, accompanied by lymphocytosis and the presence of atypical lymphocytes, while blast cells were absent. The thrombocyte analysis identified macrothrombocytes without clumping. Based on these findings, a conclusion was drawn that there is an absolute lymphocytosis suggestive of an ongoing infection process. Furthermore, suspicions were raised regarding the possibility of BA and CMV infection. To further evaluate the patient's condition, it is recommended to conduct tests for C-reactive protein (CRP) and PCR for CMV to gain more insights into the underlying pathology and confirm the suspected diagnoses.

The patient received pre-surgical therapeutic by the pediatric surgery department. This prophylactic treatment consisted of Metronidazole, with an initial loading dose of 15 mg/kg amounting to 100 mg, followed by subsequent doses of 7.5 mg/kg every 8 hours, totaling 50 mg every 8 hours intravenously. In addition to Metronidazole, the patient was also administered inj. ampicillin sulbactam at a dosage of 25 mg/kg every 6 hours, totaling 170 mg every 6 hours intravenously. These therapies were administered over the course of three days prior to the surgical procedure to minimize the risk of infection.

We planned to intra operative colangiography for this patient. The surgical procedure involved the identification and assessment of the biliary tract. It was determined that the biliary tract remained intact. However, congenital adhesions in the form of bands were discovered, which were found to be affixed to the gall bladder, common bile duct, and omentum. To address this issue, a release procedure for the adhesions was carried out. During the procedure, the liver appeared

visibly red, but upon palpation, it exhibited a nodular texture (Figure 3). The patient underwent C-arm examination with iohexol contrast. The patient initially received an ioheksol injection, but its visibility was not apparent before the release of a band. Subsequently, following the band release, the ioheksol injection became visible in the imaging (Figure 4). To further evaluate the hepatic condition, a liver biopsy was performed, with subsequent examination by a pathologist to provide a comprehensive histopathological analysis

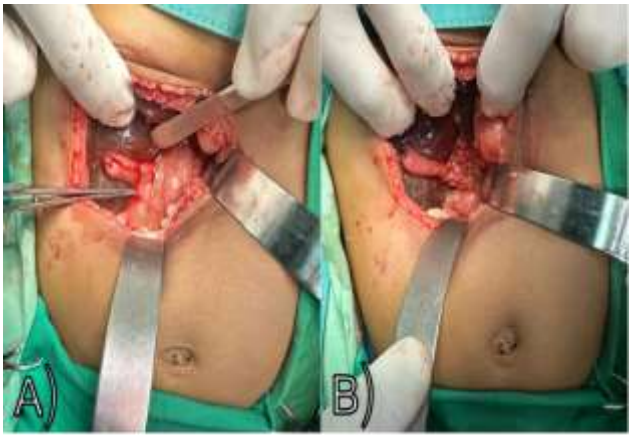

Figure 3 (A and B): Congenital adhesion bands were exposed intraoperatively, post release congenital biliary bands.

Figure 4 (A and B): Contrast iohexol evaluation, iohexol contrast have not fill the tract and iohexal contrast filling the tract.

After 2 hours of surgery procedure, the patient went to pediatric ward and stayed for 3 days (Figure 3). On the second blood test obtained Hemoglobin 9.1, hematocrit 27, leukocytes 23.9, platelets 351.000, erythrocytes 3.51, neutrophils 39.90, lymphocytes 47.90, SGOT 1.165, SGPT 553, PT 13.1, APTT 31.9, and INR 0.960. There were no major and minor side effect of this procedure.

Figure 5: Post-operative condition shows no major and minor side effect.

DISCUSSION

Jaundice is caused by an excess of bilirubin production, a decrease in elimination, or a mix of the two. While the terms "conjugated" and "direct" bilirubin are frequently used interchangeably, direct hyperbilirubinemia includes both conjugated and delta bilirubin, and an increase in either causes jaundice.[6] BA is a progressive fibrotic obstructive condition that affects the biliary system both intrahepatic and extrahepatic. This leads bile flow obstruction and the development of newborn jaundice.[1] BA is an uncommon disorder with an unknown cause and an unfavorable prognosis. For diagnostic, intraoperative cholangiography is the preferred and commonly acknowledged method, which is often performed via laparotomy or minimally invasive access. Endoscopic retrograde cholangiopancreatography (ERCP) is used in some cases, but it requires general anesthesia and has technical obstacles.[5] In this study, the patient was performed intraoperative cholangiography and we found an intact biliary tract but congenital adhesions in the form of bands and nodular hepar texture were discovered.

Exploration is imperative for both diagnosis and treatment of congenital band. Congenital adhesion bands develop as a result of embryologic factors such as the persistence or incomplete regression of the fetal vitelline circulation and the ventral mesentery theory. This could be linked to genetic abnormalities that impair development. Other conditions that contribute to band formation include intrauterine mesothelioma trauma. Congenital adhesion bands may also develop as a result of intrauterine exposure to particular infectious agents or ischemia events.[4] For the nodular hepar texture, we done liver biopsy in this case. Liver biopsy plays a crucial role in guiding diagnostic investigations and therapy for infants experiencing undiagnosed cholestasis. The interpretation of liver biopsies in infantile cholestasis presents a challenge due to the overlapping histologic features of many disorders causing this condition. Moreover, these features are dynamic and vary with age.[7]

CONCLUSION

Biliary atresia is a obstructive condition that leads to newborn jaundice. Congenital adhesion band commonly found in pediatric. Intraoperative cholangiography is an effective method to investigate and diagnose these condition.

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