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Biliary Atresia in Neonate: A Case Report

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Abstract

Biliary atresia is a rare but devastating progressive cholangiopathy and is the leading cause of liver transplantation in children. The key to a better prognosis and the success of the Kasai portoenterostomy (HPE) procedure is early diagnosis and timely surgical intervention ideally before 60 days of life. This case report aims to highlight the diagnostic process and challenges in confirming biliary atresia (BA) in a jaundice neonate. Diagnosing biliary atresia (BA) can be challenging as its early symptoms often overlap with more common benign causes of neonatal jaundice. The definitive diagnosis depends on clinical presentation, laboratory results, imaging studies (specifically the USG findings of an atrophic gallbladder or triangular cord sign) and ultimately an intraoperative cholangiogram. This case emphasizes the critical need for vigilance and a high index of suspicion in any infant with persistent conjugated hyperbilirubinemia.

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Introduction

Biliary atresia (BA) is a rare, life-threatening inflammatory obliterative cholangiopathy that affects full-term infants, characterized by the progressive fibrosis and obstruction of the extrahepatic bile ducts.[1] It represents the most frequent cause of cholestasis in neonates and infants and is the leading indication for pediatric liver transplantation worldwide.[1,2] The incidence of biliary atresia (BA) varies globally estimated at approximately 1 in 10,000 to 1 in 18,000 live births.[1]

The etiology of biliary atresia remains largely unknown although research suggests a complex interplay of genetic predispositions, viral infections and immune-mediated responses may contribute to the development of the disease.[3] The hallmark clinical presentation is persistent neonatal jaundice lasting beyond two weeks of age, coupled with acholic (pale or white) stools and dark urine.[2]

Crucially, the prognosis for patients with coupled with acholic (pale or white) stools biliary atresia (BA) is heavily dependent on the timing of surgical intervention. The standard procedure, the Kasai portoenterostomy (HPE), aims to establish bile flow by connecting a loop of intestine directly to the liver.[4] When performed within the first 60 days of life, the success rate for achieving adequate bile drainage and native liver survival is significantly higher. Delays in diagnosis lead to irreversible liver cirrhosis, necessitating liver transplantation.[4]

Due to the critical need for early intervention and the challenges in differentiating biliary atresia (BA) from other causes of neonatal jaundice, a high index of clinical suspicion and a systematic diagnostic approach are paramount.[5] This case report presents the clinical course and diagnostic workup of a neonate with biliary atresia, emphasizing the utility of

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standard diagnostic tools, particularly abdominal ultrasound findings or specific lab markers, in facilitating a timely diagnosis and optimal management.

Case Summary

A female infant aged 1 month and 20 days was brought to the emergency department of our hospital by her mother with history of convulsion (2-3 times), She had ever for seven days and whitish stools (science birth). The child had a prior history of hospitalization due to bluish discoloration of deferent part of body. On examination during admission, the infant appeared sick and febrile with heart rates of 117 beats per minute (bpm) and respiratory rates of 44 breaths per minute. The abdomen was soft but distended and the infant weighed 5kg. Ultrasonogram report revealed non-visualization of the gallbladder, mild hepatomegaly (left lobe) with increased hepatic echogenicity (Fig. 1). She subsequently underwent a modified Kasai portoenterostomy with Roux-en-Y jejunojejunostomy. Intraoperative findings included an atretic gallbladder, absent hepatic and cystic ducts, and cholestatic changes in the liver. Histopathological examination (HPE) of the liver revealed ballooning degeneration of hepatocytes, intrahepatic cholestasis with bile plugs, focal giant cell transformation, bile ductular proliferation, periportal fibrosis and focal neutrophil infiltration consistent with cholestatic disease and fibrosis. The gallbladder specimen showed markedly attenuated lining with congested vessels, hemorrhage, and fibrosis, suggestive of chronic cholecystitis. Based on those features, a diagnosis of extrahepatic biliary atresia was confirmed. Her medical history was notable for yellowish discoloration of the eyes and clay-colored stools since birth. At 29 days of age, she was admitted with these symptoms. The results of the laboratory investigations are given in the Table-I.

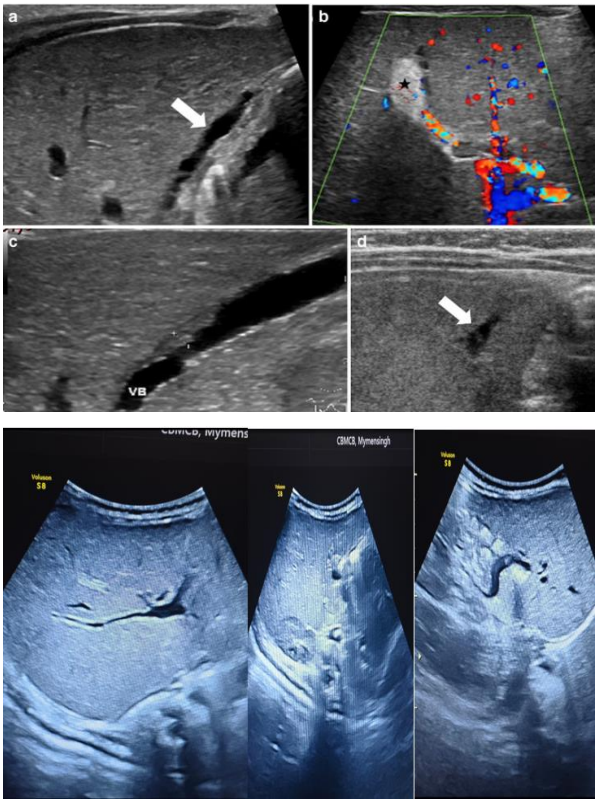

Fig. 1: Ultrasonography findings of Biliary atresia in a neonate.

Table-I: Results of the previous investigations

Investigation	Result	Normal range
Total bilirubin	16.8 mg/dl	Up to 13.3 mg/dl
Direct bilirubin	5.9 mg/dL	<0.2 mg/dL
SGPT	248 U/L	Up to 42 U/L
Serum calcium	9.9 mg/dl	8.1-10.4 mg/dl
Hb%	5.3 g/dl	10-13 g/dl
Random blood sugar	6.3 mmol/L	3.8-7.8 mmol/L
WBC	28500 /cumm	5000-15000 /cumm
Peripheral blood film	Sever normochromic normocytic anaemia with leucocytosis with thrombocytosis	
Platelets	8,50,000/cumm	150000-450000
APTT	Control 32 Second Patient:>120 Second	22.0-38.0
INR	4.50	1.0-1.3
C-reactive protein (CRP)	43.1 mg/dl	<8 mg/dl
D-dimer	<0.19	<0.50

Discussion

Biliary atresia is a serious condition in infants that can progress to cirrhosis, liver failure, and ultimately death if not treated. It is the leading cause of pediatric liver transplantation in developed countries with an incidence of one in 17,000 live births in the United Kingdom, slightly higher in regions such as Japan and China (one in 8,000).[6] There is a slight female predominance, particularly in syndromic forms. biliary atresia affects both intra- and extra-hepatic bile ducts and is marked by cholangiolar plugging, proliferation, fibrosis, inflammatory infiltrates and luminal obliteration in the extrahepatic bile ducts.[6] The cause of biliary atresia (BA) remains unclear but it is believed to be multifactorial with bile duct damage potentially arising from genetic, infectious, inflammatory and/or toxic factors.[7] A history of consistently pale stools may indicate extrahepatic obstruction.[8]

In symptomatic infants with biliary atresia, laboratory results often show elevated direct or conjugated bilirubin, mildly raised aminotransferases and a marked increase in gamma-glutamyl transpeptidase (GGT). Presymptomatic infants may exhibit mildly elevated conjugated bilirubin levels shortly after birth, which demands further monitoring. Early screening methods such as measuring conjugated bilirubin levels or using stool color cards have improved diagnosis and outcomes. The diagnosis of biliary atresia is confirmed through a combination of imaging, laboratory tests and liver biopsy to rule out other causes of cholestasis. Infants must be evaluated promptly as the success of surgical intervention decreases with increasing age at the time of surgery. The gold standard for diagnosing biliary atresia (BA) remains an intraoperative cholangiogram often followed by the Kasai procedure if bile duct obstruction is confirmed.[7-9] Infants with biliary atresia typically present with jaundice, pale stools and dark urine, despite normal meconium passage at birth. Some may have pigmented stools initially, gradually progressing to acholic stools. Without treatment, biliary atresia may lead to end-stage liver disease and death within two years. The purpose of the Kasai procedure is to improve bile flow by eliminating the atretic portal plate and establishing a channel from the bile ductules to the intestines.[2] Liver failure signs such as ascites and hypoalbuminemia are contraindications for KPE leading to referral for liver transplantation. Jaundice clearance is monitored and growth failure could signal the need for liver transplantation.[9] Around 57% of cases attain a resolution of jaundice within six months. Postsurgical treatment includes intravenous antibiotics followed by low-dose oral antibiotics for 8-12 weeks. Ursodeoxycholic acid is administered for at least 12 months to enhance the excretion of bile acids from the liver and reduce their intestinal reabsorption, thereby limiting their recirculation. Fat-soluble vitamin supplements (A, D, E, and K) are essential for preventing malnutrition, managing fat malabsorption and reducing the effects of excessive metabolic breakdown. Medium-chain triglycerides may be provided for patients with steatorrhea. Steroid use has been controversial with studies showing no significant benefit and potential risks and immunomodulatory therapies remain debated as an adjunct to surgery.[1,7,10] Preventing biliary atresia remains challenging. Epidemiological studies suggest a potential association with maternal factors such as intestinal and genitourinary infections, the use of anti-inflammatory asthma medications during the first trimester and substance abuse which may increase the risk of developing the condition. However, there is currently no clinical evidence to confirm that

addressing these risk factors can prevent the onset of biliary atresia in at-risk infants.[1,10] Research using human stem cells and organoids serves as a preclinical platform for disease modeling and drug testing with potential for diagnostic and therapeutic advances. Stem cells could be used to modulate inflammation, immune dysregulation or liver fibrosis possibly as an adjunct to KPE and tissue engineering may eventually replace liver transplantation.[1,8-10] Collaboration between academia and industry could make these emerging treatments successful, potentially turning biliary atresia into a curable condition.

Conclusion

Biliary atresia is the most common cause of cholestatic jaundice in neonates, necessitating surgery. The primary treatment is the Kasai operation, which is a surgical intervention designed to restore bile flow. It is usually done within the first 2-3 months of life, preferably before 60-90 days of age. If the Kasai surgery fails, the sole therapeutic choice for life-saving treatment is liver transplant. Liver transplantation restores liver function, reduces jaundice and improves the overall quality of life. For successful results, prompt referral and surgical intervention are essential. End-stage liver disease can be definitively treated with liver transplantation, giving patients a second chance at life. Patients who receive proper care can live active and healthy lives following transplantation. Vigilant observation is necessary. Recent advances in imaging technology have enhanced the ability to predict biliary atresia in fetus during pregnancy, particularly through high-resolution maternal ultrasound scans. This case serves as a wake-up call to paediatricians to adopt appropriate screening and investigations in suspected cases for early diagnosis and thus, save infants' lives from such potentially fatal illness.

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