International Journal of Contemporary Pediatrics
Radhakrishnan S et al. Int J Contemp Pediatr. 2024 May;11(5):612-615
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20241047

Case Report

Case report on failed Kasai-portoenterostomy for extrahepatic biliary atresia with neonatal cholestasis in one-month-old infant

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Received: 16 March 2024
Revised: 04 April 2024
Accepted: 06 April 2024

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ABSTRACT

Biliary atresia is a neonatal onset, obstructive cholangiopathy of the intrahepatic or extrahepatic biliary system, leading to the build-up of bile in the liver. This case discusses a one-month-old infant who was previously diagnosed with jaundice, presenting the symptoms of yellowish eyes and skin, pale stools, and palpable liver. The infant and her mother's liver enzyme levels were found to be higher and her histopathology studies evidenced atretic gall bladder. The liver biopsy revealed mild periportal ductular reaction and diffuse hepatocanalicular cholestasis. She was diagnosed with neonatal cholestasis- extrahepatic biliary atresia (EHBA) and underwent Kasai-portoenterostomy. She was stable and discharged with gallstone dissolution agents, antibiotics, vitamin supplements, and a barbiturates-liver enzyme inducer. After six months, she presented hepatosplenomegaly with ascites and was found to have transaminitis. She was then diagnosed with EHBA- failed Kasai, probable cholangitis, and planned for living donor liver transplantation. Antibiotics, antiviral, antifungal, anticoagulants, and immunosuppressants were prescribed on discharge. The Doppler study of allograft was performed to check the blood flow after transplantation. Acute graft rejection was monitored on day 5 with liver transplant pack reports. One year later, the infant's condition had shown improvement, evidenced by enhanced food intake, absence of symptoms, and the return of liver function tests to normal levels.

Keywords: Atretic gall bladder, Bile duct, Cholangiogram, Liver transplantation, Doppler study, Cholestasis

INTRODUCTION

Biliary atresia (BA) is a rare, progressive, and obstructive cholangiopathy that is the primary cause of pediatric liver transplantation due to the lack of effective treatments, affecting both extrahepatic and intrahepatic bile ducts and potentially leading to biliary cirrhosis and death if untreated.[1]

Infants with BA are typically at-term, normal-weight, and thriving. Early signs include dark urine, acholic stools, jaundice, high serum bile salts, and conjugated bilirubin, possibly indicating cholestasis.[2] Gamma-glutamyl transferase (GGT) levels and matrix metalloproteinase-7 (MMP-7) are crucial in diagnosing BA.[3] BA is divided into perinatal or non-syndromic cases, accounting for over two-thirds, and embryonic or syndromic cases, more common with multiple anomalies.[4] Genetically predisposed newborns may develop BA due to in-utero insults, triggering immunity, bile duct injury, bile acid buildup, and epithelial destruction, and a viral trigger is a plausible etiological factor.[5,6]

Abdominal ultrasound can detect gallbladder visualization and liver hilum cysts, but magnetic resonance imaging (MRI)-cholangiography is insufficient for infants under 3 months. Endoscopic retrograde cholangiopancreatography (ERCP) and liver biopsy may be helpful.[1] BA is primarily

treated with Kasai portoenterostomy (KPE) at diagnosis but is the most common cause of pediatric liver transplantation due to hepatic damage. Isolated transplantation without KPE may be effective for delayed diagnosis. Post-KPE, medical care includes supplementation with fat-soluble vitamins, nutrition, and preventing complications. N-acetylcysteine, an antioxidant, improves hepatic damage, fibrosis, lowers biliary obstruction, and increases survival.[7]

CASE REPORT

A one-month-old female infant with a history of jaundice was admitted to the hospital with chief complaints of abdominal distension, passing pale stools, yellowish eyes, and body since 2 weeks of age. She was afebrile, gaining adequate weight, and hemodynamically stable. General examination revealed a palpable left lobe of the liver up to 4 cm. Her family history indicates that the infant's mother had type 2 diabetes mellitus and experienced pregnancy-related-transaminitis. The patient's vitals were normal. She was provisionally diagnosed with Infantile cholestasis. Laboratory investigations from Table 1 revealed transaminitis, and increased gamma-glutamyl transpeptidase and bilirubin levels.

Histopathological findings revealed an atretic gallbladder with extrahepatic bile duct, complete ulceration of mucosa of the bile duct with a thickened and fibrotic wall containing lymphoid cell infiltration. Cystic lymph node identified and shows reactive lymphoid hyperplasia. Stage-2 fibrosis was observed with moderate portal expansion. A liver biopsy revealed mild periportal ductular reaction and diffuse hepatocanalicular cholestasis leading to difficulty in bile flow from the liver to the duodenum and also showed chronic hepatitis with moderate lymphoid cell infiltration. Urine cytomegalovirus test showed negative.

Based on the subjective and objective evidence, the patient was diagnosed with neonatal cholestasis - EHBA. The patient was planned to undergo the surgical procedure of KEP with an intraoperative cholangiogram. Intraoperative cholangiogram is a procedure where a small tube is placed into the cystic duct which drains the bile from the gallbladder and injection of dye to improve visualization of the bile duct anatomy. On day 2, during the kasai procedure, the damaged bile ducts were removed and replaced by the infant's loop of small intestine which was uneventful. The findings of the surgical procedure were non-visualization of inter extrahepatic biliary system, atretic gall bladder with white bile excretion, minimal serous ascites present with <100 ml, and normal liver and visceral regions. Postoperatively, she was initiated with oral feeds after 48 hours and opened her bowel which was greenish. Feeding was continued with the mother's milk along with top-up paladai feeds. Hemodynamics were stable and the patient was discharged. She was instructed to continue direct breastfeeding (DBF) and top up paladai feeds and review after one week with liver function test (LFT) and INR reports.

Table 1: Laboratory investigations (on the day of admission).

Parameters	Observed value	Normal value
Clinical hematology		
Haemoglobin	10.7	12-16 g/dl
WBC	11,320	4500-11000/mm^3
RDW-CV	17.2	12-15%
Neutrophils	16.7	40-60%
Liver function tests		
AST/SGOT	226	8-40 U/l
ALT/SGPT	142	8-40 U/l
ALP	369	90-180 U/l
T. Bilirubin	8.3	0.1-1.0 mg/dl
D. Bilirubin	7.56	0.0-0.3 mg/dl
GGT	293	<20 U/l
Albumin	2.7	3.5-5.5 g/dl
Biochemistry		
Potassium	5.73	3.5-5.0 mmol/l

After 6 months, the infant came to the hospital with complaints of low-grade intermittent fever, poor oral intake, abdominal distension, and breathlessness for the past one day. Physical examination showed hepatosplenomegaly and minimal ascites. She was diagnosed as EHBA with failed kasai, and probable cholangitis, and was planned for living donor liver transplantation (LDLT) - left lateral segment. Her father volunteered to donate. Pre-operatively she was prescribed antibiotics and continued for 7 days after surgery. The donor's arteries and veins were anastomosed with the recipients. She was sedated using fentanyl infusion and serum lactate levels were normalized. Doppler study of allograft was performed to detect the blood flow which showed normal. She was started with oral feeds on day 2 of surgery, developed chylous ascites on day 5, and was advised to have a fat free diet which improved the volume and color of the drain. So, she was instructed to continue for 3 weeks. Immunosuppressants and Anticoagulants were prescribed to prevent rejection of graft and clot formation respectively. Suitable antimicrobials were prescribed as per the protocol. Liver enzymes were nearly normal with ultrasonography (USG) abdomen, hepatic Doppler and plasma drug level being normal. She was instructed to review after 5 days in the outpatient department with post liver transplant pack reports. By the fifth day, the infant displayed acceptance of the graft without acute rejection, as evidenced by the reports and absence of symptoms. On one-year follow-up, the child showed improvement in oral intake and the LFT were seemed to be normal.

Table 2: Medications on the course of hospitalization.

Brand name	Generic name	Dose (mg)	Route	Frequency	Duration
Inj. Piptaz	Piperacillin+Tazobactam	300	IV	Q8h	Day 1
Inj. Meronem	Meropenem	60	IV	Q8h	Day 2-5
Inj. Dolo	Paracetamol	30	IV	Q8h	Day 2-4
Syr.Taxim-O	Cefixime	50 mg/ml	PO	1.5-0-1.5 ml	Day 6-8

Table 3: Drugs on discharge.

Brand name	Generic name	Dose (mg)	Route	Frequency	Duration
Syr.Taxim-O	Cefixime	5 ml/50 mg	PO	1.5-0-1.5 ml	2 weeks
Syr. Domstal	Domperidone	1	PO	1-1-1 ml	To continue
Zincovit Drops	Multivitamin and minerals	15 ml	PO	1 ml-0-0	3 days
Syr. Udcament	Ursodeoxycholic acid	125	PO	1-0-1 ml	3 days
Cap. Aquasol A	Vitamin-A	25000 IU	PO	½ Once a week on same days	To continue
Syr. Ossapan D	Calcium and vitamin D3	500	PO	2.5-0-2.5 ml	3 days
Cap. Evion	Vitamin-E	400	PO	Once a week on same days	To continue
Tab. Kenadion	Vitamin-K	10	PO	¼ Twice a week on same days	To continue
Syr. Gardenal	Phenobarbitone	15	PO	0-0-5 ml	3 days

DISCUSSION

Although uncommon, BA is one of the most common forms of hepatic disease in neonates and the primary rationale for liver transplantation in this age category. Untreated BA can cause fatal complications like hepatic synthetic failure or portal hypertension within the first two years.[8]

Untreated BA has a very bad prognosis. The KEP technique can restore bile flow in most newborns who undergo it, although it is rarely curative. Studies show that bile flow can be restored in 80 to 90% of newborns referred for surgery within 60 days of birth. However, children 90 days or older at the time of surgery have a success probability of less than 20%.[9] In our case, even though the patient was only 1 month old, the surgery did not cure the condition which resulted in chronic liver disease and the patient was admitted again to the hospital due to complaints of unclear jaundice.

When portoenterostomy fails or liver function gradually deteriorates following successful bile flow establishment, liver transplantation is used as a salvage procedure.[10] In our case, the patient had undergone successive liver transplantation procedure in which her father came forward as a liver donor.

CONCLUSION

Liver transplantation is critical for managing EHBA and should be considered in patients whose surgery fails to restore bile flow, who are referred late (typically at 60 days of age or later), and who eventually develop end-stage liver disease despite bile drainage. Diagnosing BA early and performing a KEP at the age of 2 months is essential for a good prognosis. Individuals with chronic acholic stool and high GGT should be assessed for BA. Recent imaging advancements have improved antenatal prediction of EHBA, notably on high-resolution maternal ultrasonography.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Radhakrishnan S, Velmani V, Mavis MS, John Felix SN, Mohamed Sulaiman G. Case report on failed Kasai-portoenterostomy for extrahepatic biliary atresia with neonatal cholestasis in one-month-old infant. Int J Contemp Pediatr 2024;11:612-5.