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Case Report on Failed Kasai Hepatico Portoenterostomy with Biliary Atresia in 3 years old child

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ABSTRACT

Biliary atresia is a rare liver disease in children where the bile ducts are blocked or not present in the liver. This case explains a 3 years old child who was previously diagnosed with Biliary Atresia. The Child admitted electively for cholangiogram and asses the possibility of interventional radiology guided Percutaneous transhepatic biliary Drainage insertion for biliary strictures. The child liver enzymes are increased and patient histopathology studies Indicates biliary atresia. The Child was diagnosed with Biliary atresia. Doppler study showed decreased peribiliary echogenicity, short segments of Centre prominent intrahepatic biliary radicals with intervening on dilated duct noted presented intrahepatic strictures. MRCP showed multifocal intrahepatic strictures, cystic dilation central part segment 3 intrahepatic biliary radicals positive. The patient underwent cholangiogram. Medications Used in the Course of Hospitalization are Prograf 1mg and 0.5mg, Mycophenolate Mofetil 250-125mg, Omnacortil 2.5mg and Sirolimus 1mg. Child was hemodynamically stable, vitals are normal, accepting feed well and discharge. Discharge medications are Sirolimus 1mg, Prograf 0.5mg, Ciplox 125mg, Omnacortil 2.5mg, Udcament 2.5ml, Cellcept 250mg.

Keywords: Biliary Atresia, Bile duct, MRCP, Cholangiogram, Doppler study, Discharge drugs.

1. INTRODUCTION

Biliary Atresia is a rare but serious congenital liver disease of newborns, in which bile ducts carrying bile from liver to the intestine are not present, blocked, or abnormally developed. It causes bile to accumulate in liver, cause liver injury, scarring(fibrosis), and ultimately cirrhosis. It usually presents between the first two to six weeks of age with ongoing jaundice, clay-colored stools, dark-colored urine, and hepatomegaly. The etiology is not specifically known, but various

hypotheses include a variety of genetic defects, abnormal in utero development, immune-mediated damage, or viral infections like cytomegalovirus or reovirus. Biliary atresia is further divided into syndromic and non-syndromic types, with syndromic types presenting with other birth anomalies like polysplenia or heart defects. The most prevalent type is type III, which is complete obstruction at the porta hepatis. Diagnosis involves a combination of blood work (indicating elevated direct bilirubin and liver enzymes), imaging tests (such as ultrasound and hepatobiliary scintigraphy), and a confirmatory biopsy of the liver, which usually reveals proliferation of bile ducts and bile plugging. Kasai portoenterostomy is the initial surgical intervention and is optimal if done prior to 60 days of life, since it ensures bile drainage and postpones liver damage. But even with early intervention, most infants still go on to develop chronic liver disease and ultimately require a liver transplantation, which is still the ultimate therapy. Early detection and prompt intervention are vital for enhancing survival and long-term prognosis in afflicted infants.

2. CASE REPORT:

A 3 years old patient was electively admitted for cholangiogram and assess the feasibility of IR guided PTBD insertion for biliary strictures. BA FK transplant in 2022. Child as antibody mediated rejection with biliary dilation. He received for it but the bilirubin didn't settle (initially received bortezomib in 2023). He was initiated on sirolimus on april 2024 following which the bilirubin began to fall slowly. It has peaked at 9 decreased gradually to a minimum of 1.3 as on june 2024 showed some enzyme fluctuation even on sirolimus on prograf 1 mg and 0.5 mg, MMF 250-125, Omnacortil 2.5 mg and sirolimus 1 mg DOPPLER 2025 – scant perbiliary echogenicity, short length of mildly prominent intrahepatic strictures MRCP- multifocal intrahepatic strictures (central greater than peripheral).

Cystic dilation central portion segment 3 intrahepatic biliary radical+. Cholangiogram, IR guided balloon dilatation of strictures and PTBD insertion (internal and external may 2025. Under SAP/GA/USG/FLURO,LHD punctured. Cholangiogram done revealed communicating ductal system with cystic dilation in one of the segmental ducts. Finding of anastomotic site strictures. Serial dilatation of strictures performed BF internal and external tube is left in place. Procedure performed uneventfully. Post operative child was commenced on prophylactic antibiotic. PTBD tube kept open for 24 hours with drained 128ml and then closed. Repeat LFT after 24 hours shown 3.13/2.53/163/190. PTBD insitu. Feeds commenced on the same day which he accepted well. Child hemodynamically stable, accepting feeds well and planned to monitor on follow up. Vitals stable

Lab parameters:

The tacrolimus (11.6 ng/ml) and sirolimus (8.51 ng/ml) levels of the patient are within therapeutic levels. Laboratory tests for liver function reveal a high total bilirubin (4.15 mg/dl) and direct bilirubin (3.19 mg/dl), reflecting cholestasis. ALP (1500 U/L), AST (219 U/L) and GGT (184 U/L) are markedly increased, indicating liver damage. Total protein (8.1 g/dl) and globulin (4.3 g/dl) are slightly increased, possibly from inflammation. In general, findings suggest liver dysfunction with a cholestatic profile.

Medications Used in the Course of Hospitalization:

The patient received tacrolimus 1 mg intravenous twice a day (BD) for 7 days, in addition to mycophenolate mofetil 250 mg IV BD for 7 days. Prednisolone 2.5 mg orally once daily (OD) and sirolimus 1 mg orally OD were also given for a total of 7 days each. Such a regimen is an example of simultaneous administration of immunosuppressive agents both intravenously and orally

Drugs on discharge:

The patient was treated with a 7-day oral regimen consisting of sirolimus 1 mg OD and tacrolimus 1 mg OD as immunosuppressants. Mycophenolate mofetil 250 mg BD was administered for immunosuppression. Prednisolone 2.5 mg OD was added as a corticosteroid. Prophylaxis for infection was provided using ciprofloxacin 250 mg BD. Ursodeoxycholic acid 2.5 ml BD was also prescribed to aid bile flow and liver function.

3. DISCUSSION:

This is the patient of a 3-year-old with a history of biliary atresia and liver transplant in 2022, presenting with antibody-mediated rejection and chronic biliary strictures. Following treatment with bortezomib in 2023, bilirubin was persistently elevated. Introducing sirolimus in April 2024 led to progressive decrease of bilirubin but with variable liver enzymes. Multifocal intrahepatic strictures with central segment cystic dilation were observed on imaging. In May 2025, cholangiogram, IR-guided balloon dilation, and PTBD placement were successfully achieved in the child. After the procedure, the child tolerated feeds and was hemodynamically stable. Immunosuppression was with tacrolimus, sirolimus, MMF, and prednisolone. The child is clinically stable with raised LFTs and cholestatic markers on close follow-up. This case describes the complexity of managing post-transplant biliary complications and the importance of multimodal management.

4. CONCLUSION:

This case points to the complicated post-transplant course of a 3-year-old child with biliary atresia and antibody-mediated rejection. After initial treatment resistance, the addition of sirolimus aided in the gradual decrease in bilirubin levels. Imaging showed multifocal intrahepatic strictures and cystic dilation, for which IR-guided balloon dilation and PTBD placement was necessary. Following the procedure, the child remained hemodynamically stable and tolerated feeds well. An immunosuppressive regimen was continued in its entirety. The case highlights the need for early intervention, imaging-guided interventions, and individualized immunosuppressive treatment in the management of post-transplant biliary complications

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