Case Report

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Obstructive Jaundice In A Neonate Secondary To Biliary Atresia: A Case Report

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Abstract

A 3-day-old male neonate presented with obstructive jaundice; a critical condition often indicative of severe underlying hepatobiliary dysfunction. Subsequent imaging and diagnostic evaluations confirmed biliary atresia, a rare congenital disorder characterized by obliteration or discontinuity of the extrahepatic biliary system, leading to bile flow obstruction. This case report details the clinical presentation, diagnostic approach, management strategies, and the short-term outcomes observed. This case provides a comparative analysis with similar cases reported in the literature, highlighting the importance of early diagnosis and the implications of surgical intervention in the prognosis of biliary atresia. Through this case, we aim to augment the existing knowledge on neonatal jaundice, focusing particularly on its etiological spectrum and management outcomes.

Keywords:- Biliary Atresia, Neonatal Jaundice, Kasai Portoenterostomy, Obstructive Jaundice in Neonates.

INTRODUCTION

Biliary atresia is a critical pediatric liver disease marked by the progressive obliteration of bile ducts, culminating in obstructive jaundice and liver failure if left untreated.[1] This condition, predominantly affecting neonates, is the leading indication for pediatric liver transplantation globally. The pathophysiology involves an inflammatory process that leads to fibrosis and obliteration of the bile ducts, although the exact etiological factors remain under investigation. [2]

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Epidemiologically, biliary atresia affects approximately 1 in every 10,000 to 15,000 live births in the United States, with variable incidence worldwide.[3] It presents typically within the first two weeks of life with persistent jaundice, acholic stools, and hepatomegaly. Given its severity and progression, understanding its clinical presentation and diagnostic markers is crucial for timely intervention.[4]

Clinically, infants with biliary atresia typically present with jaundice that persists beyond the first two weeks of life, dark urine, and pale stools. The diagnostic approach includes a combination of ultrasonography, which may show an absent or abnormal gallbladder, and a definitive diagnosis usually requires a cholangiogram or liver biopsy.[5] Notably, in this case, the diagnosis was supported by characteristic imaging findings suggestive of biliary atresia.

CASE REPORT

The patient, a 3-day-old male neonate, presented with jaundice noticed shortly after birth. He was born full-term via uncomplicated vaginal delivery with no significant prenatal or perinatal issues reported. Initial physical examination revealed jaundice and a slightly distended abdomen but no other anomalies.

Diagnostic evaluation began with liver function tests which indicated elevated direct bilirubin and alkaline phosphatase levels. An abdominal ultrasound was promptly performed, showing an absent gallbladder and abnormal bile ducts, suggestive of biliary atresia. This was further confirmed through a diagnostic cholangiogram. Given the absence of relevant lab investigations showing significant anomalies besides those typically associated with obstructive jaundice, a detailed table of lab results is omitted.

Management was promptly initiated with an exploratory laparotomy and a Kasai procedure (portoenterostomy), aiming to restore bile flow to the intestine. Postoperatively, the neonate showed initial improvement in bilirubin levels and general condition. However, the long-term outlook remains cautious, with potential considerations for liver transplantation as the disease progresses.

Test	Result	Reference Range	Remarks
Total Bilirubin	12 mg/dL	0.1 - 1.2 mg/dL	Significantly elevated
Direct Bilirubin	10 mg/dL	0 - 0.3 mg/dL	Indicative of obstructive jaundice
Alanine Transaminase (ALT)	70 U/L	5 - 40 U/L	Mildly elevated
Aspartate Transaminase (AST)	80 U/L	5 - 40 U/L	Mildly elevated
Alkaline Phosphatase	450 U/L	40 - 129 U/L	Significantly elevated
Gamma-Glutamyl Transferase (GGT)	250 U/L	5 - 55 U/L	Significantly elevated
Serum Albumin	3.5 g/dL	3.5 - 5.0 g/dL	Normal
Platelet Count	300,000 /µL	150,000 - 450,000 /µL	Normal

Table 1: Laboratory Investigations of the Neonate with Biliary Atresia

DISCUSSION

The present case of obstructive jaundice in a neonate secondary to biliary atresia offers a critical insight into a rare but serious congenital condition affecting the biliary system. Biliary atresia is characterized by progressive fibrosis and obliteration of both intrahepatic and extrahepatic bile ducts, leading to bile stasis and liver damage. Early diagnosis and surgical intervention are crucial to improve outcomes and prevent irreversible liver damage and eventual liver failure.[6]

This case underscores the complexity of diagnosing biliary atresia, especially within the first few days of life. The initial presentation of jaundice in neonates is often misattributed to more benign causes such as physiological jaundice of the newborn, which typically resolves without intervention. However, persistent jaundice after two weeks of age strongly suggests an obstructive or pathological cause, necessitating further investigation.[7]

In cases where an early diagnosis through the use of abdominal ultrasonography and a liver biopsy confirmed biliary atresia, leading to timely surgical intervention. This parallels the current case in the utilization of imaging and histopathology for diagnosis but differs slightly in the timing and specifics of the interventions provided.[8]

Additionally, literature emphasizes the role of the Kasai portoenterostomy, performed in our case. Studies suggest that the success of this procedure often depends on the age at surgery, with outcomes being significantly better if the surgery is performed before the neonate reaches 60 days of age. The survival rate with native liver after Kasai procedure decreases substantially with delayed diagnosis and surgery. Our case supports this finding, as early surgical intervention led to initial improvement in clinical symptoms. [9]

Moreover, the discussion on biliary atresia is incomplete without acknowledging the potential for complications and the need for long-term management. Postoperative cholangitis and progressive liver fibrosis are common complications that can affect the long-term survival and quality of life, even after a successful Kasai procedure. In the current case, close monitoring for signs of infection, adequate growth, and liver function was instituted, reflecting standard postoperative care recommendations found in the literature.

Comparatively, the broader implications of this case alongside similar cases highlight the necessity for improved diagnostic techniques and perhaps more critically, the need for heightened awareness among clinicians to suspect and investigate biliary atresia in any infant presenting with prolonged jaundice. This is supported by a consensus from multiple studies, including those by Sokol RJ et al. which advocate for routine screening protocols for cholestasis in neonates presenting with jaundice persisting beyond two weeks.[10]

In conclusion, the current case, supported by similar instances in the literature, highlights the importance of prompt, accurate diagnosis and timely surgical management in improving outcomes for neonates with biliary atresia. It also calls attention to the ongoing need for research into more effective management and potentially curative treatments for this devastating disease.

CONCLUSION

This case of obstructive jaundice secondary to biliary atresia in a neonate underscores the imperative for early recognition and intervention. With prompt diagnostic workup and surgical management, there is potential to significantly improve outcomes, although the prognosis remains guarded due to the progressive nature of the disease.

Conflict of interest

None

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