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Case Report

Case Report: A Rare of Late Onset Biliary Atresia! A Clinical and Pathological Challenge!

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Abstract

Biliary atresia classically presents at neonatal age, late presentation in infancy is very rare. We discuss the challenges in clinical diagnosis and henceforth its management, unusual differential diagnosis in infants with cholestatic syndrome, who may benefit from Kasai surgery with timely referrals. Here we highlight the importance of considering biliary atresia as one of rare differentials in infants with cholestasis syndromes. We present late-onset biliary atresia in a 19 weeks old baby presenting with jaundice.

This article highlights late-onset case BA, showcasing importance of BA as differential diagnosis not just in neonates, but also in infants with cholestatic syndrome. Babies should be evaluated by physicians be it a GP, pediatrician or a gastroenterologist based on clinical, laboratory, histopathological, and imaging criteria. BA could be challenging to diagnose but a timely referral and surgical intervention can improve prognosis and survival.

Keywords: *Biliary Atresia; Jaundice; Cholestasis Syndromes*

Background

Biliary atresia classically presents at neonatal age, late presentation in infancy is very rare.

We discuss the challenges in clinical diagnosis and henceforth its management, unusual differential diagnosis in infants with cholestatic syndrome, who may benefit from Kasai surgery with timely referrals. Here we highlight the importance of considering biliary atresia as one of rare differentials in infants with cholestasis syndromes.

We present late-onset biliary atresia in a 19 weeks old baby presenting with jaundice.

Case Presentation

Our patient a 19 weeks old male baby, with a history of jaundice, choluria, and fecal hypocholia, it begun at 16 weeks of age. He was a full-term 39 weaker baby, weighing 3.5 kg, and had no history of jaundice or any comorbids during neonatal period. Biochemical tests were consistent with cholestatic syndrome (baseline biochemical tests of all four cases are shown in the table to follow. Serology of cytomegalovirus was negative. Abdominal ultrasound at the hospital showed an enlargement of liver, with heterogeneous parenchyma,

distension of gallbladder, and dilatation of the intrahepatic and extra hepatic ducts both. The diagnosis was inconclusive and had an open laparotomy. We observed a thin distended gallbladder, intestinal loops adherence going up to the hilum of the liver, and a fibrotic, extra hepatic bile duct system. Cholangiography showed that there was failure to fill right at the junction of the hepatic ducts and a total lack of drainage to the small intestine. We opted for standard Kasai portoenterostomy was our treatment of choice. The infant showed drastic improvement of clinical symptoms and moving towards normalization of bilirubin levels right after a week of surgery. Baby was asymptomatic till after 3 months of follow-up. He is to keep follow up with his transfer city doctor.

BT mg/dl	BD mg/dl	AP U/L	GGT U/L
10.1	8.2	3.1	1.8

Table: *Biochemical analysis.*

Liver biopsy results: Showed a typical morphological pattern of large bile duct obstruction, with severe ductal, intracanalicular, and hepatocytic cholestasis, and cholestasis. Nodular regeneration was also observed along with gallbladder of significant luminal dilatation.

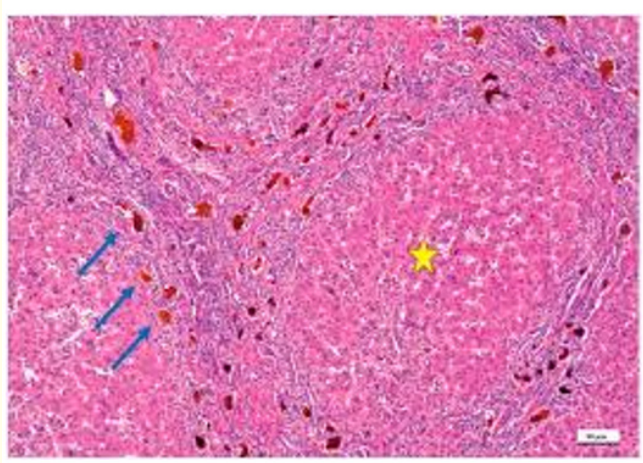

Figure: *Corran's histology.*

Discussion

Kasai portoenterostomy is standard treatment of choice followed by liver transplantation later in life, it can effectively change the prognosis of BA. Diagnosis is based on objective clinical evaluation, histopathological, laboratory, and imaging.

Studies have shown the younger the patient, better is the prognosis. BA has a higher incidence in neonates. Early onset forms with a shorter duration of disease are likely to have a favorable outcome after the Kasai procedure, even when the patient is older. Nevertheless, there are also case reports with a late diagnosis where the patient benefitted from the Kasai operation.

Our patient showed biliary dilatation at 19 weeks which is a very rare late presentation and hence puzzles the age limit for onset of it [1].

Classification of BA is broadly classified as two clinical presentations. One presentation is embryonic BA, characterized by persistent jaundice at birth, associated with non-hepatic congenital structural alterations, also called the early, fetal, or syndromic form [3,4]. The other presentation is perinatal BA, with a jaundice-free interval at birth, is present alone, and is also called the late or non-syndromic.

Conclusion

Hence conclude that late-onset case BA, showcasing importance of BA as differential diagnosis not just in neonates, but also in infants with cholestatic syndrome. Babies should be evaluated by physicians be it a GP, pediatrician or a gastroenterologist based on clinical, laboratory, histopathological, and imaging criteria. BA could be challenging to diagnose but a timely referral and surgical intervention can improve prognosis and survival [2,5].

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