

Cystic Dilatation of the Intrahepatic Biliary System Following Kasai Portoenterostomy

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Received: June 12, 2018; Published: July 25, 2018

Abstract

Cystic dilatation of the intrahepatic biliary system (CDIB) developing after Kasai's portoenterostomy (KPE) for biliary atresia (BA) has been described as an indicator of poor prognosis. We present an asymptomatic, jaundice free child with CDIB 11 years after KPE, who was treated conservatively. A review of available literature is also included.

Keywords: Biliary Atresia; Cystic Dilatation; Intrahepatic Biliary System; Cholangitis

Introduction

Cystic dilatation of intrahepatic biliary tree (CDIB) is an intractable complication of Kasai portoenterostomy (KPE) done in cases of extrahepatic biliary atresia (EHBA). There is an abundance of literature highlighting CDIB as a poor prognostic indicator. We present here the first case of CDIB post KPE, with longest survival follow up reported from India.

Case Report

A 60-day-old boy presented with cholestasis, which on work up turned out to be extra-hepatic biliary atresia (EHBA). He underwent KPE for EHBA. He had good bile flow with resolution of jaundice in the early postoperative period. He was kept on ursodeoxycholic acid as choleric therapy, steroids and antibiotic prophylaxis was given to prevent postoperative cholangitis. The first episode of cholangitis occurred 2 months after surgery and patient developed multiple liver abscesses and was treated conservatively with a course of intravenous antibiotics and supportive therapy. Again after 1-year post-KPE, he developed cholangitis with multiple liver abscesses. MRI revealed two non-communicating cysts (Tsuchida Type A) in the left lobe of liver anterior to the porta (Figure 1). Clinical improvement occurred with a prolonged course of intravenous antibiotics; however, CDIB was persistent.

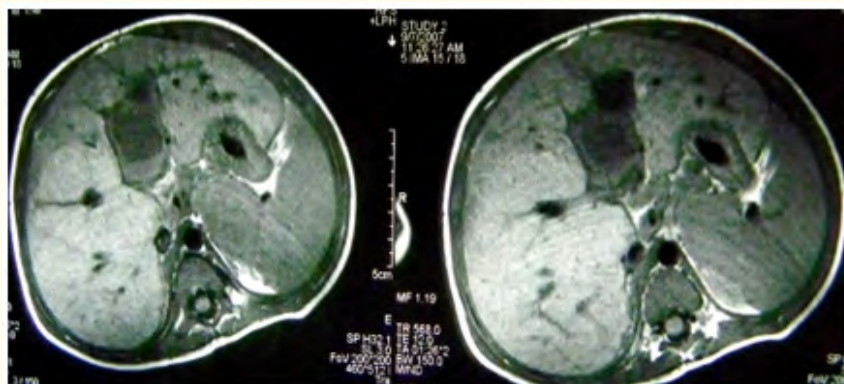


Figure 1: MRI showing two non-communicating biliary cysts.

The cyst aspirate did not grow any organism and was also negative for bile salts. HIDA scan showed normal bile flow and he was jaundice free and asymptomatic. He was closely followed with serial ultrasonography in which the CDIB was persistent for 4 years and spontaneously resolved thereafter. He continues to be asymptomatic and has been going to school. He is under close surveillance and follow up with a pediatric hepatologist.

Discussion

The development of CDIB is a known complication of KPE with a reported incidence of 8 - 25% in different reported series [1-6]. With such significant incidence of CDIB, the patients who develop recurrent cholangitis following KPE, should be examined by imaging modalities to exclude the presence of intrahepatic biliary cysts. Majority of the patients having CDIB would have jaundice (81.3%) and history of cholangitis (93.8%) before the cyst formation [3].

CDIB are usually classified in three types [7]:

- Type A: Non-communicating biliary cysts; also called bile lakes.
- Type B: Single communicating cysts (communication with intestinal loop).
- Type C: Multiple non- communicating or communicating cysts.

Watanabe, *et al.* [1] have described a simpler morphological classification based on their number, i.e. solitary or multiple (≥ 2). The cysts have also been classified on the basis of shape in literature, viz. round/oval and beaded/rosary; the shape of cysts have been linked to prognosis, with round/oval cysts having a better prognosis, than beaded dilatations, with prognosis being defined by the rate of development of recurrent cholangitis, 28% in round and 69% in beaded dilatations [8].

The exact etiopathogenesis of CDIB is unknown but cholangitis has a definite association with this entity [5,7,9]. The recurrent cholangitis is mainly due to ascending infection of biliary ducts from intestinal conduit [1,10] and is associated with worse prognosis [3,11]. It has been suggested that fibrous change in the intralobular spaces and inflammatory process around the bile ducts might be potential causes of cyst formation [7]. In fact, the liver fibrosis has been postulated as a factor helpful in predicting the development of CDIB after KPE [12]. Ductal plate malformation has been implicated for cyst formation, similar to that seen in patients with Caroli's disease and congenital hepatic fibrosis [13]. Ulu., *et al.* reported the occurrence of bile lakes in patients with BA who were not undergoing a portoenterostomy [14].

Bile lakes (Type A CDIB) in patients who underwent liver transplant (LT) have been known histopathologically to have fibrotic cyst wall and lacked epithelia [2]. It was speculated that the bile lakes arise from damaged bile ducts (due to repeated cholangitis), which fuse together after calculi formation inside them (secondary to bile stasis) [2].

The treatment in patients with post-KPE CDIB needs to be individualized. The various treatment modalities can be summarized as follows:

1. Solitary, asymptomatic cyst- Monitoring
2. Solitary cyst with cholangitis- Intravenous antibiotics
3. Solitary cyst with persistent cholangitis- PTBD [3,12,15]
4. Beaded dilatation of intrahepatic ducts- Enteric drainage
5. Multiple intrahepatic cysts with persistent cholangitis refractory to above mentioned modalities - LT [1,3].

CDIB likely changes in size according to development or subsidence of cholangitis, and may disappear after cholangitis subsides in some cases [4,5]. Our patient was monitored radiologically and clinically in follow up and did not require any invasive/surgical intervention.

Nakama, *et al.* [5] have cited PTBD and alcohol injections as treatment for solitary CDIB and re-do KPE for cysts at porta hepatitis. Kawarasaki, *et al.* have highlighted PTBD for solitary CDIB and reanastomosis as favored modality for multi-cystic CDIB [16].

Anti-reflux valve (ARV) surgery known to prevent recurrent cholangitis in patients of EHBA undergoing KPE; however, it was found to be not useful in case of cholangitis associated with CDIB [4].

No recurrence of biliary cyst has been reported after liver transplantation in patients with BA undergoing Kasai portoenterostomy [17]. This may be quoted that some local hepatic issues plays role and make LT as a definitive treatment.

Conclusion

Recurrent cholangitis following KPE should be evaluated for development of CDIB. Asymptomatic CDIB is best managed conservatively as was done in this case, but need to be monitored regularly.

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Volume 7 Issue 8 August 2018

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