Case Report

Iran J Pediatr
Sep 2009; Vol 19 (No 3), Pp:303-306

Association of Biliary Atresia with Jejuno-Ileal Atresia

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Received: *Sep 19, 2008*; Final Revision: *Dec 21, 2008*; Accepted: *Feb 08, 2009*

Abstract

Background: Jejuno-ileal atresia (JIA) is a congenital anomaly characterized clinically by bilious vomiting and abdominal distension. The incidence of JIA is between 1: 330 to 1: 3000 live births in different parts of the world. It has been associated with various congenital anomalies but the association of JIA with biliary atresia is extremely rare (0-3.2 %).

Case Presentation: We herein present a case of jejunal atresia with meconium peritonitis associated with biliary atresia. The patient was a boy who was born at 39 weeks of gestation with polyhydramnious detected on Prenatal Ultra Sonography done at 8th month of gestation.

Conclusion: It is important to explore gallbladder in cases of JIA especially when associated with meconium peritonitis.

Iranian Journal of Pediatrics, Volume 19 (Number 3), September2009, Pages: 303-306

Key Words: Biliary atresia; Jejuno-ileal atresia; Surgery; Meconium peritonitis; Cholangitis

Introduction

The incidence of Jejuno-ileal atresia (JIA) is between 1:330 to 1:3000 live births in different parts of the world[1]. About 50% of cases of intestinal atresia distal the stomach involve duodenum, while JIA comprises the other half. On the other hand, the incidence of biliary atresia (BA) is about 1:10000 to 1:16700 live births[2]. The cause of BA is unknown, although factors such as developmental malformation[3], perinatal viremia[4], toxicity of bile constituents[5], and anatomic abnormality have been implicated[6]. The association of JA with BA is extremely rare (0-3.2 %). We herein present a case of jejunal atresia with meconium peritonitis associated with BA.

Case Presentation

A boy of 39 weeks gestation was born by cesarean section. Prenatal sonography at 8th

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month of gestation showed polyhydramnios. His birth weight was 3850 gram and the baby had Apgar scores of 10 at 1 and 5 minutes after birth. Abdomen was distended. Bilious vomiting in the first day of life. Plain abdominal x-ray showed distended loops of proximal bowel, which indicated the presence of high intestinal atresia. Abdominal sonography was normal except for distended bowel loops.

He was operated on in the second day of life. There was cystic meconium peritonitis with jejunal atresia. Atresia was 30cm off from ligament of Trietz. After removing a part of proximal jejunum, end to back jejuno jejunostomy was performed. Total parentral nutrition (TPN) was started the day after operation. On the fifth post operation day the patient developed jaundice. TPN was terminated with the suspicious diagnosis of TPN-induced cholestatic jaundice. Oral feeding was started on the 7th post operation day that was well tolerated. He still had jaundice and stool became gradually acholic.

Lab tests showed total bilirubin of 10 mg/dl with direct fraction of 6.4 mg/dl; SGOT 100 IU/L, SGPT 65 IU/L and alkaline phosphatase 175 IU/L. Diisopropyl Iminodiacetic Acid (DISIDA) scan showed no evidence of bile excretion into the bowel. On the 50th day of life, total bilirubin was 22 mg/dl with direct fraction of 14 mg/dl.

The patient underwent a second operation on the 65th day of life with the diagnosis of BA. On exploration gallbladder was small. Intraoperative cholangiography could not be carried out because of the obliterated gall bladder Kasai hepatoportoenterostomy was done. Feeding started on 7th post operation day. Postoperative course was uneventful. Jaundice improved and the stool color changed to green. He was discharged with prednisolone and urodeoxycolic acid and antibiotics. Two weeks later the patient was again admitted for cholangitis. He had fever and jaundice. Stool became acholic. The patient died of septicemia due to cholangitis one month later.

Discussion

Jejuno-ileal atresia is the most frequently encountered cause of small bowel obstruction in the neonatal period[7]. It is generally considered to result from intrauterine vascular disruptions to a segment of the developed intestine [8]. A report of Japanese Society of Pediatric Surgeons showed 304 cases of intestinal atresia in Japan in 1998, with the incidence of 2.53 per 10,000 live births[9]. JIA is not usually associated with other organ system anomalies and usually occurs as an independent single anomaly[7].

Prenatal perforation of proximal jejunum in JA can cause chronic MP, that is evident by severe abdominal distention immediately after birth, and calcification on plain abdominal x-ray. The incidence of BA is much lower than JIA. There is an association between BA and polysplania syndrome as this syndrome is seen in 25% of all BA patients [10].

The association of BA with intestinal atresia is rare. The most frequent form of intestinal atresia associated with BA is duodenal atresia[11]. There have been few reports of JIA associated with BA in literature. The incidence of JIA in cases of BA is reported 0% to 3.2%[12].

De Lorimer et al reported two cases of BA in 619 patients with small bowel atresia. The association between BA and MP from perforation of small bowel atresia has been reported[13].

Ohi et al reported the one patient (0.5%) in 214 with BA who had MP from a perforation associated with JA[14]. According to Japanese Biliary Atresia Registery between 1989 to 1996, 6 in 1,198 (0.5%) BA patients had MP.

Han et al reported five out of 171 (2.9%) BA cases had MP caused by perforation of small bowel atresia [15]. They suggested that BA may be related to a dynamic, acquired inflammatory process associated with MP that starts late in uterus and progresses postnataly in contrast to the belief that the cause of BA is an early development malformation. It is suggested that meconium peritonitis-induced inflammation of adjacent preductal tissue

could lead to a severe and protracted inflammatory reaction with accompanying fibrosis, causing secondary obliteration of the extrahepatic biliary system.

Kishida et al also reported two cases of MP in 84 (2.4%) cases of BA and speculated that M.P might be one of the causes of BA [12]. It is then recommended that gallbladder be examined for MP preoperatively and during operation by sonography[15]. Our case showed also cystic meconium peritonitis due to perforation of JA and associated BA.

The problem in managing of neonates with intestinal atresia postoperatively is TPN. TPN is usually required for nutritional support in most cases of MP with intestinal atresia because of the combined effects of short bowel syndrome and the delayed return of adequate motility and absorptive capacity of the digestive tract. There is also jaundice in 20% to 30% of small bowel atresia cases. So jaundice is difficult to differentiate in these cases [13].

In our case, we started TPN in the first post operative day, in assumption that return of bowel motility would be delayed postoperatively, as is the case in intestinal obstruction after operation.

After 5 days we terminated TPN because of developed jaundice assuming TPN induced cholestasis. However, persistent jaundice and acholic stools, led to the diagnosis of BA.

Another problem that we had after Kassias operation was ascending cholangitis. As the bowel was partly resected in the first operation, we had to decide whether to save the small bowel to prevent short bowel syndrome or to make a Roux-en-Y loop long enough to prevent ascending cholangitis. We decided for a Roux-en-Y limb of 40cm in length.

We think that one reason for 2 attacks of cholangitis in our case was bacterial overgrowth due to relative short bowel syndrome.

In a report by Han et al, 2 patients died of ascending cholangitis, and one of liver failure that was exacerbated by TPN-associated liver injury. They conclude that management of these patients is more difficult than that of patients with usual form of BA, because of the necessity for a long period of TPN and the combination with short bowel syndrome[15].

Asabe et al demonstrated 13 cases of BA with JIA in Japan. Seven of these 13 patients died giving the survival rate of 46.2% [16].

Regarding BA mortality two important factors should be considered: 1) Age at surgery and 2) repeated cholangitis attacks.

In our case both factors were operated. Because of delay in diagnosis the patient was operated in 65th day of life, which places him in the group of bad prognosis regarding the age. In addition, he had recurrent attacks of cholangitis due to short bowel syndrome caused by JA.

Therefore, the survival chance of JA with BA is very low compared with that of patients with JA or BA alone.

Conclusion

Exploration of gallbladder in cases of JIA especially when associated with MP is important. In addition, in cases of jaundice after repair of JIA, one must bear in mind the possibility of BA beside sepsis and TPN induced jaundice.

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