The Turkish Journal of Pediatrics 36: 263-266 1994

IS BIRTH TRAUMA RESPONSIBLE FOR IDIOPATHIC PERFORATION OF THE BILIARY TRACT IN INFANCY?*

*Gonca Topuzlu Tekant MD**, Ümit Yiğit MD***, Melih Bulut MD****

SUMMARY: Topuzlu Tekant G, Yiğit Ü, Bulut M. (Department of Pediatric Surgery, Şişli Children's Hospital, İstanbul, Turkey). Is birth trauma responsible for idiopathic perforation of the biliary tract in infancy? Turk J Pediatr 1994; 36: 263-266.

This case report details the clinical presentation and surgical management of a neonate with idiopathic perforation of the biliary tract. A three-day-old baby girl presented with a right-upper-quadrant mass and signs of peritonitis following a prolonged, difficult vaginal delivery. At surgery, she was found to have a perforation at the junction of the cystic and common bile duct. Simple drainage of the right upper quadrant was performed, and the patient recovered uneventfully. Early presentation and the nature of delivery suggests the possibility of birth trauma as an etiological factor in this condition. *Key words: biliary tract perforation, billary peritonitis, bile ascites, hepatobiliary imaging.*

Idiopathic perforation of the biliary tract is a relatively rare condition but is the second most common surgical cause of jaundice in infancy[1]. The first reported case was by Dijkstra in 1932 and involved a three-month-old baby girl who died following five weeks of progressive jaundice and abdominal distension. Postmortem study demonstrated a perforation at the junction of the common hepatic and cystic ducts[1].

The mechanism of perforation still remains ill-understood due to the variability of the age of presentation and symptomatology[2]. The chief aim of this report was to draw attention to birth trauma as a possible etiological factor responsible for perforation of the biliary tract in infancy.

Case Report

A three-day-old white baby girl was transferred to the Department of Pediatric Surgery of Şişli Children's Hospital due to abdominal distension, dyspnea and a delay in passage of meconium. She was a full-term 3600 g neonate, the product of a prolonged, difficult vaginal delivery, with Apgar scores of four and six at one and five minutes, respectively. She had perioral cyanosis and dyspnea that improved with administration of nasal oxygen in the neonatal unit. The following day she developed marked abdominal distension and had a delay in passage of meconium.

* From the Department of Pediatric Surgery, Şişli Children's Hospital, Istanbul.
** Resident in Pediatric Surgery, Şişli Children's Hospital.
*** Chief Resident in Pediatric Surgery, Şişli Children's Hospital.
**** Associate Professor of Pediatric Surgery, Şişli Children's Hospital.

Physical examination on presentation revealed a pale, ill-appearing neonate in respiratory distress with rapid and shallow respirations. Her abdomen was grossly distended with decreased bowel sounds, and a right upper quadrant mass 3 cm x 3 cm in diameter was palpated. There was a small amount of meconium passage following rectal examination.

Laboratory findings were as follows: Hematocrit 42%, white cell blood cell count 26,600/mm^3, serum bilirubin 2.8 mg/dl (conjugated 1.8 mg/dl), SGOT 20 U/L, SGPT 13 U/L, and alkaline phosphate 145 U/L. Urine analysis was normal. Radiological examination of the abdomen showed dilated loops of bowel, and the patient was transferred to our department with an initial diagnosis of intestinal obstruction. Abdominal ultrasound suggested a cystic mass occupying the upper abdomen, closely related to the intestines. The gallbladder and extrahepatic bile ducts were not visualized.

An initial diagnosis of a duplication cyst leading to partial intestinal obstruction was made. At laparotomy through an upper transvers incision, 100 ml bile mixed with blood was evacuated from the cavity behind the right lobe of the liver, and further dissection revealed a perforation of the anterior aspect of the common bile duct at the junction with the cystic duct. The gallbladder was found to be distended with blood, and manual examination resulted in drainage of the contents from the site of perforation. The peritoneal cavity was drained with a Penrose drain through a separate incision. The patient received seven days of total parenteral nutrition (TPN) and broad-spectrum antibiotics post-operatively, followed by breast feeding. Initially small amounts of bile drained from the Penrose drain, and it was removed on the 10^{th} day when the drainage had ceased. The hepatobiliary scan using Tc-99m disofenin at two weeks demonstrated a normal gallbladder and normal excretion of radioisotope into the duodenum. The patient was discharged two weeks after surgery.

The patient was readmitted two weeks after her discharge due to bilious vomiting and abdominal distension. On laparotomy, multiple adhesions leading to intestinal obstruction were lysed. The previous site of perforation was found to have healed completely.

One year later, the patient was healthy and gaining weight.

Discussion

The clinical manifestations of idiopathic biliary tract perforation may be divided into two groups[3-5]. The acute form presents with a sudden onset of shock, generalized peritonitis, vomiting, abdominal distension and leukocytosis[6]. With leakage of bile into the peritoneal cavity, jaundice, biliary ascites and acholic stools appear. The child may die due to cardiorespiratory collapse or septic shock at this stage. However, 80 percent of cases have a subacute or chronic form of

presentation[5], where jaundice may be unnoticed within the first months of life until the clinical picture is dominated by persistent jaundice, biliary ascites and abdominal distension. Our case presented clinically in the acute form, with abdominal distension that developed rapidly after birth.

The acute cases are more difficult to diagnose preoperatively, but usually have surgery earlier due to symptoms of an acute abdomen[2,6,7]. In chronic cases, diagnosis is made on the basis of clinical presentation[8,9], abdominal paracentesis[6,10] or nuclear hepatobiliary imaging[7,11]. In our case, which presented acutely, the condition was not suspected preoperatively due to the lack of more specific symptoms (i.e., jaundice, ascites) and the suspicion of a duplication cyst on ultrasound examination.

Different etiological factors for idopathic biliary tract perforation have been proposed, including trauma[5], biliary lithiasis[12], inspissated bile[13], anomalies of the biliary tract[14], congenital weakness and vascular impairment[1,15]. Johnston[8] suggested the existence of a localized developmental weakness of the wall of the common bile duct. In that case, a sudden increase in intra-abdominal pressure during labor could lead to perforation of the area of mural weakness. Whatever the cause of spontaneous perforation, the resulting bile leak leads to the formation of a biliary pseudocyst, which in turn may leak into the peritoneal cavity[11]. The early presentation of this infant with symptoms starting from birth and the history of a prolonged, difficult vaginal delivery suggest the possibility of birth trauma as an etiological factor. In addition, the intraoperative findings in this case correlate well with the early stage of pseudocyst formation. It has been proposed that not all ruptures may occur immediately at the time of injury, and the devitalized area or clot may slough in subsequent days, with a delay in the onset of symptoms[9]. This may explain the wide variation in time intervals between trauma and the onset of symptoms.

Treatment of perforation of the biliary tract is surgical. In a recent review of 70 cases, the five that were medically treated all died. On the other hand, of the 65 surgical cases, 55 have survived[2]. However, there is controversy regarding the surgical treatment options. Some surgeons prefer invasive surgical techniques such as inserting T-tube choledochal drains, or decompressing the biliary tract by cholecystostomy[1,4,5,14,16] or by draining it into the jejunum through a Roux en "Y" anastomosis[1,4,17,18]. But due to the small size and fragile nature of an infant's common bile duct, such procedures have led to further complications of perforation, stricture formation and obstructive jaundice[5,14]. Successful results have been obtained by a more conservative surgical approach, with local drainage of the right upper quadrant with or without external decompression of the gall bladder[2,4,6,7,19,20]. In the presence of choledocholithiasis, a cholecystostomy is advocated for decompression and radiological access to the biliary tree in the

postoperative period[16]. On the other hand, nuclear imaging studies are sufficient in evaluating the patency of the biliary tract following simple drainage procedures[11].

Successful postoperative management depends on TPN, complete gastrointestinal rest and broad-spectrum antibiotics with a high biliary concentration[10,16]. It is important to realize that this condition has a high curative rate with early diagnosis and appropriate surgical intervention.

REFERENCES

1. Lilly JR, Weintraub WH, Altman RP. Spontaneous perforation of the extrahepatic bile ducts and bile peritonitis in infancy. Surgery 1974; 75: 664-73.
2. Hammoudi SM, Alauddin A. Idiopathic perforation of the biliary tract in infancy and childhood. J Pediatr Surg 1988; 23: 185-7.
3. Enell H, Cavell B, Malmfors G. Spontaneous perforation of the common bile duct. Acta Paediatr Scand 1979; 68: 625-6.
4. Lees W, Mitchel JE. Bile peritonitis in infancy. Arch Dis Child 1966; 41: 188-92.
5. Prevot J, Babut M. Spontaneous perforation of the biliary tract in infancy. In: Rickham PP, Heckler WCH, Prevot J, (eds). Progress in Pediatric Surgery Vol.1. Baltimore: University Park Press; 1971: 187-208.
6. Stringel G, Mercer S. Idiopathic perforation of the biliary tract in infancy. J Pediatr Surg 1983; 18: 546-50.
7. So SK, Lindahl JA, Sharp HL, Cook AM, Leonard AS. Bile ascites during infancy: diagnosis using Disofenin Tc 99m sequential scintiphotography. Pediatrics 1983; 71: 402-5.
8. Johnston JH. Spontaneous perforation of the common bile duct in infancy. Br J Surg 1961; 48: 532-3.
9. Masson LB, Bidbury JB, Culang S. Rupture of the extrahepatic bile ducts from nonpenetrating trauma. Ann Surg 1954; 140: 234-41.
10. Rivilla F, Diez J. Bile peritonitis in the newborn. Acta Paediatr Scand 1989; 78: 782-3.
11. Haller JO, Condon VR, Berdon WE, et al. Spontaneous perforation of the common bile duct in children. Radiology 1989; 172: 621-4.
12. Colver HD. Perforation of the biliary tract due to gallstones in infancy. An established clinical entity. Ann Surg 1964; 160: 226-31.
13. Hartong JM. Spontaneous perforation of the extrahepatic bile ducts in infancy. Am Surg 1976; 42: 795-800.
14. Ohkawa H, Takahashi H, Maie M. A malformation of the pancreatico-biliary system as a cause of perforation of the biliary tract in childhood. J Pediatr Surg 1977; 12: 541-6.
15. Northover JM, Terblanche J. A new look at the arterial supply of the bile duct in man and its surgical complications. Br J Surg 1979; 66: 379-84.
16. Megison SM, Votteler TP. Management of common bile duct obstruction associated with spontaneous perforation of the biliary tree. Surgery 1992; 111: 237-9.
17. Fitzgerald RJ, Parbhoo K, Guiney EJ. Spontaneous perforation of bile ducts in neonates. Surgery 1978; 83: 303-5.
18. Howard ER, Johnston DI, Mowat AP. Spontaneous perforation of common bile duct in infants. Arch Dis Child 1976; 51: 883-6.
19. Caulfield E. Bile peritonitis in infancy. Am J Dis Child 1936; 52: 134-60.
20. Pettersson G. Spontaneous perforation of the common bile duct in infants. Acta Chir Scand 1955; 110: 192-201.