

SPONTANEOUS PERFORATION OF THE BILE DUCT IN INFANTS: A CASE REPORT

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Abstract

Spontaneous perforation of the bile duct is a rare disease in children. To date, less than a hundred cases have been reported in English literature. A number of techniques have been applied to achieve preoperative diagnosis yet most cases are diagnosed at operation. A 3-month-old girl presented with fever, vomiting, progressive abdominal distension, jaundice and diarrhoea. Abdominal ultrasonography showed localized collection of fluid that displaced the small bowel to the right side of the abdomen. The fluid was found to be bilious on paracentesis. At laparotomy, biliary pseudocyst was found but the site of perforation was no longer identifiable. Excision of the containing wall and external drainage was carried out. 9 months after operation the child is well. A high index of suspicion should improve diagnosis and ensure early intervention.

Mots clés: Canal biliaire, perforation spontanée, enfant

Résumé

La perforation spontanée des canaux biliaires est une affection rare chez les enfants. A ce jour, moins de 100 cas ont été rapportés dans la littérature anglophone. Un certain nombre de techniques ont été mises en œuvre pour le diagnostic pré opératoire, néanmoins la plupart des cas sont de diagnostic per opératoire.

Une fille de 3 mois avait présenté de la fièvre, vomissements, distension progressive de l'abdomen, ictère et diarrhée. L'échographie de l'abdomen avait montré une collection liquidienne localisée refoulant les anses grêles vers le côté droit de l'abdomen. La paracentèse avait retrouvé un liquide bilieux. La laparotomie avait noté un pseudo kyste biliaire, mais le siège de la perforation n'était plus visible. La paroi du contenant a été excisée et un drainage externe a été réalisé. Neuf mois après l'opération, les suites étaient simples. Une forte suspicion devrait mener au diagnostic et imposer une intervention précoce.

Introduction

Spontaneous perforation of the bile duct is a rare but recognized disease entity in children.^{1,2} The aetiology is largely unknown though pancreatico-biliary maljunction has been postulated. This case report draws attention to the need for a thorough diagnostic work-up in an infant with jaundice and progressive abdominal distension. The limitations in our setting are highlighted.

Case report

A 3-month-old girl, presented with 1-month history of fever, progressive abdominal distension, vomiting, jaundice and intermittent diarrhea. The antenatal,

perinatal and neonatal periods were uneventful. There was no history of trauma. Earlier in the referring hospital, she was managed for hepatitis.

Physical examination showed respiratory distress mild jaundice and a weight of 3.5kg (birth weight was 2.9kg). The abdomen was markedly distended and tense, with the skin shiny and displaying superficial veins that drained superiorly. There was umbilical hernia with an interval diameter of 4cm, but no distinct mass was felt. Bowel sounds were diminished.

Liver function test showed mildly elevated total and conjugated bilirubin as well as alkaline phosphatase levels. Abdominal ultrasonography showed localized collection of fluid that displaced

normal caliber intestines to the right side of the abdomen. Paracentesis abdominis done afterwards yielded bilious fluid.

At laparotomy, 72 hours after the initial assessment, biliary pseudocyst with the wall attached to the anterior abdominal wall, umbilicus, greater curvature of the stomach and transverse colon was found. The liver, gallbladder, cystic duct and intestines were normal. There were adhesions in the region of the common bile duct. This was left undisturbed. The wall of the pseudocyst was excised after draining 2 litres of bilious fluid and a drain inserted in the lesser sac. Bilious drainage stopped after 18 days and the drain was removed 2 days later.

Nine months after operation the child has remained in good health. There is no jaundice and she has bile in her stools. The liver function tests are normal. Repeat abdominal ultrasonography 6-months after operation did not show any residual biliary tract problem.

Discussion

The aetiology of Spontaneous perforation of the bile duct (SPBD) is not known. Pancreatico-biliary maljunction and weakness in the bile duct have been postulated.^{1, 2} Hasegawa et al¹ noticed elongated common pancreatico-biliary channel as well as elevated bile amylase in some patients. The inconsistency of these findings led a conclusion that multiple mechanisms may be involved.

In most cases^{3, 4} there is variable postnatal symptom free interval before the onset of fever, jaundice, progressive abdominal distension, vomiting, acholic stools and failure to thrive. These symptoms were present in our patient except acholic stools. We suspect that the presence or otherwise of bile in stools may be related to the rate of bile leakage from the perforation with massive leak depriving the intestines of bile. Secondary biliary stenosis⁵ may be contributory. The rate of leakage could also explain the varying degree of rapidity of symptoms. The consistency of these symptoms in most of the reported series should raise the index of suspicion and lead to early diagnosis, but most cases including our patient were managed for another suspected pathology usually hepatitis before referral. Attention to clinical details should avoid this error.

Abdominal ultrasonography, hepatobiliary scintigraphy and abdominal paracentesis are the investigations of choice. Sonography shows either generalized ascitis or localized collection of fluid.⁴ The biliary tree is usually not dilated. Results of the scintigraphy and abdominal paracentesis demonstrate that intraperitoneal fluid originated from the biliary tract.³ Intraoperative cholangiography can be used to confirm diagnosis at surgery.⁴ Due to non-availability of scintigraphy facilities, we relied on ultrasonography and confirmed the nature of fluid by paracentesis. Use of hepatobiliary scintigraphy may

may hasten preoperative diagnosis. Ultrasonography, hepatobiliary scintigraphy and findings at operation will obviate this.

The commonest reported site of perforation is at the junction of the cystic and hepatic ducts.^{1, 2, 5} Other sites include cystic duct, common hepatic duct and common bile duct. The site of perforation was no longer identifiable at the time of operation.⁵ The presence of adhesions around the common bile duct in this patient made it the suspect site of perforation.

Definitive treatment involves surgery, which ranges from percutaneous tube drainage³ to complicated biliary procedures⁵. Factors like the site of perforation and the facilities available may influence the choice of procedure. When there are facilities to confirm diagnosis of SPBD without operative intervention, percutaneous tube drainage is the ideal treatment. The basis for this is that in most cases the perforation heals spontaneously as there is usually no distal obstruction³. In most cases however, diagnosis is confirmed at operation and the procedure of choice is external drainage with or without complete excision of wall of the pseudocyst.^{2, 4, 6} The intestines are usually not affected. The drain is left in situ for as long as there is bilious drainage, which in our patient lasted 18 days. Direct attempts to close the perforation or other biliary surgical procedures employed by some authors in cases of perforation at the common bile duct is usually associated with significant morbidity and mortality.^{2, 5}

Most of the patients managed by external drainage have no residual hepatobiliary dysfunction.^{2 - 4} Spontaneous perforation of the bile duct is a rare but potentially fatal disease in children. High index of suspicion, thorough diagnostic workup and prompt surgical intervention will improve the prognosis.

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