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Spontaneous Biliary Perforation: A Rare Cause of Infantile Cholestasis Managed Conservatively

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Abstract **Case Report**

Spontaneous biliary perforation in infancy is a rare but important cause of surgical jaundice, with management strategies ranging from conservative drainage to complex biliary reconstruction. The condition is characterized by bile leakage into the peritoneal cavity without an identifiable cause and is often associated with significant morbidity if not promptly recognized. We report the case of a 4-month-old female who presented with progressive jaundice, abdominal distension, lethargy, and failure to thrive. She was born preterm at 35 weeks with low birth weight and had prolonged neonatal hospitalization for sepsis and jaundice. On referral, she was severely malnourished with tense abdominal distension. Laboratory evaluation revealed obstructive jaundice with preserved hepatic function, while ultrasound and hepatobiliary scintigraphy confirmed biliary ascites and active bile leak. Exploratory laparotomy revealed free bile in the peritoneal cavity and a tiny calculus at the leak site; however, anatomy could not be clearly defined due to inflammation. External drainage was performed. Postoperatively, she required prolonged parenteral nutrition and drain repositioning for loculated collections. Gradual clinical and biochemical improvement was noted, with eventual clearance of jaundice, normalization of stool color, and weight gain. She was thriving at 3-month follow-up. This case highlights the diagnostic challenges of spontaneous biliary perforation in infants and supports the role of timely surgical drainage as a safe and effective initial management strategy, avoiding complex biliary reconstruction in critically ill children.

Keywords: spontaneous biliary perforation; infantile cholestasis; obstructive jaundice.

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INTRODUCTION

Spontaneous biliary perforation in children is a rarely reported entity with variable management strategies ranging from conservative approaches to complex biliary-enteric reconstructions. It is characterized by leakage of bile into the peritoneal cavity without an identifiable cause. Suggested etiopathologies include pancreaticobiliary malunion leading to reflux into inherently weak bile duct walls, along with vascular insufficiency at critical points in the biliary tree or an underlying choledochal malformation [1]. Most cases present with acute deterioration at birth, but occasionally an indolent subacute course in infancy is observed. We report a case of a 4-month-old severely ill infant referred from a rural center, successfully managed conservatively.

CASE REPORT

4-month-old female child was referred to our institute with complaints of progressively increasing jaundice and unresolving abdominal distension since 1month. It was associated with passage of acholic stools, severe lethargy and failure to thrive. The patient had history of prolonged hospitalization since birth. She was born prematurely at 35weeks by caesarean section with a birth weight of 1.2kg. Mother had an uneventful antenatal period but the antenatal scan was suggestive of intrauterine growth retardation. Following neonatal intensive care of 40days in rural hospital for management of jaundice, respiratory distress and sepsis, patient was discharged and was exclusively breast fed at home. She was taken to local hospital again at 3months of age with complaints of increasing yellowish discoloration of eyes and skin along with repeated emesis and severe abdominal distension. She was treated conservatively for

one month with intravenous antibiotics, as a case of suspected hepatitis with anaemia and received blood transfusions for the same. As there was no improvement despite maximal efforts, she was referred to our hospital for further management. On examination, patient was severely malnourished with tense distended abdomen. After detailed evaluation, deranged liver biochemistry with parameters suggestive of obstructive jaundice and preserved hepatic function were noted. Medical causes like viral infections and metabolic disorders including galactosemia were ruled out by respective laboratory investigations. Ultrasound revealed presence of gross ascites with multiple echoes and septations. Liver echogenicity was slightly altered but no intra or extra hepatic biliary tract dilatation was noted. Gall bladder was minimally distended with normal contractility and no evidence of triangular cord sign. Common bile duct was not dilated and portal vein had normal flow. Percutaneous ultrasound guided aspiration of greenish ascitic fluid was subjected to analysis, leading to the diagnosis of biliary ascites with raised bilirubin levels and high neutrophil count. HIDA Scan reported that tracer uptake was homogenous in liver, with tracer seen in ascitic fluid at 10th minute, followed by gradual increase in activity over entire peritoneal cavity demonstrating biliary leak. Decision was taken to do a mini right subcostal laparotomy. Drainage of approximately 400cc bilious peritoneal fluid was done. Liver appeared healthy. On introduction of normal saline into the patent gall bladder, an obvious leak of bile along with a tiny calculus was noted immediately from the biliary tract into the abdomen. Due to severe inflammation and periportal collection, precise biliary anatomy could not be delineated. [Figure 1]

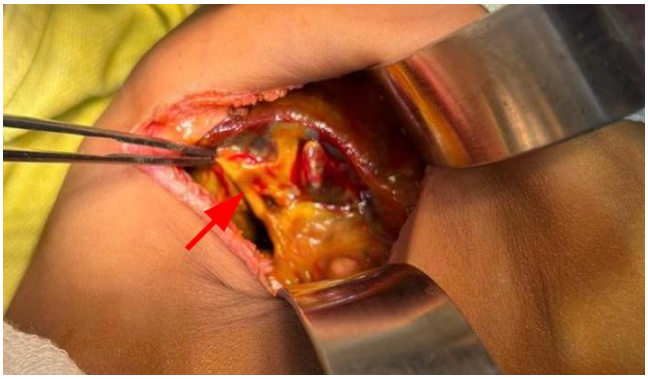

Figure 1. Intraoperative photograph showing surgical exposure of the bile duct with leak. Note the inflamed and thickened periductal tissue(arrow)

Considering the poor general condition of the patient and complex intraoperative findings, an intraabdominal drainage tube was placed after thorough lavage. Postoperatively patient was started on total parenteral nutrition for 2 weeks. On postoperative day 3, ultrasound guided repositioning of abdominal tube was done and approximately 100ml thick bilious loculated collection was drained. Improvement in vital parameters and activity of child was noted post procedure. Meticulous monitoring showed a gradual improvement in symptoms with soft abdomen and decreasing abdominal girth with passage of normal cholic stools. [Table 1].

Table 1: Clinical parameters during hospital stay

Time point	Jaundice	Abdominal distension (Girth in cm)	Drain output	Colour of stools
On admission	Involving entire upper and lower limbs	54	–	White (acholic)
Postop day 1	Bilateral upper and lower limbs	52	120 ml bilious	Not passed stools
Postop week 1	Up to arms and thigh	48	50–80 ml	Light yellow tinge
Postop week 2	Trunk	45	20–40 ml	Light yellow
Postop week 3	Face and sclera	42	10–15 ml bilious	Yellow
Before discharge	No	40	Minimal serous	Dark yellow (cholic)

Footnotes:
a Postop = postoperative.
b ml = millilitres.

Clinical jaundice clearance was confirmed by normalisation in levels of liver biochemistry parameters. As the intraabdominal drainage of fluid decreased in quantity and became serous over a period of three weeks, local removal of tube drain was done. [Table 2].

Table 2. Laboratory investigations during hospital stay

Time point	Hb (g/dl)	WBC (/µL)	Bilirubin (mg/dl) Total/Direct	SGOT/SGPT (U/L)	ALP (IU/L)	Total protein/Albumin (g/dl)
On admission	7.0	14,000	10.4 / 6.4	98 / 65	1299	6.2 / 4.3
Postop day 1	8.5	12,000	9.0 / 7.5	80 / 64	739	6.0 / 4.2
Postop week 1	10.5	10,500	4.4 / 3.3	30 / 14	536	4.9 / 2.6
Postop week 2	10.0	8,600	2.2 / 1.7	89 / 44	346	4.8 / 2.8
Postop week 3	10.2	6,000	1.4 / 1.0	42 / 15	189	4.9 / 4.1
Before discharge	10.6	4,400	1.0 / 0.7	36 / 7	187	4.8 / 3.6

Footnotes:
a Hb = hemoglobin; WBC = white blood cell count; SGOT = serum glutamic oxaloacetic transaminase; SGPT = serum glutamic pyruvic transaminase; ALP = alkaline phosphatase.
b Units: g/dl = grams per decilitre; U/L = units per litre; IU/L = international units per litre; µL = microlitre.

Followup ultrasound over the next week confirmed evidence of only minimal <1cc collection in peritoneal cavity. Gradual introduction of fat free milk was tolerated well by the patient and significant weight gain noted after which she was discharged on oral antibiotics supplemented with cholestatic agents and fat-soluble vitamins. The patient was thriving well at 3months follow-up visit.

DISCUSSION

Spontaneous biliary perforation is a rare, but important cause of surgical jaundice in infants, first described by Caulfield in 1936. According to recent literature review the incidence equals 1.5 in 10,00,000 live births with most cases presenting in infancy with an equal sex ratio. [2] It is difficult to diagnose as the etiopathology remains obscure and multifactorial. The theory of congenital weakness of walls of biliary tree, presence of mural malformations along with ischemia is supported by the fact that most common site of perforation is at the anterior wall of junction of the cystic duct and common bile duct. [3] Secondly, abnormal pancreaticobiliary malunion near sphincter of oddi leads to reflux of pancreatic contents into biliary tree. This additional inflammation over inherent mural immaturity may be contributory to perforation. Lastly a previously undiagnosed choledochal cyst may also be a pathological site. [4] Rarer sites of perforation may include any part of extra or intrahepatic biliary tree leading to a constrained leak. [5] High osmotic pressure of biliary contents lead to free fluid shift into peritoneal cavity with increasing ascites.

Clinical presentation is variable and the most common symptoms include abdominal distension, nonbilious vomiting and features of obstructive jaundice. It may be associated with umbilical or inguinal hernia and hydroceles with greenish hue. [6] There is usually an asymptomatic interval preceding the perforation. It is followed by mild constitutional upset with a period of illusion during which egress of sterile bile into peritoneal cavity occurs. Superimposed bacterial peritonitis may lead to acute abdominal pain and fever. Loculated or contained collections may have a subacute course. Secondary bile duct strictures, portal vein thrombosis and pseudocyst formation may occur with prolonged course. [7]

Preoperative diagnosis is often challenging. Liver function tests reveal characteristic picture of obstructive jaundice. There is normal bile production, but bile neither reaches gut nor blood in sufficient quantities as it escapes into peritoneal cavity. While ultrasound is initial non-invasive investigation of choice and easier for repeated monitoring and followup, Magnetic resonance cholangiopancreatography gives better anatomical and functional delineation in complex cases.[8] Guided abdominal paracentesis is diagnostic. Hepatobiliary scintigraphy is highly sensitive and specific and is virtually diagnostic when it shows extravasation of tracer into the peritoneal cavity with no activity in the duodenum. [9] Gd-BOPTA and Gd-EOB-DTPA allow hepatic dynamic imaging and provide both functional and morphological information by localizing site of bile leakage. [10] In the management of spontaneous biliary perforation, various treatment modalities have been advocated, to be used depending upon individual patient characteristics and surgeon preference. Primarily conservative management with antibiotics and simple external drainage can help in successful healing of perforation, provided there is no distal ductal obstruction. [11] Importantly, wide drainage using either percutaneous tube insertion or laparotomy with simple external drainage has been reported as the best initial treatment to manage bile leak without primary biliary reconstruction in 85% of patients. This avoids complex and difficult dissection in an inflamed operative field and minimizes risk of additional injury with resolution of symptoms evident within 2 weeks. [12] However the risk of persistent biliary fistula always exists. Diagnostic laparoscopy with percutaneous drainage tube placement also works on similar principle.[13]

A laparotomy with intraoperative cholangiogram is helpful in delineating the exact site of leak in cases where the biliary anatomy is not easily discernible due to inflammation or mass formation. Flushing of the distal common bile duct with a fine

catheter has also been recommended, to remove any distal inspissated bile plugs. [14,15] Cholecystectomy may be added as a preliminary procedure, especially in localised cystic duct perforations. In cases where the site of perforation is easily identifiable, a primary repair with or without an external biliary drainage by cholecystostomy or T tube insertion is done. For cases where the site is sloughed off due to necrosis, a T-tube may be secured with a purse-string in the proximal and the distal stumps, for external drainage of the bile via the third limb. Temporary cholecystostomy tubes followed by delayed reconstruction is also practiced. In rarer cases of choledochal cyst, excision of cyst followed by a biliary reconstruction as a primary or a secondary procedure is done. [16,17] Complex biliary enteric bypass procedures like hepaticojejunostomy are definitive, especially if there is a pancreaticobiliary malunion or a distal obstruction and are usually taken up after resolution of acute phase of illness. The post-operative complications include formation of biliary duct stricture, bile leak with persistent biliary fistula and recurrent cholangitis. [18,19] Minimally invasive alternatives include endoscopic retrograde cholangiopancreatography or percutaneous transhepatic drainage and biliary stenting. Access of the biliary tree via interventional radiology procedures for complex biliary disease is novel and presents an alternative to traditional open surgical treatment and control of spontaneous biliary perforations. [19]

Conclusion

Spontaneous biliary perforation is a rare cause of infantile cholestasis. This challenging diagnosis requires a high index of suspicion as timely management is life saving. Surgical drainage is a simple and effective method for primary treatment, particularly in children with delayed presentations, comorbidities and unstable conditions. The immediate high risks associated with major surgical procedures in these patients far outweigh the beneficial long-term outcomes.

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