

Case Report

Spontaneous biliary cutaneous fistula communicating with intrahepatic bile duct in a six years old child: a very rare case report

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ABSTRACT

Biliary cutaneous fistula or external biliary fistula is an abnormal communication between the biliary tree and the skin surface. We present here a very rare case of spontaneous biliary cutaneous fistula communicating with intrahepatic bile duct in a 6 years old child. The child suffered from chronic hepatic parenchymal disease and presented initially with abdominal wall abscess, right subdiaphragmatic collection and possible hepatic abscesses. The patient left the hospital against medical advice and returned back after 6 months with external biliary fistula in the epigastric region.

Keywords: Biliary cutaneous fistula, External biliary fistula, Abdominal wall abscess, Hepatic abscess

INTRODUCTION

Biliary cutaneous fistula or external biliary fistula is an abnormal communication between the biliary tree and the skin surface. External biliary fistulas are subdivided based on etiology into spontaneous, traumatic, therapeutic and iatrogenic. Spontaneous external biliary fistulas secondary to gall stones perforating through the abdominal wall were common in the past. These are rare in modern day medical practice; fewer than 25 cases have been reported over the past 50 years. We present here a case of spontaneous external biliary fistula in a 6 years old child. The fistula communicated with the intrahepatic bile duct and skin surface in the epigastric region. Spontaneous external biliary fistula arising from intrahepatic bile duct in pediatric age group is an extremely rare condition. Only two cases of spontaneous choledochocutaneous fistula have been reported in elderly age group patients. Underlying pathology was cholangiocarcinoma in both the cases.¹

CASE REPORT

A 6 years old female child presented (initially) to the Department of Surgery, Silchar Medical College and Hospital, Assam, India with history of fever, pain abdomen and swelling in the epigastric region. On examination, patient was pale, dehydrated, febrile and a soft, tender, non-mobile lump was palpable in the epigastric region. Local skin temperature was raised. Routine blood examination revealed leucocytosis (16,000/ μ L), anaemia (Hb-9g/dL) and raised ESR (40mm AEFH). Abdominal ultrasound examination showed debris, thick-walled collection measuring 3.5 x 3 cm in the anterior abdominal wall in the epigastric region. Surrounding fat planes were edematous. Right subdiaphragmatic debris collection was also noted indenting the liver surface. Liver showed coarse parenchymal echopattern and surface nodularity. Gall bladder and common bile duct were normal. Contrast enhanced CT scan abdomen was performed which revealed a rim-enhancing anterior abdominal wall abscess which tracked along the falciform ligament into the left hepatic intersegmental fissure. High attenuation

collection was noted in right subdiaphragmatic region. Irregular non-enhancing areas were noted in segments IVa, IVb, V and VIII of liver which suggested possibility of resolving / evolving hepatic abscesses. Irregularity of liver surface, widening of interlobar fissure, atrophy of segment IV and enlargement of caudate lobe were noted suggestive of chronic hepatic parenchymal disease (Figure 1A, 1B and 1C). Exploration was planned, but the patient left the hospital against medical advice. After a period of six months, the patient again presented to our hospital (this time) with a discharging sinus in the epigastric region. On examination, thick, greenish discharge was noted in the mouth of the cutaneous ulcer and surrounding skin was mildly indurated (Figure 2). Sinogram was performed where the contrast material was noted to collect underneath the abdominal wall and further opacify the biliary tree (Figure 3). On ultrasonography abdomen, a well-defined, thick-walled, fluid-filled tract was noted (diameter approx. 5 mm) extending from the cutaneous opening in the anterior abdominal wall upto the left hepatic intersegmental fissure. Minimal collection was noted along the tract underneath the abdominal wall. However, no evidences of subdiaphragmatic collection or intrahepatic abscess were present this time. Features of chronic hepatic parenchymal disease were persistent (Figure 4A and 4B). Gall bladder was normal; a small roundworm was noted in distal CBD lumen with no evidence of proximal biliary dilatation. Magnetic Resonance Imaging of the abdomen was performed subsequently; T2 Weighted Fat-suppressed images in axial and sagittal planes demonstrated the hyperintense fluid-filled fistulous tract between the skin surface and the left hepatic intersegmental fissure with minimal collection underneath the abdominal wall (Figure 5A and 5B). After a course of broad spectrum antibiotics the patient was posted for surgery. On laparotomy, a well-formed thick-walled fistulous tract was discovered communicating with the anterior abdominal wall and the left hepatic intersegmental fissure with surrounding gross adhesions and omental inflammation. Liver was cirrhotic (Figure 6A and 6B). No filling defect was palpable in the CBD. Deep extension of the tract into the intersegmental fissure could not be excised and hence was anastomosed with the duodenum. Superficial aspect of the tract was excised and sent for histopathological examination. Report revealed that the tract consisted of chronic inflammatory cells and prominent bile pigments. No malignant cells were present.

DISCUSSION

A fistula is an epithelium-lined tract between two epithelium-lined surfaces. Biliary fistulas are divided into two types; internal and external. Internal biliary fistulas occur between the biliary tree and various structures like bowel, bronchial tree and vessels. External biliary fistulas communicate with the skin surface. Biliary fistulas develop as a complication of chronic cholelithiasis, infection, peptic ulcer, malignancy or trauma. External

biliary fistulas are further subdivided based on etiology into spontaneous, traumatic, therapeutic and iatrogenic.¹



Figure 1A: Immediate post-contrast scan shows the rim-enhancing anterior abdominal wall abscess.

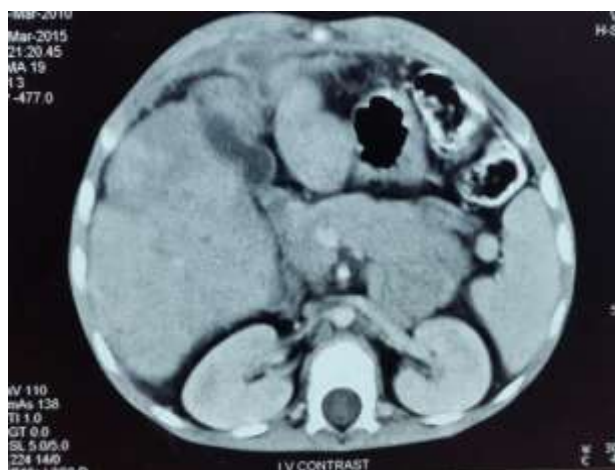


Figure 1B: Delayed scan shows atrophy of segment IVb and interlobar fissure widening.



Figure 1C: High attenuation right subdiaphragmatic collection and irregularity of superior liver surface are shown.



Figure 2: Discharging sinus in epigastrium when the child presented after 6 months.



Figure 4B: Ultrasound shows extension of the tract into left hepatic intersegmental fissure.



Figure 3: Sinogram shows contrast accumulation underneath the abdominal wall with opacification of the biliary tree.



Figure 5A: MRI (t2fs sagittal) demonstrates the hyperintense thick-walled fistulous tract from the skin to the liver surface.

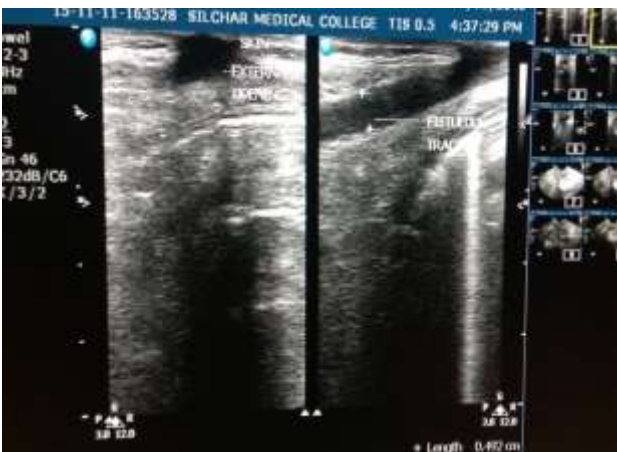


Figure 4A: Ultrasound shows the external opening and fistulous tract underneath the abdominal wall.



Figure 5B: MRI (t2fs axial) shows minimal collection underneath the abdominal wall overlying the liver surface and normally distended gall bladder.



Figure 6A: Intraoperative photograph shows liver surface in the background and the fistulous tract (held with forceps) extending into the intersegmental fissure.



Figure 6B: The firm, well-formed tract is held up for demonstration.

Spontaneous external biliary fistulas secondary to gall stones perforating through the abdominal wall were common in the past. Cholecystocutaneous fistula (an abnormal communication between the gall bladder and the skin) was first described by Thilesus in 1670. Courvoisier documented 499 cases of gall bladder perforation in 1890; 169 of these cases formed cutaneous tracts. Spontaneous external biliary fistulas almost always communicate with the gall bladder. Biliary fistula arising from intrahepatic bile duct is an extremely rare condition. Only two cases of spontaneous choledochocutaneous fistula (an abnormal communication between the bile duct and the skin) have been reported. Both the patients were in the elderly age group and cholangiocarcinoma was the underlying pathology.¹

External biliary fistulas are very rare in modern day medical practice; fewer than 25 cases have been reported over the past 50 years.² This marked decrease in incidence can be attributed to the advent of contemporary diagnostic methods, availability of broad spectrum

antibiotics, early and effective management of biliary tract disease.

Spontaneous external biliary fistulas develop as a consequence of neglected biliary tract disease. They are more frequent in women than men (F:M-3:1), likely due to higher incidence of cholecystitis in the former.^{1,2} Increased intraluminal pressure in the gall bladder due to calculus obstruction is thought to impair blood supply and lymphatic drainage, leading to mucosal necrosis and perforation, which can be acute or indolent process. A chronic perforation can lead to an internal or external biliary fistula and these arise most commonly from the fundus of the gall bladder.² The fistula usually communicates with the duodenum (77%), colon (15%) or very rarely through the abdominal wall.^{1,3} In case of an external biliary fistula, the opening is generally in the right hypochondrium. Other sites are the left hypochondrium, umbilical region, right lumbar region, right iliac fossa and gluteal region.⁴ Discharge may be purulent in case of empyema, mucoid in presence of mucocele or bilious in absence of obstruction. Small stones within the discharge often confirm the diagnosis. Fistulogram can make a definitive diagnosis of this condition.⁵ Ultrasonography, Computed Tomography and Magnetic Resonance Imaging can help in establishing the underlying pathological condition.

Management of the biliary fistula usually begins with antibiotics for controlling the inflammatory process. Fistulous tract is surgically removed thereafter. The underlying pathology must be treated properly to achieve good post-operative results. Neglected biliary fistulas are associated with complications. Chronic bilious discharge from the fistulous opening can cause skin irritation and dermatitis. Necrotizing fasciitis of the anterior abdominal wall has been reported. Chronic untreated fistula can rarely lead to dysplasia and subsequent malignant change similar to Marjolin's ulcer.⁶

CONCLUSION

Considering all the above mentioned facts we can come to the conclusion that our case is unique with respect to the following three points; Firstly, the patient was a child. Biliary fistulas usually occur in the elderly and are extremely rare in pediatric age group. Secondly, the patient did not suffer from any biliary tract pathology. Biliary fistulas are almost always a consequence of neglected biliary tract disease and are extremely unusual in their absence. Though a round worm was detected in common bile duct on ultrasonography of abdomen when the patient presented for the second time, it was not palpable on laparotomy. Therefore, it must have been a transient finding and passed out spontaneously. Thirdly, the fistula communicated with the intrahepatic bile duct. Biliary fistulas mostly communicate with the gall bladder, fundus being the usual site. Only two reported cases of biliary cutaneous fistulas communicating with intrahepatic bile duct could be found in literature review.

We put forward the hypothesis that the underlying pathology in our case could be; (1) The hepatic abscess or (2) The abdominal wall abscess which extended along the falciform ligament into left hepatic intersegmental fissure. As the patient left the hospital against medical advice during her first visit and did not receive proper treatment, either/both of the abscesses communicated with the intrahepatic bile duct and decompressed into the overlying skin surface resulting in a biliary cutaneous fistula.

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REFERENCES

1. In DS, Hyung CO, Jae HD, Lae IJ, Beom JK. Spontaneous external biliary fistula: A rare complication of cholangiocarcinoma. The Japanese Society of Internal Medicine. Intern Med. 2011;50:443-6.
2. Arshad R, Suhail K, Sartaj HS, Farzana K. External biliary fistula presenting as anterior abdominal wall abscess. Applied Medical Research. 2015;1(3):122-3.
3. Reed MWR, Tweedie JH. Spontaneous simultaneous internal and external biliary fistulae. British Journal of Surgery. 1985;72:538.
4. Sibakoti YC, Basnet RB, Poudel R. Spontaneous cholecystocutaneous fistula: A case report. Post-Graduate Medical Journal of NAMS. 2012;12(1):51-4.
5. Abderrhiam AD. Spontaneous cholecystocutaneous fistula. Sudan Medical Journal. 2009;45(1):64-6.
6. Cherry EPK, John G, Brian JD. Cholecystocutaneous fistula. Medscape. 2013.

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