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CASE REPORT

Acute obstructive jaundice and hematochezia in a 3-year-old child: Attention for massive hemobilia

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ABSTRACT

Massive hemobilia, an acute life-threatening condition, remains a diagnostic challenge in pediatric patients due to its rarity. In this study, the difficulties encountered in the diagnosis and treatment of acute massive hemobilia and acute obstructive jaundice in a 3-year-old boy are discussed in the light of the literature.

Keywords: Child, gastric heterotopia, hemobilia, intestinal bleeding, obstructive jaundice.

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INTRODUCTION

Hemobilia known as the blood loss from the hepatobiliary ducts may be caused by traumatic or non-traumatic reasons. Hemobilia is seen very rarely in childhood.[1,2] The causes of non-traumatic hemobilia reported in childhood include liver abscess, multiple aneurysms due to parasite infestations, vascular malformations, gastric ectopia, papillomatosis and gallbladder polyps, and von Willebrand's disease.[3–9] The causes of hemobilia seen in adult patients are generally non-traumatic; these include malignant or benign tumors of the hepatobiliary system, systemic lupus erythematosus, sarcoidosis, cholelithiasis, cholecystitis, choledochal cyst, gallbladder ulcer, pancreatitis, and warfarin treatment.[10–16]

Gastric heterotopia in the gallbladder is usually clinically silent and often only discovered incidentally. In here, a 3-year-old boy described who had been presented with acute massive hematochezia and obstructive jaundice was discussed the confounding issues that were encountered in the diagnosis and management of massive hemobilia with particular reference to the use of emergency procedures.

CASE REPORT

A 3-year-old male patient was admitted to the pediatric emergency department with complaints of hematemesis, melena, and anemia lasting for 2 days. In history, it was learned that he had been diagnosed with anemia 6 months ago and had intermittent abdominal pain for a year, and there was no history of trauma. The patient was consulted to the pediatric surgery due to abdominal pain. In his first clinical examination, it was determined that the patient was hemodynamically stable and had no other findings other than some pallor. In nasogastric decompression, gastric juice was clear, his stomach was tender, and dark green stools were found on rectal examination, but he was using oral iron replacement due to anemia. The patient had normocytic and normochromic anemia with hemoglobin (Hb) 6.56 g/dL and hematocrit (Hct) 19.5%; and total bilirubin 0.8 mg/dL, direct bilirubin 0.61 mg/dL, ALT 322 IU, AST 142 IU, Alk PO4 480 IU, GGT 261 IU, and others did not show any feature at admission. Abdominal ultrasound (US) revealed a swollen gallbladder and bile ducts and hepatic ducts consisting of biliary sludge. The patient was interned to the pediatrics department.

While no icterus was noted in the 1st day of admission, in the 2nd day, a tender gallbladder hydropic mass was palpable, and severe icterus and abdominal distention and abandon gastrointestinal hemorrhage through rectum were developed suddenly. The patient had severe hemorrhagic shock within the hour, hemoglobin level decreased to 5.19 gm/dL and Hct 14.9% although transfused of two units packed cells. The deep jaundice with significant elevation of bilirubin, ALT, AST, alkaline phosphatase, and amylase was developed which rapidly improved after 24 h in admission. All laboratory tests were elevated as total bilirubin 17,05 mg/dl, direct bilirubin 13,66 mg/dl, ALT 454 IU, AST 303 IU, alkaline PO4 772 IU, and GGT 484 IU. Abdominal US and Doppler US showed a huge distended gallbladder and biliary tract dilatation with biliary sludge increased further by day before (Fig. 1). However subsequent to patient hospitalization and replacement of blood, gastric juice drainage from nasogastric tube, deep occlusive jaundice, hydrops of the gallbladder, and abandon rectal fresh bleeding with clots were developed (Fig. 2). Although

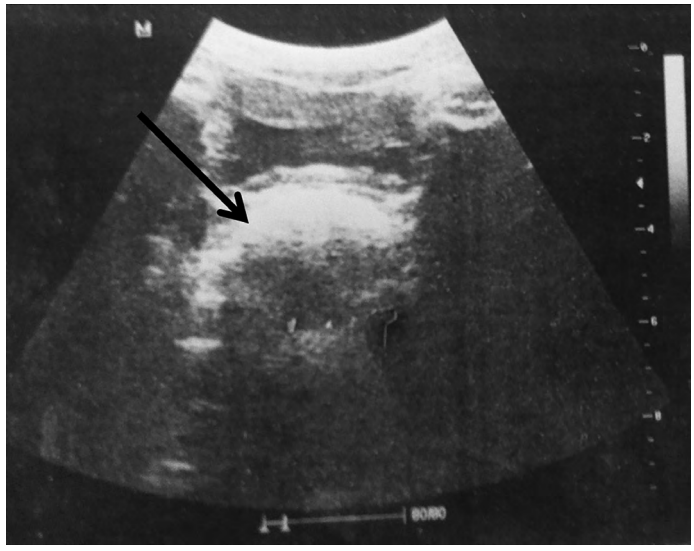

Figure 1: Abdominal ultrasound and Doppler US showed a huge distended gallbladder and biliary tract dilatation with biliary sludge.

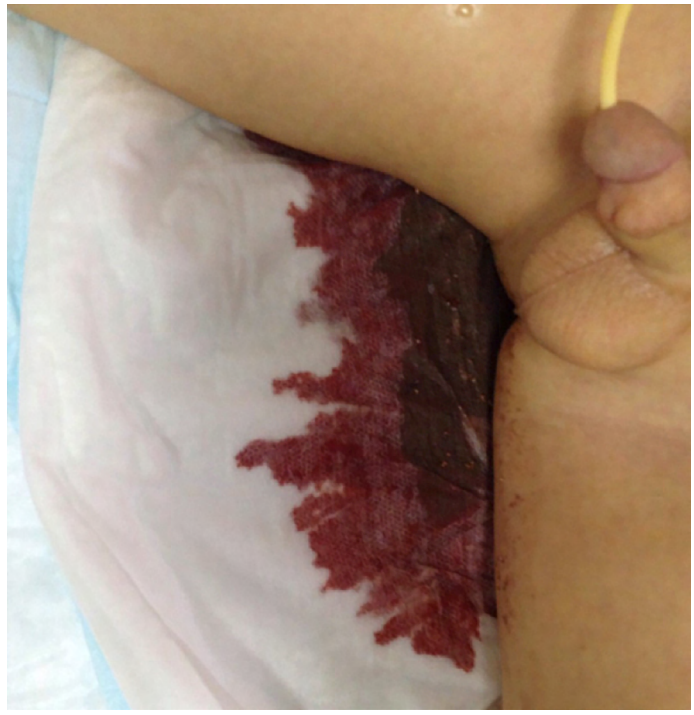

Figure 2: Abandon rectal fresh bleeding with clots.

massive blood transfusions (four units of packed cells), his hemoglobin level decreased quickly, the patient continued to have increased of lower gastrointestinal fresh bleeding. To perform an emergency, laparotomy was decided. Two units of packed cells, Vitamin K, and commenced on antibiotics were transfused during the half an hour before the laparotomy in the operating room. The patient achieved hemodynamically instable with a blood pressure 122/72 mmHg and pulse rate 155/min. A hard tender mass was palpable in the right upper quadrant and he had severe abdominal distension on the operating table; there was fresh blood and clots on digital rectal examination; Hb levels were 5.4 g/dL; and liver function tests deranged.

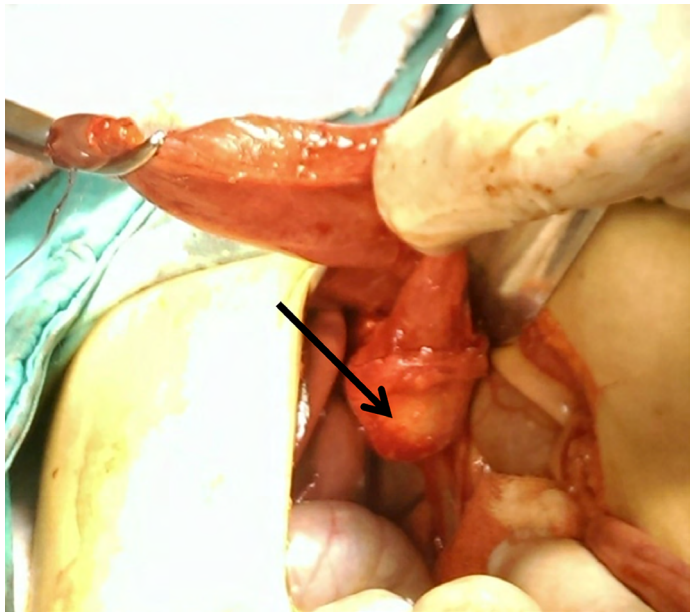

Figure 3: An thickened tissue (similar like Calot's ganglion) impacted at the corner of the cystic duct junction with gallbladder was seen during the exploration.

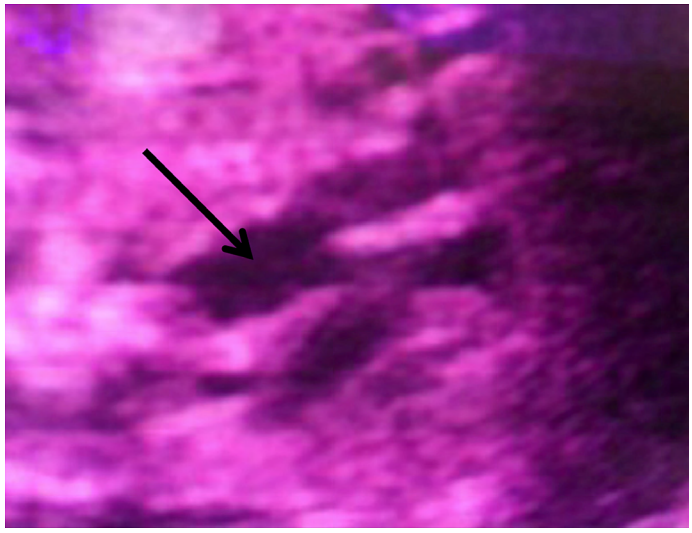

Figure 4: Intraoperatively, liver US was revealed biliary tract dilatation as same with pre-operative US, and any pseudoaneurysm of the liver was excluded.

Emergency laparotomy was performed through right upper subcostal incision. An exploration of porta hepatis revealed dense adhesions and edema of Calot's triangle, liver was normal color in full view, gallbladder and common bile duct were distended to 3 cm diameters with blood clots severely. An thickened tissue of the gallbladder impacted at the corner of the cystic duct junction with gallbladder was seen (Fig. 3). After removing clothes from the tip of the gallbladder, a thickened wall tissue in infundibulum of the gallbladder wall was palpated and gallbladder excised totally. Due to insufficiency from transcystic irrigation of common bile duct, an oblique incision in anterior wall of the common bile duct underwent. The common bile duct had dilated with a blood clot approximately 3×5 cm like a fusiform choledocal cyst, and the wall of common bile duct had thickened as 3–4 mm due to severe edematous inflammation. After removing the blood clot from the common bile duct, an arterial abandon bleeding from inner layer wall of the common bile duct was happened. A spurting arterial tip was ligated in the inner layer of the choledocus. Intraoperative liver US was revealed biliary tract dilatation of intrahepatic as same with pre-operative US, and any pseudoaneurysm of the liver was excluded from the study (Fig. 4). Luminal clearance control performed with a catheter advancing from the common bile duct toward the duodenum. Incision of the common bile duct was sutured with 6/0 PDS-II interrupted sutures as oblique without biliary t-tube and/or intra-abdominal drainage tube. Operation was completed under the intravenous bolus infusions of the blood products (as two units of packed cells and two units of fresh frozen plasma) during 2 h operation.

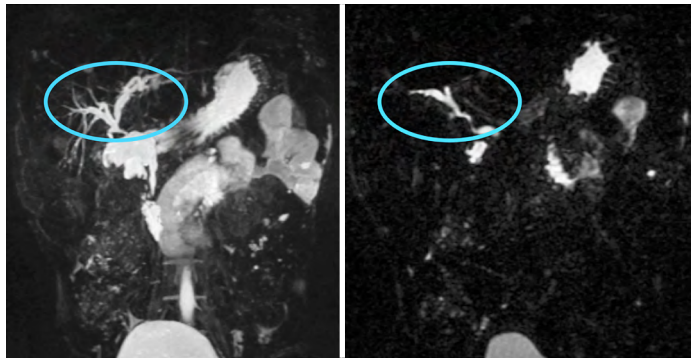

Figure 5: Computed tomography cholangiography and magnetic resonance imaging angiography at post-operative 8th days were normal demonstrated free flow in the hepatic artery, and mild dilatation in the biliary three and common bile duct.

Hemodynamic stable of patient was supplied entirely after 4 h later from the operation, and deep jaundice and bilirubin levels decreased rapidly. The patient condition improved after 24 h and his all other biochemical parameters normalized within 2 weeks. Post-operative management included prophylactic antibiotics, infusion, and electrolytes also. There was no leakage around the choledocus; and 1 week later, the patient was tolerated full oral enteral feeding. Computed tomography angiography and MR cholangiopancreatography revealed demonstrating normal hepatic artery and branches; and slightly decreased dilation was still ongoing in the biliary three and common bile duct on post-operative 8 days; and the patient was discharged home at same day (Fig. 5a, b). Histopathologic examination of the gallbladder was reported features suggestive of acute cholecystitis, and an area of ectopic gastric tissue was also seen at the site of gallbladder wall (Fig. 6). At the 8th week postoperatively, technetium 99m-pertechnetate scintigraphy demonstrated no evidence of gastric heterotopia elsewhere in the patient's body. The child was asymptomatic without any further episode of bleeding or jaundice or anemia in early and late follow-ups; and his laboratory investigations were unremarkable in 6 years follows-up.

DISCUSSION

Massive gastrointestinal bleeding is an important emergency medical and/or surgical problem of childhood. In gastrointestinal bleeding, it is necessary to plan the treatment for the cause; therefore, differential diagnosis of gastrointestinal bleeding is very important. Since

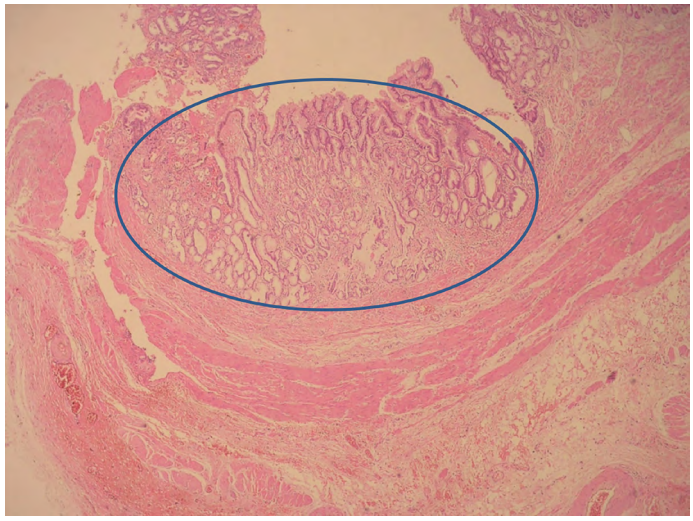

Figure 6: Histopathologic examination of the gallbladder showed features suggestive of acute cholecystitis, and an area of ectopic gastric tissue was also seen at the site of gallbladder wall (H&E ×100).

hemobilia, which is one of the upper gastrointestinal bleedings, is very rare, awareness of the causes and management of hemobilia allows patients to be treated more effectively.[17]

There is an abnormal vascular communication between the splanchnic and biliary systems in hemobilia. Hemobilia usually develops secondary to hepatobiliary trauma; it may be iatrogenic after hepatobiliary system instrumentation or may occur incidentally.[18–20] Rarely, hemobilia has been reported in cystic artery pseudoaneurysm secondary to trauma.[21]

Spontaneous fistula development between the cystic artery and gallbladder without trauma or surgical intervention is a very rare condition, infections and inflammations being the two most common causes of non-traumatic hemobilia. Lysosomal disorders such as leukodystrophy, worm infestations, vasculitic syndromes, and overdose of anticoagulant medication have been reported as other causes of hemobilia.[8,22–25] Non-traumatic hemobilia is a very rare cause of upper gastrointestinal bleeding in childhood. In the history of our patient, although there was no history of trauma, it was learned that he played by lying on a plate ball for about a year. Maybe the pressure of the ball on the liver may have caused microtraumas.

The presence of jaundice with hematochezia should raise the suspicion of hemobilia. In addition, coagulopathy should be excluded in all patients with suspected hemobilia, and full parasite screening should be performed.[5,9] The development of sudden obstructive jaundice after excessive rectal bleeding suggested that our patient may have hemobilia. Thus, the rapid development of obstructive jaundice and excessive rectal bleeding facilitated the rapid diagnosis of hemobilia in our patient.

In a case report with spontaneous cystic artery bleeding after erosion of gallbladder ulcer to cystic artery due to ectopic stomach tissues, the presence of ulcer could not be demonstrated.[16] It has been reported that excessive acid secretion by ectopic stomach tissue in the biliary tract may cause inflammation of the perivascular tissue and weakening of the vessel walls. The emergence of inflammatory processes around the vessel, causing ulcer formation from the ectopic gastric mucosa and resulting in hemobilia or rarely ruptures into the peritoneal cavity; and it has also been reported that this condition can be detected incidentally in radiological imaging studies.[16,26,27]

Gastric heterotopia is a developmental anomaly and heterotopic stomach tissues can be seen anywhere along the gastrointestinal tract, from the oral cavity to the rectum. It can also be found anywhere in the hepatobiliary system, including the gallbladder, cystic duct, common hepatic duct, common bile duct, and ampulla Vater.[28,29] Due to the proximity of embryonic endodermal cells, congenital displacement of the gastric mucosa to the hepatobiliary system is possible. Gastric progenitor cells in the gallbladder are assumed to cause gastric heterotopia.[30–35] The reason why ulceration and bleeding from heterotopic gastric mucosa in the biliary system are rare is attributed to alkalinity of bile.[32]

Gallbladder abnormalities in children can cause abdominal pain and should be included in the differential diagnosis of abdominal pain. The classic triad of the right upper abdominal pain, jaundice and gastrointestinal bleeding, which is pathognomonic for hemobilia, is reported to be seen only in 22% of adult patients.[1–19] In children, hemobilia may not always present with the classic triad of biliary colic.[3,21] This pathognomonic triad of clinical manifestations is observed very rarely, especially in children with gastric heterotopia of the gallbladder. As in our patient, it may also present with findings such as intestinal bleeding and obstructive jaundice.

The number of heterotopic gastric mucosal hemobilia cases reported to date in the literature is very few.[33] Case reports of heterotopic gastric mucosa in the cystic duct or gallbladder associated with cholelithiasis/cholecystitis in childhood are particularly rare.[30–35] Gastric heterotopia of the gallbladder was first described in 1934. In 1977, gastric heterotopia in calculous cholecystitis was detected in a 7-year-old boy, this is the first reported case of pediatric gallbladder stomach heterotopia.[30] To date, a few cases of gallbladder gastric heterotopia causing bleeding in children have been reported. The youngest of these cases is a 6-year-old boy who presented with hematemesis and melena. In this case, perforation of the gallbladder from the gastric heterotopia bed was reported.[36] Our case is quite unique as the youngest child reported with hemobilia in the English literature, with abdominal pain and signs of acute fresh sudden hematochezia and obstructive jaundice.

Gallbladder gastric heterotopia has been reported to be diagnosed incidentally, most often when it causes gallbladder obstruction, inflammation, or perforation.[28–30] They occur in patients with hepatobiliary gastric heterotopia when complications such as mucosal ulceration, bleeding, and biliary obstruction develop.[29] In the few studies of the literature reporting gastric heterotopia in the gallbladder, it is reported that the diagnosis was made incidentally in three asymptomatic children.[35] Jaundice episodes have been described several times in young men with gastric heterotopia in the gallbladder, however, in a young child like our case, a discharge from gastric heterotopic tissue and consequent obstructive jaundice was never described as a complication of gastric heterotopia of the gallbladder.

In this case report, gastric heterotopia and arteriobiliary fistula and massive hematobilia perforated from the infundibulum to the gallbladder are described. Although heterotopic gastric tissue in the gallbladder has been reported in young men, no case of gastric heterotopia in

the extrahepatic bile ducts has been reported before in the literature; and also, bleeding from the common bile duct was never described as a complication of gastric heterotopia of the gallbladder. If the child had massive hemobilia and acute obstructive jaundice due to gastric heterotopia in the gallbladder like in our case, emergency surgical exploration might be a diagnostic and therapeutic and lifesaving options.

CONCLUSION

A high index of suspicion and the use of appropriate diagnostic tools can help diagnose and treat of gastric heterotopia as this rare cause of hemobilia. Massive hemobilia may present as acute life-threatening hemorrhage even in the subset of non-traumatic hemobilia. A history of melena may be profuse or minimal, intermittent, or continuous; and/or especially stool color may be confused with stool in cases where oral iron medication is used, as in our case. Hemobilia should be included in the differential diagnosis in patients with other hepatobiliary diseases presenting with gastrointestinal bleeding.

Statement

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – AC; Design – AC; Supervision – AC; Materials – FY, HA, HY; Data Collection and/or Processing – NİÖ; Analysis and/or Interpretation – NİÖ, SM; Literature Search – NİÖ, HA, HY; Writing – NİÖ, AC; Critical Reviews – AC.

Conflict of Interest: The authors have no conflict of interest to declare.

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