

Nontraumatic Hemobilia in Children

Anjana Bairagi¹ Daniel C. Aronson¹

¹ Department of Paediatric Surgery, Inkosi Albert Luthuli Central Hospital, University of Kwazulu-Natal, Durban, South Africa

Address for correspondence Daniel C. Aronson, Department of Paediatric Surgery, Inkosi Albert Luthuli Central Hospital, University of Kwazulu-Natal, Private Bag X03, Mayville 4058, Durban, South Africa (e-mail: aronson.dc@hotmail.com).

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Abstract

Keywords

- ▶ nontraumatic
- ▶ hemobilia
- ▶ children

Nontraumatic hemobilia is a rare cause of upper gastrointestinal hemorrhage in children. In the developing world, infections and inflammation are the two most common causes. Two patients are presented illustrating the diagnostic difficulties. Following recognition of the site of bleeding the surgery was successful in each case. After a review of the literature, a diagnostic workup is proposed.

Introduction

Hemobilia is a rare cause of upper gastrointestinal bleeding in children and usually follows trauma to the hepatobiliary-pancreatic system. Hemobilia was first described in the 17th century by Francis Glisson. He described his findings of a fatal hemorrhage from the biliary tree secondary to a traumatic liver laceration. In 1948, Sandblom was the first to define hemobilia as hemorrhage arising from trauma in the biliary tract. There is a paucity of data regarding nontraumatic hemobilia, especially in children. The most common etiologies reported in adults are infections, infestations, or neoplasms. The classic triad of upper abdominal pain, melena, and jaundice (Quinke, 1871) was reported in just 22% of adult patients.¹ The advent of endoscopy and interventional radiology raises the possibility of noninvasive management. Due to its rarity, this condition continues to be a diagnostic challenge in pediatric surgical patients. We present two patients with nontraumatic hemobilia, a review of the current literature, and propose a diagnostic algorithm.

Case 1

A 4-year-old female was referred with a weeks' history of hematemesis and passing melena stools. She was transfused a unit of packed cells at base hospital before referral. The patient had a history of similar vomiting a year ago and also complained of intermittent abdominal pain since the age of 1 year. There was no history of trauma. On clinical examination, the patient was found to be hemodynamically stable with some pallor and no other contributory findings. The

patient had a normocytic, normochromic anemia of hemoglobin (Hb) 7.1 g/dL (normal 10.7–13.1 g/dL) and hematocrit of 21.8%. The remaining laboratory investigations were unremarkable. An upper gastrointestinal endoscopy revealed a normal esophagus, stomach, and D1 (duodenum). At D2 (duodenum), blood was noted at the ampulla of Vater and at the floor of the duodenum (▶Fig. 1). At abdominal ultrasound, a 4.8 × 4.2 cm heterogeneous mass in the midline with avid color uptake (suggestive of a vascular mass) and mixed flow was found. Computed tomography angiography (CTA) findings were suggestive of a large pseudoaneurysm arising from the splenic artery with an associated thrombus (▶Fig. 2). The patient was then taken to theater and an exploratory laparotomy was performed. Intraoperative findings were that of a large pancreatic cyst, closely adhered to the posterior wall of the stomach, and involving most of the pancreas except for the tail and a rim of the pancreatic head. The posterior wall of the cyst was closely adhering to the superior mesenteric vein and portal vein. Thus the cyst was deroofed (▶Fig. 3) and an extended distal pancreatectomy performed. The splenic artery was ligated and spleen was found to be grossly normal. Postoperative management included prophylactic antibiotics, somatostatin infusion, and total parenteral nutrition. A week later, the patient was tolerating full diet and was discharged home. At her 4 month outpatient clinic review, the patient was doing well with no signs of exocrine or endocrine pancreatic insufficiency. The final histology results were: pancreatic tissue with inflammation, hemosiderin, fibrosis, and granulation. These features are in keeping with a pancreatic pseudocyst.

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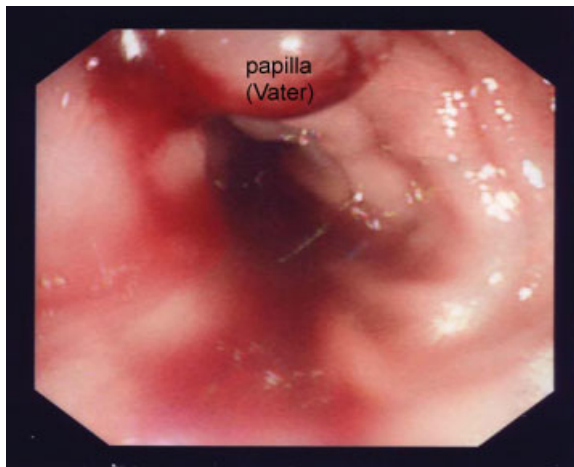


Fig. 1 Hemobilia at endoscopy. Note the blood at ampulla of Vater.



Fig. 2 CTA showing a large pseudoaneurysm arising from the splenic artery with an associated thrombus. ca, celiac axis; CTA, computed tomography angiography.

Case 2

An 11-year-old female was referred with 1 month history of jaundice, hematemesis, and melena (~1 L) over 2 days. The patient had no travel history, and a negative malaria screen. A diagnostic ultrasound done at base hospital showed an abnormal liver echo structure with dilated intrahepatic ducts and portal vein. The CT scan showed an enlarged gallbladder, and an irregular biliary tree dilatation. Upper endoscopy findings were that of bleeding from the ampulla of Vater. The patient had anemia of Hb 4.2 g/dL (normal 11.5–14.5 g/dL) and international normalized ratio 1.33. Before transfer for further management, the patient was transfused two units of blood, given freeze dried plasma, vitamin K, and commenced on antibiotics. The patient arrived hemodynamically stable with a blood pressure 122/72 mm Hg, pulse rate 102/min. A soft, nontender mass was palpable in the right upper quadrant. There was no melena on digital rectal examination. The patient



Fig. 3 The deroofed cyst containing thrombus.

arrived with Hb 9.7 g/dL and deranged liver function tests. Diagnostic CTA demonstrated multiple saccular aneurysms of the right hepatic artery, dilated intrahepatic ducts up to the common hepatic duct, a 3.1 × 1.8 cm hyper dense mass in the porta hepatis with mass effect on the proximal common bile duct (CBD) and cystic duct and lastly, a grossly dilated gallbladder containing sludge. We proceeded to perform an exploration of this porta hepatis mass. A much enlarged gallbladder was encountered which was removed via a retrograde cholecystectomy. The CBD was obstructed, dilated, and draped over a mass thought to be a right hepatic artery aneurysm. The right hepatic artery was ligated, the mass deroofed and its posterior wall left in situ as it was densely adherent to the portal vein. The CBD was excised and the distal CBD ligated flush with the superior part of the duodenum. A standard hepaticojejunostomy and enteroenterostomy was performed to restore bowel continuity. The patient had an uneventful postoperative recovery and on day 6 was discharged to base hospital. Final histology results confirmed a choledochal cyst with associated inflammatory changes and a blood clot-filled gallbladder with mild wall edema.

Discussion

Hemobilia arises when there is a fistula between the splanchnic circulation and the biliary tree. Bleeding may originate from the hepatic circulation, the vessels of the hepatoduodenal ligament, the extra hepatic biliary tree, the gallbladder, and the pancreas.

Nontraumatic hemobilia is rare in children and many patients do not present with the classic triad of biliary colic, obstructive jaundice, and intestinal bleeding.²

The most commonly reported causes of nontraumatic hemobilia in children are infection (liver abscess,^{3,4} parasitic infestation [ascariasis],^{5,6} anatomical abnormalities [gastric duplication⁷], biliary pathology [papillomatosis of the gallbladder,⁸ gallbladder polyps⁹) and bleeding disorders (von Willebrand disease).¹⁰ Other documented nontraumatic causes that have been described in adults include tumors (malignant^{11,12} or benign¹³), systemic lupus erythematosus,¹⁴ sarcoidosis,¹⁵ biliary pathology (cholelithiasis,

Table 1 Differences of the etiology of hemobilia in children and adults as described in the literature

Etiology
<i>Children</i>
<i>Ascaris lumbricoides</i> infestation
Liver abscess
Pancreatitis
Gastric duplication
Choledochal cyst
Gall bladder papillomatosis
Gall bladder polyps
von Willebrand disease
<i>Adults</i>
Tumors (malignant/benign)
Systemic lupus erythematosus
Sarcoidosis
Cholecystitis
Cholelithiasis
Gall bladder ulcer
Pancreatitis
Warfarin therapy

cholecystitis, choledochal cyst,¹⁶ gallbladder ulcer¹⁷), pancreatitis, and warfarin therapy (→Table 1).

Patients may present with symptoms of an upper gastrointestinal bleed in a chronic or an acute manner. The history of melena may be profuse or minimal, intermittent or continuous. Rapid hemobilia may present as acute life-threatening hemorrhage even in the subset of nontraumatic hemobilia. Parasitic infestations, endemic to our environment, followed by inflammation are the more common etiology.¹⁸ One should obtain a comprehensive history including patient’s previous admission, and recent travel. Hemosuccus pancreaticus (HP), sometimes called “wirsungorrhagia,” is defined as upper gastrointestinal hemorrhage from the papilla of Vater via the pancreatic duct.¹⁹ HP most commonly occurs in males aged 50 to 60 years and is associated with pseudoaneurysm formation secondary to chronic pancreatitis.¹⁹ This pseudoaneurysm communicates with the pancreatic duct through a pancreatic pseudocyst or a fistula secondary to the destruction of its walls by enzymatic injury and compression. Subsequent rupture of the pseudoaneurysm results in HP. This diagnosis is uncommon, if not rare in the pediatric population.²⁰ Pseudoaneurysm formation is a known complication of chronic pancreatitis (pancreatic cyst) as described in the first patient.²⁰ This can also be a complication of a choledochal cyst as per second described patient. While both hemobilia and HP may overlap in terms of definition and findings on diagnostic endoscopy one should bear them in mind when faced with such patients.

Hydatid infections of the liver caused by *Echinococcus granulosum*, result in inflammation of perivascular tissue and weakening of vessel walls. The subsequent pseudoaneurysm formation may rupture into the biliary tree and result in hemobilia.²¹ The most common nematode and trematodes to infect the liver are *Ascariasis lumbricoides*, *Clonoris sinensis*, and *Fasciola hepaticum*. Biliary ascariasis with associated intrahepatic pseudoaneurysm following antihelminthic treatment has been described by our institution.¹⁸ Although, exceedingly rare, the development of an ascariasis-related pseudoaneurysm was presumed after inflammatory involvement of the artery by a contiguous liver abscess that may rupture into the biliary tree.^{18,21}

When hemobilia is suspected one should exclude coagulopathy and should complete a parasitic screen. Sonographic demonstration of the “four-line sign” (nonshadowing echogenic strips with central anechoic tube representing the parasites’ digestive tract) is pathognomonic of biliary ascariasis.¹⁸ Ultrasound may also be helpful in identifying other causes of hemobilia, for example, liver abscess. When the patient is hemodynamically stable upper endoscopy is necessary to identify blood at the ampulla of Vater confirming a diagnosis of hemobilia and excluding more common causes of upper gastrointestinal bleeding. Where available, CTA may identify the source of the hemorrhage and assist in planning the management of these patients.

→Fig. 4 shows a proposed diagnostic algorithm. The advent of interventional radiology has contributed successfully to the nonsurgical management of many of these patients but in small children the size of the vasculature may limit the options for cannulation and subsequent instrumentation.

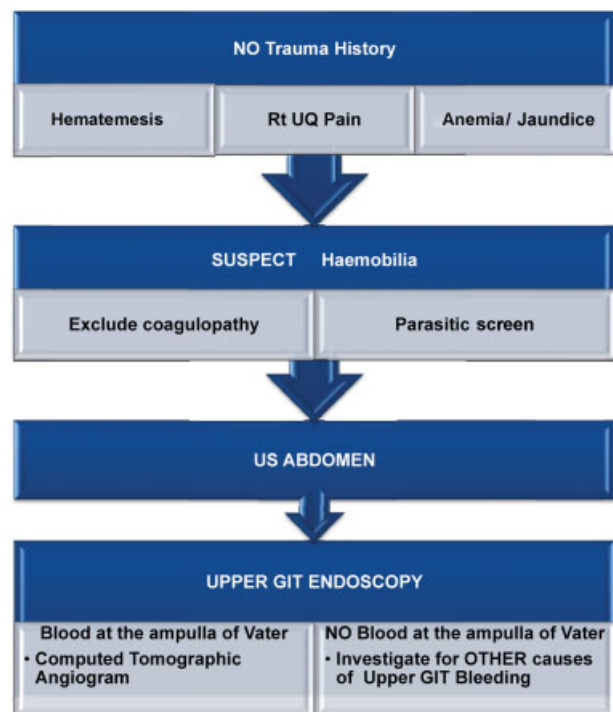


Fig. 4 Diagnostic approach for suspected non traumatic hemobilia.

Conclusion

Nontraumatic hemobilia is a rare cause of upper gastrointestinal hemorrhage in children. A systematic approach as proposed above, should ensure timely diagnosis, and guide appropriate management of these patients.

Conflict of Interest

None.

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