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HEMOBILIA FROM A RUPTURED HEPATIC ARTERY ANEURYSM IN A 16-YEAR-OLD GIRL*

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The hepatic artery is the second most common site of visceral artery aneurysm, accounting for about one-fifth of reported cases[1]. Even though selective celiac arteriography is the most useful diagnostic procedure, most hepatic artery aneurysms are not discovered until a laparotomy has been performed. The diagnosis may be suggested by the triad of biliary colic, jaundice and gastrointestinal bleeding. The majority of patients are over fifty years of age, but some cases have been described at both age extremes[2].

Of the cases reported, infectious diseases, and atheromatous and fibromuscular changes have been responsible for the pathogenesis of hepatic artery aneurysm.

In this article, we present a case of hemobilia from a ruptured hepatic artery aneurysm due to fibromuscular dysplasia which occurred in a patient who was only sixteen years of age.

Case Report

A sixteen-year-old female was admitted to Karadeniz University Hospital with a one-week history of right upper quadrant pain, nausea, vomiting and melena which occurred twice a day. There was also a history of dark urine and icteric sclera of several days' duration. There had been no recent history of systemic infection or trauma. On admission the blood pressure was 110/80 mm Hg, pulse rate 140/min, hemoglobin level 6.4 g/dl, serum total bilirubin 3.5 mg/dl, indirect bilirubin 3 mg/dl, alkaline phosphatase 40 Smogy units, and SGOT 100 mU/ml. The only positive sign was a thrill on the right upper quadrant of the abdomen. She was given 2000 cc of blood during the night to maintain her vital signs within normal limits. Gastroscopy revealed no abnormality. During this course of time hematemesis commenced and the cramping discomfort in the right upper quadrant and epigastrium continued. An emergency laparotomy was performed.

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The stomach, the entire small bowel and colon contained blood. During surgery, a pulsating mass was found in the region of the hepatoduodenal ligament. The mass was presumed to be an aneurysm of the hepatic artery. A choledochotomy was then performed and active arterial bleeding was encountered coming through a 1-2 cm communication of the aneurysm and the posterior wall of the common hepatic duct, which had small multiple perforations. The common hepatic artery was ligated and bleeding in the common bile duct instantly ceased. At surgery the gallbladder was edematous and grossly distended with blood; a cholecystectomy was performed. The gallbladder contained no calculi and the specimen revealed acute hemorrhagic cholecystitis. A T-tube was placed in the common bile duct and a penrose drain was placed in Winslow's foramen; the abdominal wound was closed in layers. During the operation four units of whole blood were administered to maintain the patient's vital signs within normal limits. T-tube drainage was clear postoperatively and a T-tube cholangiography was taken on the fifth day which illustrated the aneurysmal sac being filled from the common duct (Fig. 1).

Fig 1: T-tube cholangiography demonstrating the aneurysmal sac being filled from the common bile duct.

On the ninth postoperative-day the subject again experienced crampy right upper quadrant pain, nausea and arterial bleeding through the T-tube. Physical examination revealed nothing abnormal. The hematocrit reading was 34 percent, SGOT 120 mU/ml and serum biluribin 2.5 mg/dl. A laparotomy was performed again. When the T-tube was removed, arterial bleeding was encountered coming through the communication between the aneurysm and the common hepatic duct. The aneurysmal sac was seen and an erosion into the common hepatic duct was identified. The aneurysm was presumed to be an aneurysm of the right hepatic artery taking a retrograde blood supply from the gastroduodenal artery. The gastroduodenal artery was ligated. The bleeding stopped immediately. A biopsy specimen was taken from the communication between the aneurysmal sac and the common hepatic duct. The defect in the duct was then closed by using continuous 5-0 arterial sutures and a T-tube was inserted in the common bile duct without obstructing the flow of bile through the duct; the abdominal incision was closed in layers. Histological examination of the specimen revealed an aneurysm of the hepatic artery due to fibromuscular dysplasia (Fig.2). During

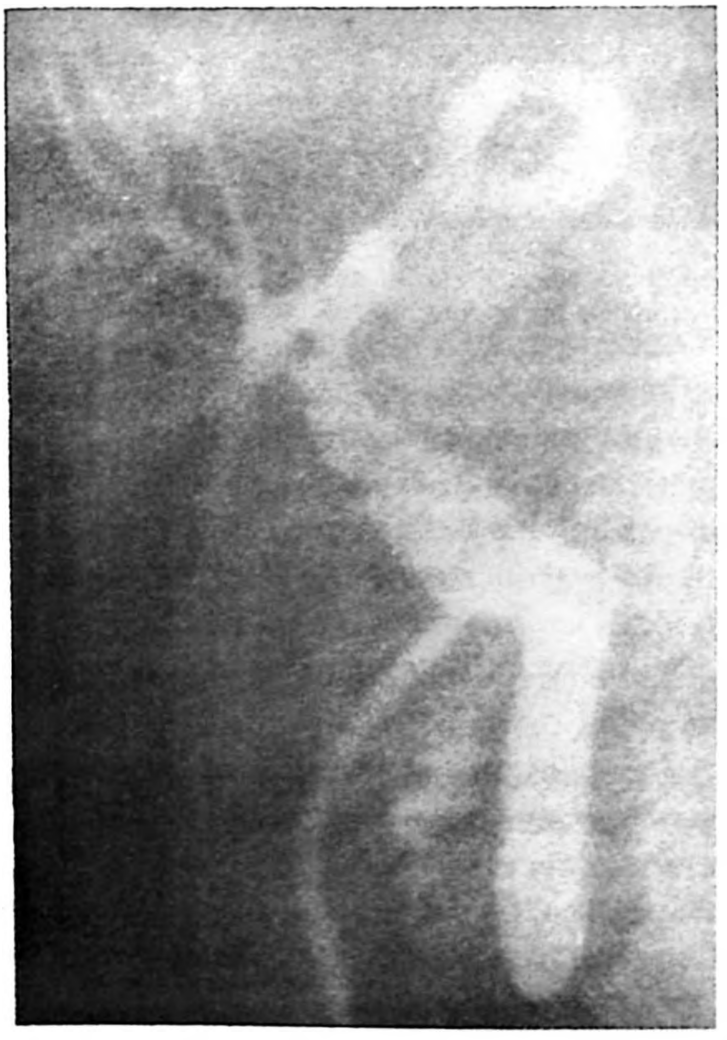

Fig 2: Spaces not having endothelial linings and areas containing fibrinoid necrosis infiltrated by mononuclear cells (HE × 12.5).

the postoperative course, slight hemorrhagic T-tube drainage was observed during the first two weeks and two units of whole blood were administered during this period. Three days following the second laparotomy, a T-tube cholangiography demonstrated no intraductal filling defects (Fig. 3), and therefore the T-tube was removed. The patient was discharged two days later.

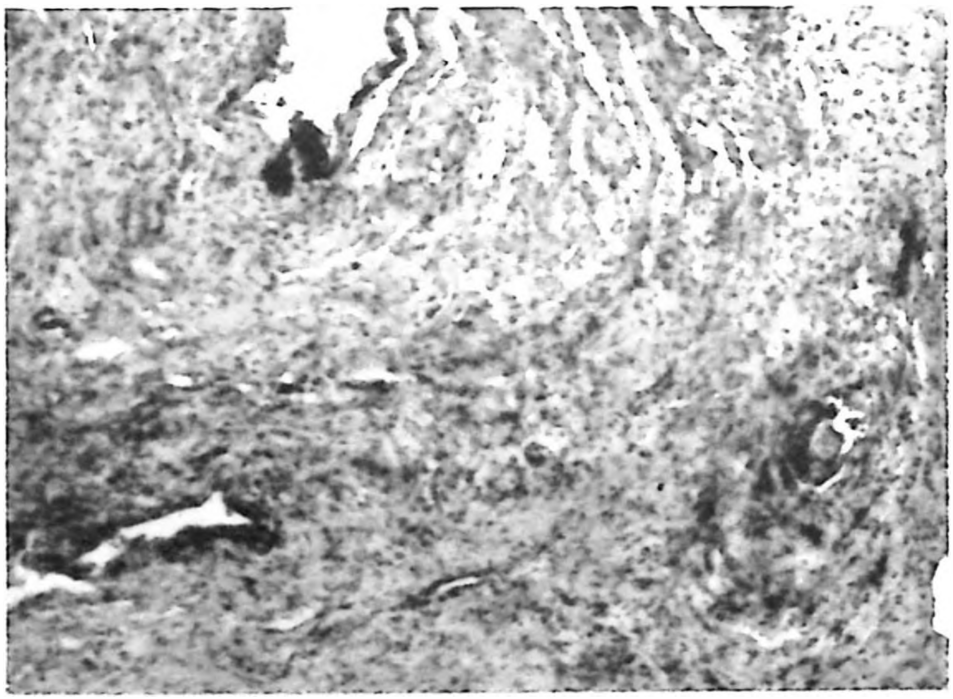

Fig 3: T-tube cholangiography after the fistulous opening was closed showing no aneurysmal sac or filling defects.

The patient did well until three months after the operation when she was readmitted to the hospital because of a two-day history of melena which occurred twice a day. On admission, her vital signs were within normal limits. The hemoglobin level was 10.5 g/dl, serum bilirubin 0.8 mg/dl, SGOT 40 mU/ml. Melena was found on rectal examination. Digital subtraction angiography showed good backflow to the liver (Fig. 4). The patient was discharged fifteen days after admission.

Three months after her last discharge from the hospital, the patient was again readmitted because of hematemesis and melena. Physical examination was normal. The hematocrit reading was 33%, hemoglobin level 8.8 g/dl, SGOT 100 mU/ml, and serum bilirubin 0.8 mg/dl. She was given two units of whole blood and was discharged a week after her admission. The patient was followed up biannually for three years postoperatively and there were no symptoms referable to either the gastrointestinal or biliary systems; her liver function tests were completely normal.

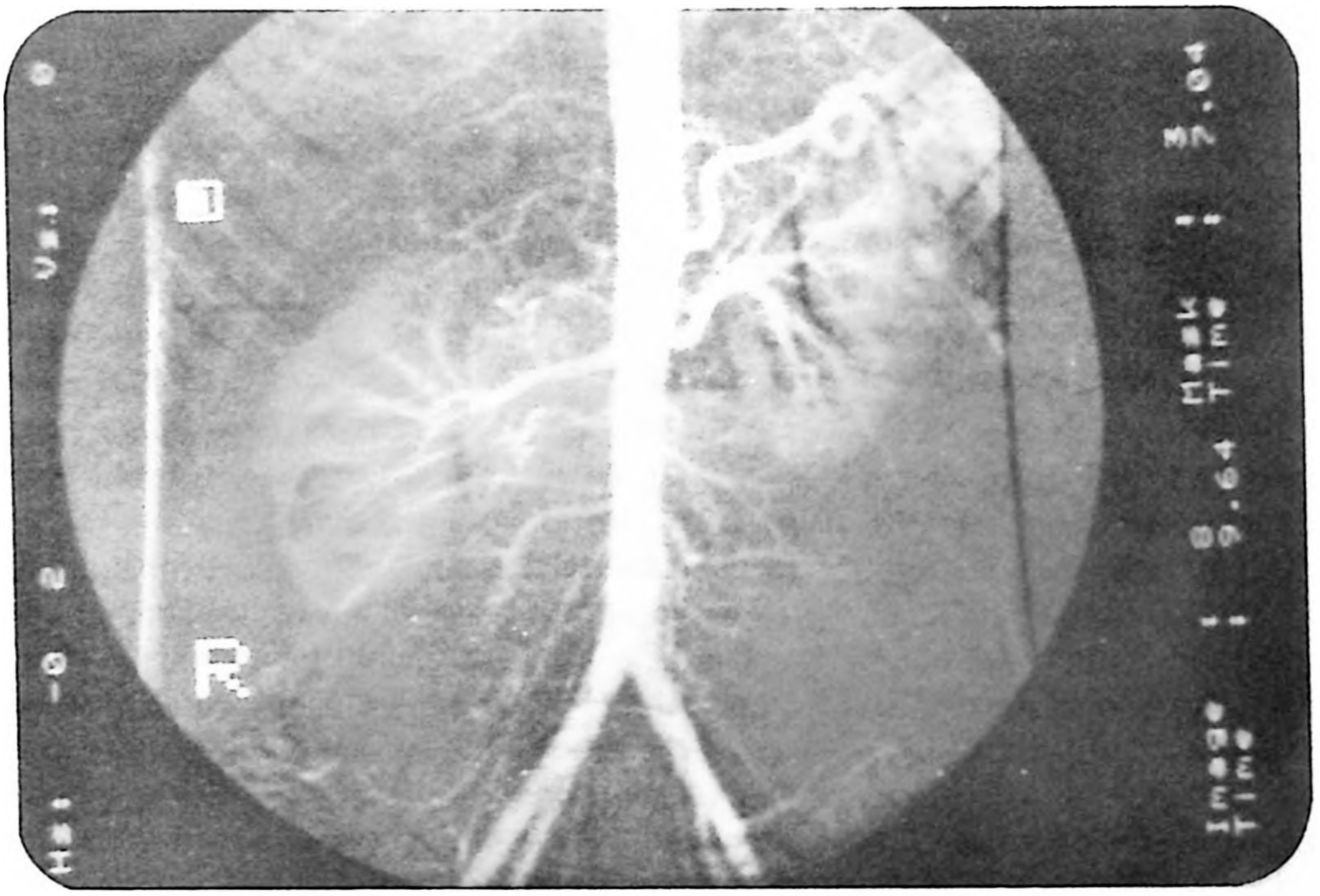

Fig 4: Digital subtraction angiography taken three years after surgery, demonstrating good backflow to the liver.

Discussion

The most common cause of hemobilia is vascular disease and accounts for 55 percent of the 355 fully documented cases reviewed by Sandblom[3]. The most common vascular lesion causing hemobilia is an aneurysm of the hepatic artery. Hemorrhage from the rupture of an aneurysm of the hepatic artery into the common bile duct was first reported by Jackson[4]. Two-thirds of hepatic artery aneurysms are located in the common hepatic artery, with most of the remaining ones being located in the right hepatic artery[5].

In a review of reported cases only 16 percent of hepatic artery aneurysms had an infectious etiology[2]. Atheromatous changes are found in one-half of the hepatic artery aneurysms[6]. Fibrodysplastic changes are found in about one-fourth of the cases[1]. Fibromuscular dysplasia may involve a wide variety of arteries, sometimes in a multicentric fashion[7]. This condition normally becomes manifest during the third or fourth decade of life, although it can also be seen in children[8]. In our case, the microscopic examination of the specimen being taken from the communication between the hepatic artery aneurysm and the common hepatic duct could support a diagnosis of fibromuscular dysplasia. Figure 3 shows some areas which did not exhibit endothelial linings but contained a few erythrocytes in

their lumens and were surrounded by some groups of cells arranged in a concentric manner being parallel to each other, which probably were smooth muscle tissue. These areas were probably related to the defect between the aneurysmal sac and the common hepatic duct. Around these areas, the connective tissue was infiltrated by scattered mononuclear cells and a small focus of fibrinoid necrosis could be seen. Additionally, there were some ductal structures covered by prismatic epithelial cells which probably resembled the mucosal invagination of the biliary tract.

The classic triad of hemobilia is gastrointestinal bleeding, right upper quadrant or epigastric pain and jaundice[9]. The bleeding into the biliary tree secondary to a ruptured aneurysm may be slow, leading to melena and secondary anemia, or rapid, resulting in hematemesis and shock. In our case, it was granted that at her first presentation, the patient was suffering from shock, but at her second and third admittance, the patient exhibited no signs of shock, however, she was suffering from melena. When the hemorrhage flows from the gallbladder through the biliary tract, and if clotting prevents emptying, the distended gallbladder may be palpated[5]. Jaundice is caused by obstruction of the biliary tree resulting from blood clots or from compression by the aneurysm[10].

Generally, physical examination is not helpful in the diagnosis of hepatic artery aneurysm because of the small size of an expansive mass and the fact that bruit or thrill does not commonly accompany such lesions. However, in our case the patient's thrill was located on the upper quadrant of the abdomen. Although we did not palpate the gallbladder on physical examination, at surgery, the gallbladder was edematous and grossly distended with blood.

At present, the most useful diagnostic test is selective arteriography of the celiac axis, the hepatic artery or the superior mesenteric artery. Other methods include barium studies of the upper gastrointestinal tract, intravenous cholangiography, duodenoscopy with retrograde cholangiography, percutaneous splenoportography and scintigraphy of the liver[4,11]. Despite the advisability of preoperative arteriography most hepatic artery aneurysms have been discovered at the time of laparotomy. The aneurysm may be identified by palpation of hepatic vessels for dilation and thrills.

The first successful surgical treatment of a hepatic artery aneurysm which ruptured into the biliary tract was performed by Kehr[12], who treated such a patient by ligation of the hepatic artery in 1903. Since then, additional methods have been employed. Today, the surgical methods that have been used in cases of hemobilia secondary to hepatic artery aneurysm are ligation of the hepatic artery or one of its branches, excision of the aneurysm with or without anastomosis, obliterative or reconstrictive endoaneurysmorrhaphy and hepatic resection which have been used chiefly in cases of intrahepatic aneurysm.

The selection of any one method depends on the findings at the time of surgery. Despite concern about potential hepatic necrosis, hepatic arterial ligation on both sides of the aneurysm may be the safest treatment in certain individuals such as critically ill patients with either massive bleeding from the aneurysm or severe intraabdominal sepsis[13,14]. In our case, the patient was critically ill during the first operation, and we chose ligation of the common hepatic artery. After ligating the common hepatic artery the bleeding stopped instantly and considering the potential necrosis of the liver, we did not want to ligate the hepatic artery distal to the gastroduodenal artery. The feasibility of this approach under such adverse circumstances is supported by recent studies of hepatic artery ligation for liver tumor[15]. Hepatic artery ligation is usually well-tolerated[11] and under certain circumstances may be the safest method in treating hepatic artery aneurysms. Extensive collateral arteries to the liver were documented by the studies of Michels[16], and Redman and Reuter[17]. In our case, despite our ligating the common hepatic and the gastroduodenal artery, the patient was readmitted twice to the hospital because of melena and secondary anemia. We were of the opinion that the aneurysm still had enough flow from collateral vessels. But since in the later period, there were no signs or symptoms of hemobilia, it was considered to have been obliterated.

Aneurysmal rupture into adjacent structures necessitates their repair after managing the aneurysm. Erosion into the common bile duct may be treated by repair of the common duct along with T-tube drainage[18] by Roux-en-Y choledochojejunostomy or by choledochoduodenostomy[19]. We sutured the fistulous opening between the choledoch and aneurysmal sac with continuous 5-0 arterial sutures and performed T-tube drainage.

Digital subtraction angiography demonstrated that our patient's liver had a good backflow from the distal segment of the hepatic artery (Fig. 4). She was totally asymptomatic and her liver function tests including the studies of enzyme and coagulation factors were completely within normal limits.

Summary

A 16-year-old girl with hepatic artery aneurysm due to fibromuscular dysplasia which caused hemobilia is presented. This most unusual and life-threatening type of gastrointestinal bleeding was successfully treated by ligation of the common hepatic and gastroduodenal arteries. The clinical aspects and operative procedures of hepatic artery aneurysm are discussed.

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