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Hemobilia and cholangitis in hereditary hemorrhagic telangiectasia

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A 54-year-old woman presented with postprandial right upper quadrant pain. Her medical history included type 2 respiratory failure due to severe congenital scoliosis, home mechanical
ventilation with tracheotomy, right chronic empyema, complicated urinary tract infection, and hereditary hemorrhagic telangiectasia (HHT). Physical examination revealed stable vital signs and positive Murphy’s sign. Laboratory tests revealed white blood cells of 8,500/mm³, hemoglobin of 7.2 g/dL, C-reactive protein of 2.15 mg/dL (reference range, <0.14 mg/dL), total bilirubin of 0.7 mg/dL (0.4-1.5 mg/dL), aspartate aminotransferase of 44 U/L (13-30 U/L), alanine aminotransferase of 64 U/L (7-23 U/L), γ-glutamyl transpeptidase of 102 U/L (9-32 U/L), alkaline phosphatase of 751 U/L (106-322 U/L), and amylase of 146 U/L (44-132 U/L). As she had allergy for intravenous iodinated contrast material, plain computed tomography (CT) scan was performed, showing high density fluid, which was suggestive of blood, in the gall bladder (FIGURE A). With a suspicion of acute cholangitis, endoscopic retrograde cholangiopancreatography (ERCP) was performed, disclosing active hemobilia (FIGURE B). Intraductal ultrasonography showed high echogenic material, which was suggestive of blood, in the dilated common bile duct (FIGURE C). Although the presence of apparent hepatic or biliary telangiectasias and arteriovenous malformations (AVMs) could not be proved, acute cholangitis by hemobilia associated with HHT was clinically diagnosed. No lithiasis was present and a biliary plastic stent was placed, resulting in clinical improvement. Her symptoms recurred; however, then ERCP was repeated two weeks later. After the stent impacted with clot was removed, massive hemobilia was drained and no further intervention was added (FIGURE D). She had had recurrent hemobilia and spontaneous improvement until her death by respiratory failure two years later.

HHT, also known as Osler-Weber-Rendu disease, is an autosomal dominant inheritance, characterized by epistaxis, mucocutaneous telangiectasias, and visceral AVMs, which lead to chronic hemorrhage and anemia. Symptomatic hepatic and biliary AVMs occur very rarely. Hepatic involvement may result in high-output heart failure, portal hypertension due to hepatic artery to portal vein shunt, hepatomegaly, and jaundice, whereas biliary diseases
include cholestasis, recurrent cholangitis, and hepatic disintegration [1]. Most cases still have difficulties in diagnosis of apparent bleeding points. Moreover, no strong correlation was observed between CT findings and the clinical subtypes of hepatobiliary lesions in HHT [2].

Although AVMs could not be proved inside the biliary tract in this case, fortunately hemobilia was followed conservatively. In cases of telangiectasias detected by angiography or endoscopy including cholangioscopy, endovascular intervention and endoscopic thermal ablation can be effective for hemostasis [3, 4]. Present therapy is intended to reduce the symptoms. Although mechanism-based therapy is not available so far, recent progress has been made using drugs that target vascular endothelial growth factor (VEGF) and the angiogenic pathway with the use of the humanized monoclonal antibody against VEGF (bevacizumab) [1, 5]. In conclusion, although rare, hemobilia should be included in the differential diagnosis of gastrointestinal bleeding in HHT.

References


Figure A. A plain computed tomography scan showed high density fluid, which was suggestive of blood, in the gall bladder (arrow).
Figure B. Endoscopic retrograde cholangiopancreatography disclosed active bleeding from the papillary orifice (arrow) and clots (arrowhead).
Figure C. Intraductal ultrasonography showed high echogenic material (arrow), which was suggestive of blood, in the dilated common bile duct.
Figure D. Massive hemobilia was drained after the stent impacted with clot was removed.