We report a very rare case of acute obstructive hemobilia caused by bleeding of a mass-forming intrahepatic cholangiocarcinoma in a 75-year old man. The prompt complete emergency diagnosis was made by ultrasound and confirmed by MDCT. The features of hemobilia and intrahepatic cholangiocarcinoma are briefly reviewed.

Key-word: Bile ducts, neoplasms.

Case report

A 75-year old man was admitted in the emergency department with complaints of acute colicky pain in the right upper quadrant, associated with jaundice and maelena. Laboratory tests showed a marked rise of transaminase (AST or GOT) at 71,7 U/L (nl < 40 U/L) and alkaline phosphatase (ALT or GPT) at 151 U/L (nl < 41 U/L). Complementary abdominal ultrasound was performed. Three of the eleven retained echogenic bile spots were confirmed in the left hepatic lobe – just distally to the main hepatic biliary tree – and caused painful jaundice.

Emergency abdominal ultrasound of the right upper quadrant (Fig. 1A) was immediately performed and showed a 6x5x3 cm moderately hypoechoic tumoral mass in the S3 hepatic segment. This mass was responsible for a proximal diffuse ductal dilatation in the S3 hepatic segment (Fig. 1B). The dilated ducts were clearly seen converging to the mass and a biliary tumor was immediately suspected. Three centimetric small satellites tumors were found in the vicinity of the main lesion.

The elective cause of the mechanical jaundice was attributed to the presence of a very long serpiginous clot extending from the main left hepatic biliary tree – just distally from the main tumor – to the bottom of the distal intrapancreatic choledocus (Fig. 1C-F). This long clot was not only responsible for the dilatation of the entire left biliary tree but also for an indirect global dilatation of the contralateral right biliary tree (Fig. 1D & 3B). This dilated right biliary tree appeared free from clot but contained echogenic retained bile.

We report a rare case of hemobilia diagnosed in a 75-year old man presenting at the emergency room with the classic triad of Quincke associating right upper quadrant pain, jaundice and maelena. The full diagnosis was that of a mass-forming intrahepatic cholangiocarcinoma was made during emergency ultrasound examination completed by MDCT. The features of hemobilia and intrahepatic cholangiocarcinoma are briefly reviewed.

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transhepatic cholangiography – account for the main part and are related to a more and more increasing practice (2, 4). Stones in the gallbladder or in the biliary tree may also cause direct mucosal damage or may erode the cystic artery leading usually to minor bleeding (2). Nevertheless acalculous cholecystitis may also lead to hemobilia due to the high pressure induced. Cholangitis, parasite diseases (ascariasis or hydatid disease) and hepatic abscesses are other possible causes of hemobilia. Malignancies of the liver (hepatocarcinoma, cholangiocarcinoma and metastases), of the pancreas and biliary tract, especially in advanced stages, may also cause hemobilia by direct invasion of surrounding vessels (2). Finally vascular diseases comprising polyanteritis nodosa, aneurysms of the hepatic artery but also coagulopathies (i.e. Bernard-Soulier syndrome, idiopathic thrombocytopenic purpura, hemophilia and treatment with anticoagulants) have been implicated in the induction of hemobilia (2-3).

Cholangiocarcinoma presenting with hemobilia is an extremely rare event and to our best knowledge there have been only three or four previous descriptions (3, 4-7) essentially in Southeast Asia were the disease is more prevalent. In the reported case the administration of Clopidrogel (Plavix) to the patient for previous coronary bypass was considered an associated predisposing factor for obstructive hemobilia.

The most common symptoms associated with hemobilia are jaundice (30%), biliary colic (52%), and gastrointestinal hemorrhage (73%) (2-4). These three symptoms are referred to as Quincke’s classic triad of hemobilia. This complete triad was found in our patient but only occurs in approximately 22% - 37.9% of symptomatic patients (2-5). Hemobilia usually concerns minor bleeding and stops spontaneously in most cases. In cases of very active bleeding, the blood flows rapidly into the duodenum and the patient usually presents with melaena or hematemesis. In contrast, a slow bleeding tends to form clots and preferentially may cause biliary obstruction because the blood does not mix with bile and forms separate layers due to the difference in gravity and surface tension (2).

The currently available diagnostic modalities for hemobilia include esophagogastroduodenoscopy, side-view endoscopy, abdominal MDCT, ultrasonography, and angiography (2). Although CT angiography is very helpful to detect the bleeding point and mass lesions, it does not indicate the amount and rate of bleeding. An endoscopy has many advantages in that endoscopy can demonstrate fresh blood around the ampulla of Vater and exclude the other bleeding sites in the gastrointestinal tract. ERCP plays important roles in the diagnosis of hemobilia. The rate and amount of bleeding can
be directly estimated and opacification of the biliary tree can reveal the causes of hemobilia, the type and site of filling defects, and degree of bile duct dilatation (2).

In the report case the patient presented with the Quincke’s classic triad but his dominant symptom was nevertheless an extremely acute colicky pain in the right upper quadrant associated with jaundice. For this reason ultrasound was first performed as the recommended initial emergent imaging test in patients with signs and symptoms of hepatic, gallbladder or biliary disease as well as right upper quadrant pain (8-9). The full imaging diagnosis was directly made by this first line imaging modality and the other imaging modalities (MDCT, MRI and PET/CT) which were performed to complete the preoperative staging didn’t reveal fundamental additional data.

Cholangiocarcinoma accounts for 3% of all gastrointestinal cancers and is the second most common primary malignancy of the liver (5, 10). It accounts for approximately 15-20% of all primary liver cancers (11). The prevalence and mortality of intrahepatic cholangiocarcinoma are continuously increasing the highest prevalence being found in Southeast Asia (10-11). On the contrary, the prevalence of cholangiocarcinoma of the gallbladder and extrahepatic cholangiocarcinoma seems to remain rather constant or decrease (10).

Recognized risk factors for cholangiocarcinoma all share the common feature of chronic biliary inflammation (10-12). Schistosomiasis and hepatolithiasis are common risk factors in eastern Asia, whereas primary sclerosing cholangitis, liver cirrhosis, alcohol-related liver disease, and diabetes are relatively common risk factors in Western countries (10). Various viral infections, anomalies and/or malformations of the biliary tract, biliary-enteric drainage procedures and environmental or occupational toxins also constitute risk factors (10, 12).

Based on gross anatomic morphologic characteristics, cholangiocarcinoma are classified into mass-forming, periductal infiltrating and intraductal grow type (10, 12). Traditionally, extrahepatic bile duct cancer has been classified as nodular, sclerosing, or papillary, corresponding to the mass-forming, periductal infiltrating, and intraductal growth types of intrahepatic cholangiocarcinoma, respectively (10, 12-13). This classification is useful for the interpretation of the imaging findings and for the differential diagnosis but also for prediction of dissemination, prognosis and planning of surgery (10).

The reported case was a typical intrahepatic mass-forming cholangiocarcinoma (MFC) where a homogeneous mass with irregular but well-defined margin is found with a frequent dilatation of the biliary intrahepatic tree at the periphery. This type of intrahepatic cholangiocarcinoma is the most common representing 60% of all intrahepatic cholangiocarcinomas (11, 13). During ultrasound MFC is homogenous well-defined mass with irregular but well-defined margin and presenting with a peripheral hypoechoic rim – as found in the reported case – in about 33% of cases. Tumors larger than 3 cm are usually hypoechoic and tumors less than 3 cm are rather hypo- or isoechoic. In the reported case the echogenicity appeared heterogeneous but predominantly hypoechoic (10, 13). Typical CT features of the mass-forming type of cholangiocarcinoma include homogeneous attenuation, irregular peripheral enhancement – as illustrated in our case – with gradual centripetal enhancement on much delayed phase – phase not performed in the reported case – (10-12). The degree of delayed enhancement is closely related to the proportion of fibrous stroma. Capsular retraction, the presence of satellite nodules – as found in our patient – and vascular encasement without the formation of a grossly thrombus are other possible features. The MR imaging features of mass-forming cholangiocarcinoma are similar to its CT features (10).

At gross examination, mass-forming cholangiocarcinoma is characterized by a homogeneous sclerotic mass with an irregular lobulated margin, typically in the absence of hemorrhage or central necrosis. The tumor is firm and whitish gray because of its large amount of fibrous stroma (13). Histologically the viable
tumor cells are usually located at the periphery of the tumor. The central portion of the tumor is composed of a variable degree of fibrosis with necrosis and scattered tumor cells. The differential diagnosis concerns tumors with abundant fibrous stroma such as hepatocarcinoma (HCC) with cirrhotic stroma, sclerosing HCC or combined HCC-cholangiocarcinoma but also various tumors with abundant fibrous stroma, embryonal sarcoma and neuroendocrine carcinoma (10, 12).

References