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CASE REPORT

Suprahepatic Gallbladder and Right Lobe Anomaly of the Liver in Patients with Biliary Cancers

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Suprahepatic gallbladder is among the rarest sites of an ectopic gallbladder (1). This condition implies that the abnormal location is simply along the retrosuperior surface of the liver in the subphrenic space (1, 2). Such a location is attributed primarily to agenesis of the right lobe of the liver. However, the presence of a suprahepatic gallbladder is not specific to an embryological problem. Some other acquired diagnoses should be differentiated; namely, this rare presentation would be associated with right lobe agenesis, abnormally mobile gallbladder (3), diaphragmatic eventration (4), inversion of the liver (5), and hypoplasia from cirrhosis, hepatic vascular injuries, and cholangiocarcinoma (6–9).

Since 1965, when this rare anomaly was first reported by Regen and Poindexter (10), only relatively few cases have been described (1–9). In addition, the combinations of suprahepatic gallbladder with biliary cancers are even less frequent. In this manuscript, two cases with such rare findings are presented to introduce our experience in differential diagnosis and to compare the hepatic anomaly in these two cases. These combined situations have received limited attention before.

CASE REPORTS

Case 1. A previously healthy 42-year-old male patient had right upper quadrant pain, intermittent fever, and chills for two weeks' duration. On physical examination in the emergency room, he appeared to have fever (38.7°C), mild icteric sclera, tenderness over the right upper quadrant, but no evidence of cirrhosis. In particular, the knocking pain over the right upper back was significant. Laboratory studies disclosed WBC 21.8 × 10^9/liter, with segments 89%, and bands 5%; elevated asparatate and alanine transminase (129 IU/liter and 62 IU/liter, respectively), alkaline phosphotase (310 mg/dl), and γ-glutamyltransferase (340 mg/dl); bilirubin was 1.6 mg/dl. Ultrasonography revealed an abnormal location of the gallbladder in the suprahepatic region, absence of the right lobe of the liver, and a hypoechoic cystic-like lesion (3 × 3.5 cm in size) over the left lobe. Computed tomographic (CT) scans revealed that the inferior vena cava was located outside the liver parenchyma and adjacent to the medial posterior surface of the remnant liver. In addition, the gallbladder was retroplaced to the liver just below the diaphragm (Figure 1). The findings are quite specific to the diagnosis of right lobe agenesis. In addition, the portal system was mildly dilated but without any evidence of thrombosis. The tender point corresponded to the hypoechoic area within the left lobe of the liver, which was suspected to be an abscess. Some fluid accumulation occurred over the gallbladder fossa, located in the subphrenic area. Therefore, percutaneous drainage was performed under sonographic guidance for both the liver abscess and the area with fluid accumulation. Only a slight amount of pus aspirated from the abscess area, while about 25 ml of frank pus aspirated from the subphrenic area, which subsequently yielded a positive culture of *E. coli*. Despite the coverage of strong antibiotics and external drainage, his fever persisted and the clinical condition became worse. Under the diagnosis of the liver abscess rupture or gallbladder rupture, he received laparotomy for abscess drainage and cholecystectomy. During the operation, the gallbladder was found to be ruptured and was therefore resected. Gallbladder adenocarcinoma was diagnosed from the resected specimen. The tumor cells had invaded beyond the serosal layers. Omental seeding of tumor was diagnosed and the omentum was widely resected. Subphrenic pus was also drained successfully. In addition, no right side feeding vasculature was found.

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Fig 1. (a) This CT film disclosed an absence of the right lobe and a hypodense area (dot arrows) in the left lobe liver; (b–d). The gallbladder (g) is located ectopically in the suprahepatic region with fluid accumulation over the subphrenic space (armed arrows); both (c) and (d) revealed that the inferior vena cava (v) was not surrounded within the liver, but just adjacent to the medial posterior surface of the remnant liver; (d) disclosed mild dilated portal vein without thrombosis (p) and splenic vein(s).

Therefore, agenesis of the right lobe was diagnosed. His clinical condition improved soon after the operation. However, he died from this cancer with diffuse metastasis one year later.

Case 2. A 55-year-old male patient experienced intermittent right upper quadrant pain for several years. His pain had become aggravated in the past month. He also suffered from progressive general weakness, poor appetite, and a weight loss of 9 kg. He was admitted through our emergency department because of fever, chills, tea-colored urine, and yellowish skin for one week. The serial studies showed hyperbilirubinemia (bilirubin 9.8 mg/dl); asparatate transaminase levels of 124 IU/liter, alanine transminase of 241 IU/liter, alkaline phosphates of 465 mg/dl, and γ-glutamyltransferase of 626 mg/dl; and leukocystosis (WBC 18.8 $\times 10^9$/liter). The tumor markers were elevated significantly: CEA 10.8 (normal < 3.5 ng/ml); CA125 49.8 (normal < 30 ng/dl); CA19-9 2675.3 (normal < 33 ng/dl). The ultrasound and CT scan (Figure 2) revealed hypoplasia of the right lobe, abnormal position of gallbladder, and markedly dilated intrahepatic duct. His common bile duct was normal in caliber; however, a suspected hypodense lesion over the hilar area was noted, which was considered to be the cause of jaundice. He was treated as having sepsis from biliary obstruction. The jaundice was relieved by percutaneous transhepatic choledochal drainage. The cholangiography showed the presence of a hilar mass with an abrupt interruption of common hepatic duct (Figure 3). Surgical exploration was carried out, but it failed to resect the tumor because of widespread tumor invasion and direct tumor encasement to the right-side portal vein and hepatic artery. The tumor also adhered to omentum with multiple intra-abdominal seedings. The pathology of biopsy from the hilar mass revealed tumor nests spreading in a desmoid background, which is characteristic of cholangiocarcinoma. His condition deteriorated soon after surgery, and the patient died within one month.

DISCUSSION

Ectopic gallbladders carry unusual images in radiology and raise problems in clinical management. The ectopic location is classified into four major types: transverse, left-sided, intrahepatic, and suprahepatic (11, 12). The last one is the rarest type and is usually

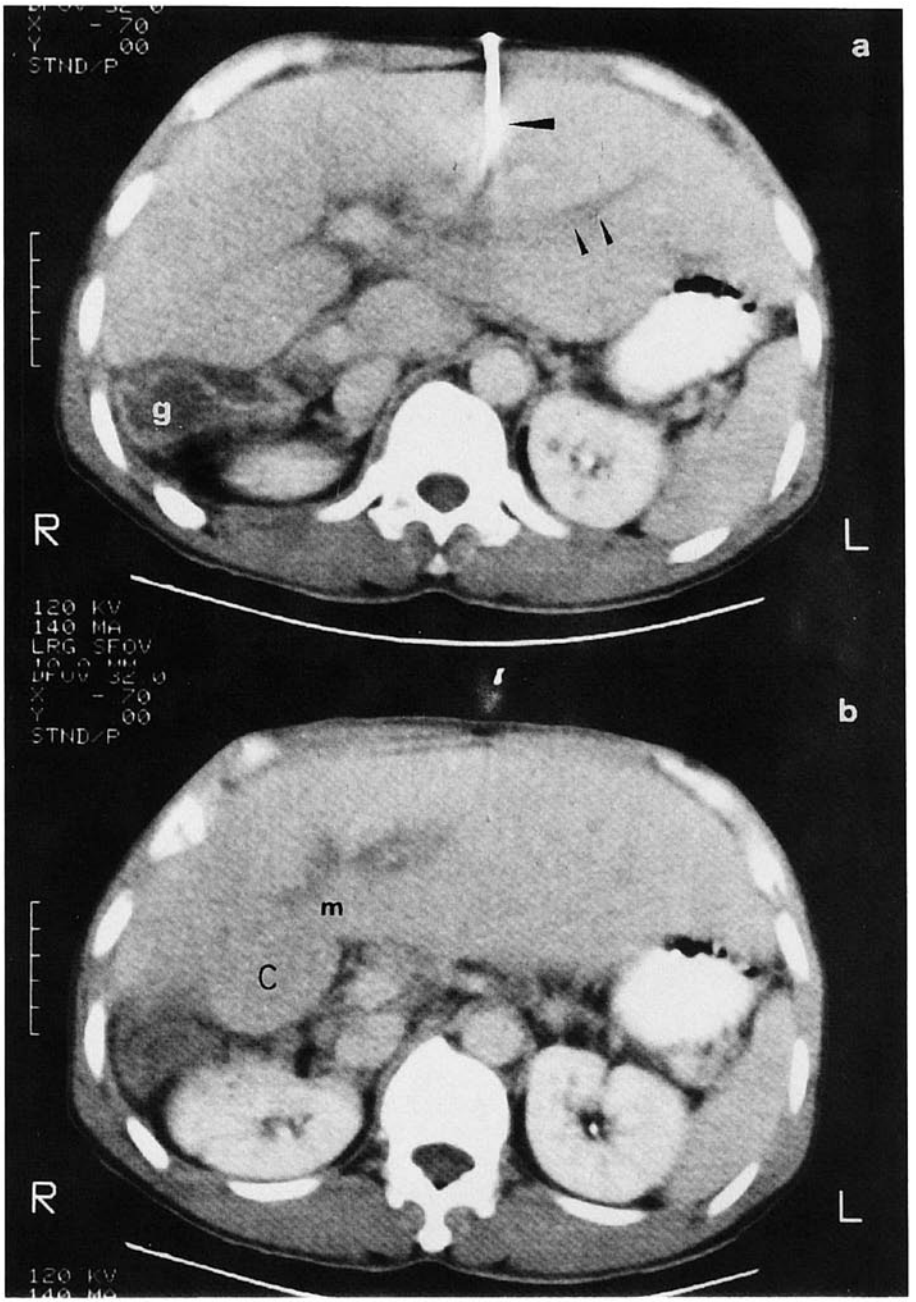

Fig 2. (a) The ectopic gallbladder (g) located at the retrosuperior surface of the liver. The percutaneous transhepatic choledochal drainage tube was inserted within the marked dilated intrahepatic duct (arrows). (b) This film revealed hypertrophy in the left lobe and caudate lobe (C) of the liver. A suspected hilar mass (m) with a relatively hypodense area and the retroperitoneal lymph nodes are shown as well.

combined with right lobe anomaly (13). The combination could be accounted for by the embryological hypothesis that the right lobe of the liver and gallbladder precursor are closely related after the seventh gestational week (14). Once the massive decrease in the volume of the right lobe occurred due to either congenital agenesis or acquired hypoplasia, the gallbladder's position would be pulled backward and upward. Therefore, for a sonographer or a radiologist, the suprahepatic position of the gallbladder is a clue to right lobe anomaly, or *vice versa.*

Radin et al reported that the suprahepatic location

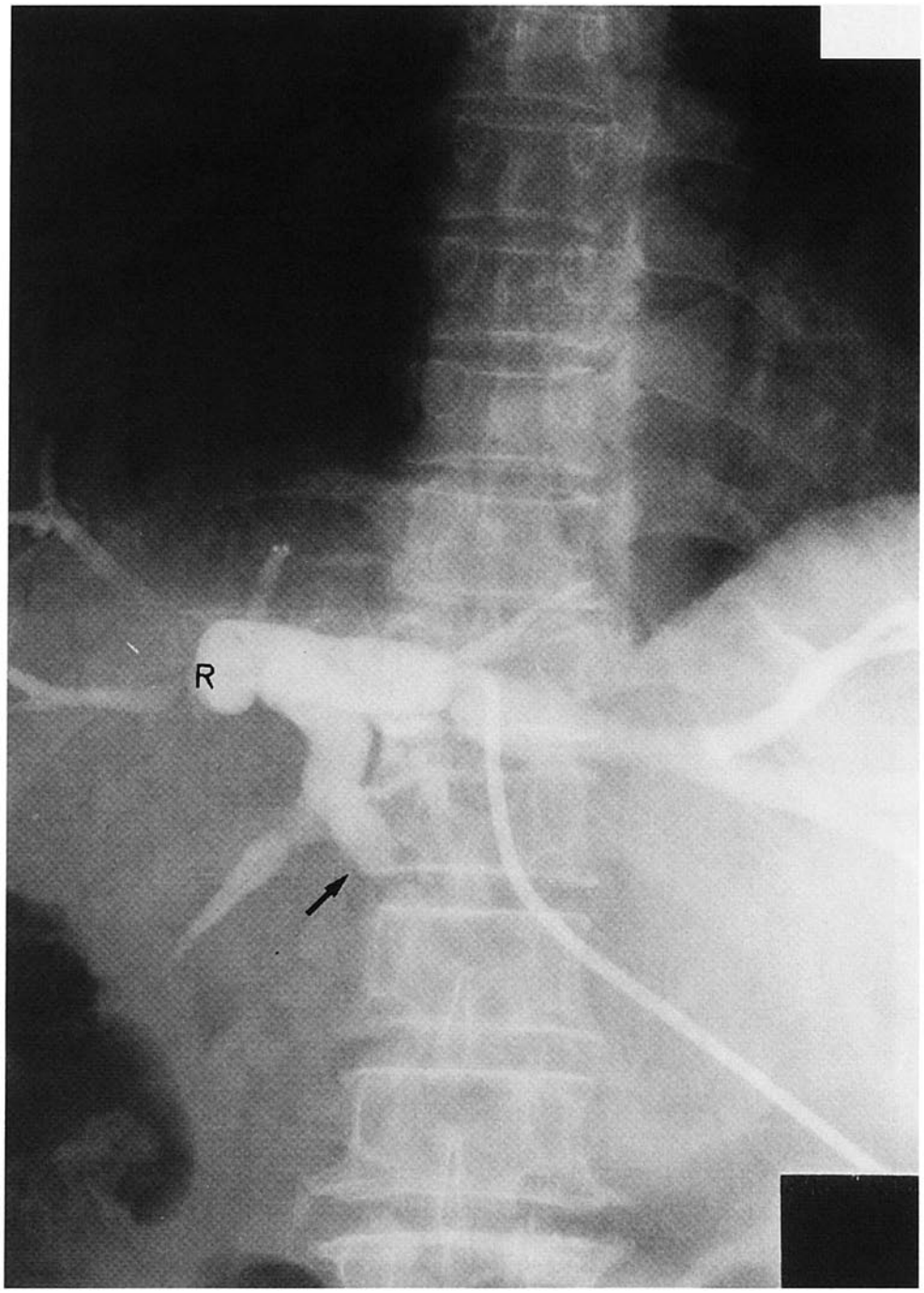

Fig 3. Percutaneous transhepatic cholangiography showed dilation of the left intrahepatic ducts and the complete absence of the sequential ductal bifurcation in right intrahepatic duct (R) due to right lobe atrophy. Abrupt interruption of the biliary trace (arrows) was found at the level of common bile duct near the hilum, where the tumor is.

is not specific to right lobe agenesis (7). Some cases of acquired hypoplasia or atrophy have been reported, including: liver cirrhosis, hepatolithiasis, and hilar cholangiocarcinoma (7–9). Even though the incidence is quite rare, detailed left-sided subcostal and higher right-sided intercostal scannings by ultrasound are suggested to survey the inversion of the liver, diaphragmatic eventration (5), and the hypermobile gallbladder (10). In addition, previous right lobe lobectomy should be traced by history.

The CT scan can normally document the presence of suprahepatic gallbladder and offer some hints in images for differentiating the exact causes. It was widely recognized as the best assessment for obtaining a diagnosis of lobar agenesis of the liver (7, 8). The main lobar fissure and the middle hepatic vein serve as the borderline between the right and the left lobe of the liver. The left hepatic vein and the remnant of ligament teres help topographic separation between medial and lateral segment of left lobe liver. The exact entity of lobar absence under CT scans can be known via the application of these landmarkers in the liver (8). Furthermore, clinical correlation and other associated radiological evidence of cirrhosis would enhance the accuracy in distinguishing agenesis of hypoplasia secondary to cirrhosis (14). CT scans with markedly dilated intrahepatic ducts were considered to be highly specific for hilar cholangiocarcinoma, which may obstruct the right bile duct and portal vein, resulting into ipsilateral atrophy and contralateral hypertrophy in a nonobstructed (left) hepatic lobe (7, 9). As in case 2, the surgeons found that the hilar cholangiocarcinoma directly invaded and encased the right portal system. In contrast, based on the absence of the right portal vein during operation and no portal vein thrombosis on CT films, the right lobe anomaly of case 1 was identified as agenesis instead of hypogenesis. Other associated CT findings of suprahepatic gallbladder are elevated right colic flexure (6), and inferior vena cava exposed from the caudate lobe at the junction of the hepatic vein (3, 15). In brief, the CT image is of great value in assessing suprahepatic gallbladder with right lobe anomaly.

According to previous investigations (1, 3, 7), the agenesis of right hepatic lobe with supraheptic gallbladder would predispose to biliary tract disease, such as cholelithiasis, gallbladder cancer, and Mirizzi syndrome. These findings can be explained by the poor drainage of the biliary system (3), as typically presented in case 1 of this manuscript. The patient had a combination of right lobe agenesis, gallbladder adenocarcinoma with rupture, and liver abscess of the left lobe. This combination would be assumed to be a long-term consequence of the suprahepatic gallbladder. Another indirect sign of the long-term sequelae of right lobe agenesis in this noncirrhotic patient was a mildly dilated portal vein, which could be accounted for by the poor portal venous input in the reduced liver size and relatively high portal flow in the extrahepatic portal system (16).

Overall, the exact cause of suprahepatic gallbladder must be differentiated and can be traced by CT scan. Surgeons should be aware of the unusual position of the gallbladder to ensure the accurate approach, because laparoscopic cholecystectomy is not indicated in the suprahepatic or intrahepatic gallbladder (2, 17). As suprahepatic gallbladder predisposes to certain biliary tract diseases, precautions should be taken against these long-term sequels, especially biliary malignancy.

SUMMARY

The suprahepatic region is a rare ectopic location of the gallbladder. It usually combines with right lobe anomaly of the liver. Here we report two unusual cases of suprahepatic gallbladder with agenesis or hypogenesis of the right lobe of the liver and biliary cancer. A patient with a gallbladder tumor was admitted to our emergency room with acute cholecystitis and liver abscess. Imaging examinations and operation confirmed the suprahepatic position of gallbladder, agenesis of the right lobe, and dissemination of gallbladder cancer. In the patient with cholangiocarcinoma, CT scans and percutaneous transhepatic cholangiography documented the presence of a hilar tumor and hypogenesis of the right lobe. Both of these patients died from biliary tract cancer soon after operation.

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