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CASE REPORT

Incidentally Detected Hilar Cholangiocarcinoma: A Case Report

Tesadüfen Saptanan Hiler Kolanjiyokarsinom: Bir Olgu Sunumu

Anu Jacob[1], George Sarin Zacharia[2]

[1]Department of Anesthesiology, Ahalia Hospital, Abu Dhabi, United Arab Emirates
[2]Department of Medical Gastroenterology, Ahalia Hospital, Abu Dhabi, United Arab Emirates

Abstract

Hilar cholangiocarcinomas, also known as Klatskin tumors, represent a rare subset of biliary tract malignancies. Klatskin tumors are often diagnosed late and are associated with a poor overall prognosis. Here, we report the case of a 51-year-old female, an intellectually disabled and non-communicating individual, who presented for an unrelated medical condition, incidentally detected to have icterus on clinical examination, which on evaluation culminated in the diagnosis of Klatskin tumor. Despite having no attributable symptoms or identifiable risk factors, she exhibited classical biochemical and imaging features consistent with the diagnosis. Though the underlying comorbidities might have masked the presentation in our case, potentially serious liver diseases can remain asymptomatic or have minimal or non-specific symptoms early in their course. Hence, clinical assessments and judicious interpretations of hepatic biochemistry hold the key to prompt diagnosis despite the advancements in modern medicine. Through this case report, we will briefly review the evaluation of cholestasis and hilar cholangiocarcinoma, also known as Klatskin tumor.

Keywords: Klatskin; Cholangiocarcinoma; Cholangiography

Primary cholangiocarcinomas arising at the bifurcation of the extrahepatic biliary tree are known commonly as Klatskin tumors. Named after Gerald Klatskin, who elaborated on 15 cases in 1965, they are relatively rare lesions, with an incidence of 0.01% to 0.46% in the autopsy series.[1] Of the 17,500 projected new cases of primary hepatobiliary cancers that occur annually in the United States, approximately 2,000 are Klatskin tumors.[2] Though clinically indifferent from other cholangiocarcinomas, hilar malignancies have a poorer prognosis. The most important factor affecting prognosis in cholangiocarcinomas is resectability, which is low in hilar lesions, translating to an overall poor prognosis. Here, we present a case of a Klatskin tumor, diagnosed incidentally in a middle-aged intellectually disabled female who presented to the hospital with an unrelated medical concern. This case highlights that a holistic approach to the patient and an astute clinical assessment can guide a physician to a diagnosis, even in rare cases. Also, it points to the need for more comprehensive care in differentially abled individuals, as they might fail to identify, comprehend, or report non-physiological situations or events.

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Correspondence: George Sarin Zacharia, M.D. Department of Medical Gastroenterology, Ahalia Hospital, Abu Dhabi, United Arab Emirates
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Case Report

A 51-year-old female with intellectual disability, cerebral palsy, and seizure disorder who is bedridden was brought to the hospital following a malfunction of an endoscopically inserted gastrostomy tube. The patient was non-communicative, and caretakers denied other active complaints. However, on physical examination, in addition to the sequelae of cerebral palsy, she was found to be deeply icteric. A hemogram revealed mild anemia, while a routine biochemical assay was consistent with cholestatic jaundice; results are summarized in Table 1. A computed tomography (CT) of the abdomen revealed marked intrahepatic biliary ductal dilation (Fig. 1).

Subsequent magnetic resonance imaging (MRI) with cholangiopancreatography (MRCP) confirmed the biliary dilatation and revealed a 3.1-centimeter mass at the hepatic hilum, consistent with hilar cholangiocarcinoma (Fig. 2). The gall bladder revealed the presence of cholelithiasis without evidence of cholecystitis, while the pancreas, extrahepatic biliary duct, and rest of the abdominal viscera were within normal limits. The carbohydrate antigen 19.9 was reported as 329 (normal: 0-37 units per milliliter), but the alfa fetoprotein was within normal limits. She was recommended further evaluation with endoscopic ultrasound, which, however, the family refused. After extensive discussions with her healthcare proxy, it was decided to proceed with interventional radiology-guided external biliary drainage. She underwent percutaneous transhepatic cholangiography (Fig. 3) with the placement of a biliary drain in the right biliary system, following which her jaundice improved. A radiology-guided gastrostomy placement was performed as well. The patient was discharged and referred to the palliative care unit by the family's wishes.

Table 1. Baseline laboratory workup

Parameter	Results	Reference range	Units
Hemoglobin	11.7	13.2-16.6	gm/dl
WBC	17.3	4.8-10.8	k/µl
Platelets	557	150-400	k/µl
Total bilirubin	16.7	0.2-1.2	mg/dl
Direct bilirubin	12.5	0-0.3	mg/dl
Aspartate transaminase	231	9-36	IU/L
Alanine aminotransferase	71	5-40	IU/L
Alkaline phosphatase	906	17-406	IU/L
Total protein	8.7	6.0-8.5	gm/dl
Albumin	2.5	3.2-4.8	gm/dl
Prothrombin time	24.7	10.4-15.7	seconds

WBC: White blood cell.

Discussion

Jaundice is one of the most encountered scenarios in medical practice, defined by elevated serum bilirubin levels. Cholestasis, as the name suggests, results from the stasis of bile, resulting in the reflux of bile into circulation and cholestatic jaundice. Depending on the mechanism and site of stasis, it may be classified as intrahepatic versus extrahepatic cholestatic jaundice. For all practical purposes, intrahepatic refers to defective biliary secretion across the canalicular membrane, while extrahepatic refers to an obstructive pathology in the biliary tree, synonymously called obstructive jaundice. Obstructive jaundice often

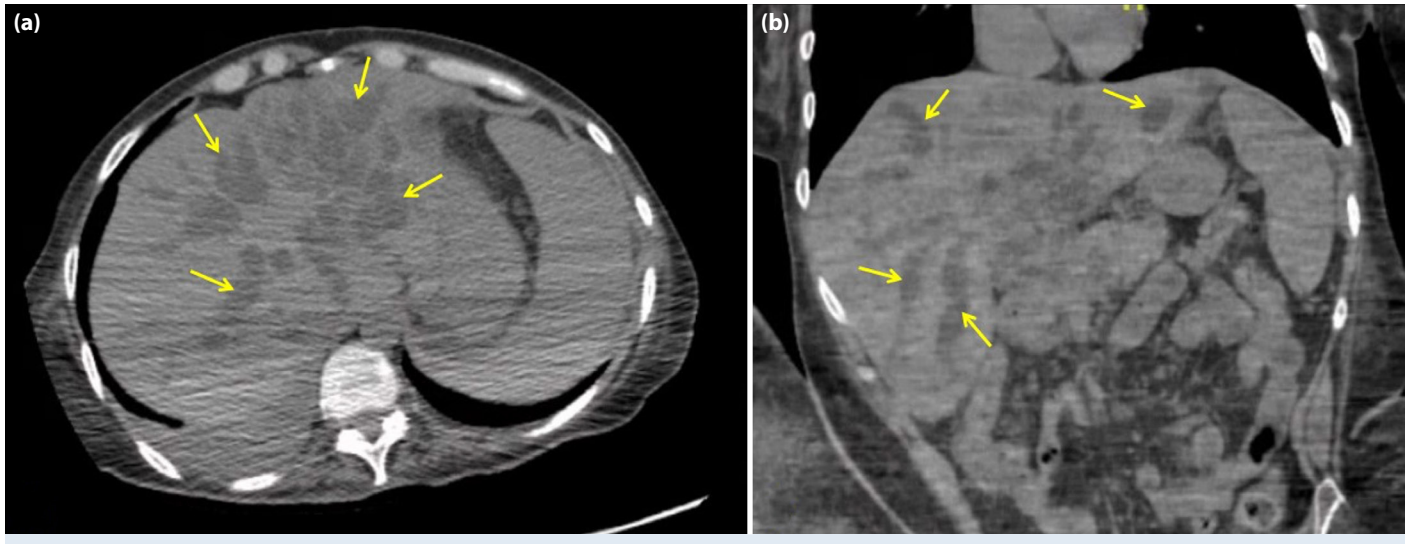

Figure 1. CT abdomen, axial **(a)**, and coronal **(b)** images, revealing hepatic bilobar intrahepatic biliary radicle dilatation (yellow arrowheads).

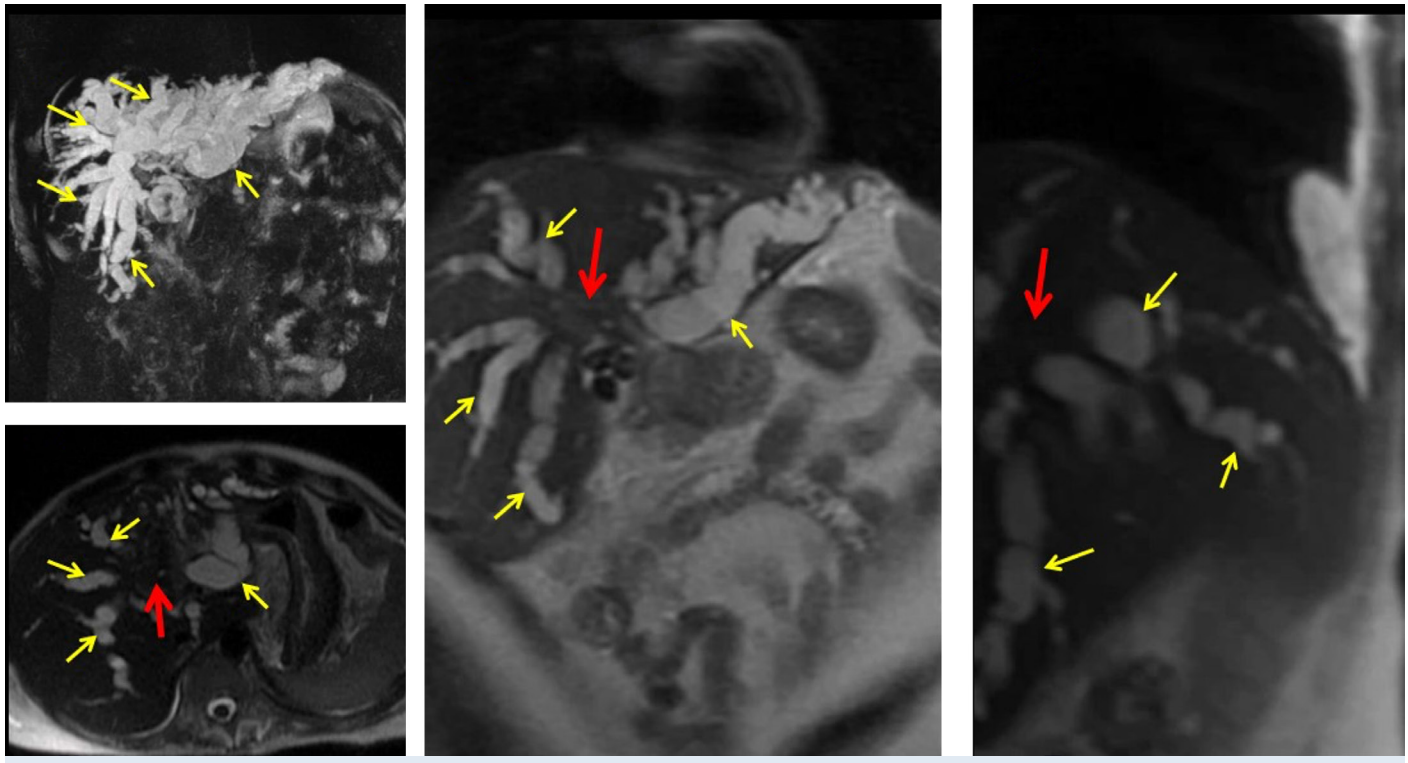

Figure 2. MRI abdomen with MRCP images revealing dilated biliary tree (yellow arrowheads) and the central occluding mass lesion (red arrowhead).

presents as upstream biliary dilatation proximal to the site of obstruction. Intrahepatic biliary radicle dilatation (IHBRD) and dilated common bile duct (CBD), defined as a common bile duct of diameter >6-7 mm, are common radiological findings in biliary obstruction. However, a dilated CBD, >6-7 mm, doesn't always mean obstruction and can be seen in choledochal cysts, post-cholecystectomy status, and advancing age.

Cholangiocarcinomas are often adenocarcinomas arising from the biliary epithelium and, depending on their location, are classified as intrahepatic, hilar, or distal. They account for 10-15% of hepatobiliary malignancies and about 3% of gastrointestinal malignancies.[3,4] The hilar or perihilar subtype is the most common and accounts for almost half of the cases of cholangiocarcinoma. Primary sclerosing cholangitis is the single most important etiological association, while the others include biliary parasitosis (Clonorchis sinensis, Opisthorchis viverrine), Thorotrast, Lynch syndrome, and biliary papillomatosis. [4,5] Intrahepatic cholangiocarcinoma can remain asymptomatic until late, but extrahepatic ones present earlier with obstructive jaundice, characterized by jaundice, pruritis, dark urine, and clay-colored or pale stools. Biochemistry reveals direct or conjugated hyperbilirubinemia, markedly elevated ALP, gamma-glutamyl transpeptidase (GGT),

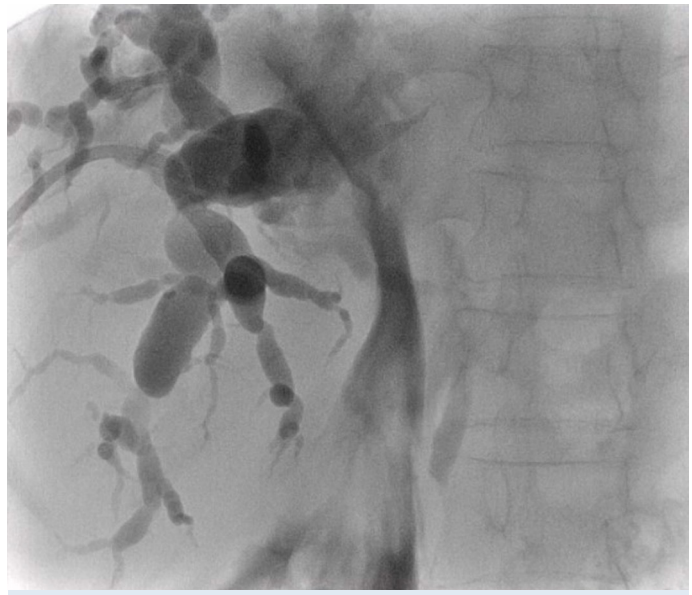

Figure 3. Percutaneous transhepatic cholangiogram showing a dilated biliary tree.

and mild to moderate elevation of ALT and AST. The best-studied tumor marker in cholangiocarcinoma is Ca19.9. An ultrasound of the abdomen reveals a dilated biliary tree, and larger tumors may be identified, as are the nodes, metastasis, or ascites. Contrast CT or MRI with MRCP is more sensitive for diagnosing smaller

lesions. Endoscopic ultrasound allows visualization and tissue sampling, especially in distal lesions. Endoscopic retrograde cholangiopancreatography allows brush cytology and biliary stenting, which is again more effective in distal cholangiocarcinoma. Tissue diagnosis can be very challenging in cases of perihilar cholangiocarcinoma, and often, diagnosis relies on imaging alone, especially when the intended treatment is palliative. [4] The Bismuth Corlette system has been conventionally used for classifying perihilar cholangiocarcinoma, and the TNM system allows tumor staging.[6] The only curative therapeutic option is surgery; however, its efficacy depends on the site and extent of the tumor, local invasion, resectability, and staging. Palliative interventions such as biliary drainage can help alleviate symptoms and improve the quality of life in patients with unresectable lesions.

Biliary cancers, including cholangiocarcinomas, have constantly challenged chemotherapy. Until 2010, gemcitabine monotherapy remained the cornerstone of treatment. The ABC02 trial and BT22 trials published during the earlier half of the last decade allowed a therapeutic evolution to gemcitabine-cisplatin combination therapy.[7,8] The advancement of cancer genetics and targeted therapeutic agents over the previous few years has revolutionized the management of all tumors, and cholangiocarcinomas are no exceptions. The most frequently identified genetic aberrations in cholangiocarcinomas include TP53, isocitrate dehydrogenase (IDH), KRAS and BRAF mutations, and FGFR2 or NTRK fusions.[9] Many of these genetic anomalies are potential targets for targeted molecules, which has widened the therapeutic landscape in cholangiocarcinoma. The FGFR2 inhibitors in development include pamigatinib, infigratinib, and futibatinib.[9] The European Society for Medical Oncology recommends IDH mutation analysis in biliary carcinomas, and the targeted therapeutic agent ivosidenib is approved as a second-line agent in the treatment of advanced or metastatic biliary tract cancers with favorable mutations.[10] The NTRK inhibitors larotrectinib and entrectinib are also evaluated in treating bilairy neoplasms.[9]

Our patient fulfills all the classical clinical, biochemical, and imaging features of hilar cholangiocarcinoma or Klatskin tumor. She had no identifiable risk factors for cholangiocarcinoma, though it might be identifiable in only up to 30% of patients.[5] Antiepileptic medications are well known to precipitate drug-induced liver injury, including the cholestatic variant. However, our patient was on Levetiracetam, which is known for its safe hepatic profile, and the presence of a dilated biliary tree is a marker of extrahepatic obstruction rather than intrahepatic cholestasis of drug-induced liver injury. Owing to the medical condition, the patient lacked decision-making capacity, and considering the multitude of comorbid ailments, the family decided not to proceed with endoscopic procedures or surgical intervention. Hence, she was palliated with percutaneous biliary drainage and post-procedure, the patient had improvement in jaundice.

Conclusion

Klatskin tumor, or hilar cholangiocarcinoma, is a relatively rare and challenging diagnosis. Astute clinical examination and interpretation of hepatic biochemistry, a high index of suspicion coupled with modern imaging modalities, might facilitate an early diagnosis. Surgery offers the only chance of cure, though many patients may not be candidates for surgical intervention.

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