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Case report

Spontaneous biloma due to an intrahepatic cholangiocarcinoma: An extremely rare case report with long term survival and literature review

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HIGHLIGHTS

- Spontaneous biloma in the presence of an intrahepatic cholangiocarcinoma is an extremely rare situation.
- Spontaneous rupture of bile vessels due an intrahepatic cholangiocarcinoma is an urgent and life-threatening complication.
- This is the first case treated with a left hepatectomy and also the first one to report such a long disease-free survival.
- Spontaneous bilomas should be considered in patients presented with epigastric pain, hepatic tumor and fluid collection.

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ABSTRACT

Cholangiocarcinomas are tumors that arise from the ductal epithelium of the intrahepatic or extra-hepatic bile ducts. Patients are usually asymptomatic or may present with weight loss, fatigue, loss of appetite and abdominal pain (intrahepatic cholangiocarcinomas) or jaundice (extra-hepatic cholangiocarcinomas). Subcapsular bile vessel rupture, due to intrahepatic cholangiocarcinoma, is an extremely rare clinical presentation, which is an emergent and potentially life-threatening complication.

We report the case of a 79-year-old female patient suffering from an intrahepatic cholangiocarcinoma that completely obliterated the left main hepatic duct. This obstruction in intrahepatic bile flow had resulted in intraperitoneal rupture of subcapsular bile vessels (not infiltrated by the tumor) of the left liver lobe and formation of spontaneous biloma. The patient was admitted for acute abdominal pain. Computed tomography (CT) and Magnetic Resonance Imaging (MRI) revealed the tumor and an upper abdominal fluid collection. Since the patient was hemodynamically stable and afebrile, a CT-guided percutaneous aspiration of the collection was undertaken, showing a biloma. A left hepatectomy was performed two weeks later and today, sixty months since the incident, the patient enjoys good health, with no signs of local recurrence or distant metastases.

Intraperitoneal rupture of bile ducts and subsequent spontaneous biloma formation, due to an intrahepatic cholangiocarcinoma which completely obstructed the left main hepatic duct, is a unique situation and this is the first time to be reported. Prompt surgical management can lead to successful treatment of this rare and difficult entity.

1. Introduction

Cholangiocarcinoma (CCA) is a rare malignant neoplasm of the biliary tree, accounting for about 10% of all primary liver tumors [1,2]. Based on its location, CCA is classified as intrahepatic and extra-hepatic (perihilar or distal) [3]. As for intrahepatic CCA (IH-

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Abbreviations

CCA	Cholangiocarcinoma
IH-CCA	Intrahepatic Cholangiocarcinoma
AST	Aspartate Aminotransferase
ALT	Alanine Transaminase
TBIL	Total Bilirubin
γ-GT	Gamma-glutamyltransferase
ALP	Alkaline Phosphatase
CEA	Carcinoembryonic Antigen
AFP	Alpha Fetoprotein
CA 19-9	Carbohydrate Antigen or Cancer Antigen 19-9
αAMS	α-Amylase
ERCP	Endoscopic Retrograde Cholangiopancreatography

CCA), progression is usually silent and most patients are diagnosed at an advanced stage, leaving no place for curative treatment, but to only about one third of patients [4]. Five-year survival after R0 resection (no macroscopic or microscopic evidence of tumor infiltration at the resection margins; curative resection) is in the range of 23–42% [3,5].

Biloma is an encapsulated bile collection outside the biliary tree [6]. The majority of bilomas are caused by trauma or iatrogenic injury. Spontaneous bilomas are very rare.

We report the unique case of a patient who presented with the clinical picture of acute abdomen, as a result of intraperitoneal rupture of subcapsular bile vessels of the left liver lobe, caused by an IH-CCA which had obstructed the left main hepatic duct, leading to spontaneous biloma formation. The work has been reported in line with the SCARE criteria [7].

2. Presentation of case

A 79-year-old female patient presented to her local hospital due to a dull epigastric pain and nausea for the last 3 days. Her past medical history included arterial hypertension, dyslipidemia, mild gastro-esophageal reflux disease and left knee osteoarthritis. Upon admission, her clinical examination was unremarkable and laboratory investigation was normal, except for elevated liver function tests (AST: 57 IU/L, ALT: 97 IU/L, TBIL: 1,3 mg/dL, γ-GT: 267 IU/L, ALP: 307 IU/L). During the next days her clinical condition deteriorated (she developed abdominal guarding and rebound tenderness at the epigastrium). Abdominal ultrasonography (U/S) and computed tomography (CT) imaging revealed a left liver lobe mass, with focal dilatation of the intrahepatic bile ducts around the lesion, findings suggestive of the presence of an intrahepatic cholangiocarcinoma. A sub-diaphragmatic fluid collection was also detected, as well as a large complex cystic-solid left adnexal mass lesion, measuring 86 × 73 mm. There was no evidence of metastatic disease.

The patient was subsequently referred to our hospital for further investigation and treatment. Anti-echinococcus antibodies and tumor markers (CEA: 1.7 ng/mL - normal upper range <5, AFP: 4.1 ng/mL - normal upper range <9) were negative and CA 19-9 was slightly elevated at 51 U/mL (normal upper range <35). Abdominal magnetic resonance imaging (MRI) revealed the presence of an ill-defined mass, of 53 mm in maximal diameter at liver segment IV, while segmental dilatation of the left intrahepatic bile ducts was seen on magnetic resonance cholangiopancreatography (MRCP), due to the presence of an intraductal polypoid mass. Two separate fluid collections under the left liver lobe that spanned towards the omentum were also detected (Fig. 1). Percutaneous drainage under CT-guidance defined the fluid as bile (BIL: 27.9 mg/dL, aAMS: 30 IU) and a drainage tube was left in place, resulting in clinical improvement. Due to the ongoing bile leak, an ERCP and stent insertion was attempted with no success, since the left bile duct was totally obstructed by the tumor. Laparotomy was decided.

At theater, after a bilateral subcostal incision with small vertical extension ('Mercedes-Benz') a left liver lobe tumor was discovered (segment IVb), with dilation and rupture of the subscapular bile ducts in segment III, which caused the biloma (Fig. 2). Typical left hepatectomy was performed. Pathological examination showed an intrahepatic cholangiocarcinoma of the mixed mass-forming/periductal-infiltrating type: the tumor had a mixed pattern of development, i.e. in part it exhibited a longitudinal growth pattern along the course of the left main hepatic duct (at a distance of 3.5 cm), and in part it expanded radially in the liver parenchyma as a well-defined mass at segment IVb (sizing 5.8 × 3.6 × 3.1 cm), extending up to the liver capsule. The site of bile leak due to capsular rupture was located at segment III. Resection margins (left hepatic duct, left portal vein and left hepatic artery) were free of invasion (R0) and lymph nodes were free of invasion (pT4N0). The ovary was found to carry a mucinous cystadenoma (cystic component) combined with a fibroma (solid component).

The patient received no adjuvant chemotherapy and today, sixty months after the procedure, she enjoys good health with no signs of local recurrence or distant metastases at routine oncological follow-up (Table 1).

3. Discussion

Cholangiocarcinomas (CCAs) are tumors that arise from the ductal epithelium of the intrahepatic or extrahepatic bile ducts [4]. They comprise the second most common primary liver tumor, accounting for 10–15% of all non-metastatic hepatic malignancies [3,4,8]. Histologically, the great majority of these tumors (around 90%) are adenocarcinomas, but other variants such as squamous cell carcinomas and clear cell carcinomas have also been described [9].

Although spontaneous rupture of the mass itself can be seen with primary liver tumors such as hepatic adenomas or

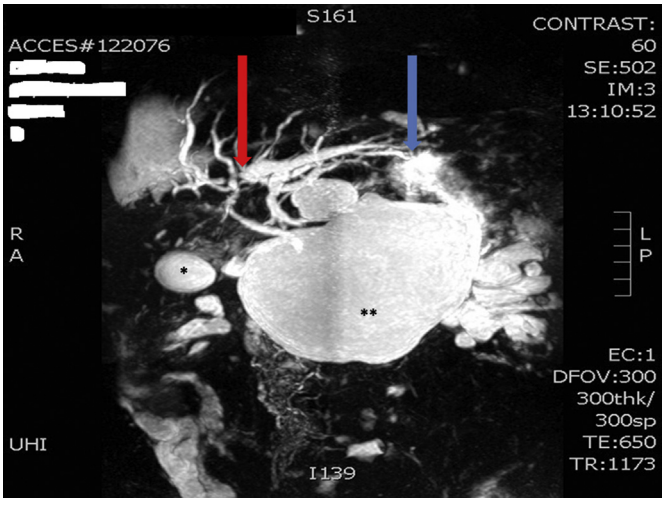

Fig. 1. Findings on Magnetic Resonance Cholangiopancreatography (MRCP): complete left main duct obliteration and peripheral bile duct dilatation, typical for cholangiocarcinoma (red arrow) and site of subcapsular bile duct rupture (blue arrow), with subsequent bile leak, leading to two distinct bilomas (the small one depicted with *, the greatest with **). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)