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Cystic Degeneration of Peripheral Intrahepatic Cholangiocarcinoma: An Atypical Presentation

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Abstract

Cholangiocarcinomas are primary malignant tumors of the liver that arises from the epithelium lining the small intra and extrahepatic bile ducts, and has numerous histologic types and growth patterns. At computed tomography (CT) and magnetic ressonance (MR), it has various appearences which motivated the present study; we present a case within an atypical presentation. The objective is to show the variety of differential diagnoses before an atypical aspect and the importance of the imaging methods for the diagnosis. Cholangiocarcinoma is still an uncommon neoplasm, and CT and MR are crucial for accurate diagnosis and for differentiation from other hepatic tumorous and nontumorous lesions.

Keywords

Cholangiocarcinoma, Peripheral Cholangiocarcinoma, Intrahepatic Cholangiocarinoma, Liver Neoplasms, Bile Ducts Neoplasms, Cystic Lesion

1. Introduction

Intrahepatic cholangiocarcinoma is a carcinoma arising from any portion of the intrahepatic biliary ductal epithelium. It is the second most common primary intrahepatic malignancy (10% - 20%) after hepatocellular carcinoma [1] [2]. According to the classification proposed by the Liver Cancer Study Group of Japan, they are classified into three types, according to the macroscopic appearance of the tumor: mass-forming, periductal infiltrative and intraductal growth [3] [4] [5] [6] [7]. Usually, peripheral intrahepatic cholangiocarcinoma presents as a low attenuation solid mass on computed tomography (CT) and as a hyperechogenic/heterogeneous mass on ultrasonography [1] [2] [8] [9]. Magnetic resonance imaging (MRI) studies related to intrahepatic cholangiocarcinoma are

still limited to case reports and small series of patients [10]-[19]; however, some authors describe it as hypointense irregular masses in T1 (*spin echo*) and hyperintense in T2 (*spin echo*) [12], associated to satellite nodules and central scar, which can still be a reliable marker for differentiation of the disease with metastatic liver tumors [11] [12] [13]. Cystic degeneration is a rare development of this tumor [20] and may mislead to other pathologies with cystic appearance such as infection and abscesses.

2. Case Report

A 60-year-old male with increased abdominal volume, asthenia, hyporexia, daily morning fever and weight loss 15 Kg in 1 month. At the clinical examination, the patient was discolored, anicteric and with diffuse abdominal pain on palpation, also had severe right limb pain. Deep venous thrombosis was ruled out. The patient was hospitalized to clarify the wasting syndrome. Admission blood tests are in **Table 1**.

HbsAg (hepatitis B surface antigen, Australia antigen); anti-HBc (Antibody to the hepatitis B core antigen); anti-HBs (Antibody to the hepatitis B surface antigen); Anti-HCV (Antibody to hepatitis C virus). **Table 1** shows anemia, lymphocytosis, elevated C-reactive protein.

Computed tomography (CT) showed multiple rounded hypodense formations scattered by the hepatic parenchyma, with a density of 15 HU and peripheral enhancement (**Figure 1**).

Magnetic resonance imaging (MRI) demonstrated heterogeneous signal liver due to multiple, some confluent, solid-cystic rounded formations with post-contrast enhancement and peripheral washout (**Figure 2**), with discrete restricted diffusion areas (**Figure 3**), besides mesenteric and retroperitoneal lymphadenopathy.

The hepatic lesion was drained, and the result was positive to neoplastic cells. After that the patient underwent hepatectomy; the material was sent to histopathological (**Figure 4**) and immunohistochemical (without images) analysis.

Table 1. Shows anemia, lymphocytosis, elevated C-reactive protein.

Erytrocyties	$3.39 \times 10^6/mm^3$	Aspartate aminotransferase	35 U/L
Hemoglobin	9.4 g/dL	Alanine aminotransferase	35 U/L
Hematócrit	27.8%	Total bilirubin	0.3 mg/dL
Platelets	$470 \times 10^3/mm^3$	Direct	0.2 mg/dL
Leucocites	$9.37 \times 10^3/mm^3$	Indirect	0.1 mg/dL
Neutrophil	74.1%	Gamma glutamyltransferase	200 U/L
Lymphocyte	13.3%	Glucose	111 mg/dL
Monocyte	7%	Potassium	5.2 mEq/L
Eosinophil	4.7%	Sodium	138 mEq/L
Basofilos	0.8%	Creatinine	1.0 mg/dL
HbsAg, anti-HBc, anti-HBs	Negative	C-reactive protein	27.4 mg/dL
Anti-HCV	Negative	Urinalysis	Without changes

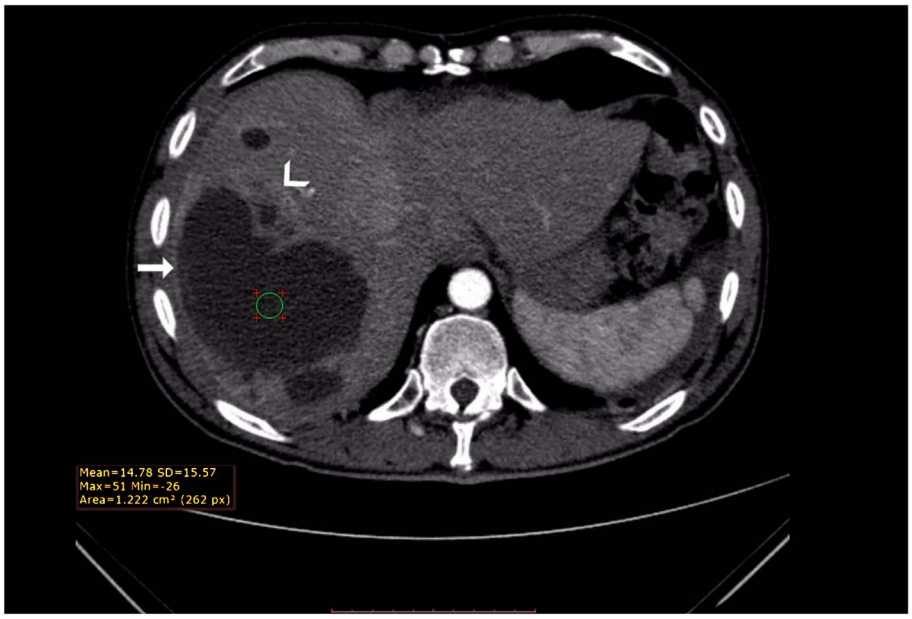

Figure 1. CT, post-contrast arterial phase: predominantly cystic hepatic nodules and masses (arrow) with peripheral enhancement (arrowhead).

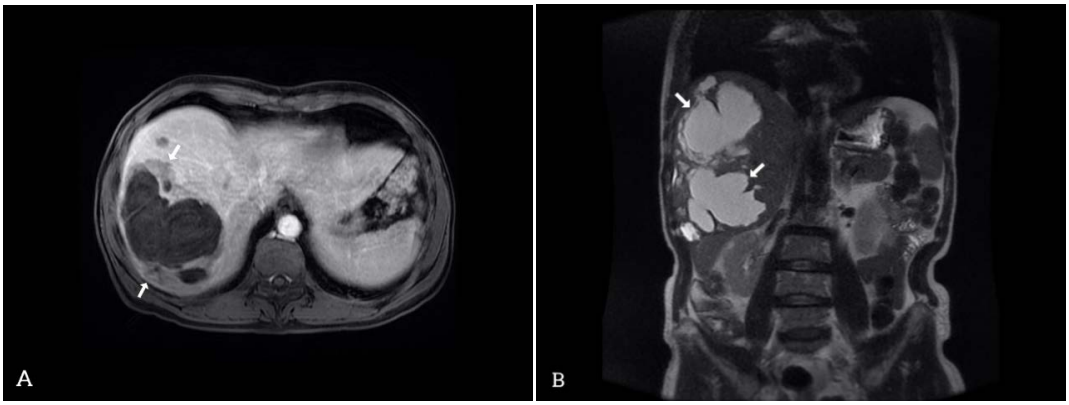

Figure 2. (A) MR, axial T1 post-gadolinium in portal phase: hepatic nodules and masses with peripheral enhancement (arrows). (B) MR, coronal T2 (*fast spin echo*): hepatic nodules and masses with high signal (arrows).

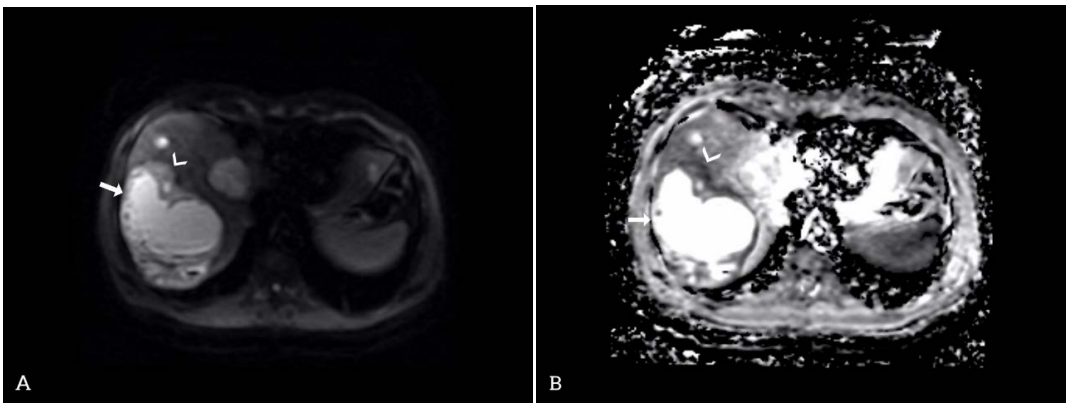

Figure 3. (A) MR, axial diffusion-weighted imaging (DWI): hepatic nodules and masses with high signal (arrow) and peripheral intermediate signal areas (arrowhead). (B) MR, axial ADC map: hepatic nodules and masses with high signal (arrow) and discrete areas of signal drop (arrowhead).

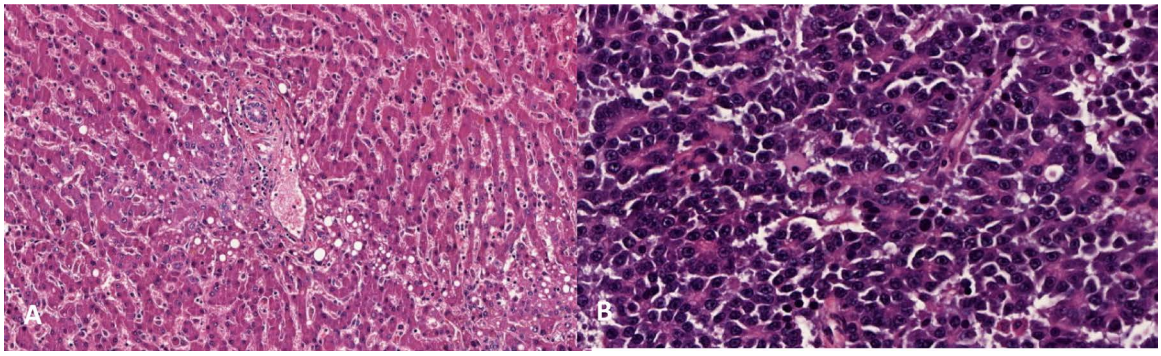

Figure 4. (Histology of the same patient): (A) normal liver; (B) Neoplastic sample (high magnification). Photomicrography of histological laminae of the lesion excision product. (A) shows liver tissue not compromised by neoplasia exhibiting marginal biliary proliferation in the portal spaces associated with steatosis zone 1. (B) shows ductal neoplasia of biliopancreatic pattern, with moderate degree of anaplasia, devoid of necrosis.

Immunohistochemical results: CEA polyclonal – positive diffuse; CA 19: positive; CK 20: positive; CK 7: positive; 34 BE 12: positive; 35 BH 11: positive. The histopatological result was cholangiocarcinoma. Days after the surgery the patient passes away.

3. Discussion

There are a few known risk factors to cholangiocarcinoma, such as chronic inflammation of the biliary tract, cholestasis, congenital abnormalities, intra-hepatic lithiasis, choledochal cyst and chronic liver diseases. As described the related patient has not shown any of these, what turned the case more challenging. From the imaging point of view, there were interesting findings, as well. The most common aspect in imaging examinations of the peripheral intrahepatic cholangiocarcinoma is a single solid mass, well rounded, with a hypovascular, lobular contour. Usually there are no signs of chronic disease in the liver. Satellite nodes of varying sizes can be found due to dissemination through the portal vein. On MRI, the tumor is hypointense in T1 and hyperintense in T2, but may be hypointense in T2 [14] [21]. On CT or MR post-contrast agent, a thin or thick annular peripheral enhancement is often seen around the periphery of the tumor in arterial phases, with gradual centripetal enhancement in the late phases [15] [21]. There may be enhancement of the whole mass a few hours after administration of the contrast medium. In some cases, the central enhancement may be heterogeneous with a central scar. Capsular retraction is relatively frequent.

In our case, we observed multiples nodules and solid-cystic masses, some with peripheral enhancement and slight restricted diffusion areas, besides intra and retroperitoneal lymphadenomegaly. Its solid-cystic aspect, uncommon to adenocarcinoma, could present difficulties for diagnosis. The hypotheses of bacterial and amoebic liver abscess, hydatidosis and peripheral cholangiocarcinoma were considered. Anatomopathological examination showed bile ducts adenocarcinoma. Therefore, the possibility of cholangiocarcinoma, although atypical-

ly, should be remembered in the presence of solid-cystic hepatic lesions. An important factor in this case, was the precocious staging, before any use of bile duct stent, or surgical approach, which could cause anatomical distortion and consequential misleading staging.

4. Conclusion

Cholangiocarcinoma is still an uncommon neoplasm, corresponding to approximately 3% of the gastrointestinal tumors. Nonetheless, it needs to be considered a differential diagnose when the medical board is against peculiars clinical symptoms, pathological background history and radiological findings that may suggest a biliary disease. When that rare neoplasm assaults a patient, it is usually diagnosed at late stage because of its insidious symptoms, and sometimes, silent signals. That is why, usually, when seen by the first time in an imaging exam, the cholangiocarcinoma already has big dimensions, especially when it is intra-hepatic and peripheric, which was the case just reported.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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