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EXTRAHEPATIC BILIARY CYSTADENOCARCINOMA MIMICKING KLATSKIN TUMOR

Cistoadenocarcinoma biliar extra-hepático mimetizando tumor de Klatskin

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INTRODUCTION

Biliary cystadenocarcinoma (BCAC) is a very rare malignant cystic neoplasm. Some authors think this particular neoplasm was conversion of biliary cystadenoma over several years of evolution. In the majority of the cases, it habitually occurs in the liver parenchyma (intrahepatic cystadenocarcinoma); sometimes can be observed extrahepatic biliary origin[3,4,6,7,8,10,11] extrahepatic biliary cystadenocarcinoma, (EBCAC), which generally leads jaundice associated with palpable mass[1,2,5,6,9]. In the past, the diagnosis was done by means of endoscopic retrograde cholangiopancreatography. However, actually the computed tomography or even nuclear magnetic resonance are preferred due non-invasive approaches[1,2,3,4,5,6,7,8,9,10].

Sometimes, EBCAC has difficulties on differential diagnosis with cholangiocarcinoma, mainly when occurs in the hepatic hilum[6]. The surgical treatment provides good prognosis; however, en-bloc biliary tract resection with hepatectomy may be necessary to get free margins[3,6,10]. To date, was not found any case reported in Brazil, nor successful operation using a en-bloc biliary resection with hepatectomy to treat it.

CASE REPORT

A 54 year-old caucasian man presented with painless obstructive jaundice and hepatic palpable mass which began one month before. Liver functions tests showed elevated bilirubin levels of 14,8 ng/dl, alkaline phosphatase of 1067 U/l, gamma-glutamyl transferase of 550 U/l, AST of 175 U/l, and ALT 143 U/l. The alpha-fetoprotein was normal while Ca19,9 was high about 345 U/l. The patient underwent a computed tomography that showed a cystic lesion with irregularity and thickened wall in conjunction with dilated intrahepatic bile ducts mainly left side with atrophy of left lobe. Subsequently, he was submitted a cholangioresonance that showed dilatation of intrahepatic biliary tree more significant at the left side biliary duct with irregularity and thickened wall close to hepatic confluence (Figure 1). The patient underwent radiological examination without signals of systemic dissemination. Therefore, main diagnosis was hilar cholangiocarcinoma or extrahepatic biliary cystadenoma or cystadenocarcinoma. Surgical treatment was indicated, and resection of the suprapancreatic biliary tree including hilar confluence and en-bloc extended left hepatectomy with caudate lobectomy was performed. A formal hilar lymphadenectomy was also realized during surgical resection (Figure 2). One self limited biliary leakage was observed, and treated by conservative management. The patient was discharged at 15th postoperative day. Definitive histological examination showed cystadenocarcinoma of left hepatic duct with free

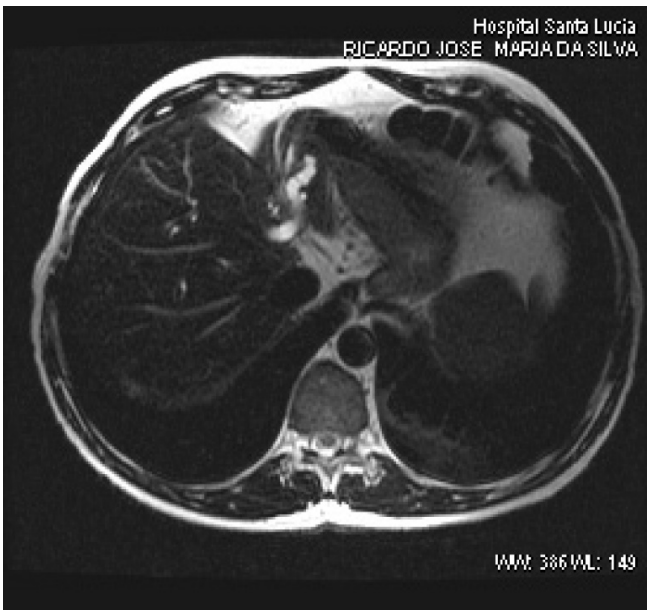

FIGURE 1 - Abdominal magnetic resonance: predominant dilatation of the left biliary tree with atrophy of left hepatic lobe

margins (Figure 3). Immunohistochemical studies showed positive reactions for carcinoembryonic antigen (CEA), cytokeratin 19 and CA 19.9. No postoperative adjuvant treatment was performed. To date, one year of follow-up, the patient is alive without tumor recurrence.

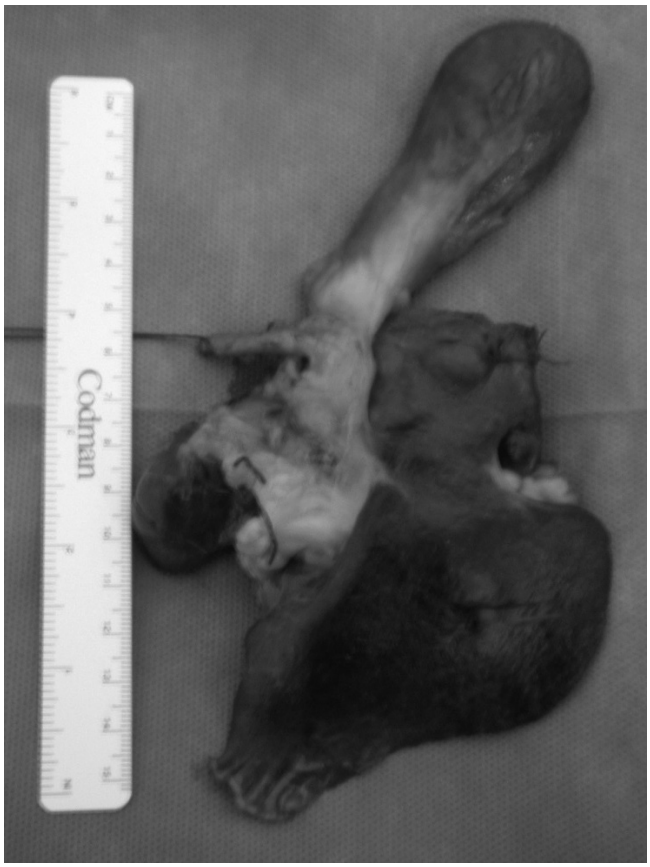

FIGURE 2 - Resection of the biliary tree including hilar confluence and en-bloc extended left hepatectomy with caudate lobectomy

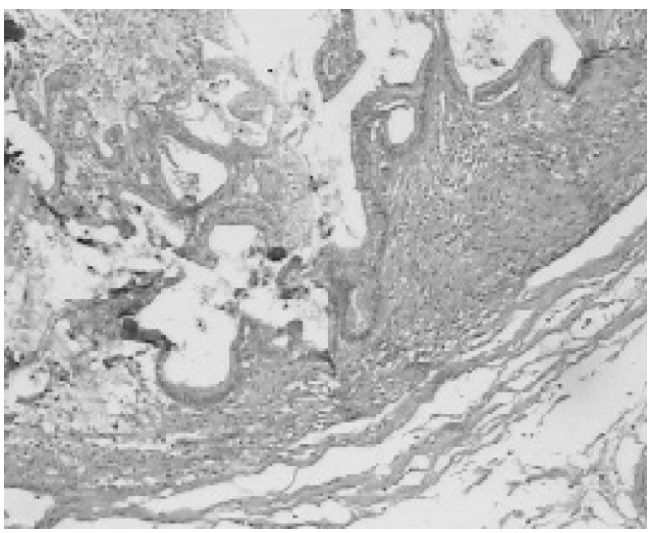

FIGURE 3 - Microscopy with multiple cyst formation with a papillary growth pattern of tumors cells that show cellular stratification with nuclear atypia and mitotic activity (hematoxylin & eosin, x 20)

DISCUSSION

BCAC is a rare malignant epithelial neoplasm of bile ducts. Takayasu et al.[9] reported an incidence of 0,41 % among all hepatic malignant epithelial tumors. Almost all of these tumors are intrahepatic while the minority arise in the major bile ducts or even gallbladder (extrahepatic origin). EBCAC arise more frequently hepatic confluence or even sectorial duct (left or right) than gallbladder[1,3,6,7,8,10]. There are few reports on literature about these uncommon neoplasms[1,2,3,4,5,6,7,8,9,10]. Especially in Brazil, no case was reported. Azambuja et al.[1] reported a single case but this tumor was an intrahepatic BCAC[1]. This tumor is more frequently observed in middle-aged people and may occur in both gender having a less favorable prognosis in man[3]. In accordance to Devaney et al.[4] there are at least two well-defined types of hepatobiliary cystadenocarcinoma. One developing exclusively in female patients, usually accompanied by an "ovarian-like" stroma, which follows an indolent course and the other, lacking the distinct cellular stroma, seen in males, follows an aggressive biological behavior that may result in the patient's death from tumor[10].

While intra-hepatic BCAC leads abdominal mass or pain, EBCAC leads generally obstructive jaundice that may eventually be accompanied by symptoms. More lately, it may lead weight loss and ascites that denotes advanced disease[1,3,6,7,8,10].

Despite its rarity, EBCAC should remind as differential diagnosis with hilar cholangiocarcinoma in patients who present high obstructive jaundice as the present case[10]. Like this tumor present more loco-regional dissemination than distant metastasis[1,10].

Histological, BCAC are characterized by papillary projections with a multilayered cell lining with foci of dysplasia and moderate mitotic activity[10]. In BCAC most tumor cells were positive on immunohistochemical staining for cytoqueratin, epithelial membrane antigen, CA 19.9 and carcinoembryonic antigen[4].

Although the diagnosis is histological and may be confirmed by immunohistochemistry, image examinations may contribute for suspect diagnosis. Computed tomography and more recently magnetic cholangioresonance may reveal multilocular cystic lesion with internal septations or nodularities. More rarely, as the case described, an unilocular lesion may be observed[6]. Some authors have suggested that presence of nodules or coarse calcifications along the wall or septa favor the BCAC[2,6]. However, as reported Tseng et al.[10], EBCAC may be confounded with hilar cholangiocarcinoma because may lead dilatation of the intrahepatic biliary ducts and atrophy of the affected lobe such as observed in present case. Different from Tseng et al.[10], the present authors observed no hepatolithiasis in this case. The real cause of atrophy of the left hepatic lobe was unknown.

Tseng et al.[10] suggested that the tumor would lead compression of the left trunk of the portal vein that could deviate hepatic flow for right lobe. As this case of the EBCAC, it presents a difficult differential diagnosis with common hilar cholangiocarcinoma because on image examinations EBCAC may be confounded due intense dilatation of intra-hepatic biliary tree. Independently of the diagnosis as EBCAC as hilar cholangiocarcinoma, the treatment has been total resection of the lesion with free margins, because partial resection has been associated a high levels of recurrence and dismal prognosis[1,2,3,4,5,6,7,8,9]. Generally, surgical resection may be associated with hepatectomy, due proximity of the hepatic parenchyma[3,10]. Radical resection is the only curative treatment and may be associated either cure or long-term survival[1,2,3,4,5,6,7,8,9]. The overall prognosis may be good since a radical resection is performed, as in the present case.

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