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Massive left lobe primary hepatolithiasis with extremely high CA 19.9

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Hepatolithiasis (HL) or intrahepatic calculi is an uncommon condition in Western countries, with a prevalence under 1%. It is much more frequent in East Asia, reaching 20% in China and Taiwan, up to 50% of which show associated cholelithiasis [1]. The authors report a European patient with primary HL of the left hepatic lobe, 12 years after cholecystectomy for acute calculous cholecystitis, now successfully treated after left lobe hepatectomy.

A 63-year-old caucasian man with a past medical history of hypertension, coronary artery disease, dyslipidaemia and urgent cholecystectomy 12 years ago (due to an acute calculous cholecystitis without ultrasonographic or perioperative evidence of bile duct dilation or choledocholithiasis, and with no cholangitis ever since) was now admitted to our ward following a 2-day course of fever (39ºC), epigastric pain, nausea, vomiting and tender right upper abdominal quadrant.
Bloodwork showed mild leukocytosis with neutrophilia and low platelets; elevated C-reactive protein, γGT, alkaline phosphatase and LDH; normal AST, ALT, bilirubin and amylase. Negative HBV, HCV and HIV serologies. CA19.9 was notably high (7500 U/mL), 200x above normal range (N≤37). A diagnosis of acute cholangitis was assumed on clinical and laboratory basis, despite the absence of jaundice. The patient began therapy with iv ceftriaxone and improved clearly over the next few days. Raised CA19.9 in the setting of acute cholangitis, however, forced us into further study directed at the possibility of underlying biliary or pancreatic malignancy.
Abdominal ultrasound disclosed multiple calcifications in the left lobe. CT displayed several left lobe intrahepatic bile duct dilations but no calcifications, thus suggesting intrahepatic cholesterol stones and cholangitis due to an obstacle before the hilum. MR-cholangiography was performed, showing marked dilation of the left intrahepatic bile duct and moderate dilation in the extrahepatic portion of the common bile duct (CBD). Neither exam showed any sign of cancer. Surgery was the therapeutic approach, and the patient underwent a left hepatectomy.
Intra-operative ultrasound study unveiled severe dilation of the left intrahepatic bile duct, which was filled with gallstones, but no choledocholithiasis and no tumour whatsoever. Both the surgical procedure and the post-operative period were uneventful.
Macroscopic liver section showed multiple dilated bile ductules containing fragile yellowish-green gallstones (Fig.1). Microscopy revealed cystic dilations of the left intrahepatic biliary tree packed with intraluminal gallstones (Fig.2), fibrosis and moderate chronic inflammatory infiltrate with lymphoid aggregates (Fig.3). Some areas of the epithelium displayed erosion and reactive changes. No biliary neoplasm was found.
Immunocytochemistry for angiogenesis or lymphocyte membrane markers are not available in our Hospital. Soon after surgery, CA19.9 values decreased abruptly and just 4 months after the left hepatectomy they were back within reference range.

Our working hypotheses were primary hepatolithiasis (HL), secondary HL and Caroli disease.
Secondary HL, due to stone migration from the CBD, was discarded because calculi in the CBD were absent in the cholecystectomy 12 years ago and in ultrasonographic studies both then and now. Caroli disease, a congenital condition characterised by intrahepatic bile duct dilation [2], was contradicted by the ultrasonography in 2000 showing normal bile duct dimension. Excluding these two entities made primary idiopathic HL our final diagnosis.
The patient is now asymptomatic, having returned to his normal life with no limitations.

This report showcases a rare entity known as primary hepatolithiasis, usually causing recurrent cholangitis in older patients of Asian descent but seldom seen in Europe [3]. The steep increase in CA19.9 related to cholangitis, although previously reported, is also very uncommon [4]. Another interesting aspect is the conspicuous inexistence, over 12 years, of recurrent episodes of acute cholangitis (a traditional finding in hepatolithiasis). These three features help to compose this most peculiar case.

References
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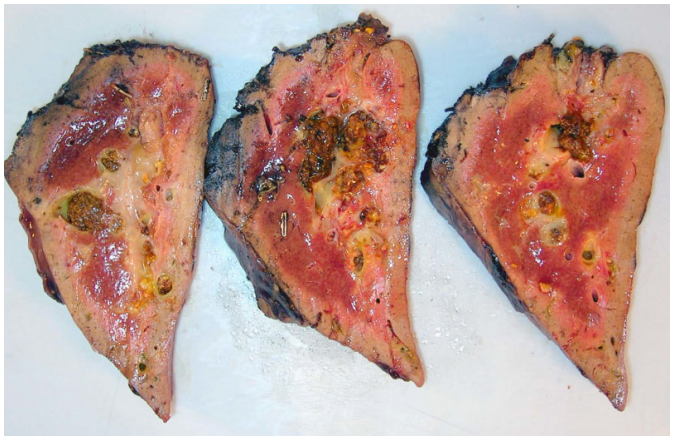

Figure 1: Macroscopic image of left liver cut sections after left hepatectomy. Several yellowish-green gallstones take centre stage, occupying grossly dilated bile ductules.

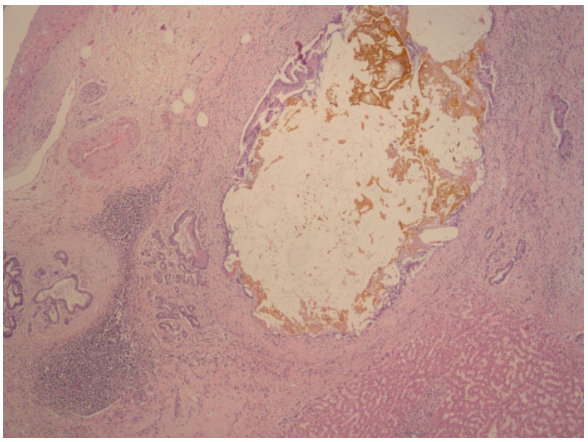

Figure 2: Left lobe liver histopathology (H&E 40x). Bile duct cystic dilation with intraluminal gallstone.

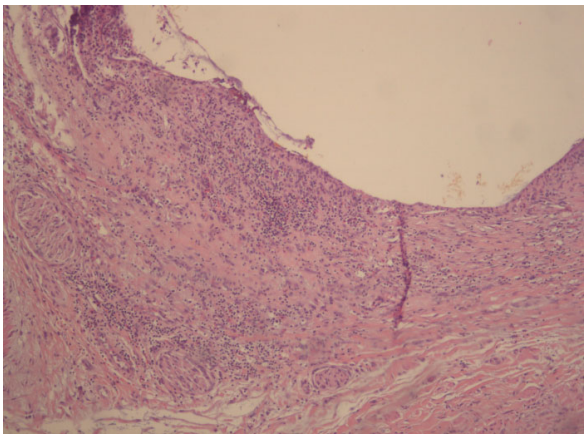

Figure 3: Left lobe liver histopathology (H&E 100x). Fibrosis and moderate chronic inflammatory infiltrate with lymphoid aggregates and areas of acute reactive changes.

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