A 56-year old man was referred for painless obstructive jaundice and pruritus. Laboratory analysis showed elevated hepatic enzymes and raised CA 19-9 levels. Sonography of the abdomen revealed dilatation of the intrahepatic bile ducts and a normal diameter of the distal part of the common bile duct.

Imaging Findings:

Multidetector CT examination including unenhanced scanning, with a detector collimation of 16x1.5 mm and triphasic contrast-enhanced CT at 25 sec (arterial phase), 45 sec (CT cholangiography) and 70 sec (portal phase), with a detector collimation of 16x0.75 mm was performed. Mild intrahepatic bile duct dilatation was detected, abruptly terminating in a level above the hepatic hilum, within an ill-defined mass, moderately and heterogeneously enhancing (Fig. 1). The proximal part of the common bile duct had a focally thickened wall, hyperdense relative to liver. The CT findings were suggestive for the presence of cholangiocarcinoma, of both infiltrating and mass-forming type. CT angiography revealed patency of the proper and main hepatic arteries (Fig. 2). Portal phase showed narrowing of the right portal vein, a finding suggestive for neoplastic infiltration (Fig. 3).

The patient underwent a right hepatectomy, cholecystectomy, resection of the extrahepatic biliary tree, and biliary-enteric anastomosis (hepatico-jejunostomy). Histopathology confirmed the CT findings (Fig. 4).
Discussion:

Background
Hilar cholangiocarcinoma or Klatskin tumour is a primary malignancy typically originating at the confluence of the right and left hepatic ducts within the porta hepatitis [1, 2]. Prognosis is poor, with a 5-year survival rate of 0%, if it is unresectable and 25-40% after curative resection. Less than 50% of patients are candidates for curative resection, because of the tendency for invasion of the adjacent organs, including the large portal veins and hepatic arteries. Imaging plays an important role in determining whether a patient is suitable for a complicated surgery, as in this patient.

Cholangiocarcinoma is classified into three types, based on macroscopic appearance: periductal infiltrating, mass-forming, and intraductal [1]. Periductal infiltrating is the commonest form involving the hilar area. More than one type may coexist in a single patient.

Imaging Perspective

Multidetector CT (MDCT), with multiphasic imaging (CT angiography and cholangiography), multiplanar reformations and three-dimensional (3D) reconstructions represents an excellent imaging modality for the assessment of the soft-tissue extent of hilar cholangiocarcinoma and its relationship with hepatic vessels [3-7].

The classical CT signs of the infiltrating type of cholangiocarcinoma include a focally, irregularly thickened ductal wall, usually hyperdense when compared to hepatic parenchyma [1-7]. Other signs include asymmetric upstream dilatation of the intrahepatic ducts with isodensity relative to water, unless stone or sludge coexists, combined with nondilated extrahepatic ducts and diffuse enhancement around hilum. These features were met in this patient, and proved in accordance with the operative and the histopathologic findings.

Tumour resectability is based on the following criteria: site and type of biliary obstruction, as proposed by Bismuth classification, relationship of the tumour and blood vessels at porta hepatis, signs of neoplastic invasion of the second-order branch of the bilateral hepatic ducts, tumour invasion of the proper, right and left hepatic arteries, invasion of both the right and left branches of the portal vein, signs of distant metastasis or retroperitoneal lymph node metastasis [3-7]. The detection rate for cholangiocarcinoma and the assessment of tumour resectability by MDCT is reported up to 100% and 75-92%, respectively [3].

CT signs for diagnosing invasion of the hepatic vessels include irregularity of the endoluminal surface, straightening, narrowing or occlusion, contour deformity and more than 50% of the vessel circumference detected in contact with the neoplasm. MDCT with angiography with 3D reconstructions allows an accurate delineation of the relationship of the tumour and hepatic vasculature providing important preoperative information of tumour resectability [3-7].

Differential Diagnosis List: Hilar cholangiocarcinoma, Inflammatory stricture, Including primary and secondary sclerosing cholangitis, Mirizzi syndrome, Xanthogranulomatous cholangitis, Biliary sarcoidosis, Hepatocellular carcinoma, Intrabiliary metastases, Biliary tract melanoma, Lymphoma, Leukaemia, Carcinoid tumour

Final Diagnosis: Hilar cholangiocarcinoma

References:

Figure 1

Description: (a) Non-contrast CT image, coronal reformation depicts a soft-tissue mass (arrow, CT density: 45HU) in the hepatic hilum and dilated intrahepatic bile ducts. Common bile duct wall is thickened (arrowhead) and hyperdense relative to liver. Origin: Ntorkou A, Department of Radiology, Ioannina, Greece
Description: (b) CT cholangiography, coronal reformation shows heterogeneous enhancement of the mass (arrow, CT density: 100 HU) and focal thickening of the wall of the proximal common bile duct (arrowhead). **Origin:** Ntorkou A, Department of Radiology, Ioannina, Greece

description: (d) CT cholangiography, transverse reformation shows enhancement and focal thickening of the wall of the proximal common bile duct (arrowhead), a finding suggestive for the presence of infiltrating cholangiocarcinoma. **Origin:** Ntorkou A, Department of Radiology, Ioannina, Greece
Description: (e) CT cholangiography, transverse reformation shows normal appearance and diameter of the distal part of the common bile duct (arrowhead). Origin: Ntorkou A, Department of Radiology, Ioannina, Greece
Description: (a) Reconstructed 3D CT angiography, transverse plane depicts normal celiac, right and left hepatic arteries. Origin: Ntorkou A, Department of Radiology, Ioannina, Greece
Description: (b) Reconstructed 3D CT angiography, coronal plane depicts patency of the right and left hepatic arteries (arrowhead). Origin: Ntorkou A, Department of Radiology, Ioannina, Greece
Description: (a) Reconstructed 3D CT portovenogram, transverse plane depicts narrowing of the right portal vein (pointer), a finding suggestive for neoplastic invasion. This was confirmed both surgically and pathologically. Origin: Ntorkou A, Department of Radiology, Ioannina, Greece
Description: (b) Reconstructed 3D CT portovenogram, coronal plane shows normal main trunk of the portal vein (arrowhead) and narrowing of the right portal vein (pointer). Origin: Ntorkou A, Department of Radiology, Ioannina, Greece
Description: Histology revealed a moderately differentiated adenocarcinoma (asterisk) in the perihilar region (H-E X200). Origin: Ntorkou A, Department of Radiology, Ioannina, Greece.

Description: The tumour invades the adjacent hepatic parenchyma (asterisk, H-E X200). Origin: Ntorkou A, Department of Clinical Radiology, Ioannina, Greece.
Description: Perineural invasion (arrow) was identified (H-E X200). Origin: Ntorkou A, Department of Clinical Radiology, Ioannina, Greece.

Description: The tumour invades the muscular wall of the right portal vein (arrow, H-E X200). Origin: Ntorkou A, Department of Clinical Radiology, Ioannina, Greece.