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Case Report

Klatskin tumor presenting as unresectable perihilar hepatic mass: A case report^{☆,☆☆}

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ABSTRACT

Cholangiocarcinoma is a rare liver tumor with three types: intrahepatic, extrahepatic, and perihilar, which alone account for about 50% of cases. The diagnosis is late with a poor prognosis. Imaging through Bili MRI and CT scan plays an essential role in the classification and staging of tumors for therapeutic management. We report the case of a 62-year-old woman, received for cholestasis syndrome and weight loss in whom CT and MRI found a hepatic hilar mass. The diagnosis of Klatskin's tumor was retained with a histological finding of adenocarcinoma. The patient underwent biliary drainage and palliative treatment as the tumor was unresectable.

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Introduction

Perihilar cholangiocarcinoma accounts for more than half of all hepatic cholangiocarcinomas (CCA), which remains a rare condition involving the bile ducts. It is the second most common liver cancer. Its incidence has increased over time and it is diagnosed at a later stage. It is presented according to its location in three types: intrahepatic, perihilar, and extrahepatic. Klatskin tumor is a particular form of cholan-

giocarcinoma characterized by a peri hilar location of the liver process. In 1965, Gerald Klatskin described tumors arising in the main bile duct, thus giving the name Klatskin tumor [1]. Functional signs are not always specific. The diagnosis is made by combining CT and MRI scan data to identify the tumor and its location. This results in the usual classification, that of Bismuth-Corlette. Histological confirmation will be done later. Treatment is discussed on a case-by-case basis. Mortality remains high.

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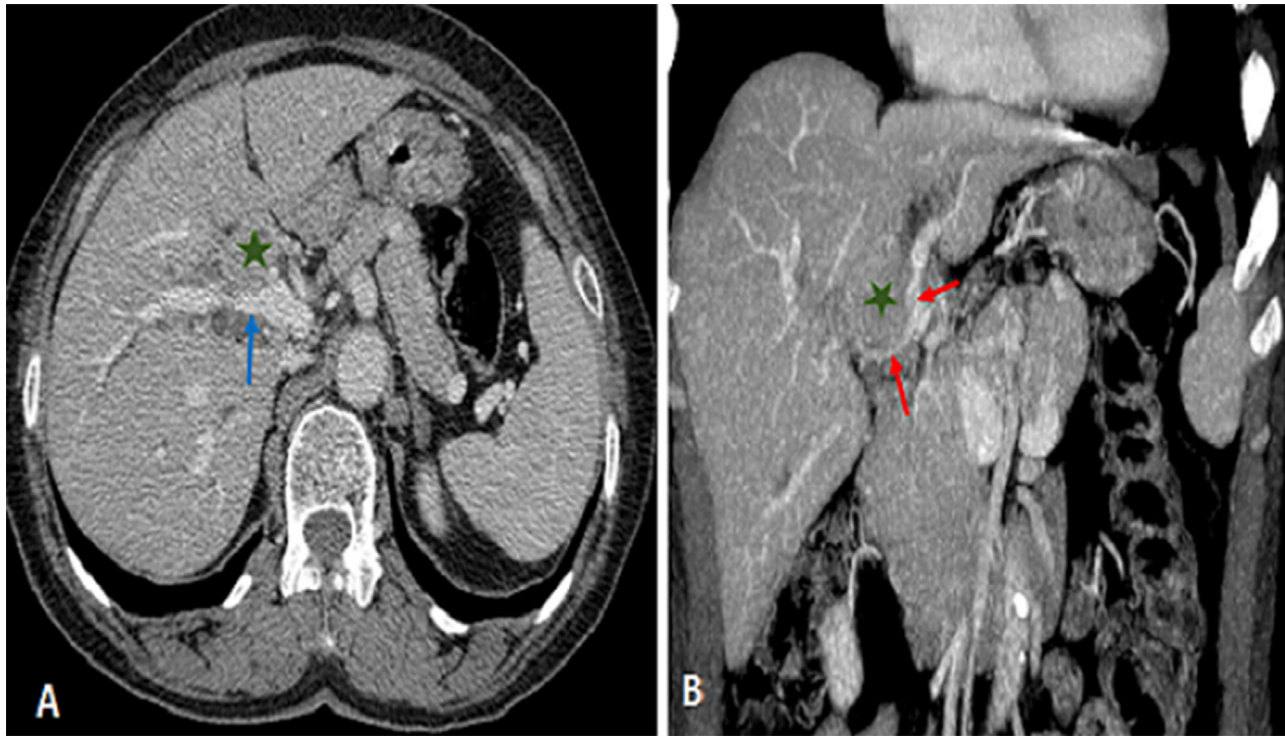


Fig. 1 – Abdominopelvic CT scan injected in axial (A) and coronal (B) slices showing an isodense hepatic hilar mass (green star), enhanced with extension to portal venous (blue arrow) and hepatic arterial (red arrow). (Color version of figure is available online.)

Case report

A 62-year-old woman who consults for progressive jaundice. She was consulted in a hospital in the area where a biological assessment revealed a syndrome of cytotoxicity and cholestasis. She had an abdominal and pelvic CT scan as part of her workup. The CT scan revealed a hepatic hilar mass, raising suspicion of a tumor etiology.

An MRI was then performed to better characterize the hilar mass and its relationship with the bile ducts.

The CT scan revealed an isodense hepatic hilar mass that was discretely enhanced after injection of iodinated contrast. Topographically it comes into intimate contact with the right portal branch and intimate contact with the right and left hepatic arteries. There is associated dilatation of the intrahepatic bile ducts (Fig. 1).

The MRI scan showed a rounded process in the hepatic hilum, with a T1 hypointense, T2 intermediate signal, and a diffusion hypersignal with ADC restriction of $0.81 \times 10^{-3} \text{mm}^2/\text{s}$, enhanced after injection of gadolinium (Figs. 2–3).

This mass infiltrates the PVB, the hilar convergence, and extends to the right and left bile ducts responsible for dilatation of the intrahepatic bile ducts.

It comes into intimate contact with the right hepatic artery which is compressed but remains patent and the right portal branch which remains patent.

The cholangio-MRI sequence shows a hepatic hilar infiltrating mass and dilatation of the intrahepatic bile ducts without dilatation of the main bile duct (Fig. 3).

The Carbohydrate Antigen CA 19.9 level was at 95 U/ml.

The diagnosis of hepatic hilar cholangiocarcinoma or Klatskin's tumor was made. It is classified as Bismuth IV and according to the Oliveira classification as B4 FA T3 PV3R HA4 N0 M0.

Histological evidence was confirmed as adenocarcinoma. The tumor was unresectable due to vascular relationships, so the patient underwent biliary drainage and was put on palliative treatment.

Discussion

Hepatic hilar cholangiocarcinoma or Klatskin's tumor is a rare tumor of late diagnosis and postoperative histological evidence. It has a poor prognosis. In the United States between 2011 and 2013, the incidence of cholangiocarcinoma was 9.8 cases per 1,000,000 people per year with an identical incidence in females and males. In 50% of cases, it was a perihilar cholangiocarcinoma compared to 25% for intra and extrahepatic cholangiocarcinomas (Fig. 4A). Its peak is in the 6th decade [2].

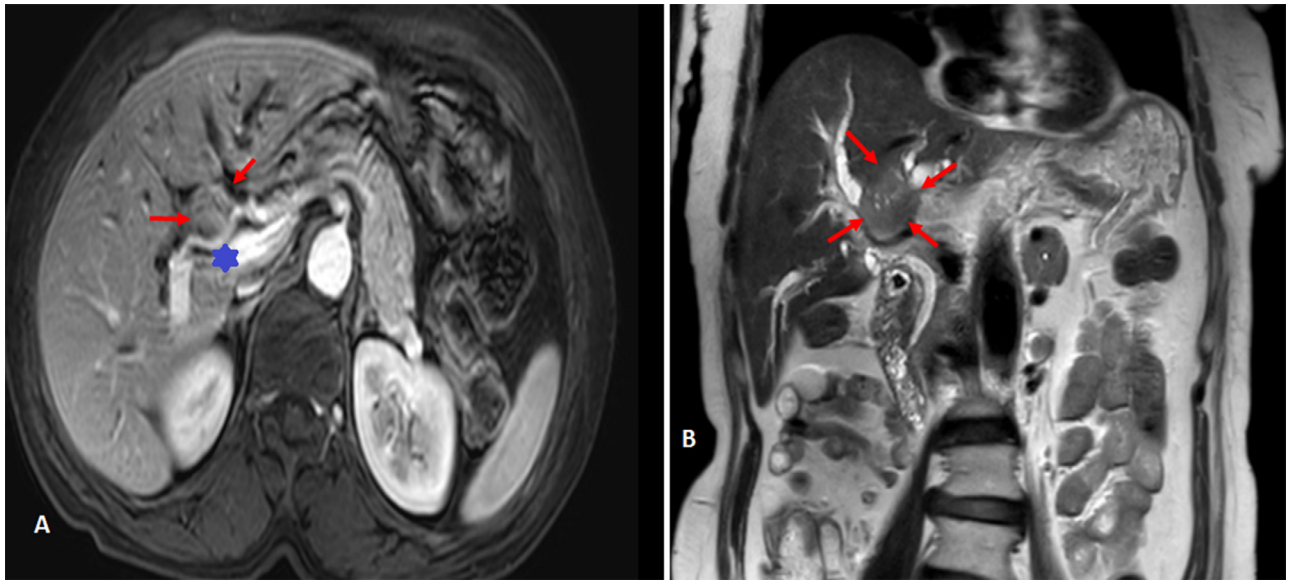


Fig. 2 – MRI T1 FS-Gadolinium axial section (A), and T2-weighted coronal section (B) showing a hepatic hilar mass (red arrow) with intermediate T2 signal, enhanced after gadolinium injection with extension to hepatic artery (blue star). (Color version of figure is available online.)

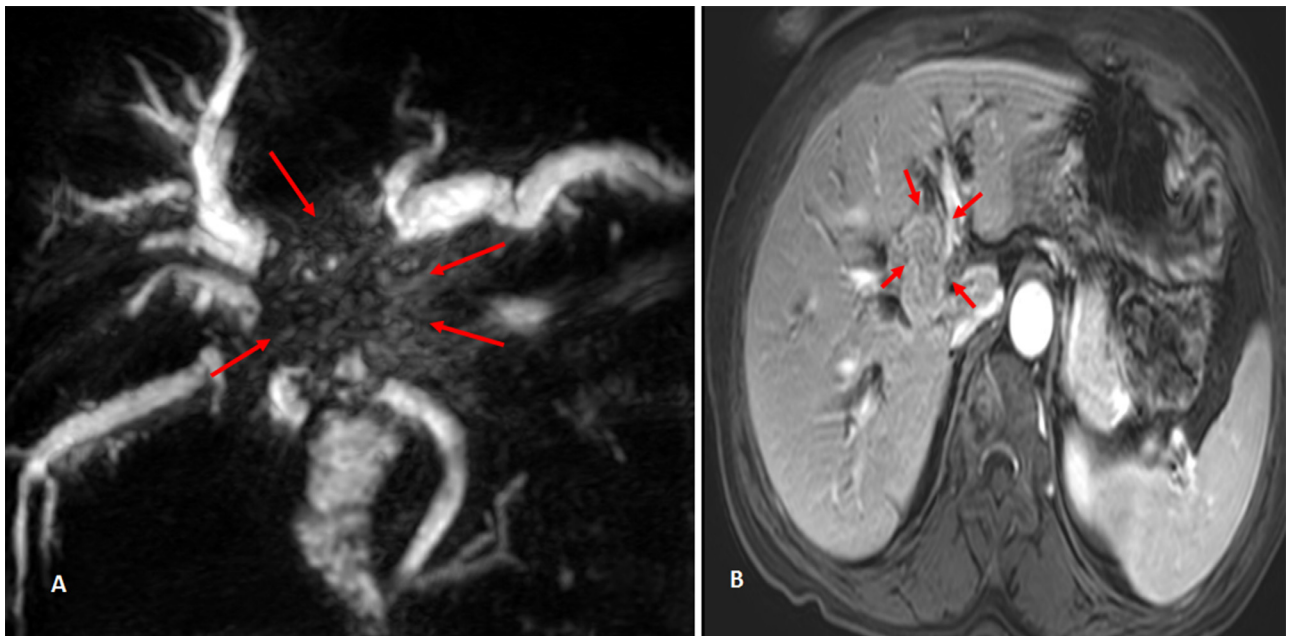


Fig. 3 – cholangio MRI (A) and T1FS gadolinium sequence (B) reveals an infiltrating mass of the hilar convergence and the right and left bile ducts (red arrow) with discrete dilatation of the intrahepatic bile duct. (Color version of figure is available online.)

Certain risk factors have been identified, in particular primary hepatobiliary diseases such as primary sclerosing cholangitis, genetic disorders, chronic liver diseases such as cirrhosis, smoking, chronic hepatitis, exposure to toxic substances, polycystic fibrosis (Caroli disease), lithiasis or non-lithiasis cholecystitis, and biliary inflammatory diseases [3].

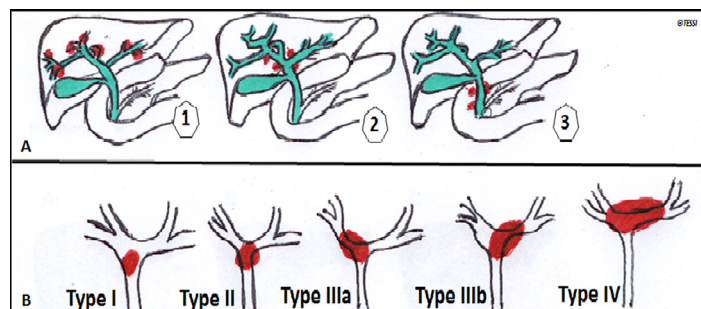
Castano Llano R, in his study found jaundice with abdominal pain as frequent symptoms. In order of frequency, pruri-

tus (66%), abdominal pain (40-50%), weight loss (40-50%), and fever (20%). It should also be noted that the triad of cholestasis syndrome, abdominal pain, and weight loss is suggestive of pancreatic or hepatobiliary cancer [4].

The blood test will include a liver test, namely alkaline phosphatase, bilirubin, gamma glutamyl transpeptidase. The search for tumor markers is systematic: Carbohydrate Antigen CA 19-9, CA 125, and Carcinoembryonic Antigen CEA. However,

Table 1 – Oliveira et al, classification of perihilar cholangiocarcinoma (BTFFPVHANMDV).

	Items description			
B: Bismuth and Corlette	Type 1	Type 2	Type 3	Type 4
T: Tumor	T1: 1cm	T2: 1-3cm	T3: 3cm	
F: Form	A. Mass	B. Periductal infiltrating: sclerosing	C. Intraductal growth: Polypoid	Mixed
PV: portal vein extension is the same as biliary extension	PV1	PV2	PV3R-PV3L	PV4
HA: hepatic artery extension is the same as the biliary extension	HA1	HA2	HA3R – HA3L	HA4
N: lymph nodes	N0 or N1			
M: metastases	M0 or M1			
D: liver diseases	Cholangitis Sclerosing Primary			
V: Volumetry				

**Fig. 4 – (A) Anatomic variants of cholangiocarcinoma. (1) Intrahepatic cholangiocarcinoma (2) Perihilar cholangiocarcinoma (Klatskin tumor), (3) Extrahepatic cholangiocarcinoma (B) Classification of Klatskin tumor by Bismuth-Corlette**

the levels are not specific and are increased in other hepatic cancer and inflammatory diseases. It can be as high as 85% in CCA, 30% for CEA, and 40-50% for CA 125. An increase of 68IU/L over baseline for CA19-9 has a specificity of 98% and a sensitivity of 90% compared to CCA [5,6].

The first line imaging remains ultrasound by demonstrating dilatation of the bile ducts, the presence of a perihilar mass (rarely) or tissue formation within the bile ducts, the presence or absence of lithiasis. It can detect an abnormality of the hepatic vascular pedicle on Doppler and the vascularisation of the mass [5].

The CT scan highlights the hypodense hilar mass of variable enhancement, but above all the vascular relationships of this mass with the hepatic pedicle, allowing classification. It also allows the lymph node extension and metastases to be assessed [5]. CT scan will also allow tumor staging through the TNM classification but also according to the 8th edition of the American Joint Cancer Committee/Union Internationale Contre le Cancer (AJCC/UICC). There is also the MSKCC (Memorial Sloan-Kettering Cancer Center) classification

Hepatic MRI with cholangio-MRI allows rapid, non-invasive, and non-radiating exploration of the bile ducts, parenchyma, and liver pedicle. [4,5]. It allows a better assessment of the tumor size in the form of a mass in T2 or intermediate T hypersignal, in diffusion hypersignal with an ADC, with a more marked enhancement at the late stage, the invasion of the bile ducts and in particular thanks to the Bili MRI

sequence (cholangiography) to assess the extension and invasion of the bile ducts. Angiography may be associated with vascular extension [5,7].

New techniques also exist, in particular endoscopic ultrasound, which allows the visualization of the bile ducts, the assessment of the gallbladder, the vascular pedicle, the adenopathies, and the taking of biopsies. We can mention percutaneous flexible cholangioscopy.

PET scan can also be performed to assess glucose uptake by these tumors.

Three types of tumors are found on imaging: perihilar or Klatskin tumors, intrahepatic cholangiocarcinoma, and extrahepatic cholangiocarcinomas (Fig. 4A). Extension to the bile ducts will be according to the Bismuth-Corlette classification (Fig. 4B):

- Type I: tumors below the confluence of the left and right hepatic ducts;
- Type II: tumors reaching the confluence but not involving the left or right hepatic ducts;
- Type III: tumors occluding the common hepatic duct and either the right (IIIa) or left (IIIb) hepatic duct;
- Type IV: tumors that are multicentric or that involve the confluence and both the right and left hepatic ducts.

Oliveira's classification is important in surgical management and highlights extension to the bile ducts, tumor size, and shape, extension to the proper hepatic artery and its

branches and to the portal trunk and its branches, metastases, lymph node extension, chronic liver disease, and residual liver volume (Table 1) [8]. The dominant histological type is adenocarcinoma in 95% of cases. Other types are squamous cell carcinoma, carcinoma in situ, mucinous adenocarcinoma, clear cell adenocarcinoma [3,5].

Differential diagnosis were hepatocellular carcinoma, Liver metastases, pancreatic cancer, cholangitis, primary sclerosing cholangitis, fasciola hepatica infection mimicking as CCA, cholecystitis or choledocholithiasis, Biliary strictures, IgG4-associated cholangiopathy. [6].

Fig. 4

Medical and surgical treatment exists but the curative treatment of choice remains surgery. It will depend on the vascular relationships of the mass, and the Oliveira classification is used to better guide the surgeon. Palliative treatment is for advanced cases. The criteria for resectability are the absence of distant hepatic metastases, the absence of retropancreatic or latero-celiac adenopathies, the absence of invasion of the common hepatic artery and the portal trunk, the absence of dissemination of the disease, or extra-hepatic invasion of neighboring organs [3].

The surgical technique varies according to the Bismuth-Corlette classification. A right hepatectomy is performed for type I, type II, and IIIa tumors involving the common hepatic duct, the right hepatic biliary trunk. Left hepatectomy is indicated for type IIIb involving the left biliary branch. For type IV invading the right and left bile ducts a central bisegmentectomy or a right and left trisegmentectomy is performed. When the tumor extends to the lower part of the main bile duct, a duodenopancreatectomy is associated. Postoperative complications exist [9, 10, 11, 12].

Liver transplantation is the treatment of choice for R0 tumors in terms of resectability and vascular pedicle invasion. Palliative treatment is usually given in the absence of transplantation [11,12].

Percutaneous radiofrequency ablation is not successful. Portal embolization may be associated depending on the therapeutic objectives [3]. Other therapeutic alternatives exist, notably radiotherapy, adjuvant chemotherapy, and photodynamic therapy [12].

Survival varies between 5-10% over 5 years [3]. Geng Z-M et al found a 5-year survival of 30% in patients without adenopathy, 15% in patients with regional metastatic adenopathy, and 12% in patients with metastatic para-aortic adenopathy [10]. For Soares KC et al, surgical resection with R0 negative margin remains the best treatment for long postoperative survival. Surgical resection will depend on the tumor ratios with morbidity between 40-70% and mortality between 5-15% [12].

Conclusion

Perihilar cholangiocarcinoma or Klatskin's tumor remains a rare pathology of the like cholangiocarcinoma in general. It remains the most frequent type of CCA with about 50%. It is often diagnosed late with a poor prognosis. Imaging is dominated by Bili MRI combined with CT for optimal classification and tumor staging. This will guide the therapeutic management with surgery as the treatment of choice. Survival at 5 years remains poor and varies between 10-15%.

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