

Klatskin's tumor as a cause of uncertain jaundice: Case presentation

Summary

Introduction: Klatskin tumor is the most frequent cholangiocarcinoma among tumors of the main bile duct.

Objective: To present a patient diagnosed and treated with the diagnosis of a type II Klatskin tumor.

Case presentation: 66-year-old mestizo male patient who came to our service for jaundice, abdominal pain, itching, weight loss, coluria and acholia.

Discussion: hilar cholangiocarcinoma usually occurs in male patients over 60 years of age, where its clinical picture is characterized by jaundice, abdominal pain and weight loss, where the most important elements for its diagnosis is the presence of a bile duct dilated intrahepatic disease, accompanied by a collapse of the extrahepatic and gallbladder. Surgery is the only effective treatment, since it is a poorly radiochemosensitive tumor.

Conclusions: Hilar cholangiocarcinoma continues to be a difficult entity to diagnose, which delays its initial treatment, thus increasing the morbidity and mortality rate and its poor prognosis. Surgery continues to be the treatment of choice for this entity since it is a non-chemosensitive tumor.

Keywords: klatskin tumor, cholangiocarcinoma, perihilar cholangiocarcinoma

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Héctor Alejandro Céspedes Rodríguez

Department of General Surgery, University of Medical Sciences of Camaguey, Cuba

Correspondence: Héctor Alejandro Céspedes Rodríguez, College of Medical Sciences Farongamagui, University Hospital "Manuelasquens Domench". Kamagui, Cuba, Email hcespedesr03@gmail.com

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Introduction

Klatskin tumor is the most frequent cholangiocarcinoma of the main bile duct,¹ this is located at the level of the confluence of the bile duct, this being according to the Logmare classification, those that occupy the upper third of the extrahepatic bile duct, which represent 50% of the tumors in this location.^{2,3} The first report of this entity was made by Alteimeier in 1957, as carcinoma of the liver, in 1965 Klatskin reported 13 cases and since then in the United States, and this eponym is universally accepted.^{1,2,4} It appears in patients over 60 years of age, generally male, and is characterized by an extrahepatic cholestatic syndrome. Given the low incidence and experience in our center, this presentation is made.

Presentation of the case

66-year-old, white, male patients with a history of high blood pressure. That he was admitted to the General Surgery service of the Manuel Ascunce Domenech University Hospital, for presenting painless jaundice, itching, acholia, coluria and weight loss. Physical examination, no palpable gallbladder was found (no Cruvoisier-Terrier sign), only abdominal excoriations from scratching and verdinic jaundice of the skin and mucous membranes. Hemochemistry shows a cholestatic pattern GGT 137IU, FA 789IU, Direct Bilirubin 234 and Total 278mmol/l. Abdominal ultrasound shows marked dilation of the intrahepatic bile duct, not demonstrating dilation of the extrahepatic, atrophic gallbladder. Contrast CT describes a hypodense lesion at the level of the confluence with marked dilation of the intrahepatic bile duct and collapse of the extrahepatic one. During the intraoperative cholangiography is performed (Figure 1), which shows stenosis at the level of the confluence (Figure 2.) it is decided to perform resection of the extrahepatic bile duct with an intrahepatoduchojejunostomy (Hess procedure) in Loop of Braun, with jejunojejunostomy. The patient

is discharged after ten days. The biopsy reports well-differentiated adenocarcinoma.

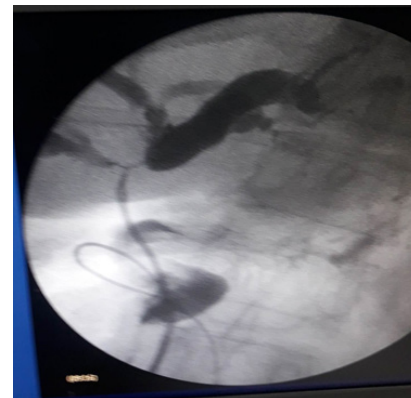


Figure 1 Stenosis is observed at the level of the hepatic confluence.

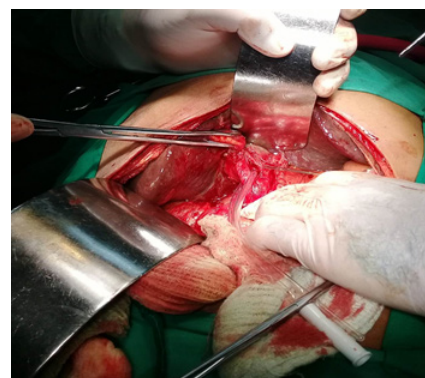


Figure 2 The atrophic gallbladder and the stenosis at the level of the confluence are observed, with great dilatation of the left hepatic.

Case discussion

The klatskin tumor or perihilar cholangiocarcinoma is a malignant tumor originating in the epithelium of the bile ducts in the hilar zone. They have a low incidence but a high mortality rate due to the level of compromise found during diagnosis. Its incidence varies worldwide, being more prevalent in East Asia due to the presence of parasitic infections. It corresponds to less than 2% of all malignant tumors, with an incidence of between 1 and 2 new cases per 100,000 inhabitants per year.^{1,2} The most frequent age of presentation is after the sixth decade of life,¹⁻³ being slightly more common in men than in women (1,3/1).^{1,4} The most common histological type is well-differentiated adenocarcinoma.^{1,3,4} Etiology of cholangiocarcinoma is unknown and in most cases the appearance is sporadic;⁴ however, some risk factors for the development of this type of tumor have been associated. Among these factors we find: primary sclerosing cholangitis whose inflammatory process leads to the development of fibrosis and stenosis of both the intrahepatic and extrahepatic routes where approximately 40-50% of these patients are also carriers of Ulcerative Colitis, polycystic liver disease, parasitic diseases by *Clonorchis sinensis* and *Opisthorchis viverrini*. Cholangiocarcinoma is considered to be any tumor that is located at the level of the bile duct.³

In the clinical picture, constant abdominal pain located at the level of the right upper quadrant (40-50%) and non-painful jaundice due to biliary obstruction (90%) usually preceded by itching (66%).¹ The patient may also present weight loss (40-50%), fever (20%), acholia or coluria. As in the case of our patient, the presenting symptoms are nonspecific.¹⁻⁵ Patients may report pain in the right upper quadrant, weight loss, anorexia, itching, acholia, choluria, and jaundice; the latter will not appear initially if the biliary obstruction is not complete or affects only the segmental ducts.² Laboratory data show a pattern of obstructive jaundice, with hyperbilirubinemia and with increased levels of alkaline phosphatase, gamma-glutamyl -transpeptidase.³⁻⁵

The differential diagnosis of cholangiocarcinoma includes entities such as carcinoma of the gallbladder, Mirizzi syndrome (impaction of a lithiasis in the gallbladder neck), benign focal strictures of the bile duct ("malignant masquerade") and biliary metastases of other neoplasms (pancreas, stomach, breast, lung, or colon).¹ It was previously believed that the only curative therapy for cholangiocarcinoma was complete resection of the tumor with negative histologic margins, however resectability rates were low and many of the patients had recurrences,^{6,7} nowadays it is believed that the most appropriate therapeutic modality for the management of these patients is multimodal therapy based on complete resection of the tumor with negative margins combined with adjuvant or neoadjuvant therapy.⁸ Surgical treatment is the therapy of choice for the management of hilar cholangiocarcinoma and resection of the bile duct will be performed with or without resection. Hepatic action plus lymphadenectomy and biliodigestive anastomosis. The extent of resection depends on the grade of the cholangiocarcinoma according to the Bismuth-Corlette classification.

They have been described as possible adjuvant or neoadjuvant therapies to external radiation therapy or brachytherapy, systemic or arterial chemotherapy, chemo radiation or photodynamic therapy. The use of laparoscopic surgery has been considered for the management of these patients; however, it is still an experimental method and although it may favor early discharge, the benefits remain controversial, and careful selection of the patient must be carried out, so it has not yet been done. it is considered a reliable treatment.⁹ Biliary drainage has been used as preoperative therapy, which has been considered of great importance in patients with liver failure; it is also believed to be the most appropriate method to minimize the risk of dissemination

and inflammatory reactions.¹⁰ Since most patients are diagnosed in late stages of the disease, palliative treatment has become one of the main therapies used. It is indicated in patients presenting with cholangitis and pruritus but also in those with high-grade jaundice and abdominal pain. Endoscopic stent drainage is the preferred palliative modality; however, in advanced stages, better results are obtained by percutaneous stenting.¹¹

In our patient, the presenting symptoms (pain in the right upper quadrant, pruritus, low-grade fever, asthenia, anorexia, choluria, and acholia) suggested a nonspecific obstruction of the bile duct. In the absence of predisposing factors that initially made us suspect biliary tumor pathology, it was essential to perform imaging techniques and a histological study to make the diagnosis. The only potentially curative treatment for cholangiocarcinoma is complete surgical resection of the tumor with histologically disease-free margins.^{1,3,4} Performing complete resection of the extrahepatic bile duct including the gallbladder, with regional lymphadenectomy, to perform biliodigestive anastomosis. We performed Hepp's principle hilar plate descent, we performed a Hess procedure (bilateral Braum loop intrahepaticotojejunostomy).

In the case of hilar cholangiocarcinoma, the surgical technique of choice is resection of the affected bile duct, partial hepatectomies (the caudate lobe is usually invaded), lymphadenectomy, and biliodigestive anastomosis.^{1,4,6} Radiation therapy, chemotherapy, and Liver transplantation has not shown significant utility in improving life expectancy and quality of life in patients with this neoplasm.^{1,2,5}

Conclusions

Hilar cholangiocarcinoma continues to be a difficult entity to diagnose, which delays its initial treatment, thus increasing the morbidity and mortality rate and its poor prognosis. Surgery continues to be the treatment of choice for this entity since it is a non-chemosensitive tumor.

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None.

Conflicts of interest

The authors have declared that they have no conflicts of interest with the publication of this document.

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