

CASE REPORT

Hilar Cholangiocarcinoma Diagnosed and Treated Early, in Prejaundice Phase

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Hilar cholangiocarcinoma, Klatskin tumor or proximal bile duct cancer, is a tumor growing in the right hepatic duct, left hepatic duct or at their confluence. It is a relatively rare but devastating disease. The tight stricture of the biliary ducts and the development of obstructive jaundice are the main characteristics of the disease. In the early phase, symptoms are nonspecific and jaundice is not present, leading to delayed diagnosis and denying the possibility of curative treatment. We present the case of a 74 years old woman who was referred to us with ambiguous symptomatology and without jaundice. The ultrasound and CT scan showed dilation of the left biliary tree, without increase of the cholestatic enzymes. Magnetic resonance cholangiography depicted a tumor in the left hepatic duct (3X3 cm.) with enlargement of the bile ducts above. The surgical treatment consisted of left hepatectomy and hilar lymph nodes dissection. The pathology findings showed a cholangiocarcinoma with a few hilar nodes involvement. Our approach was potentially curative. Unfortunately these situations are seldom because in the majority of cases the patients have obstructive jaundice at presentation and the tumors are unresectable. We consider that a magnetic resonance cholangiography made when we suspect a bile duct tumor, leads us to an early diagnosis and gives us the possibility of a potential curative surgical treatment.

Key words: hilar cholangiocarcinoma, jaundice, magnetic resonance cholangiography, left hepatectomy

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Introduction

Hilar cholangiocarcinoma, first described by Klatskin, is a tumor growing in the right hepatic duct, left hepatic duct or at their confluence [1]. It is a relatively rare but devastating disease. It is highly malignant, surgical excision being possible in only 5% of cases and has a poor prognosis – 90% mortality in 1 year. The main feature is represented by fibrosis which is responsible for the tight stricture of the biliary ducts and the development of obstructive jaundice. Before the appearance of jaundice the clinical signs are uncharacteristic and not alarming, thus explaining the delayed diagnosis. In the early stages the diagnosis is difficult in the absence of jaundice. However, it is known that in cases involving the left hepatic duct the jaundice may be of low intensity or even absent. The biliary obstruction may be suggested by the elevation of cholestatic enzymes and imagistic methods. The period between the debut of first symptoms and the appearance of jaundice is stretching from weeks to months. The confusion with benign conditions delays surgery and at the time when jaundice is installed, many cases are advanced and inoperable.

Case report

We present here the case of a 74 years old woman who had at admission anorexia, nausea, bloating and upper abdominal pain. The onset of symptoms was one month earlier and the severity of clinical manifestations increased since

then. Previously she had cholecystectomy, hysterectomy and breast lumpectomy for benign conditions. She also presented diabetes, arterial hypertension and dyslipidemia, being under chronic treatment.

Abdominal ultrasound showed enlargement of the left intrahepatic ducts without elevation of the cholestatic enzymes. CA19-9 and carcinoembryonic antigen tests was not performed as it was not available at the moment. The CT scan revealed a slight hepatomegaly and enlarged left hepatic ducts but no tumor in the liver, no pathologic lymph nodes and no ascites. The bile duct measured 10 mm visible down to the pancreas. The next step was a magnetic resonance cholangiography, which revealed the presence of a mass at the confluence of the hepatic ducts, amputating the initial segment of the left hepatic duct with enlargement of the left branches in segments 1,2,3,4, suggesting a cholangiocarcinoma (figure 1).

After a short preoperative preparation the patient underwent surgery. Through a Mercedes – Benz incision we entered the abdominal cavity and found a 3 cm operable tumor in the left liver lobe and left hepatic duct (figure 2). We performed a left hepatectomy and hilar lymph node dissection (figure 3,4). The specimen was sent to the Pathology Department (figure 5). The postoperative course was uneventful, the drains removed postoperative at day 5 and the patient was discharged on postoperative day 8 in good condition, carrying the T tube for external biliary drainage. The T tube was removed after 3 month. The histopathology examination showed a cholangiocarcinoma with involvement of the lymph nodes.

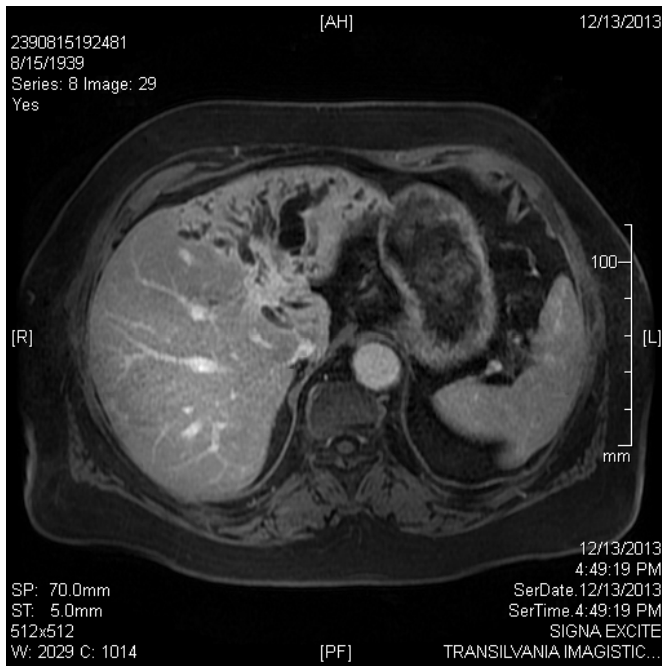


Fig. 1. Magnetic resonance cholangiography showing the tumor in the left hepatic duct. Enlargement of the left biliary tree above the tumor.

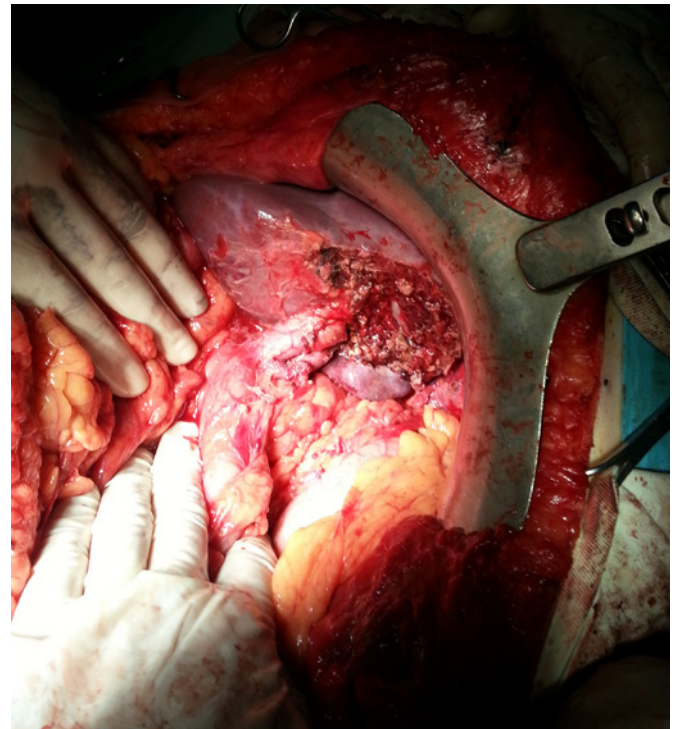


Fig. 4. Aspect after left hepatectomy.

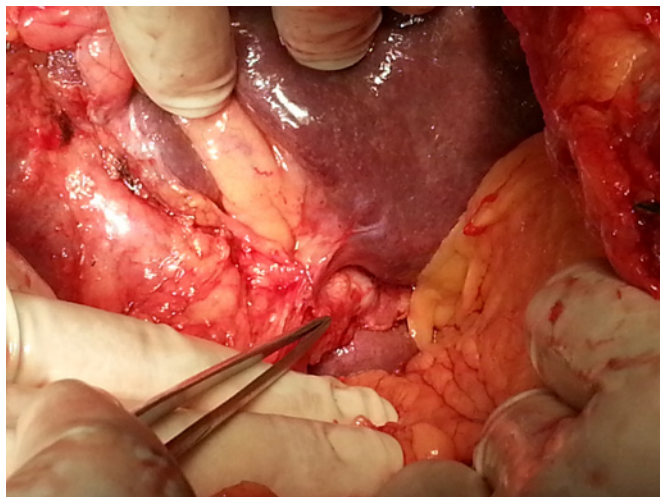


Fig. 2. Intraoperative view: the 3 cm sized tumor.

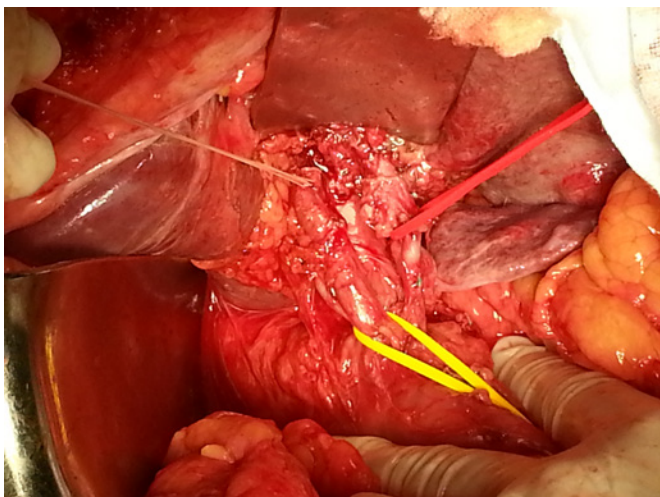


Fig. 3. The left hepatic structures with hilar lymph nodes dissection.

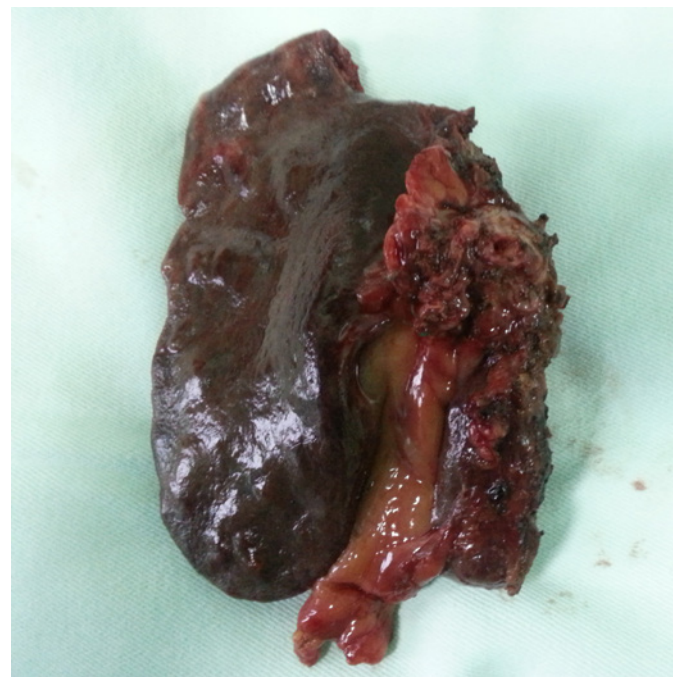


Fig. 5. The specimen.

Discussions

Hilar cholangiocarcinoma (proximal bile duct cancer) is represented by malignant tumors that occur in the right hepatic duct, left hepatic duct or at the confluence of the hepatic ducts (Klatskin tumours). Gerald Klatskin was an american physician who described for the first time the adenocarcinoma of the biliary confluence as a clinically independent tumor, leading to stricture of the hepatic ducts [1]. Although it is a rare malignancy, 4500 new cases are reported every year in USA [2]. Among the biliary tree

malignant tumors, 40 -60% are hilar cholangiocarcinomas [3]. They are sclerosing, grow slowly and advance locally, distant metastases being unusual. Due to the hepatic ducts strictures, jaundice is the main symptom, developing progressively in the absence of pain or fever [4]. But before the patient notices the jaundice there are a few uncharacteristic signs as: nausea, bloating and loss of appetite. These symptoms are more likely digestive and can delay the proper diagnosis, as they drive the attention on the investigation of the digestive tract. This prejaundice period lasts only a few weeks and a diagnosis in this phase is uncommon. When jaundice is already installed the lesions are advanced and in many cases beyond surgical resection (unresectable). In the prejaundice period the obstacle involves one hepatic duct or produces partial stricture. It is known that in cases involving the left hepatic duct jaundice may be of low intensity or even absent [5]. We noticed an elevation of the cholestatic enzymes (gamma glutamyl transferase and alkaline phosphatase), pointing out toward a biliary disease. The next investigation is the ultrasound which may show the enlargement of the intrahepatic biliary ducts. Computed tomographic scan, magnetic resonance cholangiography, ERCP, transhepatic cholangiography, PET/CT are used in case of suspicion of hilar cholangiocarcinoma [6]. Today, the magnetic resonance cholangiography is considered the golden standard for diagnosis of Klatskin tumors [3], allowing assessment of the local involvement and the operating plan. The method reveals the extension of the tumor in the biliary ducts and its surroundings and in the lymph nodes, as well as the distant metastases [7], with a sensitivity of 94% and specificity of 100% [8]. In conclusion, magnetic resonance cholangiography has the potential to replace the traditional techniques for imaging the pancreato-biliary system [9].

The surgical treatment is the only cure for the Klatskin disease. The resection of the tumor is the goal of the procedure but it may be very risky and of great magnitude. Anyway, this is today the only approach which can lead to a better 5-year survival [10], while the oncologic treatment has disappointing results [6]. Over the past 20 years, due to refinement in the hepatic resections the outcomes of the surgical treatment has significantly improved. After Bismuth-Corlette and the anatomical classification [11], the radical operations with intention for cure are: right hepatectomy for type I, II and IIIa tumors, situated in the right and common hepatic duct [12]; left hepatectomy for type IIIb tumors situated in the left hepatic duct [13] and bisegmentectomy or trisegmentectomy for type IV tumors with involvement of the hepatic confluence [14]. The results for trained teams with great experience, in high-volume centers, show a 25-79% resectability [15]. Some centers always begin the procedure with a laparoscopy to rule out carcinomatosis or an extensive tumor spread which would preclude the resection [16]. Laparoscopy can spare 40% of patients an unnecessary laparotomy.

The 5 year survival depends on the tumoral extension in the lymph nodes: it is 30% for cases without lymph node involvement, 15% for patients with regional lymph node involvement and 12% in case of paraaortic extension [17]. In case of a planned curative resection, the lymph nodes in the hepatoduodenal ligament, around the hepatic artery and behind the head of the pancreas should be removed, and dissection beyond this limit is not requested [18].

In the 2nd Department of Surgery of the Emergency County Hospital Tg.Mures, between 2008-2013 were admitted 10 patients with proximal bile duct cancers. Eight patients had jaundice and unresectable tumors at laparotomy and underwent only ductal stenting [19]. In one case the tumor was removed as palliation by limited resection of the involved hepatic duct. In another case the tumor was resectable, in a patient without jaundice, in which the diagnosis was reached at magnetic resonance cholangiography. In this case we were able to do a curative resection (left hepatectomy and skeletonization of the hepatoduodenal ligament)

Conclusions

Proximal bile duct cancer (hilar cholangiocarcinoma, Klatskin tumor) is rarely diagnosed in the prejaundice phase when the symptoms are uncharacteristic, mimicking a benign disease and when surgical treatment is potentially curative. In case of elevated cholestatic enzymes and enlargement of the hepatic ducts, especially in one hepatic lobe, the investigation of choice is represented by magnetic resonance cholangiography. This method is able to point out the exact location of the obstruction. The potential curative treatment of proximal bile duct cancer is only surgical consisting in major hepatectomies. In advanced stages, after jaundice appears, these tumors are not resectable, being possible just palliation.

References

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