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Cholangiocarcinoma of an Ectopic Outlet of the Main Bile Duct at the Duodeno-Jejunal Angle – A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors.. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

An ectopic outlet of the common bile duct is a rare congenital disorder of the biliary tract. These abnormalities pose a diagnostic problem, and their clinical presentation is not specific. As patients are usually asymptomatic at early stages, the diagnosis is often incidental and at an advanced stage, when clinical manifestations become noticeable. Imaging tests such as hepatic MRI and CT are essential for positive and differential diagnosis, assessment of extension, and treatment planning.

We report a case of a locally advanced tumour in an ectopic biliopancreatic outlet of the main bile duct at the duodenojejunal angle, in a young patient presenting with clinical cholestasis syndrome. The diagnosis was made by an abdominopelvic CT scan and confirmed by Bili-MRI and a biopsy of a liver nodule in the third segment. The biopsy showed metastasis from adenocarcinoma on extemporaneous examination. A double hepatojejunal and gastrojejunal bypass on a Y-shaped

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loop and a retrograde cholecystectomy with subhepatic redon drainage were performed. The patient's postoperative course was uneventful.
In the presence of congenital anomalies, the surgeon must remain vigilant because of the potential for accidents during the operation.

Keywords: The main bile duct; duodenojejunal angle; cholangiocarcinoma; hepatojejunal and gastrojejunal bypass ; surgery.

1. INTRODUCTION

The anatomical variations of the bile ducts are generally well known, but due to their rare occurrence, they can cause difficulties for surgeons during operations. These abnormalities include aberrant or accessory bile ducts, an aberrant cystic duct, abnormal junction of the bile ducts with the pancreatic duct, as well as bilio-pancreatic outlet abnormalities in the duodenum. Knowledge of embryology and normal or abnormal anatomy of the bile ducts is very important. These congenital abnormalities pose a diagnostic problem and are often discovered incidentally during surgery. Advanced radiological technologies, especially magnetic resonance imaging (MRI) and computed tomography (CT), have improved recognition of these abnormalities significantly.

We report a case of a patient diagnosed preoperatively with cholangiocarcinoma, with ectopic biliopancreatic access to the main bile duct in the duodenojejunal angle, after presenting with clinical cholestasis syndrome.

Congenital abnormalities should not be neglected by the surgeon in order to avoid accidentally missing lesions of the extrahepatic bile ducts.

2. CASE DESCRIPTION

The patient was a 40-year-old woman with no significant medical history. She was admitted with clinical cholestasis syndrome associated with epigastric pain evolving over 1 month, associated with bilious vomiting and generalised pruritus, without evidence of gastrointestinal haemorrhages or other associated signs. All these symptoms developed in the context of apyrexia and no significant health problems overall. The patient was haemodynamically stable with a blood pressure of 125/80 mmHg, pulse rate of 87 bpm, and temperature of 37.3°C and had generalised mucocutaneous jaundice and excoriations over the body. There was no abdominal tenderness, mass, or palpable gallbladder, and the pelvic examination was also unremarkable.

Abdominal ultrasonography showed moderate dilation of the intrahepatic bile ducts, the common bile duct dilated to 16 mm in diameter without any visible obstruction, and a distended gallbladder with thickened wall and alithiasis. The abdominopelvic CT scan (A) revealed an aberrant connection of the bilio-pancreatic junction at the level of the duodenojejunal angle (main bile duct-Wirsung), with the lesion having its epicentre located at the level of the Treitz angle. This process measured 60 × 40 × 80 mm, was very limited, and enhanced moderately and evenly after injection. This process appeared to invade Winslow's pancreas, invaded the lower common bile duct, and was responsible for significant dilation of the main bile duct, intrahepatic bile ducts, and Wirsung's ducts. It also showed a direct contiguity, without separating the fatty border from the jejunal loops, with the aorta (on a circumference of approximately 90°) and with the left renal vein. A distended gall bladder and three hepatic nodules suggestive of secondary lesions were also noted.

Bili-MRI (B) revealed a distended gallbladder with micro-stones, an abutment of the main bile duct on the left side at the level of the duodenojejunal angle, dilation of the intrahepatic bile ducts, and a main bile duct measuring 20.5 mm in diameter. This dilation took place upstream of a tissue mass formation and budding within the termination of the main bile duct measuring 83 × 47 × 37 mm. The lesion showed progressive enhancement after gadolinium injection, with hypersignal diffusion.

The patient had a haemoglobin level of 8.5 g/dL and abnormal liver function tests (ASAT = 90 IU/L; ALAT = 135 IU/L; gamma-GT = 346 IU/L; PAL = 740 IU/L; bilirubin total = 38 mg/L; conjugated bilirubin = 32.5 mg/L).

In the supine position, under general anaesthesia, the patient underwent a right subcostal laparotomy extended to the left,

confirming the diagnosis by the presence of a locally advanced tumour at the duodenojejunal angle. The tumour was 10 cm in size, solid, fixed, with the main bile duct dilated to 2 cm (C, D) and there were three metastatic nodules of 1 cm each at the segment 3 of the liver. The operative procedure included a biopsy of a liver nodule from segment III, which showed liver metastases from an adenocarcinomatous proliferation. The surgery also included a double hepatojejunal and gastrojejunal bypass on the handle mounted in Y, retrograde cholecystectomy, biopsy of the greater omentum, and sub-hepatic Redon drainage. The postoperative period was uneventful, and the patient was discharged from hospital on day 7 after drain removal and referred to the oncology department for palliative chemotherapy. Follow-up at 12 months was normal. Histopathological examination of the gall bladder showed the presence of chronic cholecystitis without specific signs of malignancy and congestive adipose tissue free from tumour proliferation.

3. DISCUSSION

There are many anatomical variations which may involve the location of the hepatic artery, the existence of accessory hepatic ducts, and/or the presence of ectopic opening of the common bile duct [1].

Normally, the Wirsung's pancreatic duct and the main bile duct terminate in the 2nd part of the duodenum at the level of the major papilla [2]. The distal bile duct enters the second part of the duodenum on the posterior medial wall in 75% of cases, but can extend to its full length. Several variations are known to occur with respect to its connections. It can enter into the first part of the duodenum (1.5%) causing intestinal reflux and spontaneous aerobilia. It can occur at the lower duodenal angle (19.5%), in the third part of the duodenum (1.5%), and exceptionally, in the fourth part of the duodenum [3]. Ectopic main bile duct access is a rare congenital biliary disorder [4] and is found in less than 5% of the general population [2]. Its frequency varies from 0 to 3.2%. It is more common in Southeast Asia, mainly in women. The vast majority of these cases are associated with cystic dilation of the common bile duct. The incidence of biliopancreatic junction abnormalities is 43% [5]. The aetiology involves developmental abnormalities which are not well identified occurring during embryogenesis [4].

These variations are important for the surgeon, particularly those concerning the right hepatic duct (which are most frequent and can lead to confusion with the cystic duct) [3].

Clinically, bile duct abnormalities can be asymptomatic for a long time. Biliary stasis is responsible for cholangitis and the formation of intrahepatic stones or abscesses. Intra- or extrahepatic cholangiocarcinomas occur in 7-14% of cases [2].

Previous reports have shown various complications of ectopic papillae, including gallstones, choledocholithiasis, obstructive cholestasis, cholangitis, pancreatitis, and peptic ulcer disease (6). However, we did not find reports of a tumour of the ectopic main bile duct at the duodenojejunal angle, as described in our case.

The diagnostic approach to suspected bile duct pathology is largely based on CT and magnetic resonance imaging. CT and magnetic resonance imaging have similar utility in assessing tumour size and detecting satellite lesions [7]. However, CT is the gold standard for identifying extrahepatic metastases and determining compliance. Meanwhile, MRI is the gold standard for assessment of local extension and detection of proximal and distal extrahepatic cholangiocarcinomas and liver metastases. Imaging must combine hepatic MRI sequences with injection and angiography sequences. The sensitivity rate for detecting tumours is 95%, and it can assess bile duct extension with 90% reliability [8].

Ultrasound endoscopy contributes to the diagnosis and assessment of locoregional extension of extrahepatic cholangiocarcinomas and vesicular carcinomas. It allows the sampling of lymph nodes as well as of the lesion when it forms a mass [8].

Retrograde cholangioscopy with endoductal sampling can confirm or rule out the diagnosis of malignancy in indeterminate biliary stenosis [8]. However, ultrasound is insufficiently validated and not very accessible, and should only be used in clinical trials [8].

With regard to serum tumour markers, the carbohydrate antigen (CA) 19.9 has a sensitivity and a specificity of about 80%; carcinoembryonic antigen and CA 125 are less sensitive (30%–50%) and not much more specific [8].

Treatment primarily concerns the abnormalities associated with the biliopancreatic junction and its associated complications. During ERCP, an endoscopic sphincterotomy can be performed to extract a stone; in the case of tumour stenosis due to a non-operable, spreading, bladder cancer, a prosthesis can be placed [9]. However, the risk of perforation is very high due to the lack of a sphincter structure [4].

If there is a cholangiocarcinoma of the main bile duct, curative treatments can be used for resection or for palliative treatment (in cases of peritoneal carcinoma or metastasis). Distal extrahepatic cholangiocarcinomas require cephalic duodenopancreatectomy with standard lymphadenectomy and resection of the main bile duct in a healthy area, according to extemporaneous examination. The aim of palliative treatment is to maintain and improve the quality of life. These can be bilio-digestive bypass or endoscopic or radiological biliary drainage with palliative chemotherapy [8].

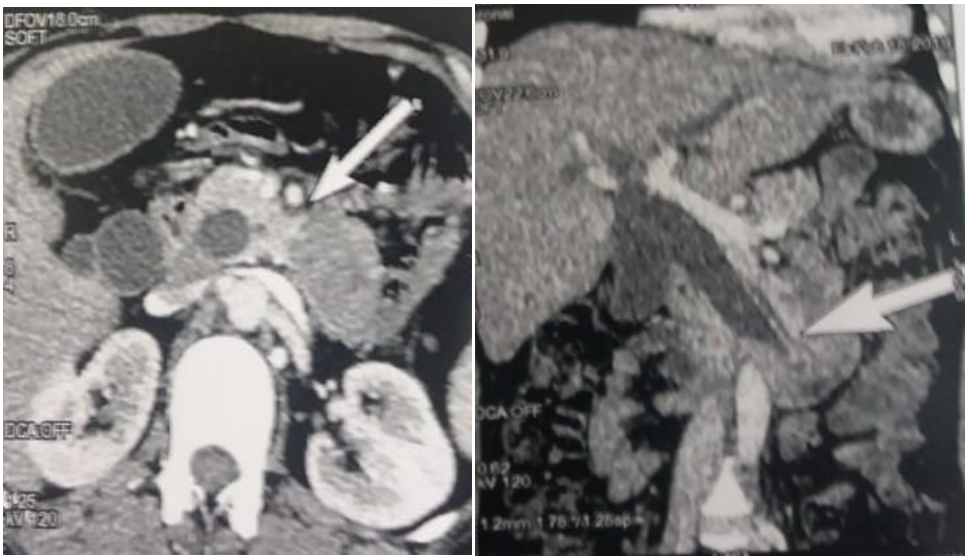

Fig. 1(A). Abdominal CT scan showing an aberration at the level of the duodenojejunal angle of the biliopancreatic junction, with a tissue lesion whose epicentre is located at the level of the duodenojejunal angle

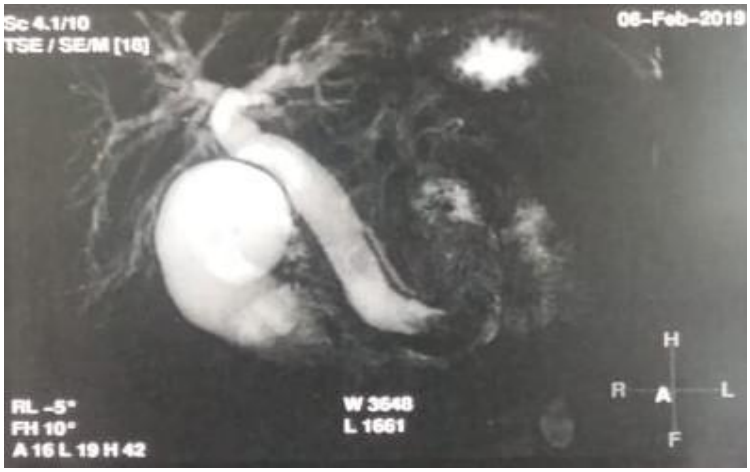

Fig. 1(B). Magnetic resonance imaging (MRI) showed a distended gallbladder, presence of microcalcifications, and a main bile duct termination on the left side at the duodenojejunal angle, with dilatation of the intrahepatic and main bile ducts

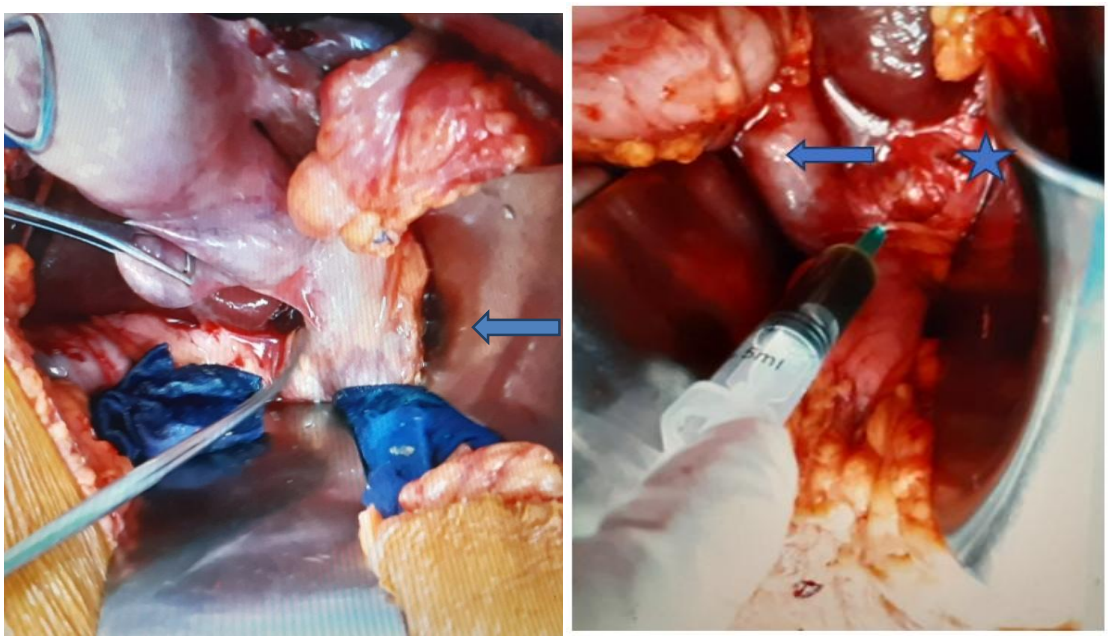

Fig. 1(C,D). Intraoperative image showing a main bile duct dilated to 2 cm, oblique down, and to the left with an ectopic connection at the level of the duodenojejunal angle

4. CONCLUSION

A biliopancreatic tract opening at the duodenojejunal angle is an exceptional congenital malformation; these anomalies are often asymptomatic.

Through medical imaging because of the presence of a complication or suspicion of bile duct pathology, we discovered the existence of bile duct abnormalities and their ectopic connection.

Therapeutic management is variable depending on the location and associated complications. These abnormalities should not be neglected in order to avoid accidental intraoperative damage of the extrahepatic bile ducts.

PATIENT CONSENT

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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