Journal of Gastrointestinal Cancer (2019) 50:1048–1050
https://doi.org/10.1007/s12029-019-00208-x

CASE REPORT

A Case Report of a Primary Neuroendocrine Tumour of the Proximal Common Bile Duct

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Published online: 9 February 2019
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Introduction

A neuroendocrine tumour (NET) of the extrahepatic bile ducts is rare accounting 0.2 to 2% of all gastrointestinal carcinoid tumours [1]. Localization in the proximal part of the common bile duct (CBD) is rare [2]. Signs and symptoms are not specific, may be similar to bile duct stones. Preoperative diagnosis is difficult [3]. The aim of this report is to describe a CBD NET not diagnosed preoperatively to highlight the difficulties encountered in this kind of situation despite the contribution of imaging in this rare localization: the junction of the common hepatic duct and cystic duct.

Case Report

A 39-year-old man with no past medical history consulted for epigastric pain, jaundice and weight loss in the last 8 months. He did not present hormone-related symptoms such as flushing, stomach aches, diarrhoea or hypoglycemia. Physical exam was normal. Laboratory data showed mild elevation of serum total bilirubin (57.7 μmol/l), direct bilirubin (38.4 μmol/l), γ-glutamyl-transferase (911 u/l), alkaline phosphatase (250 u/l) and liver enzymes (ALT, 146 u/l; AST, 59 u/l). Chromogranin-A was not elevated and CEA and CA 19-9 were not elevated. Abdominal ultrasonography showed no gallbladder or CBD stones. An increased diameter of the CBD to 11 mm with distended intrahepatic bile ducts. Abdominal CT scan showed a mass of 18 × 24 mm, located in the proximal part of the CBD, with dilatation of the intrahepatic and extrahepatic bile ducts. This tumour has an intense enhancement in the arterial phase (Fig. 1). Magnetic resonance imaging showed a severe stenosis of the junction of the common hepatic duct, cystic duct and the CBD with no vascular invasion measuring 17 mm (Fig. 2). The diagnosis of a benign extrahepatic tumour has been retained and a resection of the CBD, including the gallbladder, was decided upon. At laparotomy, we found a tough tumour located in the CBD, next to the cystic duct insertion (Fig. 3). There was no evidence of any hepatic, lymph nodes or peritoneal dissemination. The patient underwent a cholecystectomy, CBD resection 2 cm above and below a tumour with lymph node excision. Then a choledochoduodenal anastomosis was performed. The postoperative course was uneventful and the patient was discharged after 3 days. Pathological examination concluded to a well-differentiated neuroendocrine tumour G1 of 2 cm was retained. Surgical margins were free of a tumour and the exam of three lymph nodes did not reveal any metastases. After 1 year of follow-up, there are no signs of recurrence.

Discussion

Diagnosis of NET localised in the proximal CBD mass rare and difficult. Our case highlights the difficulties encountered

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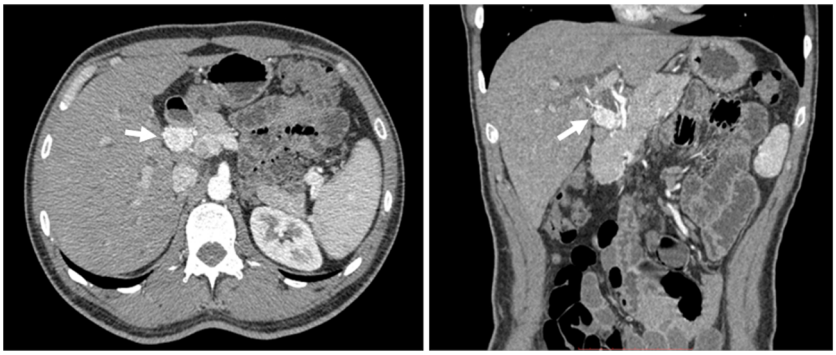

Fig. 1 Abdominal computed tomography showed a high-density mass of 18 × 24 mm, located in the hepatic hilar area (white arrows)

in this kind of situation and this is despite the contribution of imaging. In addition, this CBD NET present a rare localization: the junction of the common hepatic duct and cystic duct.

NET involves the embryonic neural crest cells called Kulchitsky cells [1]. The gastrointestinal tract is the most common site of the NET. This tumour involves frequently pancreas, stomach and bowels [4]. However, intrahepatic or extrahepatic bile duct, gallbladder and even Vater ampulla localizations were also reported [5]. Concerning the CBD, it is frequently found in the distal part (19.2%), followed by the middle part (17.9%), the cystic duct (16.7%) and the proximal part (11.5%) [2]. Like in our case, NET is generally symptomatic even with small size. This is due to bulk-related symptoms [6]. Typically, they are jaundice and pain. Rarely, in 6 to 10% of cases, associated symptoms related to hormonally active NET are reported [7]. As in our case, the preoperative diagnosis of CBD NET is difficult because of the absence of detectable serum markers and the lack of hormonal symptoms [2]. In another way, technological advances and the availability of many diagnostic imaging modalities make this diagnosis feasible but remain difficult. They provide essentially sufficient information to retain resectability of the tumour. This difficulty is due to similar radiological findings with other CBD tumours such as cholangiocarcinoma that presents 80% of CBD tumours [8] and gastrointestinal stromal tumour (GIST). Radiological imaging shows often a well-defined margin, exophytic and hypervascular mass without calcifications [8]. Cholangiocarcinoma appears as an irregular wall thickening with local invasion. In this case, a well-defined mass was diagnosed with intense enhancement in the arterial phase. The intrahepatic bile duct dilation was slight compared to the mass size. GIST appears as a sharply well-limited mass with variable contrast enhancement. Diagnosis can be made preoperatively using biopsy during an endoscopic retrograde cholangiopancreatography [3]. Treatment strategies are not well established. Aggressive surgical resection is the cornerstone of treatment. It is considered to be the only curative procedure [9]. However, these strategies are essential for the gastroenteropancreatic NET and not for CBD NET. The procedure can be an extrahepatic bile duct resection in case of proximal or medial CBD NET. It can be a duodenopancreatectomy in case of distal CBD NET. These procedures are associated with prophylactic portal lymphadenectomy. In this case, the tumour was localised in the junction

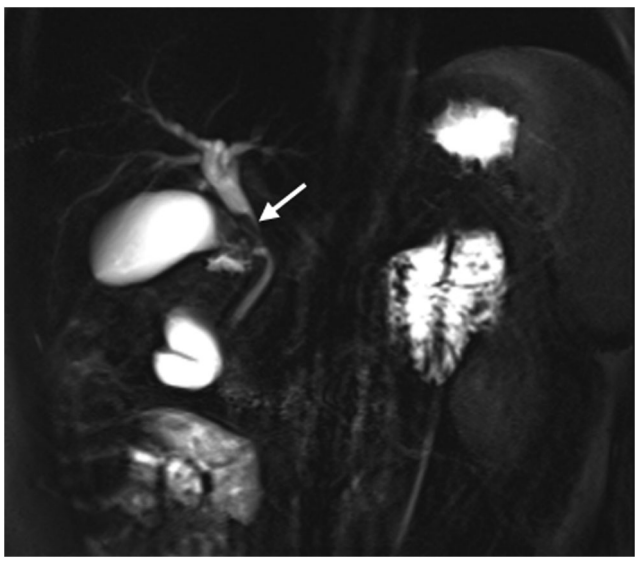

Fig. 2 Magnetic resonance cholangiopancreatography showed a tumour compressing the CBD with dilatation of the intrahepatic bile duct (white arrow)

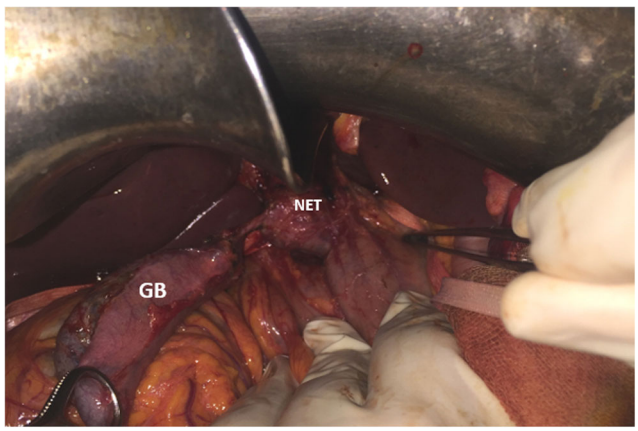

Fig. 3 Intraoperative view showing a well-limited mass of 2 cm situated at the junction of the common hepatic duct and cystic duct (GB gallbladder, NET neuroendocrine tumour)

of the common hepatic duct and cystic duct, like in this case. The lymph node resection was necessary despite small tumour size. Local lymph nodes and liver metastasis were reported respectively in 19.2% and 16.7% of cases [2]. If they exist, a high-grade NET will be suspected. Actually, no long-term follow up data are available for CBD NET [9]. A multidisciplinary approach may improve prognosis which is actually poor even after large surgical resection and despite the progress of other therapeutic approaches.

Conclusion

CBD NET are rare. They are generally considered low-grade malignancies. A few cases are reported in the literature. It must be suspected in case of a well-circumscribed hypervascular mass in the arterial phase.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflicts of interest.

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