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Neuroendocrine tumor of the extrahepatic bile duct: A case report

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ABSTRACT

INTRODUCTION: Neuroendocrine tumors (NETs) of the extrahepatic bile ducts are extremely rare neoplasms arising from endocrine cells and have variable malignant potential. They most commonly occur in young females and usually present with painless jaundice.
PRESENTATION OF CASE: Here we present the case of an asymptomatic 57-year-old woman with NET of the common bile duct that was incidentally discovered on abdominal ultrasound during a medical examination. She was admitted to our hospital with a diagnosis of hepatic hilar tumor. Computed tomography revealed the tumor surrounding the hepatic hilum and duodenum. Magnetic resonance cholangiopancreatography revealed a filling defect of the common bile duct with morphology suggestive of external compression. Endoscopic ultrasound confirmed a submucosal tumor of the duodenal bulb measuring 30 × 20 mm in size. The patient qualified for surgery with a preoperative diagnosis of submucosal tumor of the duodenal bulb. Intraoperative examination revealed that the tumor location involved the common bile duct and/or cystic duct with no signs of invasion to other organs or metastatic lymph nodes. Excision of the biliary ducts and tumor was followed by Roux-en-Y anastomosis. Histological results showed NET grade 1.
DISCUSSION: Preoperative diagnosis of NETs is difficult because of their rarity. A definitive diagnosis is usually established intraoperatively or after histopathological evaluation.
CONCLUSION: For these tumors, surgical resection is currently the only treatment modality for achieving a potentially curative effect and prolonged disease-free survival.

1. Introduction

Neuroendocrine tumors (NETs) of the extrahepatic bile ducts are extremely rare with <200 reported cases in the literature since Pilz first described it in 1961 [1]. NETs are neoplasms with variable malignant potential and arise from endocrine cells. The most common sites for primary NETs are the gastrointestinal tract (73.7%) and bronchopulmonary system (25.1%) [2], with only 0.1%–0.4% occurring in the extrahepatic bile ducts [3,4]. A diagnosis is rarely made during preoperative examinations, and a definitive diagnosis is established intraoperatively or after histopathological evaluation. Here we summarize the clinical course and radiological findings of a patient with an extrahepatic bile duct NET along with a literature review in line with the SCARE criteria [5].

2. Presentation of case

A 57-year-old woman with no past medical and surgical history was referred to our hospital for further examination of positive occult blood in her stools and a hepatic hilar tumor that was diagnosed on abdominal ultrasound during a medical examination. She had no drug and family history. She was admitted to the department of gastroenterology of our hospital in June 2014. On admission, physical examination was unremarkable, and there was no jaundice. Colon biopsy revealed ulcerative colitis; computed tomography (CT) revealed a tumor surrounding the hepatic hilum and duodenum (Fig. 1). Positron emission tomography revealed no abnormal accumulation of 2-deoxy-2-[F-18] fluoro-D-glucose in the upper abdomen. Magnetic resonance cholangiopancreatography showed a filling defect of the common bile duct with a morphology corresponding to an external compression by the lesion (Fig. 2). Endoscopic ultrasound (EUS) confirmed the diagnosis of a 30 × 20-mm submucosal tumor of the duodenal bulb (Fig. 3). We did not perform the EUS-guided fine needle aspiration because of the incidence of high false-negative results in submucosal

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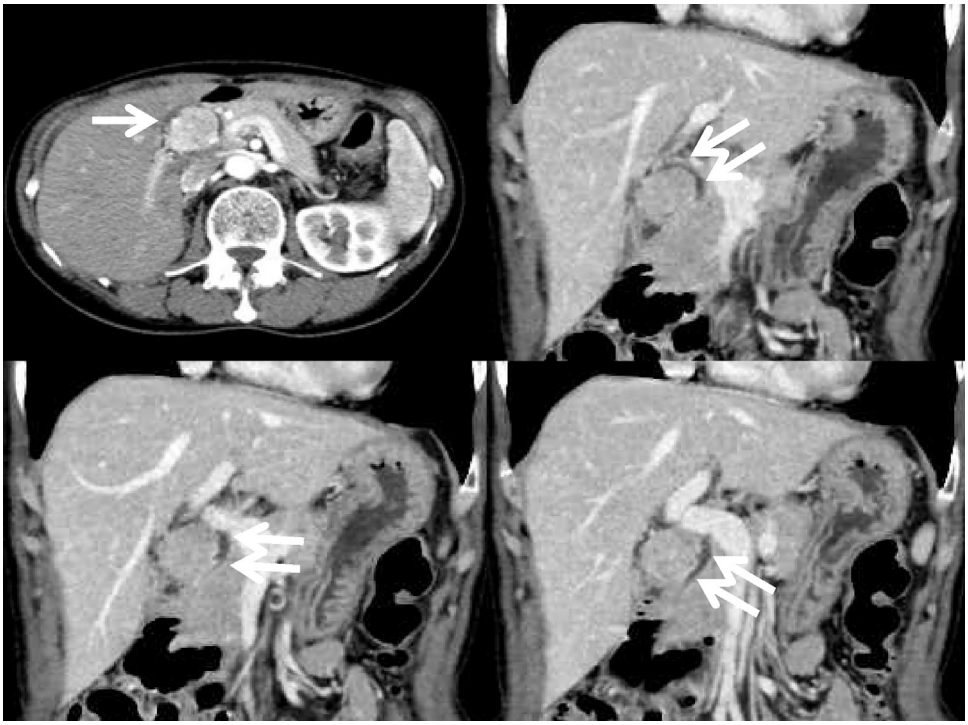

Fig. 1. Computed tomography revealed a tumor, with a 3-cm diameter, surrounding the hepatic hilum with the compression of the common bile duct from the outside. The tumor was enhanced in the early phase (single arrow). The extrahepatic bile duct was visible (double arrows).

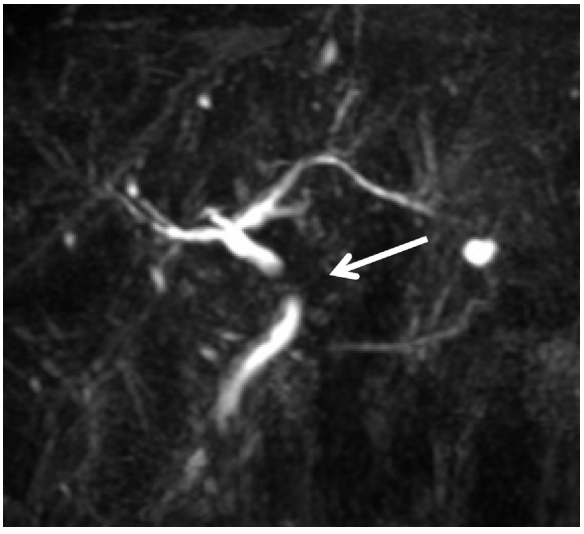

Fig. 2. Magnetic resonance cholangiopancreatography showed a filling defect of the common bile duct with no dilatation of the intrahepatic bile duct.

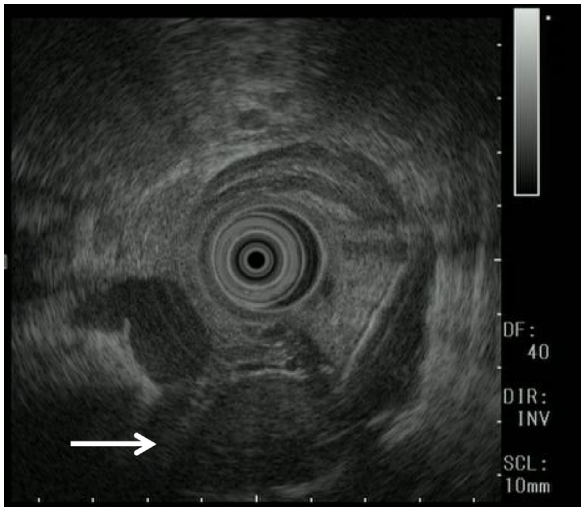

Fig. 3. Endoscopic ultrasound showed a 30 × 20-mm submucosal tumor of the duodenal bulb.

neoplasm. Our preoperative diagnosis was a submucosal duodenal tumor, and benign or malignant biliary tumors were included in the differential diagnosis. Intraoperative examination revealed that the tumor was located around the common bile duct and/or cystic duct, with no signs of invasion to other organs or metastatic lymph nodes. We resected the extrahepatic bile duct. The diagnosis of an intraoperative frozen section was an extrahepatic bile duct NET, and the resection margins and lymph nodes near the tumor were free of tumor. In our treatment strategy for biliary malignancies or other tumors, prophylactic pancreaticoduodenectomy was not performed in case resection margins, and lymph nodes were free of tumor. Therefore, only prophylactic portal lymphadenectomy was added. Histological examination revealed that the tumor, which originated from the intramuscular layer of the common bile duct and was located within the muscle layers, compressed the lumen of the bile duct (Fig. 4). The resection margins and lymph nodes were free of the tumor. Immunohistochemical analysis showed that neoplastic cells were positive for chromogranin A, synaptophysin, and CD56 (Fig. 5). The proliferative index (Mib-1) was <2%, and histological analysis showed NET grade 1 (WHO Classification 2010) [6]. The patient's postoperative clinical course was uneventful; she was discharged on postoperative day 32. She remains well and disease free for 34 months after surgery, with follow-ups and annual CT scans.