CASE REPORT
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Primary neuroendocrine tumor of the perihilar bile duct: A case report

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ABSTRACT

Neuroendocrine tumors (NETs) arising from extrahepatic bile ducts are very rare. We present a patient with perihilar NET who was operated on with a preoperative diagnosis of Klatskin tumor. A 58-year-old female patient was admitted with abdominal pain and jaundice. Laboratory data showed elevated serum bilirubin levels and liver function tests. Computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) findings were consistent with perihilar bile duct tumor. The patient was operated on with a diagnosis of Klatskin tumor. She underwent right hepatectomy, resection of the extrahepatic bile duct, portal lymphadenectomy and Roux-en-Y hepaticojejunostomy. The final pathologic examination of the resected specimen demonstrated a well differentiated neuroendocrine tumor (Grade 1). NETs originating from perihilar bile ducts are extremely rare, and preoperative definite diagnosis is very difficult. It should be kept in mind that NET may be one of the rare causes of perihilar bile duct obstruction.

Keywords: Neuroendocrine tumor, perihilar bile duct, resection

INTRODUCTION

Neuroendocrine tumors (NETs) can be found throughout the gastrointestinal tract and pancreas. However, NETs arising from extrahepatic bile duct are very rare. NETS arising from perihilar region is exceedingly rare with a few cases reported in the literature (1). Preoperative differential diagnosis of perihilar NET is difficult and in general, it is diagnosed with postoperative histopathologic evaluation of the resected specimen. We herein report a case of perihilar NET with a preoperative diagnosis of perihilar cholangiocarcinoma.

CASE REPORT

A 58-year-old female patient was admitted with abdominal pain and jaundice. Physical examination revealed only a mild tenderness on the right upper quadrant. Laboratory data showed elevated serum total and direct bilirubin levels (6.29/5.42 mg/dL) and liver function tests. Serum levels of CEA and CA19-9 were normal. The patient was referred to computed tomography (CT). Both right and left intrahepatic bile duct dilatation were revealed on contrast- enhanced abdominal CT and also a-two-cm diameter of an isodense mass was detected in the perihilar region (Figure 1). Magnetic resonance cholangiopancreatography (MRCP) was performed with a prediagnosis of perihilar cholangiocarcinoma. Dilated intrahepatic bile ducts, obstruction of common hepatic duct due to tumor mass and normal intrapancreatic distal common bile duct were seen on MRCP images (Figure 2). MRCP findings were consistent with perihilar bile duct tumor.

The patient was operated on with a diagnosis of perihilar cholangiocarcinoma (Klatskin tumor). She underwent right hepatectomy, excision of the caudate lobe, resection of the extrahepatic bile duct, regional lymphadenectomy and Roux-en hepaticojejunostomy. During the postoperative period, the patient developed collections and abscess in the operative field which were managed with percutaneous drainage procedures and prolonged antibiotic therapy. The patient was discharged after a postoperative course of 36 days.

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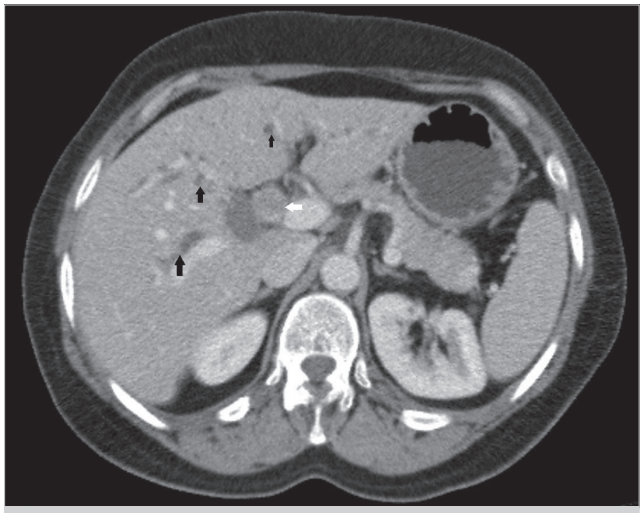

Figure 1. Contrast-enhanced axial CT image shows dilated left and right intrahepatic bile ducts (black arrows) and an isodense tumor mass (white arrow) localizated in common bile duct.

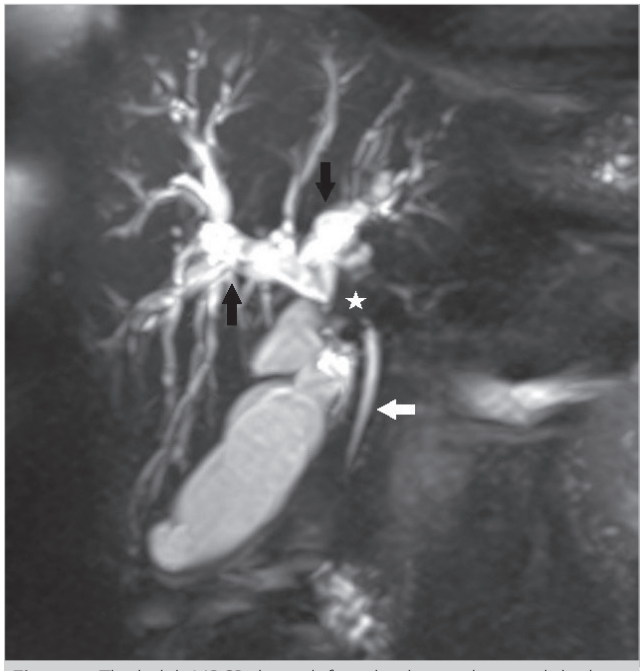

Figure 2. Thick slab MRCP shows left and right intrahepatic bile ducts dilatation (black arrows), perihilar extrahepatic bile duct tumor (star) and normal caliber of intrapancreatic terminal common bile duct.

The final pathologic examination of the resected specimen demonstrated a well differentiated (Grade 1) neuroendocrine tumor in accordance with the World Health organization (WHO) 2010 classification. The maximum diameter of the tumor was 1.2 cm. Proximal and distal surgical margins were free of tumor cells. However, there were tumor cells in the lateral margin and subepithelial stroma of the hepatic duct. There was only one metastatic local lymph node.

The patient did not receive any modality of adjuvant therapy. After a 15-month-follow-up period, the patient was doing well with normal physical examination findings and completely normal liver function tests. Ga67 PET BT showed no evidence of distant metastasis or local recurrence.

DISCUSSION

Neuroendocrine tumors are classified as as NET Grade 1 (G1), NET Grade 2 (G2), and neuroendocrine carcinoma (NEC) according to the 2010 WHO classification system (2). This paper presents a case of NET G1 arising from an extremely rare localization.

The precise diagnosis of perihiler NET is frequently not possible preoperatively. Differential diagnosis based on radiologic findings is difficult although BT reveals hypervascular, well-circumscribed lesions (3). The differential diagnosis includes cholangiocarcinoma, metastatic tumors and lymphoma (4). There are no specific hormonal symptoms and serum markers (3). Preoperative histopathologic diagnosis is usually not possible. Moreover, as the most frequent cause of malignant perihilar bile duct obstruction is adenocarcinoma, histopathologic diagnosis is not generally needed for resectable cases when a malignant obstruction is suspected. Therefore, the definite histopathologic diagnosis is generally possible after the evaluation of the resected specimen. In this case, our preoperative diagnosis was perihilar cholangiocarcinoma and the treatment plan was resection of the tumor.

Data regarding the optimal management and prognosis of these tumors are not sufficient in the literature. However, resection when technically possible should be performed as it is frequently impossible to distinguish NETs from cholangiocarcinomas. In addition, G1 and G2 NETs are slowly growing tumors and resection has been shown to be beneficial (5,6).

Surgery is the mainstay of treatment and should be considered in all patients if technically feasible in gastroenteropancreatic neuroendocrine tumors (GEP-NET). Curative surgical resection of the primary lesion should be performed in patients with localized GEP-NET (7).

In G1 and G2 NETs, curative intended surgery has been considered, even in patients with liver and/or lymph node metastases (8). Debulking surgery may also be performed for liver metastases in certain circumstances (8). In our patient, resection of the tumor with right hepatectomy and regional lymphadenectomy was performed as recommended in the literature (6-8).

As perihilar NETs are extremely rare, data regarding long-term follow-up of these tumors are lacking. However, as G1 and G2 NETs are slowly growing tumors, better survival when compared to perihilar cholangiocarcinoma may be expected. Similarly, our patient showed complete clinical and biochemical healing after a follow-up of 15 months.

CONCLUSION

In conclusion, NETs of the perihilar extrahepatic bile duct are extremely rare and are difficult to diagnose perioperatively. Surgical resection is the only therapy that offers a chance of cure. Perihilar NETs should be kept in mind in the differential diagnosis of a suspected perihilar malignant obstruction.

Informed Consent: Informed consent was obtained from patient who participated in this case.

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OLGU SUNUMU-ÖZET
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Safra yollarının primer nöroendokrin tümörü: Bir olgu sunumu

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ÖZET

Ekstrahepatik safra kanallarından kaynaklanan nöroendokrin tümörler (NET'ler) çok nadirdir. Preoperatif Klatskin tümör tanısı ile opere edilen perihiler NET'li bir hastayı sunuyoruz. Elli sekiz yaşında kadın hasta, karın ağrısı ve sarılık şikayeti ile başvurdu. Laboratuvar incelemesinde serum bilirubin düzeylerinde ve karaciğer fonksiyon testlerinde artış mevcuttu. Bilgisayarlı tomografi (BT) ve manyetik rezonans kolanjiyopankreatografi (MRCP) bulguları; perihiler safra kanalı tümörü ile uyumluydu. Hasta Klatskin tümör tanısı ile ameliyat edildi. Hastaya sağ hepatektomi, ekstrahepatik safra kanalı rezeksiyonu, portal lenfadenektomi ve Roux-en-Y hepatikojejunostomi yapıldı. Rezeke edilen numunenin patolojik incelemesinde, iyi farklılaşmış bir nöroendokrin tümör (evre 1) gösterildi. Perihiler safra kanallarından kaynaklanan NET'ler oldukça nadirdir ve preoperatif kesin tanı çok zordur. NET'in perihiler safra yolu tıkanıklığının nadir sebeplerinden biri olabileceği unutulmamalıdır.

Anahtar Kelimeler: Nöroendokrin tümör, perihiler safra yolu, rezeksiyon

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