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Case Report

Primary Neuroendocrine Tumor of the Portal Hepatic Duct in Adolescence: A Case Report

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ABSTRACT

Background: It is not uncommon for Neuroendocrine Tumors (NETs) to occur in the gastrointestinal tract, pancreas, and lungs, but NETs originating from the biliary system are relatively rare, accounting for 0.67% of gastroenteropancreatic system tumors. At present, less than 10 cases of extrahepatic biliary neuroendocrine tumors have been reported in adolescent.

Case presentation: This report presents a case of a 16-year-old boy with a NET of the portal hepatic duct. Obstructive jaundice was judged from the laboratory examination and imaging findings. After the evaluation and multidisciplinary discussion, he underwent resection of the common bile duct, common hepatic duct, and gallbladder and Roux-en-Y hepaticojejunostomy. Pathological examination revealed a NET with the tumor invading the wall of the bile duct and surrounding adipose tissue. The patient had no NETs in any other organs or tissues. This case was a primary NET, not a metastatic tumor.

Conclusion: The preoperative diagnosis of primary neuroendocrine tumor of the portal hepatic duct is difficult, and this tumor is often confused with hilar cholangiocarcinoma. Surgical treatment is the only effective way to cure it. Once the diagnosis has been made, surgical resection should be performed as soon as possible.

Keywords: Neuroendocrine tumor, Portal hepatic duct, Extrahepatic biliary, Case report

Abbreviations: NETs: Neuroendocrine Tumors

INTRODUCTION

The detection rate of NETs (3.65/100,000) has increased in recent years with improvements in medical technology and detection methods [1]. However, it has been shown that the incidence of extrahepatic biliary NETs is extremely low, accounting for approximately 0.67% of NETs in the gastroenteropancreatic system [2]. To date, only 8 cases in adolescents have also been reported. The youngest of these cases was reported by a Mexican scholar, a case of a 12-year-old girl diagnosed with an extrahepatic biliary NET. Currently, clinicians are uncertain about the epidemiology, diagnosis and prognosis of extrahepatic biliary NET as it is poorly reported. Nevertheless, surgical excision is considered to be the best treatment for this disease.

CASE PRESENTATION

A 16-year-old boy presented to our hospital with jaundice and pruritus lasting 1 week. He denied any history of other medical condition. Physical examination revealed no other abnormalities. Laboratory tests indicated obstructive jaundice. Liver biochemistry data revealed the following: total bilirubin, 276.1 μmol/L; direct bilirubin, 187.5 μmol/L; alanine aminotransferase, 211 U/L; aspartate aminotransferase, 222 U/L; alkaline phosphatase, 617.5 U/L; gamma-glutamyl transpeptidase, 117.2 U/L; cancer antigen 19–9 (CA 19–9), 168.0 U/mL. No significant abnormalities were observed for the other tumor markers. Tests for hepatitis B virus surface antigen, hepatitis B virus surface antibody, hepatitis B virus e antigen, and hepatitis C virus antibody were negative. A contrast-enhanced abdominal computed tomography scan revealed the hilar bile duct to be locally enhanced, with nodule-like thickening and luminal narrowing (Figure 1a), significant stenosis was not observed at either the left or right hepatic duct openings, and the intrahepatic bile ducts above were visibly dilated, without enlargement of the gallbladder or any readily visible nodules in the liver (Figure 1b), considering bile duct cancer in the hilar bile duct. Computed tomography angiography showed that the origin and direction of extension of the common hepatic artery and its branches were normal, with no readily visible stenosis or dilatation.

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The main trunk of the portal vein was not widened, and there were no obvious filling defects in it. The opening and course of the left, middle, and right hepatic veins were normal, with no obvious signs of stenosis or dilatation (Figure 1c). Preoperatively, the boy was diagnosed with a malignant tumor of the hilar bile duct (Bismuth-Corlette II was considered) without metastasis to other organs or tissues.

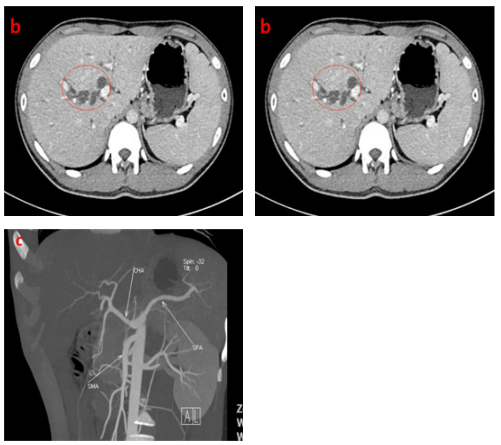

Figure 1: (a) Contrast-enhanced abdominal computed tomography scan showed the hilar bile duct to be locally enhanced, with nodule-like thickening and luminal narrowing. (b) The intrahepatic bile ducts above were obviously dilated, no significant stenosis was observed at the left or right hepatic duct openings. (c) Computed tomography angiography showed that the origin and direction of extension of the common hepatic artery and its branches were normal, with no obvious stenosis or dilatation.

After the evaluation of multidisciplinary discussion, he underwent resection of the common bile duct and common hepatic duct with gallbladder and Roux-en-Y hepaticojejunostomy. Intraoperatively, we found that the tumor was located at the bifurcation of right and left hepatic ducts, and it was approximately 1.2 cm in diameter. Extrahepatic bile duct resection from the hepatic hilar to the pancreas together with cholecystectomy and lymphadenectomy resection were performed (Figure 2a). The left and right hepatic duct margins were sent for intraoperative frozen pathology examination, and a small number of moderate to severe atypical dysplastic cells were visible in the right hepatic canal margins (Figure 2b). For this reason, we removed 0.5 cm right liver tube, and the freezing results showed no abnormality. Then, the right anterior and right posterior branches and the left hepatic bile were integrated into a larger bile duct opening, and Roux-en-Y hepaticojejunostomy was performed with the jejunum approximately 15 cm downstream of the ligament of Treitz (Figure 3a). Histologic examination showed low differentiation of the Neuroendocrine Tumor (NET) of the hilar bile duct (G3) (Figure 3b).

The tumor infiltrates the bile duct wall and involves the surrounding fibrous adipose tissue. We tested 19 lymph nodes, all of which were negative. Immunohistochemistry indicated that CD31 (intravascular tumor plugs were detected), CD56 (+), CEA (−), CK19 (+), D2-40 (no tumor plugs were detected in the lymphatic vessels), Desmin (−), Ki67 (20%), S100 (nerve invasion were be detected), Syn (+), and CgA (−). Postoperative recovery was regular. The patient was placed on a fluid diet on the 3rd postoperative day. No complications were observed, and the patient was discharged on the 8th postoperative day. The patient came back to the hospital 3 months later and recovered well without any special discomfort, and the rechecked CA199 was within the normal range.

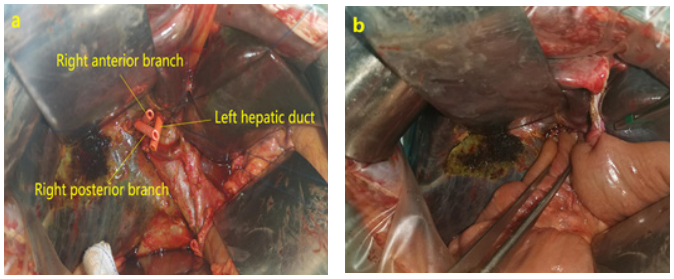

Figure 2: (a) Extrahepatic bile duct resection from the hepatic hilar to the pancreas together with cholecystectomy and lymphadenectomy resection. (b) Roux-en-Y hepaticojejunostomy was performed.

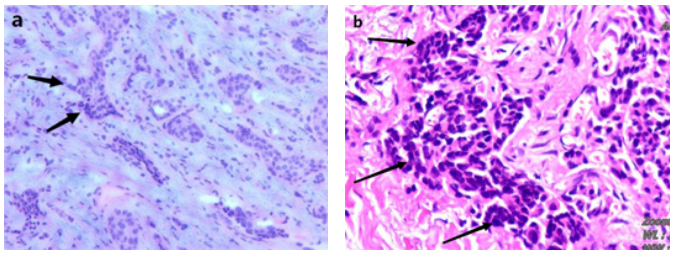

Figure 3: (a) Frozen pathology examinations indicated that a small number of moderate to severe atypical dysplastic cells were visible in the right hepatic canal margins, (hematoxylin and eosin, ×200). (b) Histologic examination showed low differentiation in neuroendocrine tumors,(hematoxylin and eosin, ×400).

RESULTS

NETs are unique tumors with typical histological, clinical, and biological characteristics. They include multipotent cells that can secrete large amounts of hormonal substances and vasoactive peptides. These include gastrin, somatostatin, vasoactive intestinal peptide, insulin, and glucagon [3, 4]. However, extrahepatic biliary NETs rarely secrete hormonal substances or vasoactive peptides that cause the corresponding clinical symptoms. NETs are caused by Kulchitsky cells in the Lieberkuhn crypt, which are visible in the gastrointestinal tract but very rare in the biliary system, which explains the low incidence of NETs of the extrahepatic bile duct. Because extrahepatic biliary NETs are rare and there is a lack of large case studies reported, Michalopoulos et al. performed a retrospective study in which they reported approximately 38 cases of extrahepatic biliary NETs from 1961 to 2013 in patients with a median age of 47 years (range 6–79 years) and a female predominance (61.5%) to date, only 8 cases in adolescents have also been reported [5]. The youngest of these cases was reported by a Mexican scholar, a case of a 12-year-old girl diagnosed with an extrahepatic biliary NET. She underwent resection of the common bile duct, cholecystectomy and end to side Roux-en-y hepaticojejunostomy, and portal lymphadenectomy [6]. NETs of the extrahepatic bile ducts can produce clinical symptoms, but these symptoms are usually related to obstruction of the bile ducts due to the increasing size of the tumor and compression of the surrounding tissues. They are rarely due to hormones or vasoactive intestinal peptides secreted. The most common clinical symptoms of NETs of extrahepatic bile are jaundice (60.3%) and pruritus (19.2%). Hormone and vasoactive peptide-associated symptoms were only reported in 9% of cases [3].

It has been reported that NETs of the extrahepatic biliary occur in

the hilar bile duct (11.5%), common bile duct (17.9%), cystic duct (16.7%), and distal common bile duct (19.2%), and the surgical approach to tumors at different sites varies [7]. In general, the surgical procedure for NETs of extrahepatic biliary is basically the same as that for adenocarcinoma of extrahepatic bile duct. In our case, the tumor was located at the bifurcation of right and left hepatic ducts, and postoperative pathological examination showed that the tumor had infiltrated the bile duct wall and involved the surrounding adipose tissue. Fortunately, we performed complete resection of the tumor and resection of the surrounding fibrous adipose tissue and lymphatic tissues, which theoretically achieved a good curative effect. The imaging presentation of NETs of extrahepatic biliary is similar to that of cholangiocarcinoma, and accurate preoperative diagnosis is difficult due to the lack of detectable serum markers and clinical hormone-related symptoms in patients. One study showed that only 4 of 87 patients were diagnosed with neuroendocrine or extrahepatic biliary tumors before surgery, and 2 of them had significantly elevated serum 5-hydroxytryptamine levels. In the other two patients, intraoperative biopsies were performed for pathological diagnosis during endoscopic retrograde cholangiopancreatography [8].

DISCUSSION

Currently, surgery is the most effective treatment for extrahepatic biliary NETs. Palliative treatment including systemic chemotherapy, growth inhibitor analogs, targeted therapy, and peptide receptor radionuclide therapy may also be able to control the progression of the disease to some extent, but most of these treatments are applicable to NETs of the gastroenteropancreatic system and not to extrahepatic biliary NETs. After all, NETs of the extrahepatic bile duct are rare. Only a few cases have been reported, and there is a dearth of systematic and large samples of data on the effects of clinical drug therapy.

CONCLUSION

Extrahepatic biliary NETs are relatively rare, and surgical treatment is the most effective treatment. Currently, fewer than 10 cases of extrahepatic biliary NETs have been reported in adolescents. We report a case in a 16-year-old boy. In our case, the primary tumor was surgically removed, we also report the patient's pathological findings and his postoperative recovery, and we will continue to follow the patient's condition long term.

COMPLIANCE WITH ETHICAL STANDARDS

Conflict of Interest

The authors declare that they have no conflict of interest.

Ethical Approval

All procedures performed in study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Author's Contributions

FHW participated in the acquisition of clinical data and drafted the manuscript. XL and KXZ revised the manuscript. FHD carried out the pathological examination and interpretation. LZ, HZ and BZ participated in the surgery as well as in the pre-operative imaging analysis. All authors have read and approved the final manuscript.

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Informed Consent

Written informed consent from the patient was obtained for publication of the case details.

REFERENCES

1. Lawrence B, Gustafsson BI, Chan A, Svejda B, Kidd M, Modlin IM. The epidemiology of gastroenteropancreatic neuroendocrine tumors. Endocrinol Metab Clin North Am. 2011; 40(1):1-8.
2. Radu S, Fung AW, Mayo SC. Primary Neuroendocrine Tumor of the Extrahepatic Biliary Tract. J Gastrointest Surg. 2021; 25(11):3007-3009.
3. Modlin IM, Lye KD, Kidd M. A 5-decade analysis of 13,715 carcinoid tumors. Cancer. 2003; 97(4):934-959.
4. Michalopoulos N, Papavramidis TS, Karayannopoulou G, Pliakos I, Papavramidis ST, Kanellos I. Neuroendocrine tumors of extrahepatic biliary tract. Pathol Oncol Res. 2014; 20(4):765-775.
5. Fernández-Ferreira R, Medina-Ceballos E, Soberanis-Piña PD, Conde-Flores E, Arroyave-Ramírez AM, Izquierdo-Tolosa CD, et al. Neuroendocrine Tumor of the Common Bile Duct: Case Report. Case Rep Oncol. 2021; 14:1785-1791.
6. Zhang L, Wan D, Bao L, Chen Q, Xie H, Xu S, et al. Neuroendocrine carcinoma in the extrahepatic biliary tract: a case report and literature review. Medicine. 2018; 97(29).
7. Raspanti C, Falco N, Silvestri V, Rotolo G, Bonventre S, Gulotta G. Neuroendocrine tumor of the common bile duct: case report. G Chir. 2016; 37(6):275.
8. Walter T, Brixi-Benmansour H, Lombard-Bohas C, Cadiot G. New treatment strategies in advanced neuroendocrine tumours. Dig Liver Dis. 2012; 44(2):95-105.