

A rare cause of bile duct obstruction in adolescence: Neuroendocrine tumor

Orçun Yalav¹, Abdullah Ülkü¹, Haluk Demiryürek¹, Figen Doran²

¹Department of General Surgery, Çukurova University Faculty of Medicine, Adana, Turkey

²Department of Pathology, Çukurova University Faculty of Medicine, Adana, Turkey

ABSTRACT

Extrahepatic bile duct is one of the rare places for neuroendocrine tumors which comprise 0.2-2 % off all neuroendocrine tumors of gastrointestinal tract. The aim of this paper is to report a case of a 16-year-old boy with a neuroendocrine tumor of extra hepatic bile duct. Laboratory and imaging findings is supported obstructive jaundice. After a pre-operative detailed evaluation, the common bile and common hepatic duct with gallbladder were resected and Roux-en-Y hepaticojejunostomy was performed. After a 36 months follow up, no locoregional recurrence or metastatic disease was observed. Pre-operative diagnosis of neuroendocrine tumors are extremely difficult and often confused with cholangiocarcinoma. Treatment modality of this rare entity depends on the location of the tumor and aggressive surgery is still the best choice of treatment. It should be kept in mind that disease-free survival for patients with neuroendocrine tumor depends on success of surgery.

Keywords: Neuroendocrine tumor, bile duct obstruction, surgical treatment

INTRODUCTION

Neuroendocrine tumor (NET) of extrahepatic bile tree are responsible for only 0,2-2 % off all neuroendocrine tumors of gastrointestinal tract (1). To the best of our knowledge, sixty-three patients with NET arising from extrahepatic bile duct are reported whereas only 5 of those are in adolescence (2). The current study reports a case of a NET originating from extrahepatic bile duct and adds the sixth case to the literature.

CASE PRESENTATION

A 16-year-old boy was admitted with a two week history of jaundice. There was no abnormality on physical examination except for icteric sclera. Laboratory tests was indicated obstructive jaundice (AST=156 IU/L, ALT=148 IU/L, Total Bil. / Direct Bil. =8.2/6.5 mg/dL, ALP=1063 IU/L, GGT=275 IU/L). Computed tomography and magnetic resonance cholangiopancreatography showed markedly dilatation of bilobar intrahepatic bile ducts (Figure 1). At the exploratory laparotomy, a polypoid mass in the middle of choledocus was palpated and no metastasis on the liver. The common bile and

common hepatic duct with gallbladder were resected and Roux-en-Y hepaticojejunostomy was performed. The patient was discharged one week after surgery. He is still in disease-free period for 40 months.

Histopathological findings

The excised specimen was exhibited immunohistochemical features of carcinoid tumor. Chromogranin (Figure 2) and Synaptophysin stains highlighted the cords of cells. The tumor was completely submucosal in location.

DISCUSSION

The cause of development of NET on extrahepatic bile duct is unclear thus the biliary tree rarely includes neuroendocrine cells. Chronic inflammation may be responsible for intestinal metaplasia due to increased inflammatory cells which may lead to development of a neuroendocrine tumor (3). The measurement of 5-hydroxy indole acetic acid in 24-hour urine and serum analysis of Chromogranin A (CgA) are helpful in diagnosis (4). Radiographic images of NETs of the bile

Address for Correspondence: Orçun Yalav, Department of General Surgery, Çukurova University Faculty of Medicine, Adana, Turkey

E-mail: drorcun2@yahoo.com

Received: May 15, 2012

Accepted: November 09, 2012

© Copyright 2014 by The Turkish Society of Gastroenterology • Available online at www.turkjgastroenterol.org • DOI: 10.5152/tjg.2014.3565



Figure 1. Black arrow indicates bilobar intrahepatic bile duct dilatation and red arrow shows location of obstruction on MRCP

ducts often confused with cholangiocarcinomas. Histological and immunohistochemical examination (Neuron-Specific Enolase, CgA and Synaptic Membrane Protein) are required to confirm the definitive diagnosis. Surgical excision is regarded as the main treatment. Removal of the tumor within the negative surgical margins should be aimed during surgery. Liver metastases also should be treated surgically or by other methods such as chemoembolization or radiofrequency ablation which may improve survival (5).

Currently, there is not enough information about NET of the bile ducts. Further studies in larger study samples are needed to clarify the ethiology, treatment and survival rates of the patients with NET. To our opinion, every additional report is helpful to improve the surgeons' awareness of this rare entity.

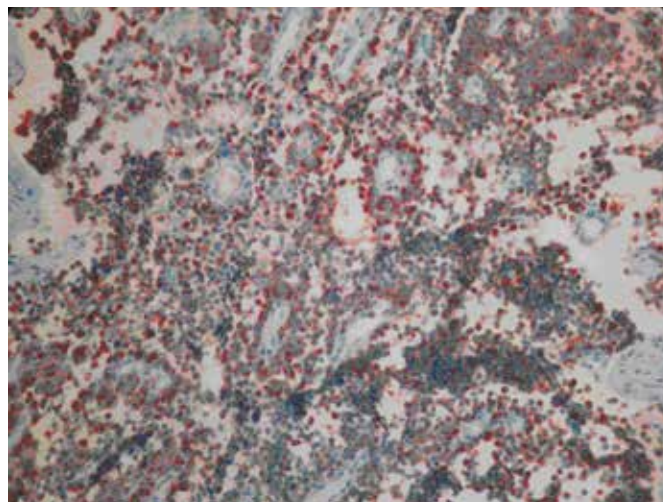


Figure 2. Chromogranin X400. The round, spindle shaped and polyhedral cells were irregularly distributed.

Conflict of Interest: No conflict of interest was declared by the authors.

REFERENCES

1. Felekouras E, Petrou A, Bramis K, Prassas E, Papaconstantinou I, Dimitriou N. Malignant carcinoid tumor of the cystic duct: a rare cause of bile duct obstruction. *Hepatobiliary Pancreat Dis Int* 2009; 8: 640-6.
2. Tonnhofer U, Balassy C, Reck CA, Koller A, Horcher E. Neuroendocrine tumor of the common hepatic duct, mimicking a choledochal cyst in a 6-year-old child. *J Pediatr Surg* 2009; 44: 23-5.
3. Barron-Rodriguez LP, Manivel JC, Mendez-Sanchez N, Jessurun J. Carcinoid tumor of the common bile duct: evidence for its origin in metaplastic endocrine cells. *Am J Gastroenterol* 1991; 86: 1073-6.
4. Sippel RS, Chen H. Carcinoid tumors. *Surg Oncol Clin N Am* 2006; 15: 463-78.
5. Sarmiento JM, Heywood G, Rubin J, Ilstrup DM, Nagorney DM, Que FG. Surgical treatment of neuroendocrine metastases to the liver: a plea for resection to increase survival. *J Am Coll Surg* 2003; 197: 29-37.