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Gangliocytic Paraganglioma With Atypical Immunohistochemical Features Presenting as Extrahepatic Biliary Obstruction

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Abstract

Gangliocytic paraganglioma is a rare benign tumor of upper gastrointestinal tract that most commonly involves the second part of duodenum. The tumor is detected incidentally on imaging in most of the cases. However, presentation with extrahepatic biliary obstruction is extremely rare. We recently encountered a 50-year-old male patient who was evaluated for extrahepatic biliary obstruction and was found to have a periampullary mass on imaging. The patient underwent pylorus-preserving pancreaticoduodenectomy along with liver biopsy and hepatoduodenal lymph node dissection. On histopathological examination, a tumor was detected in the periampullary region of duodenum, which was confirmed to be gangliocytic paraganglioma on immunohistochemistry along with atypical histological and immunohistochemical features.

Keywords

duodenum, extrahepatic biliary obstruction, gangliocytic paraganglioma, periampullary region

Introduction

Gangliocytic paraganglioma is a rare tumor of gastrointestinal tract that usually has a benign course with rare local metastases and a single report of a tumor-associated death.[1] The tumor occurs almost exclusively in the second part of duodenum predominantly in the periampullary region.[2] The histopathological hallmark of the tumor is the presence of 3 cell types—epithelioid, ganglion, and spindle cells.[2] Mitoses, nuclear pleomorphism, and infiltrative margins are indicators of aggressive behavior requiring radical surgery.[3] It is usually detected incidentally as an asymptomatic submucosal mass on radiological imaging.[4] Clinically, the patient may sometimes present with abdominal pain due to mass effect or bleeding due to mucosal ulceration. However, extrahepatic biliary obstruction is an extremely rare presentation of gangliocytic paraganglioma.[5] Few cases have been reported in literature. We report another case of gangliocytic paraganglioma in the duodenum at the level of ampulla of Vater causing extrahepatic biliary obstruction, which was removed by pancreaticoduodenectomy.

Case Report

A 50-year-old man presented with complaints of pruritis for 5 months and jaundice for 2 months with high-colored urine and clay-colored stools associated with loss of appetite and weight. He also had intermittent episodes of malaena. There were no other comorbidities except for past history of pulmonary tuberculosis, which was treated with antitubercular therapy for 6 months. On examination, he had icterus and tenderness in epigastrium and right hypochondrium. Biochemical parameters revealed deranged liver function tests with significantly elevated serum alkaline phosphatase (437 IU/L, normal = 30-120 IU/L) and direct bilirubin (1.3 mg/dL, normal = 0.1-0.4 mg/dL). Endoscopic ultrasound revealed a suspicious mass (1.5 × 1.3 cm) in the distal common bile duct (CBD), dilated CBD (1.7 cm), and pancreatic duct (0.5 cm) along with distended gallbladder showing internal echoes. Side viewing endoscopy revealed a bulky, friable, and ulcerated papilla. Endoscopic biopsy confirmed the presence of a tumor with morphological features favoring paraganglioma. Contrast-enhanced computed tomography of the abdomen also revealed a contrast enhancing mass (1.6 × 1.3 cm) at the level of distal CBD

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