

CASE REPORT

Synchronous Adenocarcinomas of the Papilla Major and Minor

Sriram Parthasarathy¹, Biju Pottakkat¹, Mutheeswaraiyah Yootla¹,
Sudhkar N Reddy¹, Kumaraswamy M Reddy²

Departments of ¹Surgical Gastroenterology, and ²Pathology,
Sri Venkateswara Institute of Medical Sciences. Tirupati, Andhra Pradesh, India

ABSTRACT

Context Synchronous malignancies affecting both the major and minor papilla are rare and are usually of endocrine origin.

Case report A 60-year-old female presented with a two-week history of progressive jaundice and pruritus. Evaluation revealed features of a periampullary malignancy. A Whipple pancreaticoduodenectomy was done. There was a growth in the ampulla of Vater and another in the region of the minor duodenal papilla. On histopathology, both lesions were adenocarcinomas.

Conclusion Although rare, synchronous adenocarcinomas can occur in the major and minor papilla.

INTRODUCTION

Malignancy affecting the minor papilla in the region of the accessory pancreatic duct is rare. Although there are a few reports of synchronous endocrine tumors in the major and minor papilla, synchronous carcinomas in this region are extremely rare. We herein report a patient who presented with synchronous adenocarcinomas of the major and minor papilla.

CASE REPORT

A 60-year-old woman presented to our outpatient clinic with a two-week history of

progressive jaundice and pruritus. She had also been suffering from fever and chills, consistent with cholangitis, for five days preceding her outpatient visit. A history of passing clay-colored stools was also present. She had had a laparoscopic cholecystectomy for a calculus cholecystitis 18 months earlier. Examination revealed icterus. No mass was palpable per abdomen. Her serum bilirubin was 4.0 mg/dL (reference range: 0.1-1.2 mg/dL) and serum alkaline phosphatase was 546 IU/L (reference range: 44-147 IU/L). Ultrasonography revealed bilobar intrahepatic biliary dilatation and dilatation of the common bile duct up to 16 mm extending to its distal end. There was a calculus 6 mm in diameter at the lower end of the common bile duct. Her symptoms were attributed to the common bile duct stone and an endoscopic extraction of the stone was planned.

On side viewing endoscopy, an ulcerated growth was detected in the ampulla of Vater. The plan for endoscopic stone extraction was dropped because of the presence of the growth. Tissues were taken for biopsy during endoscopy. Histopathology of these tissues revealed an adenocarcinoma. A contrast-enhanced computerized tomogram of the abdomen was performed to assess the stage of the disease. A contrast-enhanced computerized tomogram revealed dilatation of the common bile duct and the pancreatic duct. The common bile duct stone was clearly delineated in the contrast-enhanced

computerized tomogram (Figure 1). No mass was detected. There was no evidence of metastasis. Chest X-ray was normal.

The patient was scheduled for a laparotomy and a planned pancreaticoduodenectomy. After opening the abdomen, Kocherization of the duodenum was performed. On palpation, a button like growth was palpable in the perampullary region. The growth was completely localized to the perampullary area. There was no lymphadenopathy. Neither liver nor peritoneal metastasis was detected. A classical Whipple's pancreaticoduodenectomy was performed. The common bile duct and the pancreatic duct were dilated and a stone was present at the lower end of the common bile duct. After pancreaticoduodenectomy, reconstruction was done in the following order: pancreaticojejunostomy, hepaticojejunostomy and gastrojejunostomy.

Gross examination of the specimen revealed a 22x25 mm firm ulcerated growth in the ampulla of Vater. A 15x12 mm hard nodule was seen and felt, 2 cm proximal to the ampulla of Vater in the region of the minor papilla (Figure 2). The mucosa overlying the nodule in the minor papilla was normal. Microscopic examination of the ampullary growth revealed malignant cells with pleomorphic and vesicular nuclei. Papillary projections with a variable degree of dysplasia were visible on histopathology. A moderately differentiated adenocarcinoma was diagnosed. The tumor infiltrated the

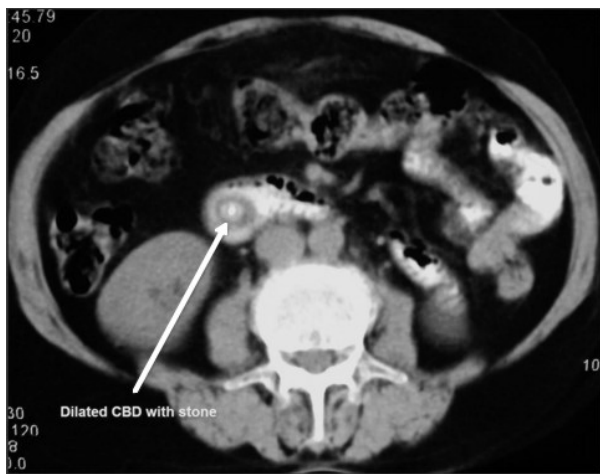


Figure 1. Contrast-enhanced computerized tomogram showing the dilated common bile duct with a calculus.

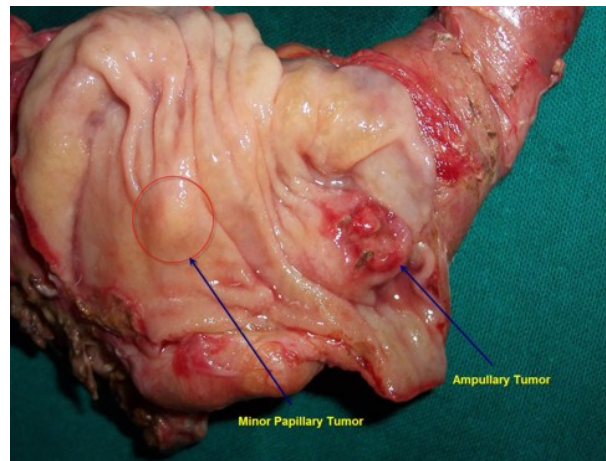


Figure 2. Pancreaticoduodenectomy specimen showing an ulcerated growth in the ampulla of Vater and a nodule in the minor papillary region.

pancreatic lobules superficially. There was ulceration of the duodenal mucosa due to involvement by the tumor. The minor papillary nodule also showed moderately differentiated adenocarcinomatous growth on histopathology. Histological examination of the lymph nodes which were removed at surgery did not reveal any metastasis. The tumor was staged as T3, N0, M0 (AJCC stage II).

The patient had an uneventful post-operative recovery and was discharged on the 10th post operative day. Post-operative adjuvant therapy was not scheduled because of the early stage of the disease. Eight months after the operation, there is no evidence of any recurrent disease and she is doing well.

DISCUSSION

Carcinoma of the ampulla of Vater is part of the spectrum of perampullary carcinomas, and they usually present with obstructive jaundice. Malignancy in the region of the minor papilla is rare. Although the majority of minor papillary malignancies are tumors of endocrine origin, primary adenocarcinomas have also been previously reported [1]. An association between minor papillary tumors and pancreas divisum is reported in the literature [2] and these patients may present with acute pancreatitis. Synchronous tumors affecting both the ampulla of Vater and the minor papilla are rare. Most of the cases reported are somatostatinomas [3, 4].

Only one report is available establishing the synchronous occurrence of adenocarcinomas in the major and minor papilla [5]. In this report, the patient presented with acute pancreatitis and jaundice and, although the ampullary growth was seen on side viewing duodenoscopy, the minor papillary tumor was demonstrated only after examination of the resected specimen. Our patient presented with obstructive jaundice and cholangitis. The common bile duct stone detected on ultrasonography was initially thought to be the cause of the jaundice and cholangitis. The side viewing endoscopy revealed the ampullary tumor, but it could not detect the tumor at the minor papilla. Similar to the previously published report, in our patient, the minor ampullary lesion was also detected only after examination of the resected specimen.

The pathogenesis of this synchronous pattern needs further study. It is not possible to differentiate a synchronous tumor from a metastatic deposit by conventional histopathology. Application of molecular diagnostic techniques would enable us to differentiate between these two patterns based upon the genetic differences. Synchronous carcinogenesis in the periampullary region may be a result of a malignant change in the pluripotent cells as proposed in synchronous somatostatinomas. Although the incidence is rare, in patients with a periampullary malignancy, awareness of this synchronous pattern may help surgeons and pathologists to specifically look at the minor papillary region for any associated malignancy.

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Correspondence

Biju Pottakkat

Department of Surgical Gastroenterology
Sri Venkateswara Institute of Medical
Sciences

Tirupati, Andhra Pradesh
India

Phone: +91-877.228.7777 ext 2418

Mobile: +91-990.883.7029

Fax: +91-877.228.6803

E-mail: bijupottakkat@rediffmail.com

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