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CASE REPORT | BILIARY

Utility of Endoscopic Retrograde Cholangiopancreatography in the Treatment of Intraductal Papillary Neoplasm of the Bile Duct

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ABSTRACT

Intraductal papillary neoplasm of the bile duct is a rare bile duct tumor that displays a range of cytoarchitectural atypia and is recognized as a precursor of invasive carcinoma. We present a 71-year-old woman with a recent diagnosis of lung adenocarcinoma, who presented with acute cholangitis secondary to an obstructive intraductal papillary neoplasm of the bile duct. The patient underwent endoscopic retrograde cholangiopancreatography, which identified the lesion, and on biliary sweep, the polyp presented externally and she underwent successful polypectomy with resolution of the infection.

INTRODUCTION

Intraductal papillary neoplasm of the bile duct (IPNB) is a rare bile duct tumor often compared with intraductal papillary mucinous neoplasm of the pancreas due to their similarities.[1] IPNB accounts for 9%–38% of all bile duct carcinomas and occurs most often in patients aged 50 to 70 years.[2–8] The greatest incidence occurs in Far Eastern countries and is predicted to be due to high-risk causes of hepatolithiasis and clonorchiasis in these endemic areas.[1] Between 40% and 80% of resected IPNB contains invasive components.[1,4,7,9,10] We present a case of an IPNB that caused acute cholangitis, in the setting of a newly diagnosed lung adenocarcinoma, which was removed via endoscopic retrograde cholangiopancreatography (ERCP).

CASE REPORT

A 71-year-old woman presented with a sudden onset of abdominal pain associated with nausea and vomiting. Medical history was significant for hypertension, hyperlipidemia, and newly diagnosed lung cancer. The patient had a recent admission for 1 day of nausea, vomiting, and abdominal pain. Liver function tests were notable for aspartate aminotransferase of 72 U/L, alanine aminotransferase of 61 U/L, alkaline phosphatase of 106 U/L, total bilirubin of 1.2 mg/dL, and direct bilirubin of 0.59 mg/dL. An abdominal ultrasound showed a dilated common bile duct (CBD), with findings suggestive of mild intrahepatic biliary duct dilation as well.

A follow-up abdominal computed tomography (CT) with intravenous contrast showed the previously noted intrahepatic and extrahepatic biliary dilation with abrupt cut off of the CBD at the level of the pancreatic head, along with soft-tissue attenuation within the CBD. No definitive pancreatic head mass was identified. Of note, a right lower lobe pulmonary nodule measuring 7 mm was also seen. A magnetic resonance cholangiopancreatography was recommended. A chest CT with intravenous contrast was done to further evaluate the pulmonary nodule, which showed a mass-like right apical consolidation worrisome for primary lung cancer, with additional nodules at the apex of the right lower lobe and the medial right lung base. The magnetic resonance cholangiopancreatography showed a 1.4 cm CBD dilation with a 1.4 × 1 cm enhancing lesion within the distal CBD consistent with neoplasm, which the surgery team favored to be a distal cholangiocarcinoma. The surgery team also believed that endoscopic ultrasound/fine-needle aspiration had

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limited utility for a resectable CBD mass. A diagnostic laparoscopy did not show any evidence of carcinomatosis. A CT-guided right upper lobe lung biopsy showed adenocarcinoma.

The symptoms resolved, and the patient tolerated diet and was subsequently discharged with outpatient follow-up. An outpatient positron emission tomographic/CT scan showed normal fluorodeoxyglucose uptake in the liver and gallbladder/biliary tree; however, there was fluorodeoxyglucose uptake in the right apical lung field (standardized uptake value 4.7, 4.0 × 2.7 cm) and in the superior segment of the right lower lobe (standardized uptake value 2.1, 1.8 × 1.5 cm). Subsequently, the patient presented to the emergency department again with sudden onset of abdominal pain as earlier. Vital signs on this admission showed a temperature of 102.6°F. Labs were notable for lactic acid of 3.74 mmol/L, aspartate aminotransferase of 519 U/L, alanine aminotransferase of 322 U/L, total bilirubin of 1.4 mg/dL, direct bilirubin of 0.72 mg/dL, and white blood cells of 11.1 K/μL. Abdominal CT showed stable, moderate intrahepatic biliary dilation and a soft-tissue filling defect in the CBD. Abdominal ultrasound showed a distended gallbladder and a 1.0 cm dilated CBD.

The patient was started on antibiotic coverage with piperacillin-tazobactam, and the blood cultures came back positive in 4/4 bottles for Gram-negative rods concerning for a biliary source. The patient was taken for an urgent ERCP with a plan for sphincterotomy and possible biopsy of the lesion and stent placement. ERCP showed that the entire main bile duct was moderately dilated with a mass, causing obstruction (Figure 1). Biliary sphincterotomy was performed and the bile duct was swept with a stone extraction balloon. This caused the polypoid mass to present externally. The base of the polyp was able to be identified, and a subsequent snare polypectomy of the whole polyp was performed (Figure 2).

Biopsy of the mass came back showing IPNB, low-grade, predominately intestinal type. The cauterized margin was focally positive for low-grade dysplasia, and review of the deeper sections showed no high-grade dysplasia or carcinoma. After the procedure, the patient's cholangitis resolved, and a surgical intervention, such as a Whipple procedure, the definitive treatment option for IPNB, was deferred until outpatient follow-up and repeat ERCP. An outpatient repeat ERCP demonstrated an ampullary adenoma and abnormal cholangiogram concerning for possible bile duct growth of previously removed IPNB. SpyGlass cholangioscopy demonstrated abnormal-appearing distal CBD mucosa worrisome for recurrence of the intraductal papillary mucinous neoplasm; however, biopsies were only consistent with intestinal-type mucosa. Given the patient's history of lung carcinoma, she is currently considering endoscopic ampullectomy vs surveillance of the ampullary adenomata.

DISCUSSION

IPNB is a rare disease, and the World Health Organization has only recently recognized IPNB as a distinct pathologic entity.[4] IPNB in North America has remained a relatively rare disease, and therefore, literature has remained limited unlike in Asia where the incidence is greater.[11] Patients with IPNB should all be considered for treatment because they frequently cause recurrent cholangitis and obstructive jaundice.[1] Moreover, although IPNB is considered premalignant, pathologic diagnosis

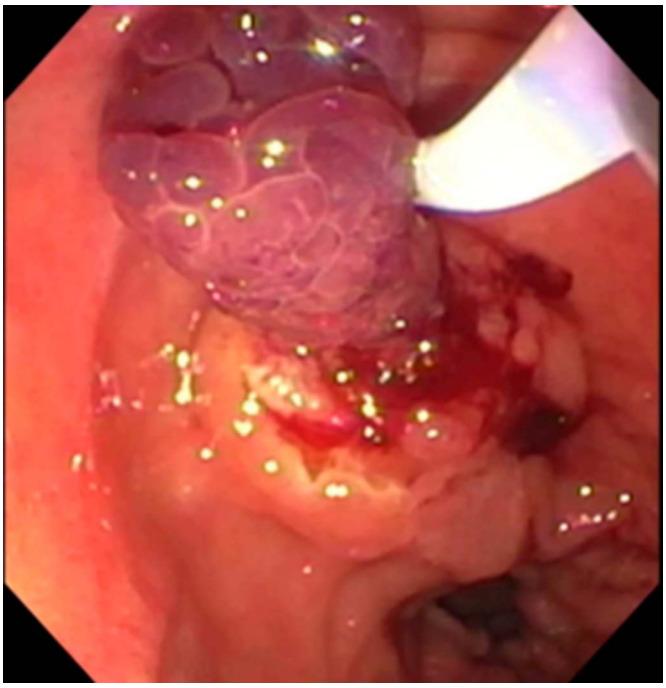

Figure 1. Intraductal papillary neoplasm of the bile duct swept externally during endoscopic retrograde cholangiopancreatography.

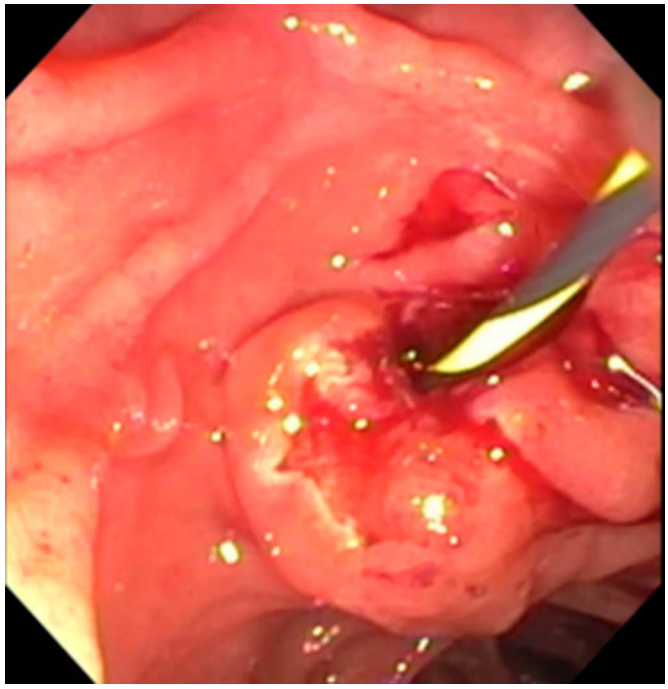

Figure 2. Intraductal papillary neoplasm of the bile duct removal after polypectomy.

by preoperative biopsy cannot always accurately reflect the maximum degree of cytoarchitectural atypia.[1] As a result, IPNB should be treated similarly to intrahepatic cholangiocarcinomas and extrahepatic bile duct carcinomas, and thus undergo surgical intervention such as major hepatectomy with or without extrahepatic bile duct resection or pancreaticoduodenectomy, also known as Whipple procedure.[1]

In the study by Rocha et al, it was concluded that most IPNB, 74%, contained a component of invasive carcinoma.[4] Moreover, they found that the median survival was 62 months and factors associated with a worse survival included the presence and depth of tumor invasion, margin-positive resection, and expression of mucin 1 and carcinoembryonic antigen.[4] With our patient, after the polypectomy, the biopsy ultimately came back with positive margins necessitating further workup, repeat ERCP, and the potential need for further surgical intervention.

In our review of the literature, IPNB is a rare disease with limited information, and we present a case of an IPNB removed by ERCP and polypectomy. Furthermore, we highlight the potential utility of ERCP in the management of IPNB.

DISCLOSURES

Author contributions: N. Patel wrote and edited the manuscript. A. Goodman edited the manuscript and is the article guarantor.

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Informed consent was obtained for this case report.

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REFERENCES

1. Ohtsuka M, Shimizu H, Kato A, et al. Intraductal papillary neoplasms of the bile duct. *Int J Hepatol*. 2014;2014:459091.
2. Barton JG, Barrett DA, Maricevich MA, et al. Intraductal papillary mucinous neoplasm of the biliary tract: A real disease? *HPB (Oxford)*. 2009;11(8):684–91.
3. Ohtsuka M, Kimura F, Shimizu H, et al. Similarities and differences between intraductal papillary tumors of the bile duct with and without macroscopically visible mucin secretion. *Am J Surg Pathol*. 2011;35(4):512–21.
4. Rocha FG, Lee H, Katabi N, et al. Intraductal papillary neoplasm of the bile duct: A biliary equivalent to intraductal papillary mucinous neoplasm of the pancreas? *Hepatology*. 2012;56(4):1352–60.
5. Onoe S, Shimoyama Y, Ebata T, et al. Prognostic delineation of papillary cholangiocarcinoma based on the invasive proportion: A single-institution study with 184 patients. *Surgery*. 2014;155(2):280–91.
6. Yeh CN, Jan YY, Yeh TS, Hwang TL, Chen MF. Hepatic resection of the intraductal papillary type of peripheral cholangiocarcinoma. *Ann Surg Oncol*. 2004;11(6):606–11.
7. Jung G, Park KM, Lee SS, Yu E, Hong SM, Kim J. Long-term clinical outcome of the surgically resected intraductal papillary neoplasm of the bile duct. *J Hepatol*. 2012;57(4):787–93.
8. Yang J, Wang W, Yan L. The clinicopathological features of intraductal papillary neoplasms of the bile duct in a Chinese population. *Dig Liver Dis*. 2012;44(3):251–6.
9. Wang X, Cai YQ, Chen YH, Liu XB. Biliary tract intraductal papillary mucinous neoplasm: Report of 19 cases. *World J Gastroenterol*. 2015;21(14):4261–7.
10. Yeh TS, Tseng JH, Chen TC, et al. Characterization of intrahepatic cholangiocarcinoma of the intraductal growth-type and its precursor lesions. *Hepatology*. 2005;42(3):657–64.
11. Bennett S, Marginean EC, Paquin-Gobeil M, et al. Clinical and pathological features of intraductal papillary neoplasm of the biliary tract and gallbladder. *HPB (Oxford)*. 2015;17(9):811–8.