Minor Papilla Adenoma Management in Patients with Pancreas Divisum and Familial Adenomatous Polyposis

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Abstract
Several case reports on endoscopic resection of minor papilla adenomas exist in the literature. However, there are no reported cases of endoscopic resection in patients with minor papilla adenomas with associated familial adenomatous polyposis (FAP) and pancreas divisum. We report a case of a minor papilla adenoma in a patient with FAP and pancreas divisum. The case demonstrates a new association between these disease processes. Defining pancreatic ductal anatomy prior to endoscopic intervention is essential. In addition, we demonstrate the safety and feasibility of endoscopic management of minor papilla tumors in patients with FAP and associated pancreas divisum.

Introduction
Tumors of the minor papilla are uncommon and, to date, are only described in case reports. In contrast, tumors of the major papilla account for approximately 5% of gastrointestinal (GI) neoplasms. Duodenal adenomas may occur as sporadic lesions or in patients with familial adenomatous polyposis (FAP). In the subset of patients with FAP, up to 90% of adult patients will develop duodenal adenomas. For many years, surgical resection was considered the mainstay of therapy for adenomas involving the major or minor papilla. In more recent years, advances in endoscopic papillectomy of ampullary adenomas has been shown to have a high success rate, low complication rate, and, importantly, a low recurrence rate. We report a case of endoscopic treatment of a minor papillary adenoma in a patient with FAP and associated pancreas divisum, a condition not previously reported with suggested algorithmic work-up.

Case Report
A 36-year-old woman with personal and family history of FAP, status post-total abdominal proctocolectomy with rectal mucosectomy and ileoanal pouch anastomosis (IPAA) performed at the age 18 after she was diagnosed with colon cancer, was found to have an adenoma involving her minor papilla. Her baseline upper endoscopy exam for evaluation of gastric and duodenal polyps was performed at age 30. On her initial endoscopy she had multiple 3–4-mm polyps in her duodenum, which were managed with endoscopic resection. No duodenal polyps were associated with the major or minor papilla at this time. In addition, she had multiple 3–4-mm fundic gland polyps in her stomach.

On follow-up surveillance endoscopy 2 years later, she was found to have a 1-cm adenoma involving her minor papilla. A biopsy of this polyp was consistent with a tubular adenoma without high-grade dysplasia. At EUS, the lesion was confined to the mucosal layers and was without duodenal or duodenal wall involvement. A suspicion of pancreas divisum at EUS prompted a non-invasive MRCP, which confirmed pancreas divisum anatomy (Figure 1).
During ERCP, a pancreatic spincterotomy was performed, followed by placement of a 5F x 5-cm dorsal pancreatic duct stent (Figures 2 and 3). Next, endoscopic snare resection of the duodenal lesion was performed, followed by fulgation of the remaining area of adenomatous-appearing tissue with APC (Figure 4). Six months later at follow-up endoscopy, biopsies of the minor papilla were negative for adenomatous tissue.

**Discussion**

There are several case reports on endoscopic resection of minor papilla tumors; however, this case represents the first report describing endoscopic minor papilla resection in a patient with FAP and pancreas divisum anatomy. Periampullary and duodenal tumors are reported to be present in up to 80% of patients with FAP, with the cancer risk 100-fold greater than that of the general population. No known association of FAP and pancreas divisum exists in the literature. Prior to the acceptance of endoscopic papillectomy as a safe and feasible technique, surgical removal was the standard of care. However, surgical removal has considerable morbidity and mortality. In contrast, endoscopic resection is a safe and effective treatment of these patients in the absence of ductal involvement and is the standard of care in most high-volume advanced endoscopy centers. Complications of endoscopic papillectomy are similar to those encountered with ERCP, including pancreatitis, perforation, bleeding, sedation complications, and cholangitis.
of adenomatous involvement of the ampulla. Complementing our endoscopic evaluation, performance of a secretin-enhanced magnetic resonance cholangiopancreatography (MRCP) has become a valuable adjunct given its non-invasive nature and its improved sensitivity in the diagnosis of many pancreaticobiliary disorders, including pancreas divisum. ERCP remains the gold standard for diagnosis of pancreas divisum, but it is invasive and associated with many complications including pancreatitis. Following EUS examination and review of diagnostic imaging, ERCP with sphincterotomy of the minor papilla is usually performed. The preferred method at endoscopy is en bloc resection of a given lesion, though piecemeal resection is sometimes employed when lesions are larger than 2 cm. Resection in lesions of this size or greater may leave residual tissue. Argon plasma coagulation is often then performed to ablate residual tissue, as was the case in our patient.

Complete adenoma resection is the goal of papillectomy, but recurrence rates after endoscopic snare papillectomy have been reported at 0–26%. Our patient demonstrated tumor-free margins on histological evaluation and surveillance endoscopy, with biopsy demonstrating no tumor recurrence at the resection site. Follow-up for our patient will include endoscopy with side-viewing duodenoscopy every 6 months for a minimum of 2 years, and endoscopic surveillance performed every 3 years thereafter, given her history of FAP.

We report the first case of minor papillary adenoma involvement in a patient with FAP and previously undiagnosed pancreas divisum anatomy. Based on our experience, MRI/MRCP in addition to EUS examination of the ampulla to exclude ductal involvement seems a prudent, non-invasive cross-sectional imaging procedure given the high degree of potential complications involving the major and minor endoscopic papillectomy. Our case demonstrates that endoscopic resection appears to be a feasible and safe alternative to surgery for management of minor papillary adenomas in patients with FAP and pancreas divisum anatomy.

Disclosures

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