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CASE REPORT | ENDOSCOPY

Endoscopic Papillectomy for Major and Minor Papillary Adenoma in Familial Adenomatous Polyposis

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ABSTRACT

Ampullary adenomas can occur sporadically or in the patients with adenomatous polyposis syndrome, including familial adenomatous polyposis (FAP) and *MUTYH*-associated polyposis. The potential for malignant transformation is high in the setting of FAP. Although endoscopic resection of major papillary adenoma has been well described, minor papilla adenomas are exceptionally rare. This is the second documented case of an endoscopic papillectomy in a patient with FAP with simultaneous ampullary adenoma of the major and minor papilla.

INTRODUCTION

Adenoma involving the ampulla of Vater can occur sporadically or in the setting of adenomatous polyposis syndromes, including familial adenomatous polyposis (FAP) and *MUTYH*-associated polyposis. FAP is a well-described form of hereditary colorectal cancer, with an estimated prevalence of 1 per 100,000 individuals.[1] The incidence of ampullary tumors is 200-300-fold higher among patients with FAP compared with the general population. Cumulative risk of cancer reaches as high as 10% by age 60 years. Peripapillary carcinoma is the second most common cause of cancer death after colorectal cancer in FAP.[2]

Treatment options for ampullary adenomas include local surgical excision, pancreatic duodenectomy, and endoscopic papillectomy. Despite excellent surgical outcomes, the morbidity and mortality rates (between 4% and 15%) with surgery cannot be discounted.[3] Although endoscopic resection of major papillary adenoma has been well described, minor papilla adenomas are exceptionally rare.[4] We report a unique case of endoscopic papillectomy in a patient with FAP with simultaneous ampullary adenoma of the major and minor papilla. To date, this is only the second such case described after Adler.[5]

CASE REPORT

A 25-year-old woman with known personal and family history of FAP was referred for an endoscopic mucosal resection of a 1.5-cm duodenal polyp, which further revealed a biopsy-confirmed tubular adenoma without dysplasia of both major and minor papillae (Figure 1). Endoscopic ultrasound showed no invasion into the deeper layers of mucosa and no pancreatic divisum on MRCP. Conventional endoscopic retrograde cholangiopancreatography (ERCP) was performed using the wire-guided cannulation technique. Contrast was injected into the common bile duct and pancreatic duct (PD), and no intraductal invasion was seen. Biliary and pancreatic sphincterotomies were not performed before resection. The submucosa of the major papillae was then injected with a combination of 1:10,000 epinephrine, methylene blue, and normal saline to better visualize the lesion. This was followed by an en bloc removal of the lesion using hot-snare papillectomy. The margins of the papillectomy site were ablated with argon plasma coagulation (APC) at 20 W for any possible residual adenoma. On the same examination, the minor papilla adenoma was also resected by performing saline solution lift injection, hot snare, and APC ablation of the papillectomy site (Figures 2 and 3). Temporary plastic stents were deployed in the main PD, common bile duct, and minor PD (Figures 4 and 5). Histopathology revealed tubular adenoma, with tumor-free margins and absence of invasive carcinoma in the

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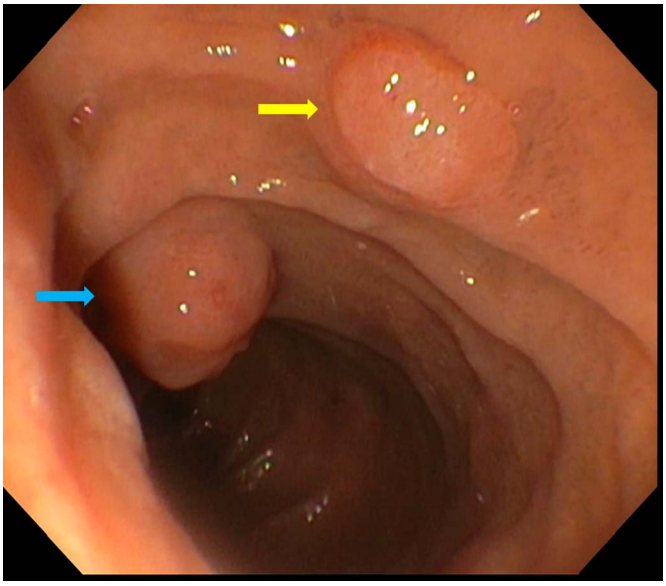

Figure 1. Major (blue arrow) and minor (yellow arrow) papillary adenoma.

major and minor papillae (Figure 6). She was discharged the same day without any complications and was asked to follow up for stent removal and surveillance endoscopy. Unfortunately, the patient moved and was lost to follow-up, but she was notified again for stent removal.

DISCUSSION

FAP and its variants follow an autosomal dominant pattern of inheritance because of a germline mutation in the tumor suppressor gene, adenomatous polyposis coli (*APC*), located on chromosome 5q21-q22.[6] They develop a wide variety of extracolonic manifestations, including polyps in the second and third portions of the duodenum and periampullary region. Papillary adenomas follow the adenoma-carcinoma sequence, similar to the one described for colorectal tumorigenesis.[7]

Routine duodenal surveillance using a side-viewing endoscope with random biopsy samples from normal-appearing papilla may harbor adenoma in 12%–54% of patients with FAP.[2] Although there is uniform agreement on resection of papillary adenoma, the unresolved issue remains between the optimal methods of excision. The conventional method of treatment has been radical pancreaticoduodenectomy, which confers a definitive approach because of the otherwise high recurrence rate and consequent progression to carcinoma. Nevertheless, it confers high mortality and morbidity, especially among patients with a previous major abdominal surgery.[2] In our case, the patient was offered surgical excision but refused, given her young childbearing age and the probability of needing a colectomy in the future. All these factors have favored less invasive endoscopic interventions such as snare papillectomy with frequent endoscopic surveillance. Ampullary adenoma without ductal extension is currently considered an indication for endoscopic papillectomy for tumors of the major papillae.[4]

Suzuki et al.[8] first described endoscopic papillectomy in 1983. There is no standardized technique for endoscopic removal of ampullary adenomas because of limited data. However, preoperative diagnosis and staging are imperative to determine the most appropriate therapeutic approach. EUS can be used as an adjunct for staging ampullary neoplasms. In addition, an MRCP before the procedure is valuable in excluding other pancreaticobiliary disorders such as pancreatic divisum.[9] EUS and ERCP not only helps in pretreatment staging but also helps

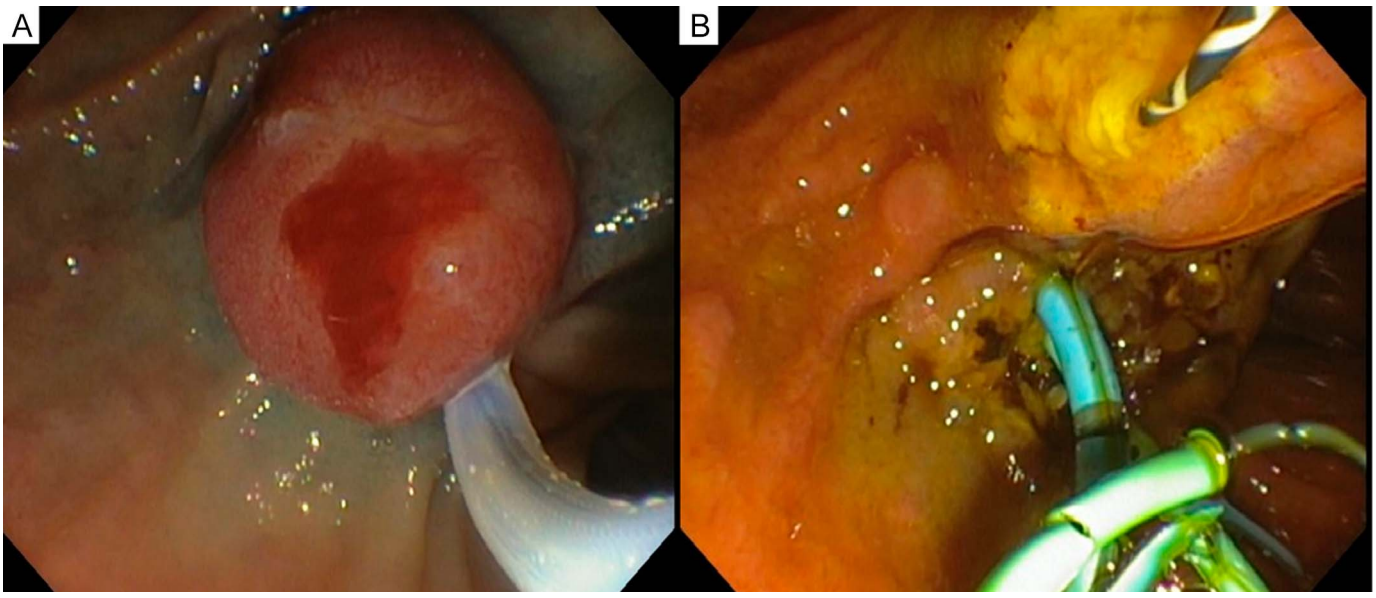

Figure 2. Endoscopic papillectomy of the (A) major duodenal papilla and (B) cannulation of the minor papilla.

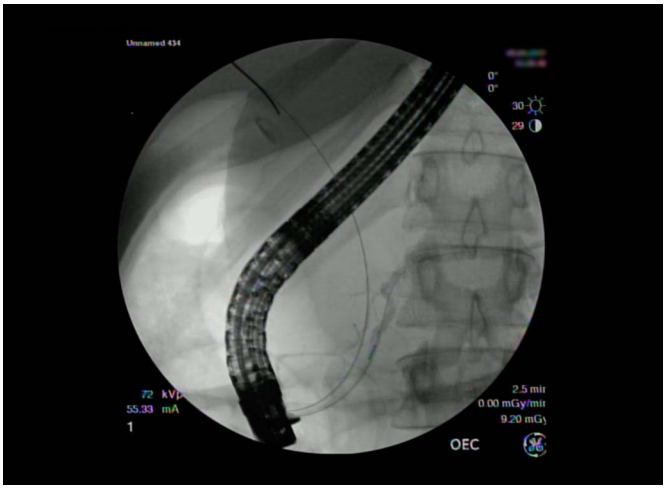

Figure 3. Cannulation of common bile duct and pancreatic duct using the double-wire technique.

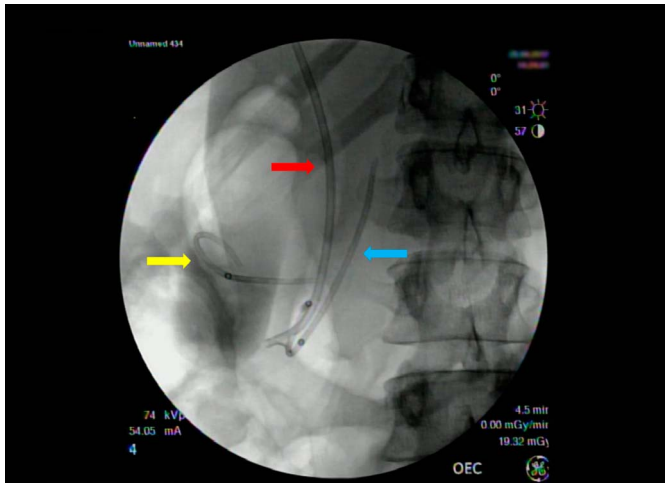

Figure 5. Fluoroscopic image showing a stent in the minor pancreatic duct (yellow arrow), common bile duct (red arrow), and main pancreatic duct (blue arrow).

in deploying prophylactic pancreatic stents to decrease the risk of post-ERCP pancreatitis. Pancreatic stents also minimize the risk of stenosis of the pancreatic orifice.[10] Papillectomy can be achieved either in an en bloc or piecemeal fashion. En bloc resection not only increases the likelihood of complete removal but also provides clear margins for histopathological evaluation.[8] After papillectomy, Nd: YAG laser or APC could be used to ablate residual tissue.[11,12]

Endoscopic papillectomy is a safe and effective procedure, but recurrence rates between 0 and 30% have been reported.[8] However, there exists a distinct difference in the recurrence rates among patients with FAP. A retrospective study by Ma et al looked at 26 patients with FAP who underwent EA, and adenoma recurrence was observed in 14 patients (58.3%), with most recurrences seen in adenomas >10 mm.[3] The adenoma recurrence rate is significantly higher in patients with FAP compared with sporadic adenoma.[13,14] These reports suggest an underlying lifelong genetic risk of ampullary adenomas to recur among patients with FAP. Thus, follow-up endoscopic surveillance plays an important role, especially in patients with FAP. Our patient had tumor-free margins without high-grade dysplasia on histopathology. Thus, our patient entails surveillance with side-viewing endoscopy 1–6 months after the index procedure, followed by examination every 3–12 months for a minimum of 2 years and every 3 years thereafter, given the history of FAP.[15]

In conclusion, we report the second case of a simultaneous minor and major papillary adenoma in a patient with FAP. Our case demonstrates endoscopic resection to be a safe first-line therapeutic approach, with minimal complications in expert hands.

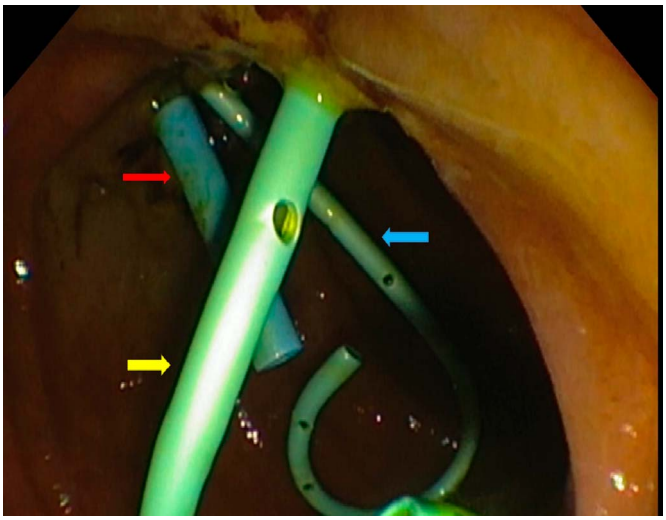

Figure 4. Stent in the minor pancreatic duct (yellow arrow), common bile duct (red arrow), and main pancreatic duct (blue arrow).

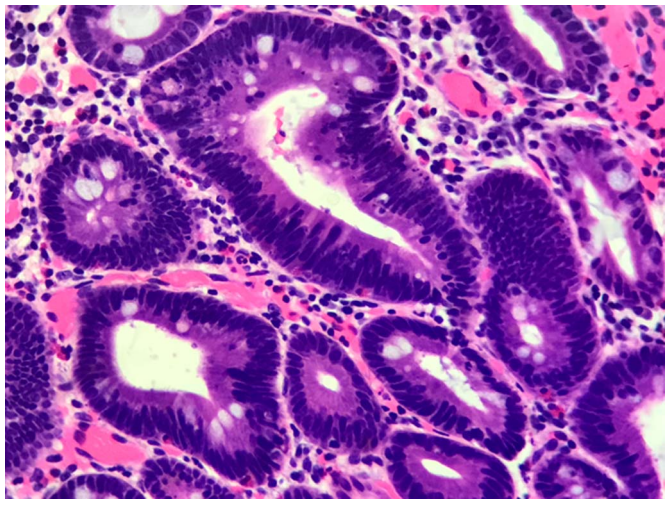

Figure 6. Histopathology shows that nuclei are elongated, hyperchromatic, and pseudostratified, suggestive of adenoma (hematoxylin and eosin stain 200×).

DISCLOSURES

Author contributions: M. Ahmed wrote the manuscript and searched the literature. J. Philipose searched the literature, edited the manuscript, and is the article guarantor. A. Hunton edited the manuscript. S. Andrawes was the study mentor and critically revised the manuscript.

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Informed consent was obtained for this case report.

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REFERENCES

1. Wennstrom J, Pierce ER, McKusick VA. Hereditary benign and malignant lesions of the large bowel. *Cancer*. 1974;34:850–7.
2. Gluck N, Strul H, Rozner G, Leshno M, Santo E. Endoscopy and EUS are key for effective surveillance and management of duodenal adenomas in familial adenomatous polyposis. *Gastrointest Endosc*. 2015; 81:960–6.
3. Ma T, Jang EJ, Zukerberg LR, et al. Recurrences are common after endoscopic ampullectomy for adenoma in the familial adenomatous polyposis (FAP) syndrome. *Surg Endosc*. 2014;28:2349–56.
4. Matsui T, Matsubayashi H, Hotta K, Sasaki K, Ito H, Ono H. A case of carcinoma in an adenoma of the duodenal minor papilla successfully treated with endoscopic mucosal resection. *Endosc Int Open*. 2016;4: E252–4.
5. Adler DG. Endoscopic ampullectomy of simultaneous major and minor papilla adenomas in a patient with attenuated familial adenomatous polyposis. *Gastrointest Endosc*. 2016;84:866–7.
6. Burt RW, DiSario JA, Cannon-Albright L. Genetics of colon cancer: Impact of inheritance on colon cancer risk. *Annu Rev Med*. 1995;46:371–9.
7. Brosens LA, Keller JJ, Offerhaus GJ, Goggins M, Giardiello FM. Prevention and management of duodenal polyps in familial adenomatous polyposis. *Gut*. 2005;54:1034–43.
8. Suzuki K, Kantou U, Murakami Y. Two cases with ampullary cancer who underwent endoscopic excision. *Prog Dig Endosc*. 1983;23:236–9.
9. Lapp RT, Hutchins GF. Minor papilla adenoma management in patients with pancreas divisum and familial adenomatous polyposis. *ACG Case Rep J*. 2013;1:47–50.
10. Chathadi KV, Khashab MA, Acosta RD, et al. The role of endoscopy in ampullary and duodenal adenomas. *Gastrointest Endosc*. 2015;82:773–81.
11. De Palma GD. Endoscopic papillectomy: Indications, techniques, and results. *World J Gastroenterol*. 2014;20:1537–43.
12. Bleau BL, Gostout CJ. Endoscopic treatment of ampullary adenomas in familial adenomatous polyposis. *J Clin Gastroenterol*. 1996;22:237–41.
13. Catalano MF, Linder JD, Chak A, et al. Endoscopic management of adenoma of the major duodenal papilla. *Gastrointest Endosc*. 2004;59:225–32.
14. Irani S, Arai A, Ayub K, et al. Papillectomy for ampullary neoplasm: Results of a single referral center over a 10-year period. *Gastrointest Endosc*. 2009;70:923–32.
15. Pandolfi M, Martino M, Gabbrielli A. *Endoscopic Treatment of Ampullary Adenomas*. Jop: Italy, 2008, pp 1–8.