The Korean Journal of Internal Medicine: 19:257-260, 2004

A Carcinoid Tumor of the Ampulla of Vater Treated by Endoscopic Snare Papillectomy

Dae-Keun Pyun, M.D., Gyoo Moon, M.D.[2], Jimin Han, M.D., Myung-Hwan Kim, M.D., Sang Soo Lee, M.D., Dong-Wan Seo, M.D. and Sung-Koo Lee, M.D.

Department of Internal Medicine, University of Ulsan Collage of Medicine, Asan Medical Center, Seoul, Korea ; Department of Internal Medicine, Song-do Hospital[2], Seoul, Korea

Here, a case of a patient with incidental finding of a carcinoid tumor of the ampulla of Vater, who was treated with endoscopic snare papillectomy, is reported. A 62-year-old male was admitted to our hospital due to a carcinoid tumor of the ampulla of Vater, which was found during follow-up endoscopy after an endoscopic mucosal resection of early gastric cancer. No lymphadenopathy or visceral metastasis was found on an abdominal CT scan, In-111 octerotide scan and EUS. The ampulla was then en bloc removed by endoscopic snare papillectomy. The resected specimen revealed a 0.7×0.5×0.1 cm sized carcinoid tumor. All margins of resection were negative for tumor. After six months of follow-up, there was no evidence of recurrence and metastasis, either endoscopically or radiologically. To our knowledge, this case is the first report of an ampullary carcinoid tumor treated by endoscopic snare papillectomy in Korea.

Key Words : Carcinoid tumor, Ampulla of Vater, Cholangiopancreatography, Endoscopic Retrograde.

INTRODUCTION

Carcinoid tumors of the ampulla of Vater are extremely rare[1]. The natural history of this disease entity has not been well established, but it has been postulated that the prognosis is generally good[1, 2]. In contrast to carcinoid tumors arising in the jejunum and ileum, the clinical and laboratory findings of carcinoid syndrome are absent in patients with a carcinoid tumor of the ampulla of Vater, such as duodenal carcinoid tumors[3, 4]. However, in small numbers of carcinoid tumors of the ampulla of Vater can show more aggressive behaviors, such as distant metastasis. Therefore, the standard treatment for this entity has been complete surgical removal[5]. There have been several reports of endoscopic treatment of small carcinoid tumors arising from the duodenum[6, 7], but there has been no report of pretreatment diagnosis and intentional endoscopic treatment of carcinoid tumors of the ampulla of Vater in Korea.

The ampulla of Vater is a complex structure, and is the confluent portion of common bile duct, pancreatic duct and contains the sphincter of Oddi. This may explain the reason why an ampullary carcinoid often clinically manifests as obstructive jaundice or acute pancreatitis, and an attempt to remove the tumor may result in more frequent procedure-related complications. Here, a case of a carcinoid tumor of the ampulla of Vater, diagnosed before treatment and intentionally treated by endoscopic snare papillectomy, is reported.

CASE REPORT

A 62-year-old male was admitted with an incidental finding of a carcinoid tumor of the ampulla of Vater. The patient had undergone an endoscopic mucosal resection (EMR) of early gastric cancer 1 year earlier. Every 3 month following the EMR, he has undergone surveillance gastroscopic examination with forward-viewing endoscopy. During the latest endoscopic examination, subtle abnormalities were noted on the covering mucosa of the ampulla of Vater. Biopsies were taken from the lesion, which was histologically confirmed as a carcinoid tumor. He denied weight loss, diarrhea, flushing, or any respiratory

• Received : April 12, 2004
• Accepted : June 16, 2004
• Correspondence to : Gyoo Moon, M.D., Department of Internal Medicine, Song-do Hospital, 366-144 Shindang 3-dong, Jung-gu, Seoul, 100-453, Korea Tel : 82-2-2231-0900, Fax : 82-2-2233-8528, E-mail : sun6230@kornet.net

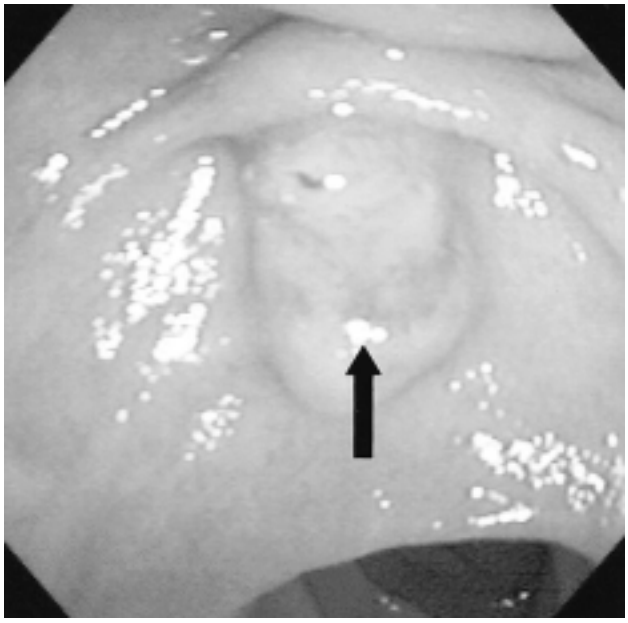

Figure 1. Endoscopic view of carcinoid tumor of the ampulla of Vater. Side-viewing duodenoscope showed a rather prominent, but preserved configuration of the ampulla, and depressed erosion (arrow) with hyperemia on the covering mucosa.

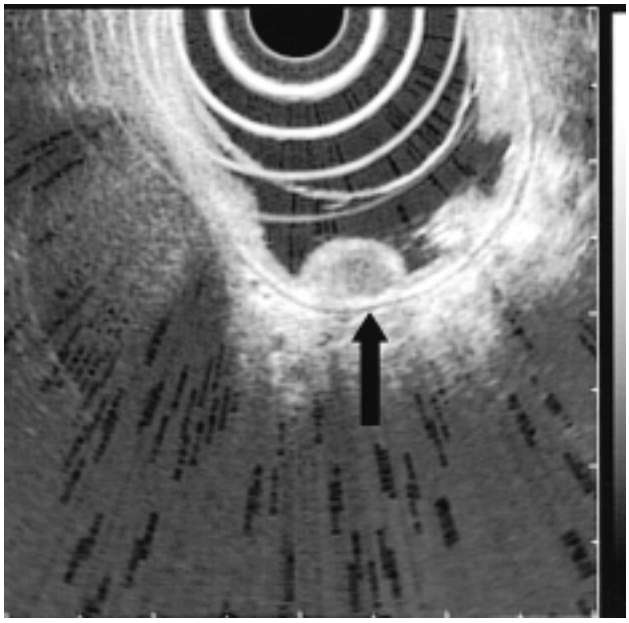

Figure 2. EUS showed a 0.7×0.3 cm sized, hypoechoic lesion (arrows) confined to the mucosal layer. The proper muscle layer was intact.

difficulties or obstructive biliary symptoms.

He underwent ERCP for a more detailed evaluation of the ampulla of Vater. An examination with a side-viewing duodenoscope showed prominent, but a preserved configuration of the ampulla, but depressed erosion with hyperemia on the covering mucosa (Figure 1). There was no definite ulcer or mass. Contrast agent was injected into common bile and pancreatic ducts, but no abnormality within the lumen of either the bile or pancreatic ducts were seen. Following the ERCP, EUS was performed to determine the depth of invasion and regional lymph node status. This revealed a hypoechoic, ovoid shaped lesion less than 1 cm in diameter arising from the mucosal layer (Figure 2). There was no evidence of proper muscle layer invasion and regional lymphadenopathy. An abdominal CT scan was negative for lymphadenopathy or visceral metastasis. An In-111 octreotide scan revealed no definitive metastatic lesion. A chest X-ray was also negative. The patient then underwent endoscopic snare papillectomy for the removal of the carcinoid tumor of the ampulla of Vater (Figure 3). After elevation of the tumor by submucosal injection of saline solution, the ampulla was grasped by snare and a papillectomy performed using blended electrosurgical current (cut to coagulation ratio 4:1) at a setting of 50 watts employing a electrosurgical unit (UES-20, Olympus Optical Co., Tokyo, Japan). The tumor was resected en bloc. Then, plastic biliary (7 Fr, 3 cm) and pancreatic (5 Fr, 3 cm) stents were inserted to prevent possible cholangitis and

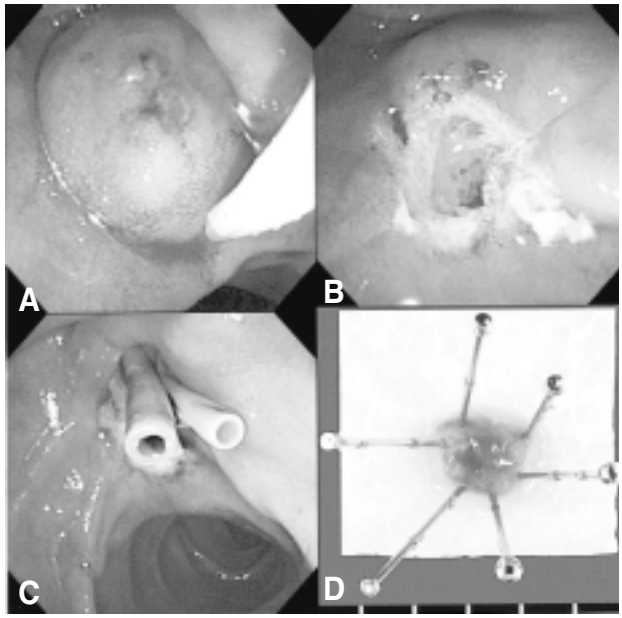

Figure 3. Endoscopic snare papillectomy of a carcinoid tumor of the ampulla of Vater. (A) Endoscopic view showing snaring of the carcinoid tumor of the ampulla of Vater after a submucosal injection of saline solution. (B) Endoscopic view immediately after papillectomy using blended electrosurgical current (C) Biliary (7 Fr, 3 cm) and pancreatic (5 Fr, 3 cm) stents were inserted to prevent possible cholangitis and pancreatitis. (D) Gross finding of the resected specimen.

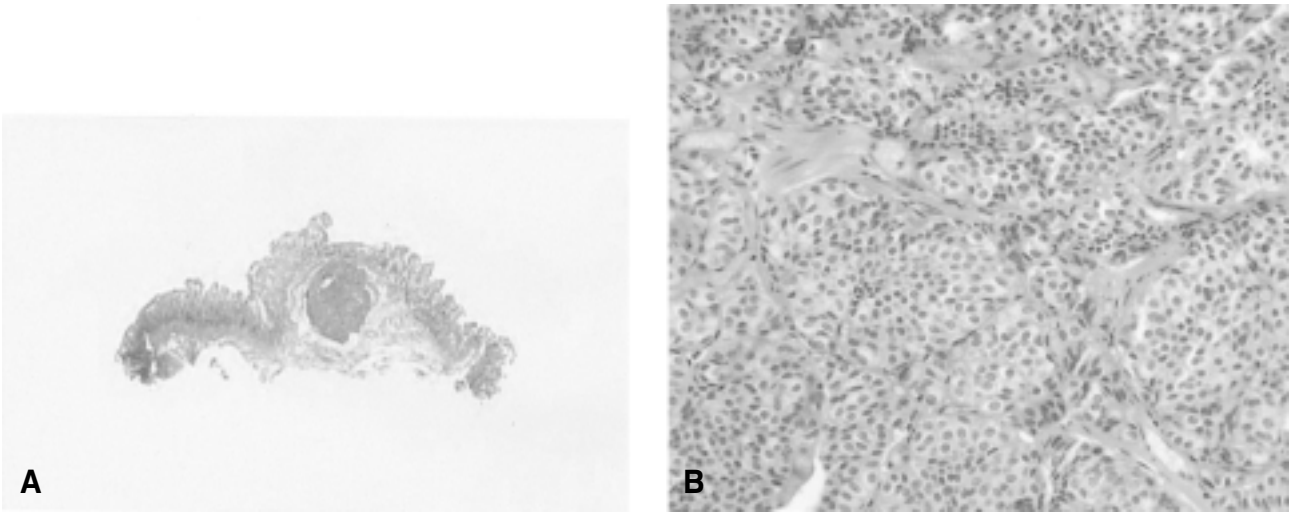

Figure 4. Pathological findings of a carcinoid tumor of the ampulla of Vater. (A) Under low power magnification, the typical histological appearance of an intramucosal tumor is seen.The tumor is a discrete, though not encapsulated, mass of multiple nests of small cells in the deep mucosa (hematoxylin and eosin, ×10). (B) The nests of a carcinoid tumor have a typical endocrine appearance, with small round cells having small round nuclei and pink to pale blue cytoplasm (hematoxylin and eosin, ×40).

pancreatitis. There was no complication after endoscopic snare papillectomy, i.e., cholangitis, pancreatitis, bleeding or perforation.

Pathologic analysis of the resected specimen revealed a 0.7 ×0.5×0.1 cm sized carcinoid tumor of the ampulla of Vater within the mucosal layer (Figure 4). All margins of resection were negative for tumor. Immunohistochemical staining of the tumor was strongly positive for synaptophysin and chromogranin. The patient had an uneventful hospital course and was discharged on the 5th post-papillectomy day following the removal of the biliary and pancreatic stents.

Two months after the endoscopic snare papillectomy, a follow-up duodenoscopic examination was performed, which revealed no evidence of tumor recurrence, either macroscopically or microscopically. Six months after the endoscopic snare papillectomy, there was no evidence of local recurrence and metastasis of the tumor, either endoscopically or radiologically.

DISCUSSION

Carcinoid tumors of the ampulla of Vater are very rare[1]. According to a review of 90 reported ampullary carcinoid cases, the mean patient age was 52 years[8], and affected patients were more often male[8]. Jaundice was the most common presenting symptom of a carcinoid tumor of the ampulla of Vater and was present in 59% of patients[8]. It is often apparent at an early stage because of the location; obstructive symptoms may arise early when the tumor is relatively small[3].

Despite frequent regional lymph node metastasis, the prognosis of carcinoid tumors of the ampulla of Vater has generally been considered good. In one study, the 5 year survival was 90%, with only 4 (6%) patients dying of a metastatic disease or progressive tumor[1, 2]. Even though there are reports on the benign nature of this tumor, they have malignant potential[4]. Unlike carcinoid tumors of the duodenum, the size of the tumor seems to have no prognostic implications for carcinoid tumor of the ampulla of Vater[8]. In one review of 73 cases of carcinoid tumors of the ampulla of Vater, thirty one were larger than 2 cm in diameter and 48% (15/31) of these had metastasized. Interestingly, 40% (17/42) of patients with tumors less than 2 cm also had a metastatic disease[9]. Hence, the tumor size was not a reliable predictor of aggressiveness; therefore, complete resection of the tumor is mandatory regardless of the size.

In our case, careful examinations, including EUS and abdominal CT scan, were performed before determining the therapeutic options. The size of the tumor was small (less than 1 cm) and there was no evidence of muscularis propria invasion and regional lymph node metastasis on EUS. An In-111 octreotide scan performed to evaluate occult metastases showed negative findings. Since the majority of carcinoid tumors express receptors for somatostatin, the detection rate using an In-111 octreotide scan ranges from 80 to 90%, and most investigators agree that this radionuclide scan and conventional imaging, such as CT, are complementary[9, 10].

The treatment of choice for carcinoid tumors of the ampulla of Vater is complete resection, and the standard treatment modality for the tumor has been surgery, such as pancreati-

coduodenectomy or local excision of the tumor. In one review of 90 patients with an ampullary carcinoid, 52 were treated with a pancreaticoduodenectomy and 22 with local excision of the tumor[8]. Local excision was generally performed in patients with tumors less than 2 cm in diameter, while a pancreaticoduodenectomy was performed in patients with tumor larger than 2 cm. Three of 52 patients who underwent a pancreaticoduodenectomy died of postoperative complications. On the other hand, 21 of the 22 patients who underwent local excision are alive, with no evidence of recurrence after long-term follow-up, and only one patient died of local recurrence 20 months following the local excision.

Either a pancreaticoduodenectomy or local excision, via a duodenotomy, may be selected depending on the size of the tumor[1, 11, 12]. Although a pancreaticoduodenectomy enables complete resection of the tumor, this procedure has the disadvantage of relatively higher morbidity[1]. Local excision showed satisfactory results in tumors less than 2 cm[1]. As a result, local excision may be an option for the treatment of a carcinoid tumor of the ampulla of Vater if the size of the tumor is small and there is no evidence of regional lymph node or distant metastasis. Compared to local surgical excision, endoscopic snare papillectomy may be much less harmful to the patient, since it does not require a laparotomy and duodenotomy. In the management of an ampullary adenoma, endoscopic therapy seems to be successful and may be a reasonable alternative to surgical resection[14].

A carcinoid tumor of the ampulla of Vater originates from the deep mucosa[7]. Most reports describe it as a round or oval mass, with intact overlying duodenal mucosa, with negative biopsies[1]. Only a few reports present it as a prominent papilla, with a shallow ulcer on the surface[3, 13]. However, a carcinoid tumor of the ampulla of Vater can show mucosal change, theoretically because it originated from the deep mucosa, not the submucosa. In our case, depressed erosion, which may be misinterpreted as papillitis, was found. Without a biopsy, this subtle mucosal change may be misdiagnosed as papillitis. Thorough examination, including the ampulla, is essential in all patients undergoing esophagogastroduodenoscopy, and biopsy of suspicious mucosal changes should always be carried out to diagnose the tumor in early stage.

In conclusion, endoscopic snare papillectomy may be one therapeutic option in the management of carcinoid tumors of the ampulla of Vater in selected patients, such as those with a small sized and localized in the mucosa or submucosal layer, without regionally or distant metastasis. Further experience with more cases will be needed to establish the exact indication for endoscopic snare papillectomy.

REFERENCES

1) Hatzitheoklitos E, Bchler MW, Friess H, Poch B, Ebert M, Mohr W, Imaizumi T, Beger HG. *Carcinoid of the ampulla of Vater: clinical characteristics and morphologic vfeatures. Cancer 73:1580-1588, 1994*
2) Klein A, Clemens J, Cameron J. *Peripampullary neoplasm in von Recklinghausen's disease. Surgery 106:815-819, 1989*
3) Kim MH, Lee SK, Seo DW, Won SY, Lee SS, Min YI. *Tumors of the major duodenal papilla. Gastrointest Endosc 54:609-620, 2001*
4) Makhouf HR, Burke AP, Sobin LH. *Carcinoid tumors of the ampulla of Vater: a comparison with duodenal carcinoic tumors. Cancer 85:1241-1249, 1999*
5) Zyromski NJ, Kendrick ML, Nagorney DM, Grant CS, Donohue JH, Farnell MB, Thompson GB, Farley DR, Sarr MG. *Duodenal carcinoid tumors: how aggressive should we be? J Gastrointest Surg 5:588-593, 2001*
6) Ricci JL. *Carcinoid of the ampulla of Vater: local resection or pancreaticoduodenectomy. Cancer 71:686-690, 1993*
7) Albizzatti V, Casco C, Gastaminza M, Speroni A, Mauro G, Rubio HW. *Endoscopic resection of two duodenal carcinoid tumors. Endoscopy 32:S42, 2000*
8) Clements WM, Martin SP, Stemmerman G, Lowy AM. *Ampullary carcinoid tumors: rationale for an aggressive surgical approach. J Gastrointest Surg 7:773-776, 2003*
9) Anthony LB, Martin W, Delbeke D, Sandler M. *Somatostatin receptor imaing: predictive and prognostic considerations. Digestion 57(Suppl 1):50-53, 1996*
10) Krenning EP, Kooij PP, Pauwels S, Breeman WA, Postema PT, de Herder WW, Valkema R, Kwekkeboom DJ. *Somatostatin receptor: scintigraphy and radionuclide therapy. Digestion 57(Suppl 1):57-61, 1996*
11) Yoshikane H, Tsukamoto Y, Niwa Y, Goto H, Hase S, Mizutani K, Nakamura T. *Carcinoid tumors of the gastrointestinal tract: evaluation with endoscopic ultrasonography. Gastrointest Endosc 39:375-383, 1993*
12) Soga J. Carcinoid tumors: *an analysis of Japanese series. J Exp Clin Cancer Res 4:233-244, 1985*
13) Sivak MV. *Clinical and endoscopic aspects of tumors of the ampulla of Vater. Endoscopy 20:211-217, 1988*
14) Catalano MF, Linder DL, Chak A, Sivak MV Jr, Raijman I, Geenen JE, Howell DA. *Endoscopic management of adenoma of the major duodenal papilla. Gastrointest Endosc 59:225-232, 2004*