

CASE REPORT

Endoscopic resection of an ampullary carcinoid presenting with upper gastrointestinal bleeding: A case report and review of the literature

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Abstract

Ampullary carcinoid is a rare tumor that can present with gastrointestinal bleeding, obstructive jaundice or pancreatitis. Some of these tumors are associated with Von Recklinghausen disease. The usual surgical options are a biliary-enteric anastomosis, Whipple procedure or rarely a local resection. The mean survival does not appear to be much different after a pancreaticoduodenectomy versus local surgical excision. We report a very rare case of a non-metastatic ampullary carcinoid causing upper gastrointestinal bleeding, which was managed by endoscopic ampullectomy.

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Key words: Carcinoid; Ampulla of Vater; Ampullary tumor; Ampullectomy; Gastrointestinal bleeding

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INTRODUCTION

Despite being rare, carcinoids are the most common neuroendocrine tumors of the gastrointestinal (GI) tract^[1,2]. Carcinoid of the ampulla is an extremely rare tumor (0.05%) for which the correct pre-operative diagnosis is very low (14%) and very rarely they present with an ulceration of the overlying duodenal mucosa^[2]. Less than 3% of ampullary carcinoids patients experience carcinoid syndrome^[3]. These tumors generally have an indolent course, but more aggressive behavior in the

form of distant metastases has been reported. We report a case of an ampullary carcinoid that manifested as an upper gastrointestinal bleeding and was managed with endoscopic snare resection of the ampulla of Vater.

CASE REPORT

A 71-year old man underwent upper endoscopy for the work up of a recent history of epigastric pain, melena and a 4 g decrease in hemoglobin. The esophagogastroduodenoscopy (EGD) was unremarkable except for a prominent ampullary area. A sideviewing scope showed an approximately 1.5 cm mass with an overlying superficial ulcer (Figure 1A). Biopsy specimen revealed a carcinoid tumor involving the muscularis propria (Figure 2A and B). The tumor lacked mitotic activity and psammoma bodies (sometimes associated with duodenal somatostatinomas). The tumor was positive for chromogranin, synaptophysin and somatostatin, and negative for gastrin, insulin, glucagon and human pancreatic polypeptide. The patient denied flushing, diarrhea, weight loss or wheezing. His liver enzymes were normal. A CT and an octreotide scan were unremarkable. EUS revealed no regional lymph nodes and tumor free pancreatic and common bile ducts. A serum serotonin level was normal at 80 ng/mL (22-180 ng/mL) and urinary 5-HIAA was 6.1 mg/24 h (normal up to 6 mg/24 h).

The patient declined aggressive surgical options but agreed to undergo an endoscopic ampullectomy with its modest risk of complications. After submucosal saline injection in the ampullary area, a snare assisted ampullectomy was performed with blended current (cut to coagulation ratio 5:1) at a setting of 150:30 watts using an electrosurgical unit (ERBE ICC-200, Tubingen, Germany). The tumor was removed piecemeal. A 5 Fr, 3 cm pancreatic stent was placed to prevent pancreatitis. The procedure was uneventful except for mild oozing which promptly stopped with local infiltration of 4 cc of 1/10000 epinephrine. The patient remained stable except for an asymptomatic transient elevation of transaminases and was discharged the next evening. The pancreatic stent was removed two weeks later and there was no residual tumor seen at this examination (Figure 1B). He did not have any further episodes of GI bleeding in the last three years. His hemoglobin and liver related tests were normal. Subsequent follow-up endoscopies using a side-viewing scope showed no residual tumor and biopsies (using jumbo

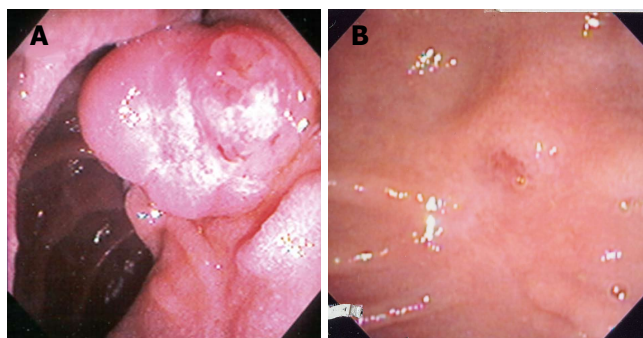


Figure 1 Endoscopic view showing an ampullary tumor (A) and the same area weeks after endoscopic resection of the tumor (B).

forceps) of the area remained negative for tumor cells. A follow-up EUS and two CT scans were also unremarkable.

DISCUSSION

Neuroendocrine neoplasms only rarely occur at the ampulla of Vater, and consist mostly of carcinoids and occasionally of poorly differentiated endocrine carcinomas (small cell carcinomas)^[4]. Up to 26% of patients may have associated Von Recklinghausen disease^[5]. There is a higher incidence (46%) of finding metastatic disease at diagnosis in carcinoids of the ampullary area. Jaundice (53%), pain (24%), pancreatitis (6%) and weight loss (3.6%) are common presenting symptoms^[6] but upper GI bleeding from ulceration is rare^[7]. Carcinoids have an intact overlying mucosa which may explain a high rate of false negative biopsies^[2,6]. Examination using a side-viewing scope and obtaining deep excavating biopsies is essential towards establishing the diagnosis. The possibility of the bile duct involvement should be ascertained with ERCP and/or EUS. In detecting smaller tumors and for local angio-invasion, EUS could be extremely sensitive^[8]. CT scan and MRI have a low sensitivity for the primary lesion^[8]. Somatostatin receptor scan is 86% sensitive for duodenal carcinoids of ≥ 1 cm, in detecting both primary and metastatic disease^[9]. A capsule endoscopy should be considered to rule out synchronous small bowel lesions.

Re-classified by WHO, carcinoids are now called neuroendocrine tumors. Ampullary carcinoids range from highly differentiated (probably benign) to well differentiated endocrine carcinomas and poorly differentiated carcinomas. The overall survival appears to be excellent at 90% for well differentiated tumors^[2], but poor when these are not so well differentiated^[10].

Choosing the best treatment option for ampullary carcinoids could be challenging. Although tumor size is not a good prognostic predictor^[2,10], most experts still recommend either a Whipple or a pylorus preserving pancreatico-duodenectomy (PPPD) for tumors over 2 cm in size^[11]. Metastatic disease has been found in 48% and 40% of the patients with tumor sizes of more than 2 cm and less than 2 cm, respectively^[12]. Thus, some experts recommend a Whipple pancreatico-duodenectomy for all size tumors. On the other hand long-term survival has been achieved with local resection only^[2,13]. In a study of

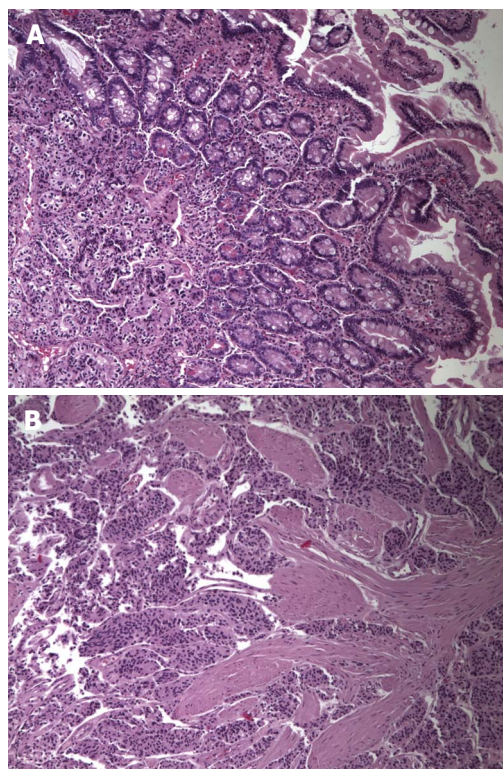


Figure 2 Histological examination of the resected specimen revealing carcinoid in the mucosa and submucosa, composed of discrete solid nests of round tumor cells with central nuclei and occasionally with gland-like lumina (A) and solid nests of tumor cells involving the muscularis propria (B) (HE, x 100).

90 patients with ampullary carcinoids, 52 had a PPPD (majority > 2 cm), while 22 underwent a local resection (majority < 2 cm). Post-operative mortality was 3/52 in the PPPD group compared to 0/22 in the group with local resection^[14]. Therefore, less radical approaches should be considered in highly differentiated, slow growing tumors. In relatively high surgical risk patients with a small, non-metastatic tumor, local excision seems to be a reasonable operative choice, whereas the least invasive approach should be reserved for those who are not surgical candidates. An endoscopic ampullectomy which seems to be an effective treatment in the management of ampullary adenomas, could also be a viable option in selected patients with tumors *in situ* (Tis)^[15] and carcinoids of the ampulla of Vater, without vascular invasion.

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