

# Somatostatinoma of the Ampulla: An Incidental Postoperative Finding Following Colorectal Cancer Resection

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## Abstract

Somatostatinoma of the gastrointestinal tract is a rare finding, especially arising from the ampulla of Vater. We present a man recently diagnosed with rectal adenocarcinoma who was found on imaging to have an ampullary mass. Histology and immunohistochemical staining of the biopsied tissue were consistent with a nonfunctioning somatostatinoma. The patient had neurofibromatosis, which have been reported in patients with ampullary somatostatinomas. Our case highlights the importance of gastrointestinal findings in those with underlying genetic conditions.

## Introduction

Somatostatinomas are rare neuroendocrine tumors that were first described in 1977.<sup>1</sup> The annual incidence of these rare tumors has been estimated at 1 in 40 million people.<sup>2</sup> The majority of somatostatinomas are found in the pancreas, but can occur in other areas of the gastrointestinal tract.<sup>3</sup> Presenting symptoms vary depending on tumor size and its ability to produce somatostatin.

## Case Report

A 59-year-old man with a history of neurofibromatosis and recently diagnosed rectal adenocarcinoma presented after a small bowel mass was found on abdominal imaging. An initial screening colonoscopy 6 months prior to presentation identified a 5-cm rectal mass that biopsies confirmed as invasive adenocarcinoma. A abdominal/pelvic computed tomography (CT) scan at that time showed cholelithiasis, small liver cysts, a mildly dilated pancreatic duct, and the rectal mass. He underwent further staging with a chest CT and pelvic magnetic resonance imaging (MRI) at our institution. Several months later, after completing neoadjuvant chemotherapy and radiation, the patient underwent a low anterior resection with a diverting ileostomy. His final colon pathology revealed stage 3 rectal adenocarcinoma (T3N1c). During his post-operative hospitalization, his course was complicated by nausea and vomiting. Abdominal/pelvic CT scan revealed moderately dilated biliary ducts, a slightly dilated pancreatic duct, and a 2.5-cm lobulated and enhancing mass at the level of the ampulla (Figure 1). Aminotransferases and total bilirubin were within normal limits.

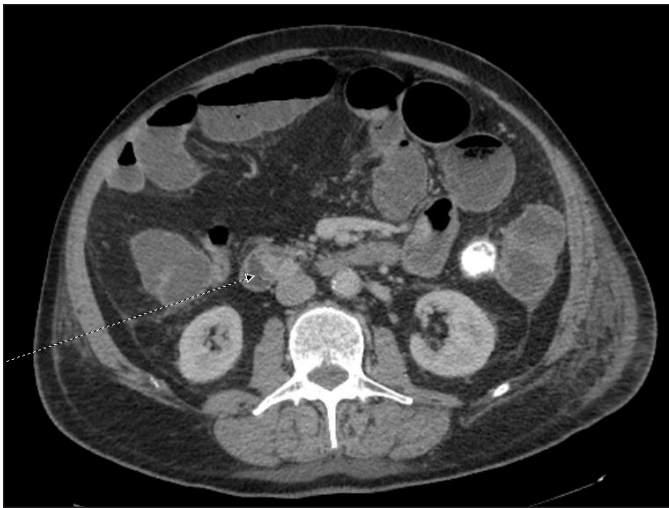
Esophagogastroduodenoscopy (EGD) revealed a large, 2.5-cm mass at the major papilla spreading across at least 1 duodenal fold, and multiple biopsies were obtained with cold forceps (Figure 2). Endoscopic ultrasound (EUS; linear echoendoscope at 7.5 MHz) demonstrated the 2.5-cm, ill-defined mass (Figure 3), as well as a slightly dilated common bile duct (maximum diameter of 10.2 mm) and dilated pancreatic duct (6.5 mm in the head and 6 mm in the body and tail). Fine-needle aspiration (FNA) was performed on the ampullary mass with a 22-gauge needle, with 2 passes under EUS guidance. The biopsied tissue obtained during the EGD demonstrated an acinar architecture with associated psammomatous calcifications and positive staining for chromogranin

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**Figure 1.** Abdominal/pelvic CT demonstrating the ampullary mass (arrow).

and synaptophysin, consistent with a grade 1 ampullary somatostatinoma (Figure 4). Diagnosis of a nonfunctioning somatostatinoma likely originating from the ampulla of Vater was made. The patient is currently on adjuvant chemotherapy for his rectal adenocarcinoma, and has been referred to surgery for consideration of transduodenal ampullectomy or Whipple procedure following completion of chemotherapy.

## Discussion

Fewer than 100 cases of periampullary somatostatinomas have been reported in the literature, with even fewer arising from the ampulla itself, making this a rare endoscopic finding. The largest proportion of these tumors are found in the pancreas (68%), but are also commonly encountered in extrapancreatic locations, including the duodenum (19%), ampulla of Vater (3%), and other areas of the small intestine (3%).<sup>3,4</sup> The masses can be functioning or nonfunctioning regarding secretion of somatostatin. Function-



**Figure 2.** Endoscopic image of ampullary mass.

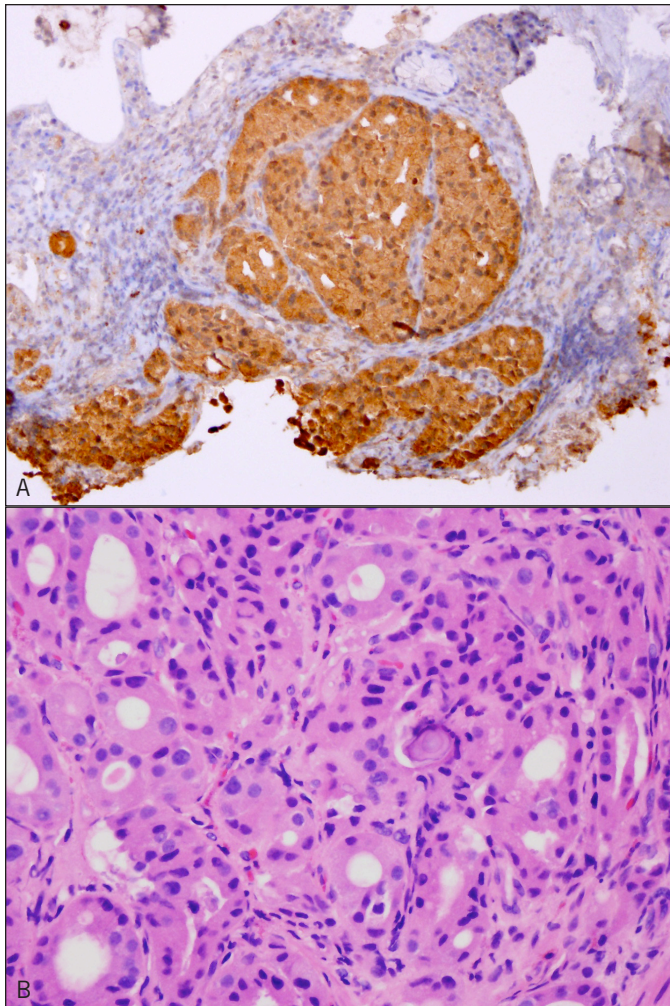


**Figure 3.** EUS-guided FNA of the ampullary mass.

ing tumors are typically found in the pancreas, and can lead to somatostatin syndrome characterized by diabetes mellitus, cholelithiasis, and steatorrhea. An extrapancreatic somatostatinoma can be functioning as well, but this is dependant on size, with such somatostatinomas typically being larger than 4 cm in size.<sup>3</sup> These tumors have been associated with neurofibromatosis type 1, as well as multiple endocrine neoplasia type 1.<sup>5,6</sup> Duodenal and ampullary somatostatinomas have been associated with neurofibromatosis type 1 and are typically metastatic when found.<sup>4,6</sup> Since duodenal somatostatinomas are usually not functional, they are typically diagnosed on EGD with biopsy or EUS with FNA after the patient develops obstructive symptoms of the gut or biliary tract.<sup>7</sup>

A characteristic finding of somatostatinoma on histological examination is the appearance of psammoma bodies due to calcium salt deposition, which appear in the somatostatinomas of extrapancreatic derivation, particularly those that are ampullary in origin.<sup>4,5,8,9</sup> Immunohistochemical staining is also used for diagnostic purposes, and the majority of somatostatinomas will have positive staining with chromogranin A, synaptophysin, and somatostatin.<sup>3,10</sup>

The mainstay of treatment for these lesions is similar to other neuroendocrine tumors of the small intestine or pancreas, with resection being the treatment of choice. It has been suggested that tumors smaller than 1 cm can be considered for endoscopic resection, but tumors larger than this typically require surgical excision via transduodenal ampullectomy or Whipple procedure.<sup>3</sup> Radiation and chemotherapy are options for this patient population, but systemic chemotherapy is only weakly active against metastatic neuroendocrine tumors. Due to the rarity of the disease, long-term studies demonstrating efficacy of these treatment modalities is lacking.<sup>3,11</sup>



**Figure 4.** Biopsied tissue during EGD demonstrating neoplastic cells with (A) cytoplasmic staining with anti-somatostatin antibody (x200 magnification), and (B) neoplasm with pseudoglandular architecture composed of cells with abundant granular eosinophilic cytoplasm. Intraluminal calcification with concentric laminations represents a psammoma body (H&E staining, x400 magnification).

## Disclosures

Author contributions: Both authors wrote the manuscript. JB French is the article guarantor.

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## References

- Harris GJ, Tio F, Cruz AB. Somatostatinoma: A case report and review of the literature. *J Surg Oncol.* 1987;36(1):8–16.
- Colović RB, Matic SV, Micev MT, et al. Two synchronous somatostatinomas of the duodenum and pancreatic head in one patient. *World J Gastroenterol.* 2009;15(46):5859–63.
- Kim JA, Choi WH, Kim CN, et al. Duodenal somatostatinoma: A case report and review. *Korean J Intern Med.* 2011;26(1):103–7.
- House MG, Yeo CJ, Schulick RD. Periampullary pancreatic somatostatinoma. *Ann Surg Oncol.* 2002;9(9):869–74.
- Garbrecht N, Anlauf M, Schmitt A, et al. Somatostatin-producing neuroendocrine tumors of the duodenum and pancreas: Incidence, types, biological behavior, association with inherited syndromes, and functional activity. *Endocr Relat Cancer.* 2008;15(1):229–41.
- Relles D, Baek J, Witkiewicz A, Yeo CJ. Periampullary and duodenal neoplasms in neurofibromatosis type 1: Two cases and an updated 20-year review of the literature yielding 76 cases. *J Gastrointest Surg.* 2010;14(6):1052–61.
- Kimura R, Hayashi Y, Takeuchi T. Large duodenal somatostatinoma in the third portion associated with severe glucose intolerance. *Intern Med.* 2004;43(8):704–7.
- Mahajan SK, Mahajan LA, Malangoni MA, Jain S. Somatostatinoma of the ampulla of Vater. *Gastrointest Endosc.* 1996;44(5):612–4.
- Mao C, Shah A, Hanson DJ, Howard JM. Von Recklinghausen's disease associated with duodenal somatostatinoma: Contrast of duodenal versus pancreatic somatostatinomas. *J Surg Oncol.* 1995;59(1):67–73.
- Burke AP, Federspiel BH, Sobin LH, et al. Carcinoids of the duodenum: A histologic and immunohistochemical study of 65 tumors. *Am J Surg Pathol.* 1989;13(10):828–37.
- Koc O, Duzkoylu Y, Sari YS, et al. Duodenal somatostatinoma: A case report and review of the literature. *J Med Case Rep.* 2013;7:115.

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