Endoscopic Papillectomy for Gangliocytic Paraganglioma

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Introduction

- **Neuroendocrine tumors (NET)** most commonly occur in the small bowel; however, NETs at the ampulla of Vater are rare and more aggressive, irrespective of size.
- **Gangliocytic paragangliomas (GP)** are a more benign type of NET that must be properly differentiated from other similar appearing NETs, which may carry a worse prognosis.

Case Description

77-year-old man presented with ongoing right flank pain.

Labs - AST 17 U/L, ALT 20 U/L, Alkaline Phosphatase 84 U/L, Total Bilirubin 0.3 mg/dL

Computed Tomography - duodenal mass in the periampullary region measuring 11 x 8 mm, with dilation of common bile duct at 10 mm

Clinical Course

- Esophagogastroduodenoscopy (EGD): subepithelial lesion at the ampulla
- Endoscopic ultrasound (EUS): 15 x 12 mm hypoechoic subepithelial lesion arising from the submucosa (Figure 1.)
- A fine needle biopsy was performed and immunohistochemical staining was positive for synaptophysin, chromogranin and pankeratin, consistent with neoplasm with neuroendocrine differentiation (Figure 1.)
- Subsequent DOTATATE PET scan showed uptake in the ampullary region with no uptake anywhere else.
- The case was discussed in multidisciplinary conference, and the decision was made to proceed with endoscopic papillectomy given concerns for NET.
- An endoscopic retrograde cholangiopancreatography (ERCP) with endoscopic papillectomy was performed (Figure 2.), and a biliary and pancreatic duct stent was placed.
- The pathology was consistent with gangliocytic paraganglioma (Figure 2.), and the patient had no adverse events with removal of stents after four weeks.

Endoscopic Ultrasound

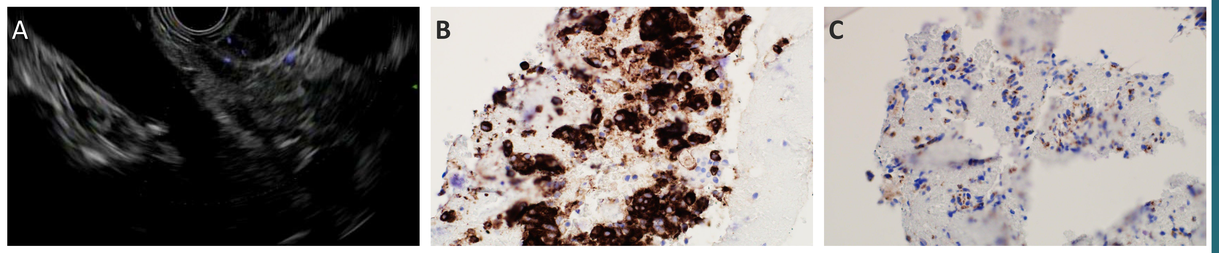

Figure 1. Hypoechoic subepithelial lesion arising from the submucosa, measuring 15 x 12 mm (A), with fine needle biopsy staining synaptophysin (B) and chromogranin (C).

Endoscopic Retrograde Cholangiopancreatography

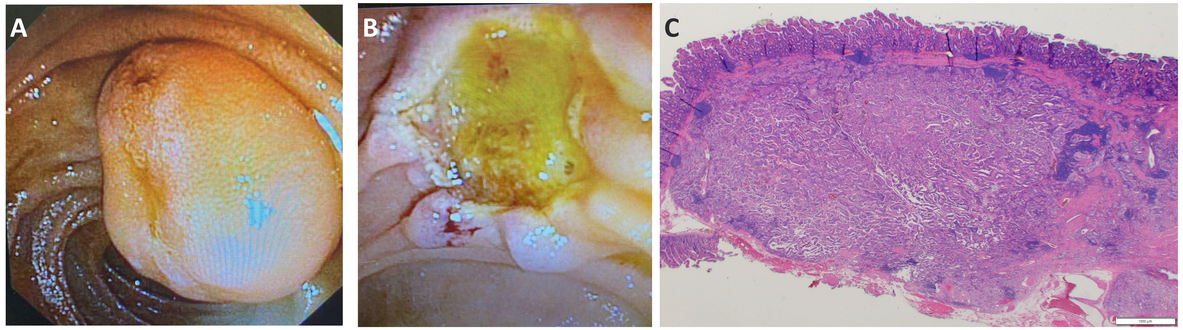

Figure 2. Ampullary lesion prior to (A) and after endoscopic papillectomy (B), with histopathology consistent with gangliocytic paraganglioma (C).

Discussion

Ampullary Neuroendocrine Tumors (NET)

- Etiology - enterochromaffin cells of crypts of Lieberkuhn
  - Usually appear as small submucosal masses and intact duodenal mucosa; consequently, deeper biopsies guided by EUS are required
  - Unlike other NETs, ampullary NETs do not express gastrin.
  - Carcinoid syndrome typically is absent since tumors are nonfunctional
- Presentation - jaundice and abdominal pain, since tumor may develop at the conjugation of the pancreatic and biliary ducts
- Histopathology - expression of chromogranin A, neuron specific enolase and synaptophysin.
  - Chromogranin A levels reflect tumor load and can be a used for monitoring treatment response and detect relapse.

Gangliocytic Paragangliomas (GP)

- Etiology - generally regarded as a benign type of neuroendocrine tumor
  - Typically arise in the second part of the duodenum
- Presentation - gastrointestinal bleeding and abdominal pain.
- Histopathology - varies significantly but typically embody epithelioid, spindle-shaped, and ganglion-like cell types
  - While GPs are predominantly benign, rare reports of spread to regional lymph nodes are present.

In this case, endoscopic papillectomy was safe and effective for the diagnostic and therapeutic management of Endoscopic Papillectomy for Gangliocytic Paraganglioma.

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