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## Learning Objectives

- Gastric neuroendocrine tumors (gNETs) are rare malignancies which arise from enterochromaffin-like cell (ECL) precursors within the gastric mucosa.
- There are 4 classifications of gNETs primarily based on size, number of lesions, serum gastrin level, tissue invasion, proliferation index and immunohistochemistry.
- Applying a combination of factors, gNETs are typically classified into one of these four categories with relative ease.
- The following case highlights an exceedingly rare hybrid presentation of gNET containing features of all four classification subtypes.

## Patient Presentation

A 78-year-old female with a past medical history of hypertension, diabetes mellitus, chronic kidney disease and gastroesophageal reflux disease (GERD) initially presented with worsening symptoms of GERD. The patient denied any alarm symptoms but did endorse persistence of reflux symptoms despite proton pump inhibitor use.

### Physical Exam:

Vitals: T 98.8F, BP 126/70, HR 70, RR 16, 96% on RA

General: AAOx3, NAD

Skin: Warm, dry, no jaundice

Cardio: RRR, Normal S1/S2

Respiratory: CTAB

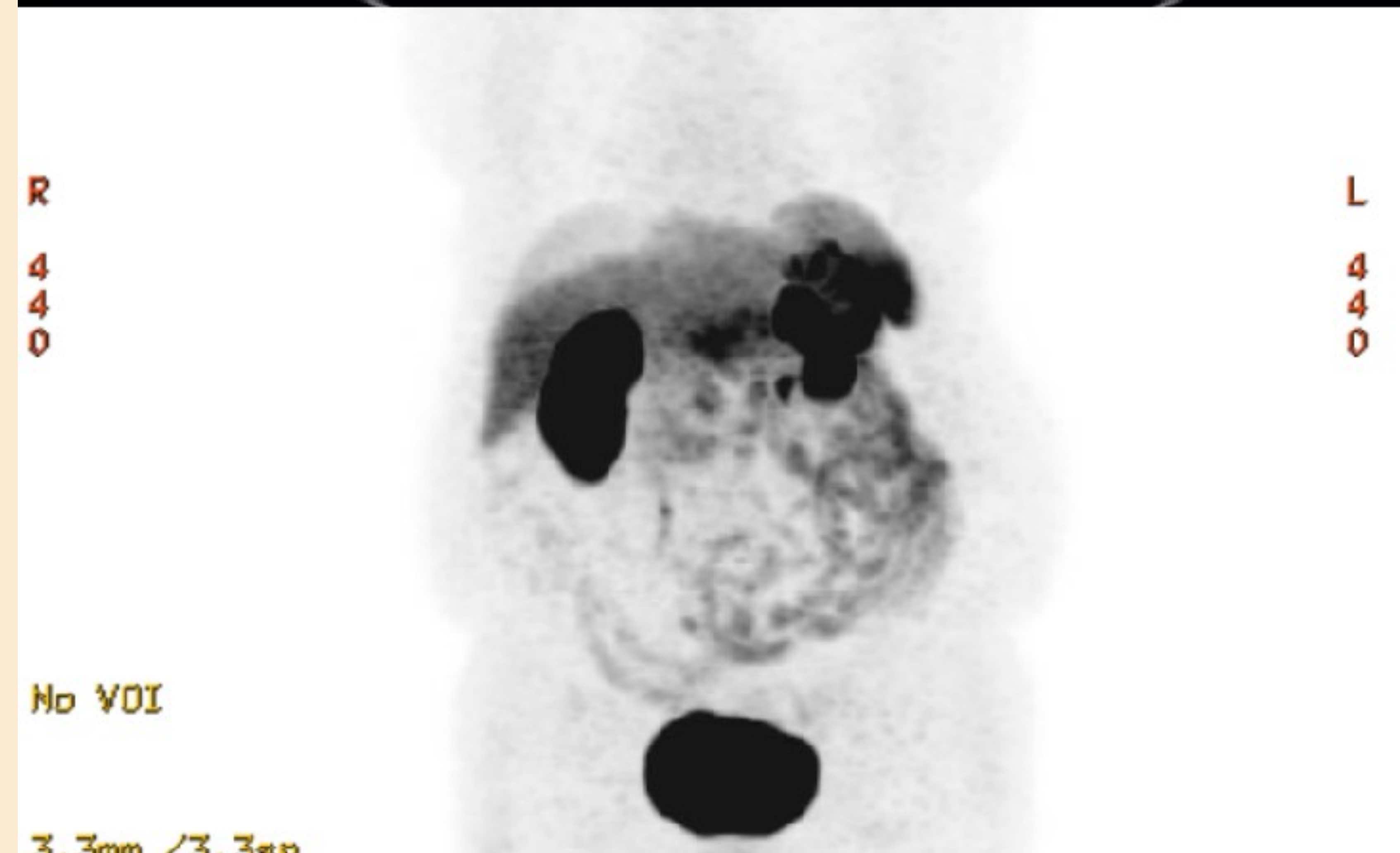
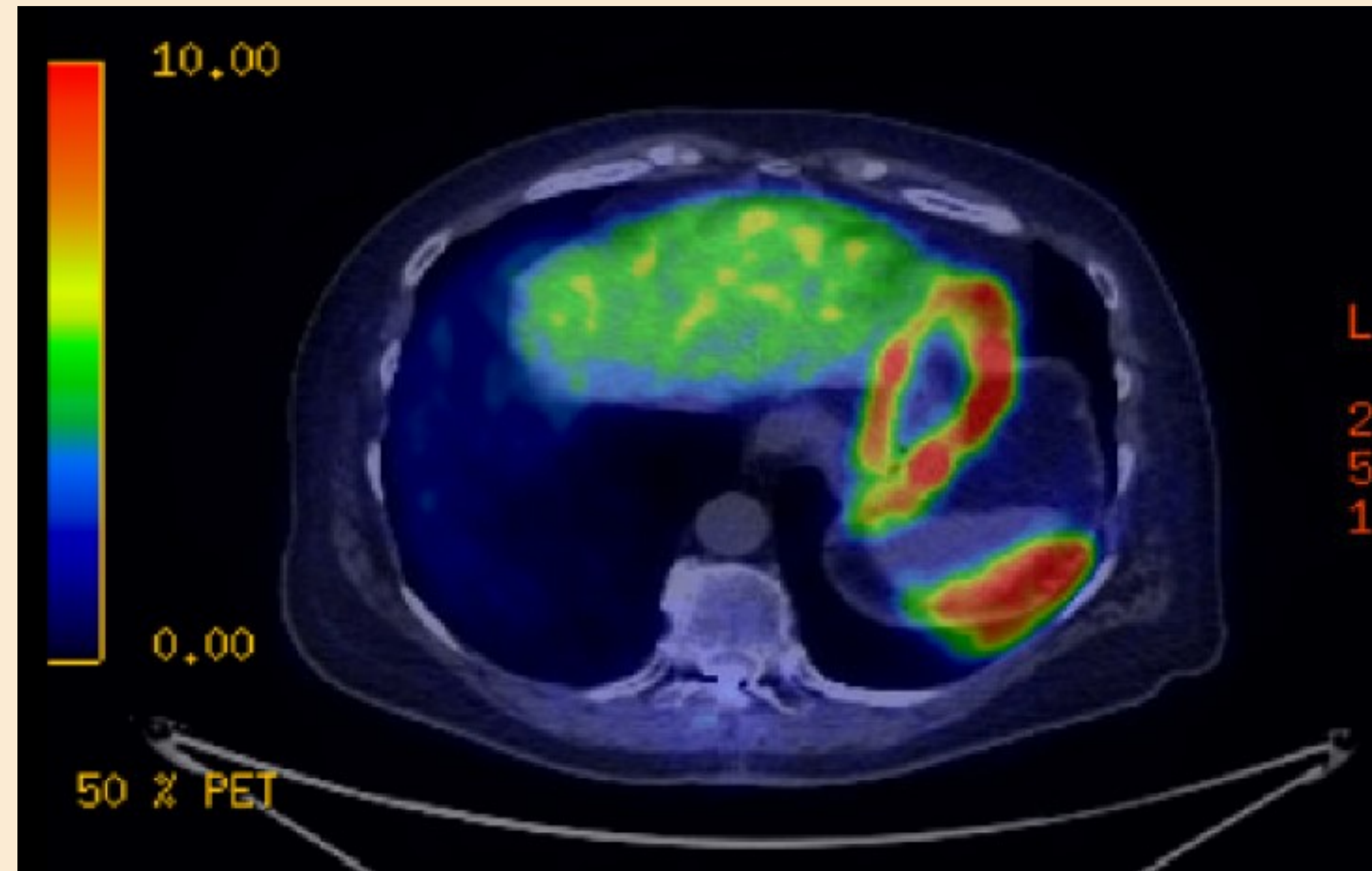
GI: Soft, non tender, no guarding or rebound tenderness

MSK: Normal range of motion, all compartments compressible

## Lab Values

134	102	30	<del>13.4</del>	ALT/AST: 20/22
4.5	28	1.6	<del>8.0 184</del>	Tbili: 0.7
			42.0	Dbili: 0.3
				ALP 80

## DOTA-TATE PET CT



DOTA-TATE PET/CT scan showing heterogenous uptake in the gastric body concerning for focal gNET.

## Clinical Course

- Screening EGD with EUS showed polyps which were removed with mucosal resection. Initial pathology illustrated well differentiated gNET.
- She underwent a DOTA-TATE PET/CT scan which showed heterogenous uptake involving the gastric body reflecting focal gNET without evidence of metastatic disease.
- The patient underwent total gastrectomy and esophagojejunostomy with upper GI series revealing no evidence of anastomotic leak.
- On subsequent pathology, at least 46 semi pedunculated gastric polyps were identified of which the greatest tumor dimension was 0.6 cm with a mitotic rate less than 2 mitoses/2mm<sup>2</sup>.
- Pathology and immunohistochemistry revealed well differentiated gNET, grade 1 and 2, staining positivity for Chromogranin A (CgA), synaptophysin, CD56, and CAM 5.2 with a Ki67 proliferation index > 6%.
- Patient's serum gastrin was within normal limits at 51 pg/ml and 24-hour urine 5-HIAA revealed a normal level of 5.7 mg/24hr.
- She was referred to a geneticist for massive parallel sequencing also known as next generation sequencing (NGS).

## Take Home Points

- This case displays a rare, hybrid presentation of gNET with features of all four classification subtypes.
- The positive synaptophysin, normal serum gastrin and increased Ki67 index are characteristic of gNET Type 3 and 4 which carry a poor prognosis.
- Type 3 and 4 however, are typically large (>2cm), single tumors. This patient had many, smaller tumors with positive CgA staining commonly seen in Type 1 and 2.
- Hybrid gNETs are extremely uncommon neoplasms which create both a diagnostic and therapeutic challenge requiring a truly multidisciplinary effort.

## References

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