

# Extensive Hepatic Lesions from a Primary Duodenal Neuroendocrine Mass in a Young Male

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## BACKGROUND

Neuroendocrine neoplasms (NEMs) are rare, accounting for 0.5% of newly diagnosed malignancies. NEMs can be found throughout the body with two-thirds arising in the GI tract and incidence increasing among duodenal NEMs. These malignancies have a female predominance and generally occur in the 6<sup>th</sup> decade of life.

## CHIEF COMPLAINT

A previously healthy 29 y/o man presented to the emergency department following acute onset right upper quadrant abdominal pain.

## CASE PRESENTATION

- In the ED, patient was hemodynamically stable and satiating >95% on room air. Physical exam was notable for tenderness to palpation in the right upper quadrant with no overlying skin changes. Initial labs (Figure A) notable for minor leukocytosis, anemia, and LFT derangements (AST 55 U/L, ALT 173 U/L, ALP 127 U/L, and TBIL 0.8mg/dL).
- Stat CT A/P showed multiple well-circumscribed liver lesions, largest measuring 8.6 x 7.8 cm, with portal venous invasion and duodenal thickening (Figure B & C).
- Stat CT Chest was negative for thoracic metastatic disease.
- Patient was admitted to inpatient medicine for further work-up.

## LABS, IMAGING, WORK-UP

### ADMISSION LABS

WBC	12.10
HGB	11.7
HCT	38
PLT	280
NA	135
K	4.7
CL	99
CO2	25
GLU	123
BUN	16
CR	1.11
AST	55
ALT	173
ALK PHOS	127
BLIRUBIN (TOTAL)	0.8
ALBUMIN	4.5

Figure A. Labs at time of ED presentation with minor leukocytosis, anemia, and LFT derangements

### IMAGING

#### CT Abdomen/Pelvis w/Contrast

#### IMPRESSION

1. Multiple large heterogeneously enhancing liver lesions are highly concerning for metastatic malignancy of unknown primary. The patient's right upper quadrant pain is likely explained by distortion/stretching of Glisson's capsule.
2. Free fluid layering in the pelvis and about the liver/spleen measures up to 50HU and therefore likely represents proteinaceous fluid or blood product.
3. Circular filling defect within the SMV just peripheral to the portal confluence could represent non-occlusive thrombus or mixing artifact.

#### CT Chest w/Contrast

#### IMPRESSION

No evidence of thoracic metastatic disease.

Figure C. Imaging completed in evaluation of abdominal pain

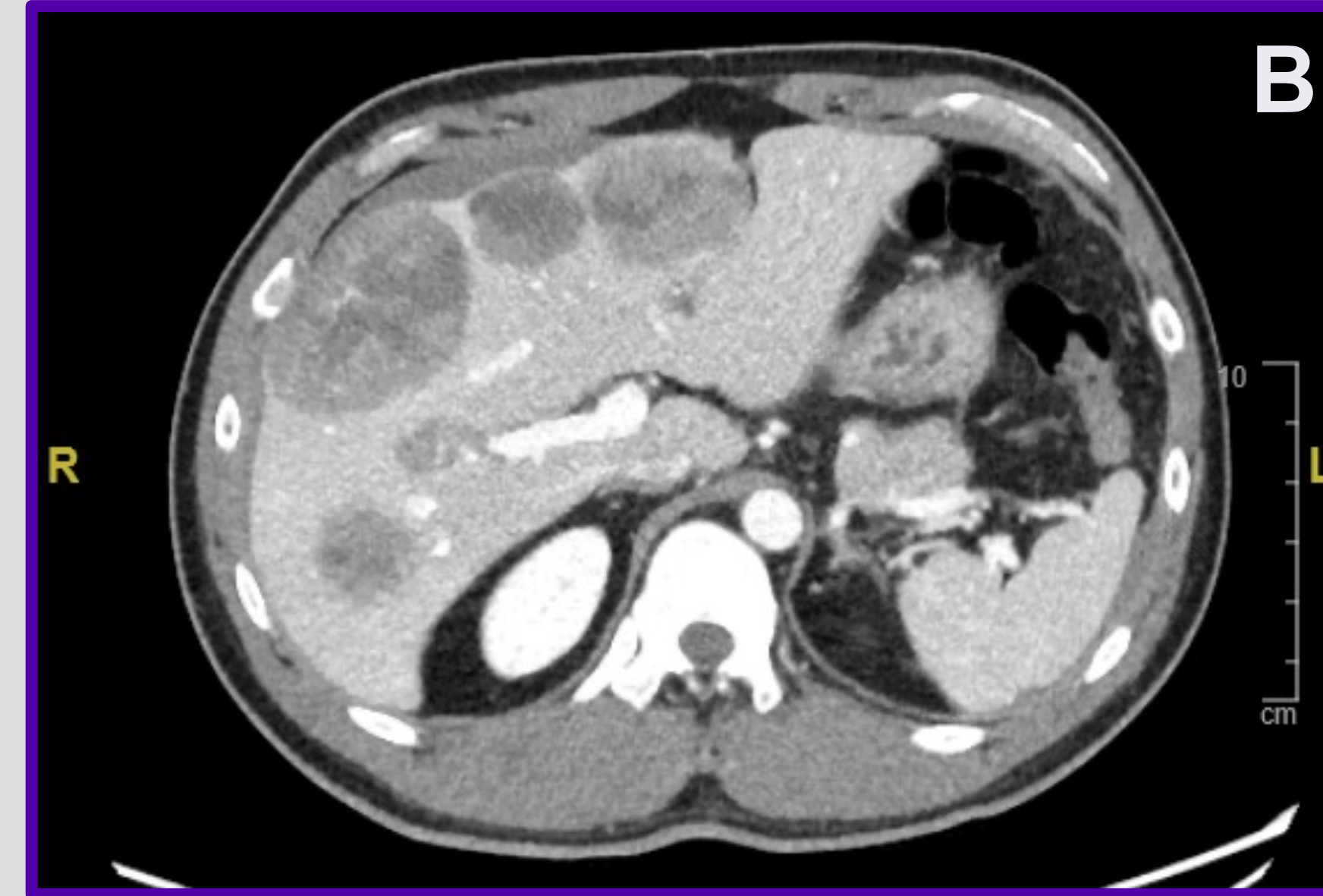


Figure B. CT A/P with multiple large enhancing liver lesions concerning for metastatic malignancy of unknown primary

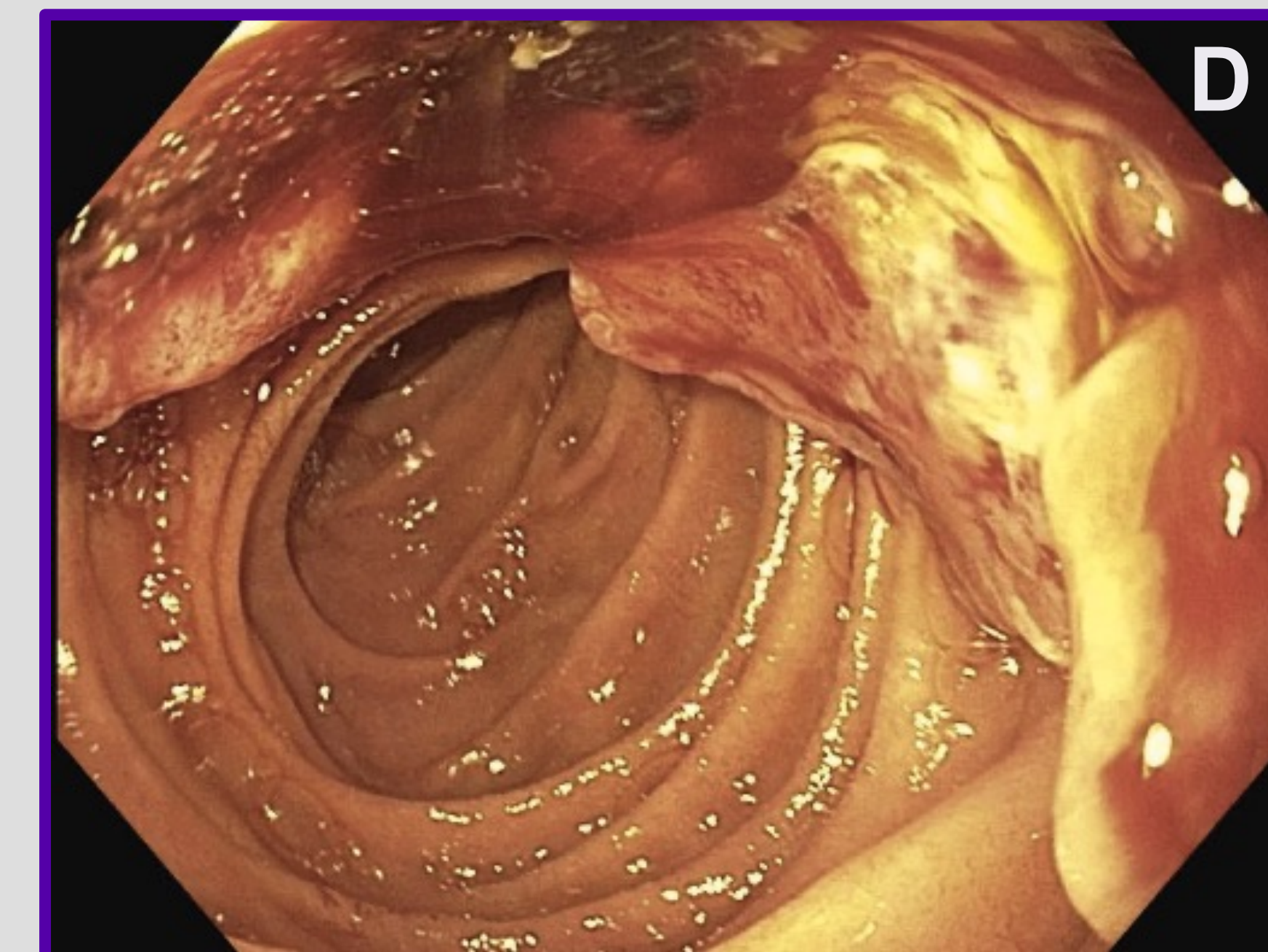


Figure D. A large fungating mass found in the second portion of the duodenum

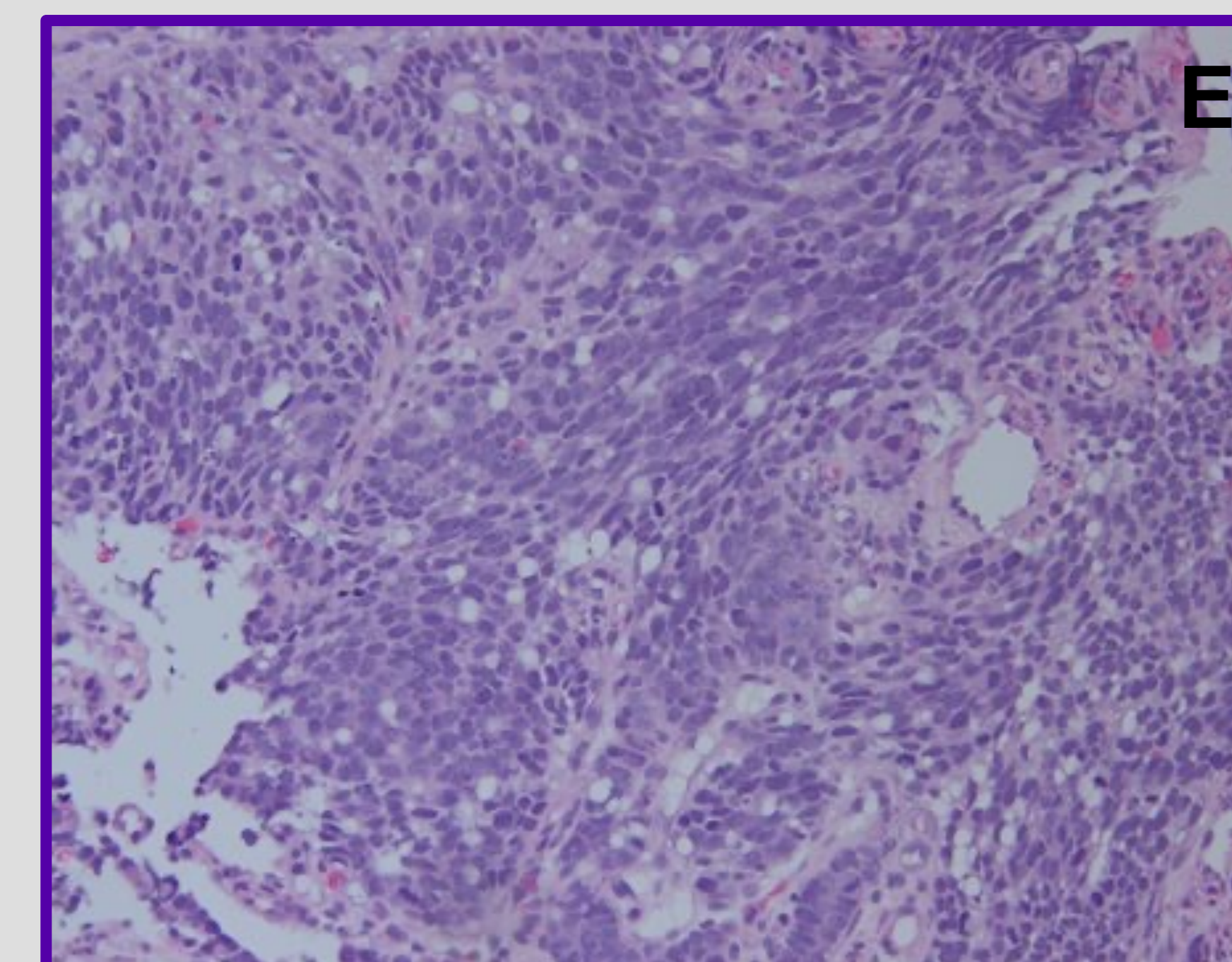


Figure E: Duodenal Mass, H&E stain, 20x, High-grade poorly differentiated epithelial malignancy composed of nested cells with indistinct nucleoli, moderate eosinophilic cytoplasm, and brisk mitosis

## CASE RESOLUTION & DISCUSSION

- Evolution of case:
  - A laboratory evaluation was pursued. CA 19-9 and CEA were negative. There was elevation of AFP 1,577 ng/mL and LDH 817 U/L, but negative hCG. A1AT, ANA, AMA, ASMA, ALKM, and ceruloplasmin were all negative.
  - Liver biopsy showed high grade dysplasia with atypia of possible enteric origin.
  - An EGD was performed that revealed a fungating mass in the second portion of the duodenum (Figure D). Pathology favored a high-grade poorly-differentiated neuroendocrine carcinoma (Figure E).
  - Patient was discharged with initiation of palliative cisplatin/irinotecan.
- Gastrointestinal NEMs have been increasing in prevalence due to improved awareness. When carcinoid symptoms are absent, detection is dependent on imaging and endoscopy after the patient presents with complications from metastasis or mass effect.
- Following biopsy, GI NEMs are classified by WHO 2010 criteria into 3 groups based on mitotic count and Ki-67 index. Group 3 tumors are classified as poorly-differentiated neuroendocrine carcinomas with Ki-67 >20% with higher rates of angioinvasion and metastatic potential. Lymph node metastasis occurs in 60% of duodenal NETs, while liver mets occur in <10%. In our case, the patient's Ki-67% index was >90%, characterizing it as a Group 3 NEM.
- Characterization is important given the impact on prognosis and treatment. Group 3 NEMs have the overall worst prognosis. Since >90% of duodenal NEMs do not cause a clinical syndrome and are not associated with metastatic disease, surgical or endoscopic resection is preferred. However, if there is metastatic disease or high-grade histologic features, chemotherapy is the mainstay of treatment.

## CONCLUSION

While overall rare, neuroendocrine neoplasms, especially in the GI tract, have been increasing in prevalence. Diagnosis with the help of imaging and endoscopy is essential to ensure proper histologic characterization and appropriate treatment initiation.

## REFERENCES

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