

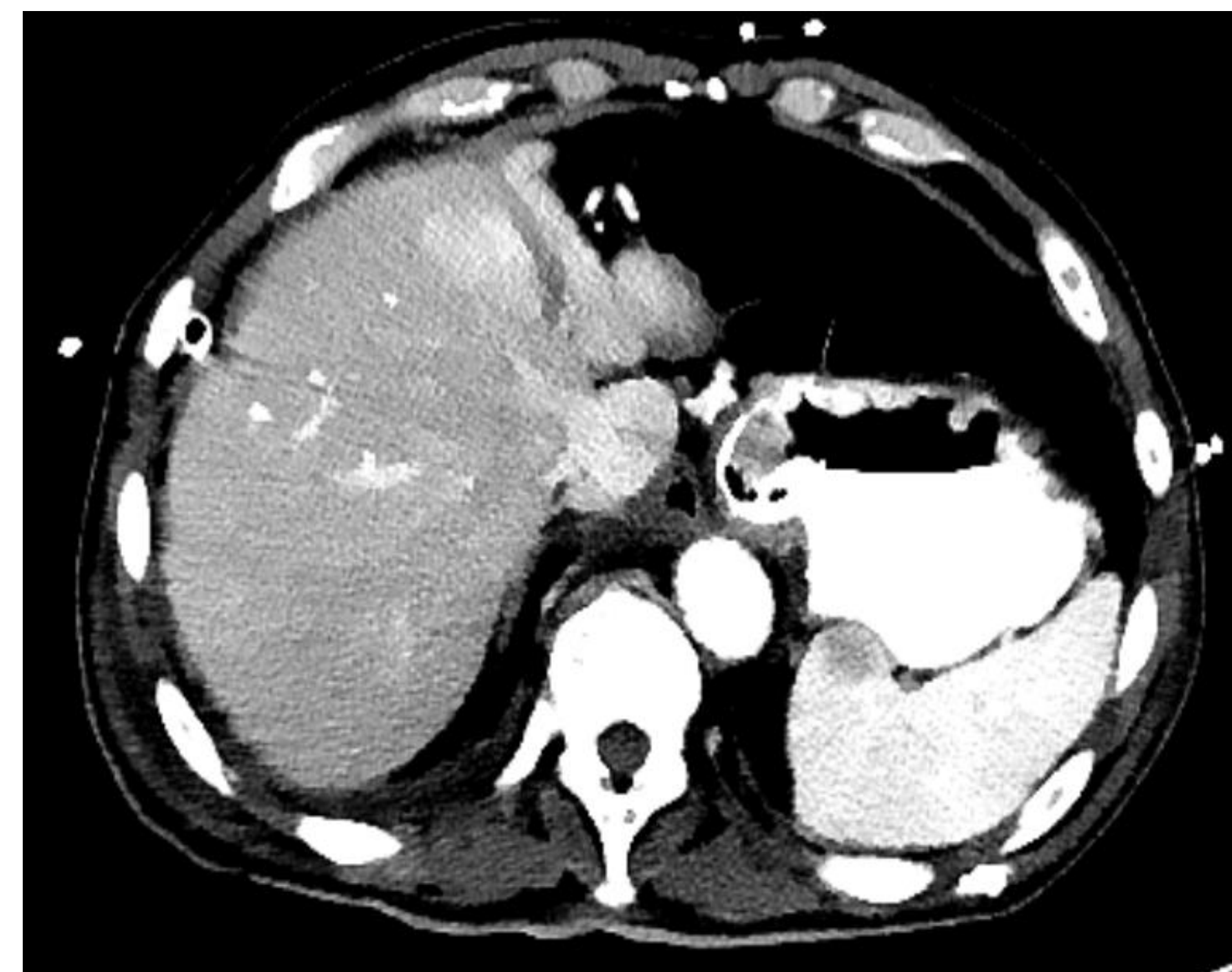
INTRODUCTION

- Zollinger Ellison syndrome (ZES) is characterized by gastric hyperacidity, resulting from increased gastrin production.
- This leads to symptoms such as abdominal pain, heartburn, and diarrhea.
- Given the overlap in the symptoms with idiopathic Peptic Ulcer Disease (PUD) and Gastro-Esophageal Reflux Disease (GERD), the diagnosis can be delayed and complications such as gastric obstruction, hemorrhage, or even perforation can be the initial presentation.
- Here we present an unusual case of esophageal perforation that was ultimately diagnosed with ZES.

CASE PRESENTATION

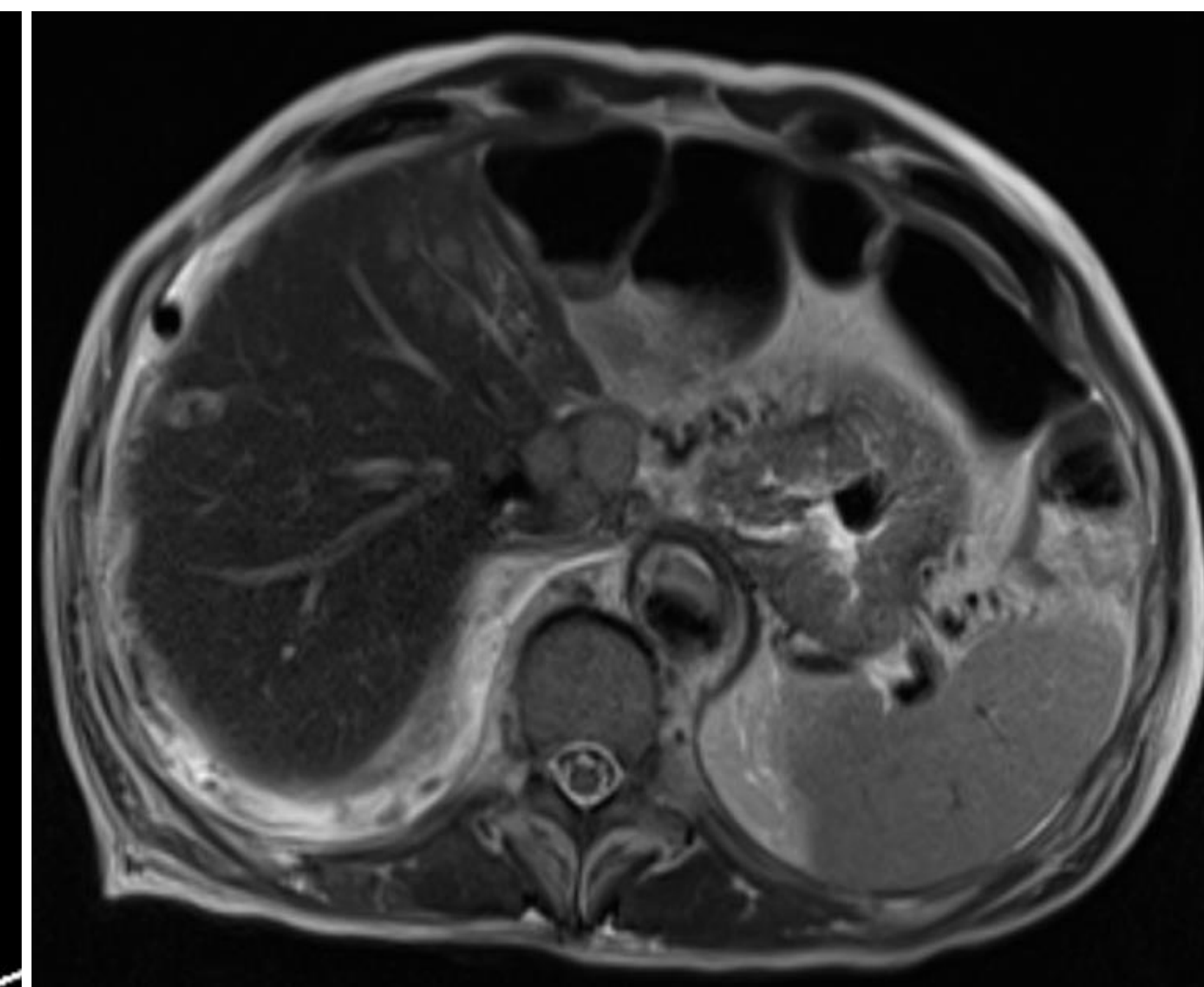
- A 65-year-old male with a history of perforated gastric ulcer s/p Graham patch repair two months prior presented with worsening abdominal pain, vomiting, and inability to swallow secretions.
- He had a positive H. pylori stool antigen test during the prior hospitalization s/p course of eradication therapy.
- CT chest showed pneumomediastinum and an esophageal perforation.
- An upper gastrointestinal endoscopy, done to place an esophageal stent, revealed multiple ulcers in the first portion of the duodenum.
- Robotic-assisted drainage of the posterior mediastinum was then pursued.
- Abdominal imaging (CT/MRI) demonstrated enhancing masses in multiple liver segments as well as periportal adenopathy (See CT and MRI abdomen).
- FDG PET/CT showed increased uptake by the left liver lesion and multiple porta hepatis adenopathy, concerning for metastatic disease.
- The primary tumor could not be clearly delineated.
- A percutaneous liver biopsy revealed a metastatic, grade 1 well-differentiated neuroendocrine tumor (see histopathology).
- Serum gastrin levels were elevated to 433 pg/mL.
- Serum ionized calcium and parathyroid hormone were within normal limits.
- Family history was negative for multiple endocrine neoplasia 1 (MEN1).
- A diagnosis of metastatic, sporadic ZES with an unknown primary site was made.
- The patient was discharged on high-dose lansoprazole and somatostatin analog with outpatient follow-up for DOTATATE scan and definitive treatment.

CT ABDOMEN



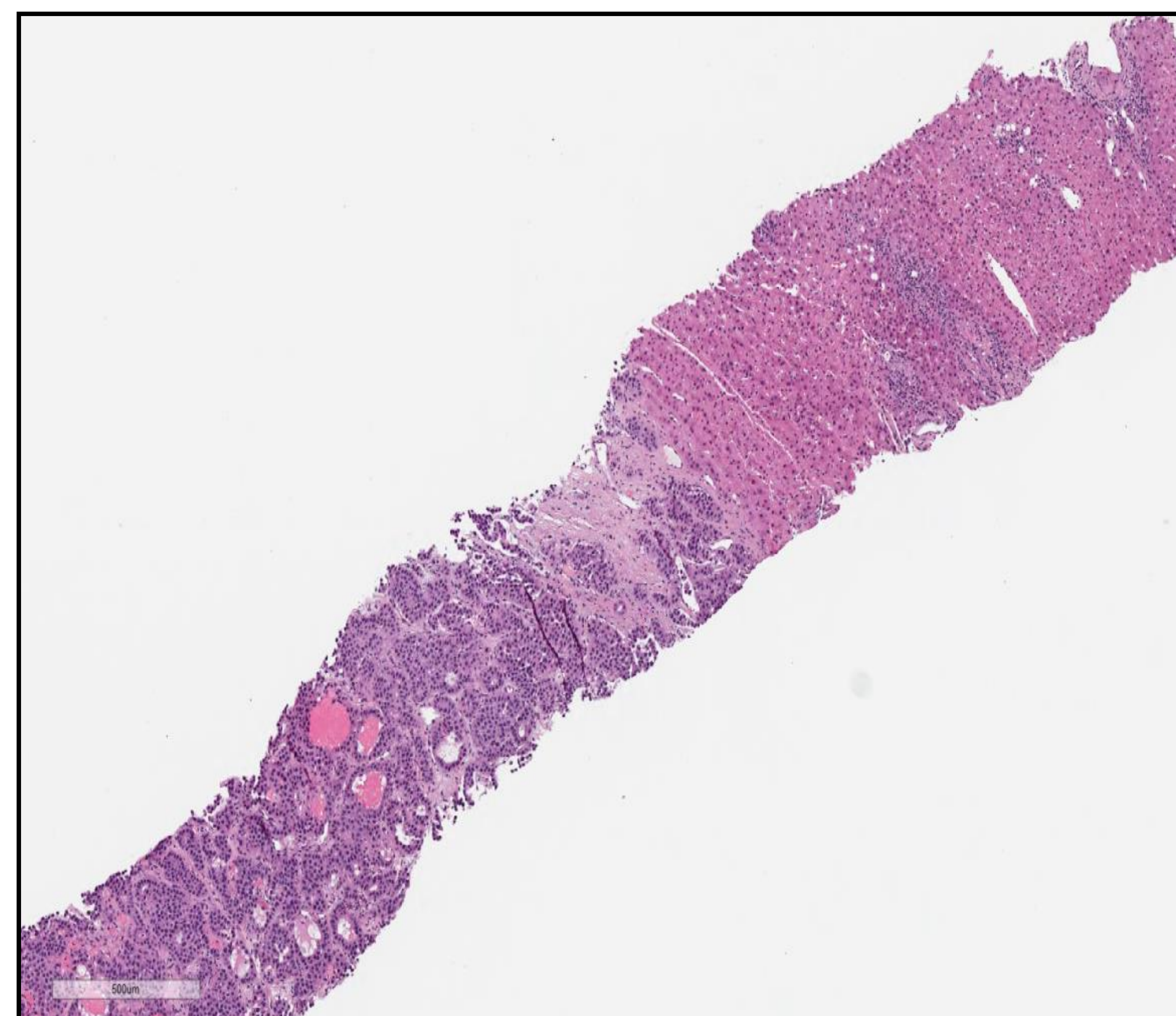
CT abdomen showing 3.5 cm mass inseparable from segment II of the liver

MRI ABDOMEN

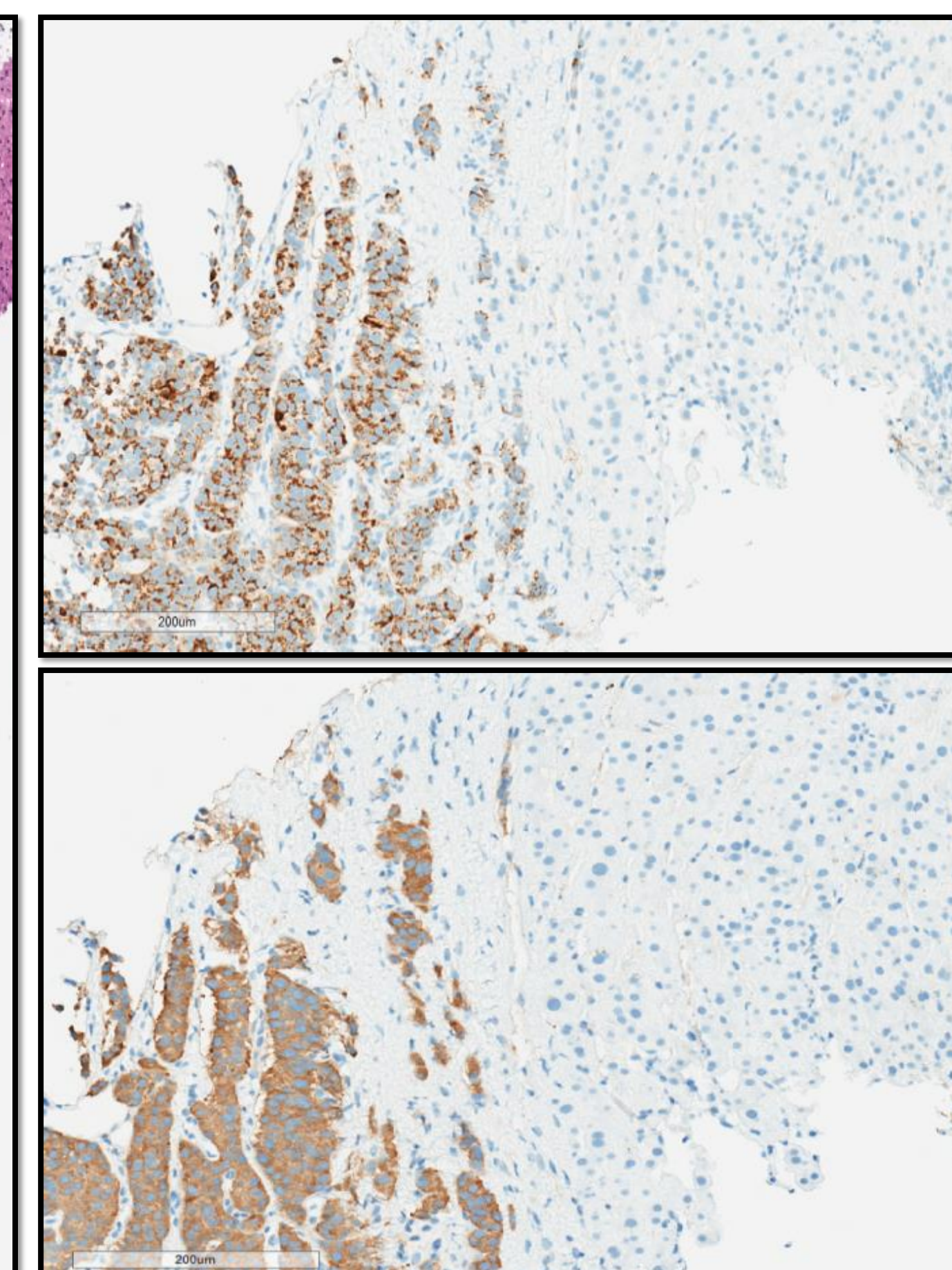


MRI abdomen showing multiple enhancing masses in segments I, II, and IVA with subtle washout and associated periportal adenopathy most compatible with metastatic process

HISTOPATHOLOGY



This image of a liver core needle biopsy shows the transition from liver parenchyma (upper right) to the tumor (lower left). The nested pattern of the tumor is suggestive of possible neuroendocrine origin



Chromogranin and synaptophysin immunohistochemical stains positively highlight the tumor cells (brown) and do not stain the liver parenchyma, confirming neuroendocrine tumor

DISCUSSION

- Early diagnosis of ZES is crucial for several reasons.
- Firstly, about 60% to 90% of gastrinomas are malignant with metastasis to the lymph nodes, liver, or distant organs. Metastasis to the liver has a direct effect on overall survival with 15% 10-year survival in the setting of metastases but those without liver metastases have a 95% 20-year survival¹.
- Delayed diagnosis can result in complications from gastroesophageal reflux and PUD.
- PUD complications including bleeding, perforation, penetration occur in 10-15% of the patients with ZES².
- Peptic ulcer perforation is seen in 4-6 % of the patients with ZES².
- Most perforations involved the duodenum with one report of jejunal involvement and no reports of gastric perforation to date.
- Esophageal symptoms can remain masked in ZES patients due to other, more compelling clinical problems.
- In the recent years, there is evidence of an increase in esophageal involvement, with several cases of esophagitis and Barrett's esophagus
- To our knowledge, two cases of esophageal perforation were documented in the literature. Of the reported cases, several were secondary to emesis with one being spontaneous³.
- This case is unique in that our patient developed complications rarely described in literature i.e. gastric perforation and esophageal perforation.

REFERENCES



CONTACT INFORMATION

Mohammed Rifat Shaik MD

shaikrifat13@gmail.com