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Clinical vignette: VIPoma as a cause of persistent diarrhea

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A VIP in the ICU

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Learning Objectives

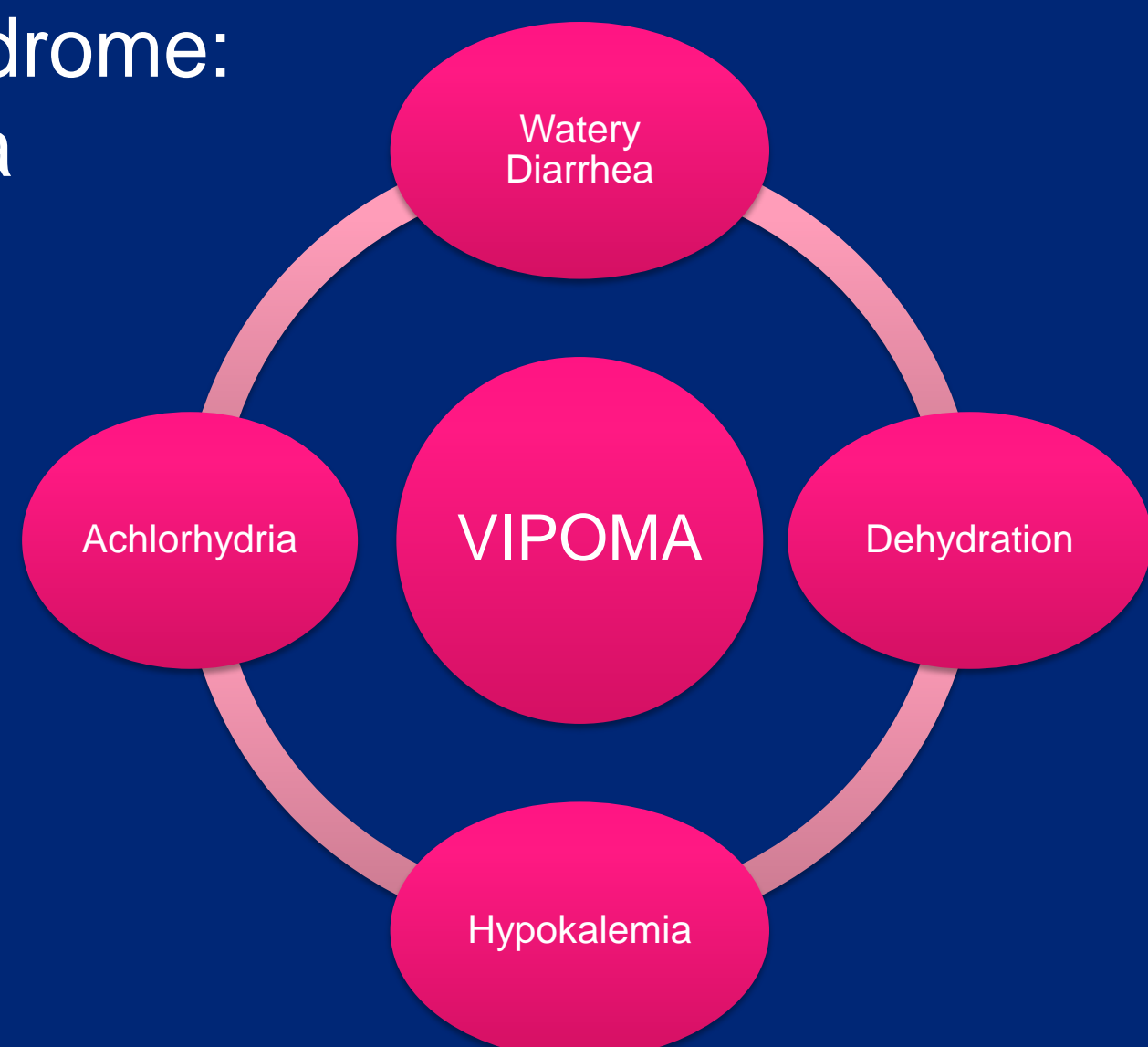
- Recognize the signs and symptoms of a patient presenting with a VIPoma
- Understand additional etiologies of diarrhea in an inpatient setting
- Review pathology involved in diagnosis of a neuroendocrine tumor

Patient Presentation

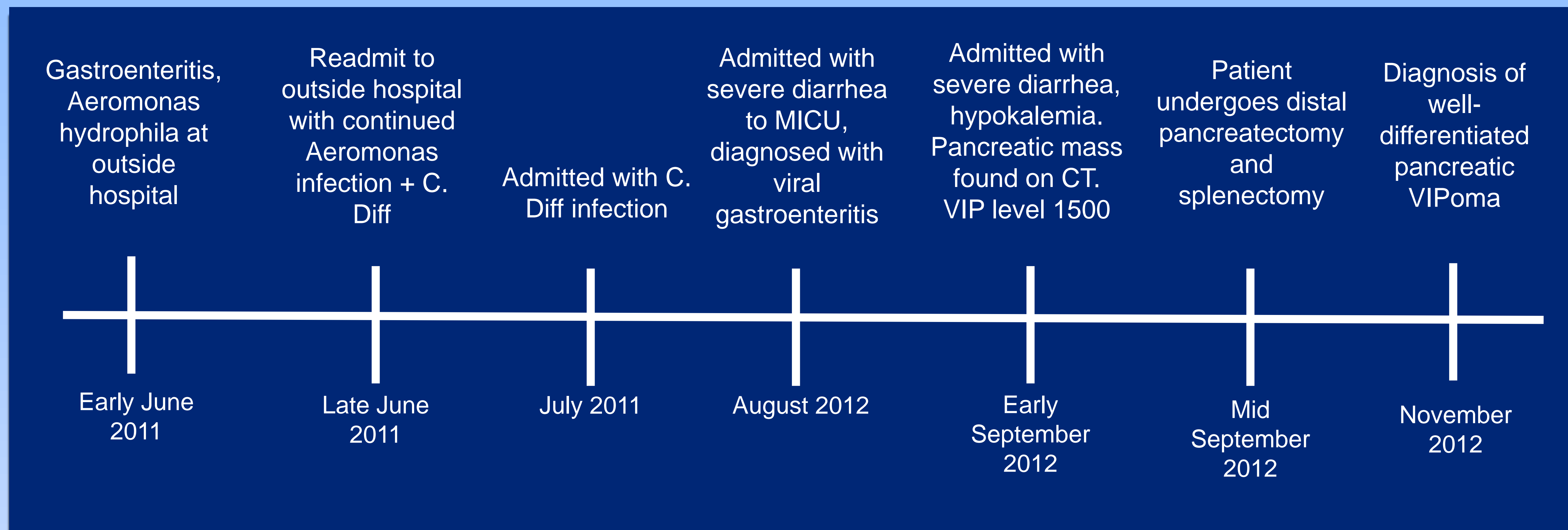
46 year old male who was initially admitted to the ICU with severe hypokalemia (1.5) secondary to profound diarrhea. The patient had previously been admitted one year prior for gastroenteritis and an episode of C. diff colitis with similar presentation (see timeline above). On the current presentation, the patient had had one week of non-bloody, large volume diarrhea, weakness, and vomiting. On physical exam had only hyperactive bowel sounds and mild right lower quadrant pain without any positive signs for appendicitis. The remainder of the exam was normal. His diarrhea persisted over several days of the hospitalization leading the team to look for other etiologies of his severe diarrhea.

Introduction

- VIPoma is a rare (1 in 10 million) endocrine tumor in non-beta islet cells of the pancreas that produces an unregulated amount of vasoactive intestinal peptide
- VIPomas typically present as the WDHA syndrome:
 - Watery Diarrhea
 - Dehydration
 - Hypokalemia
 - Achlorhydria



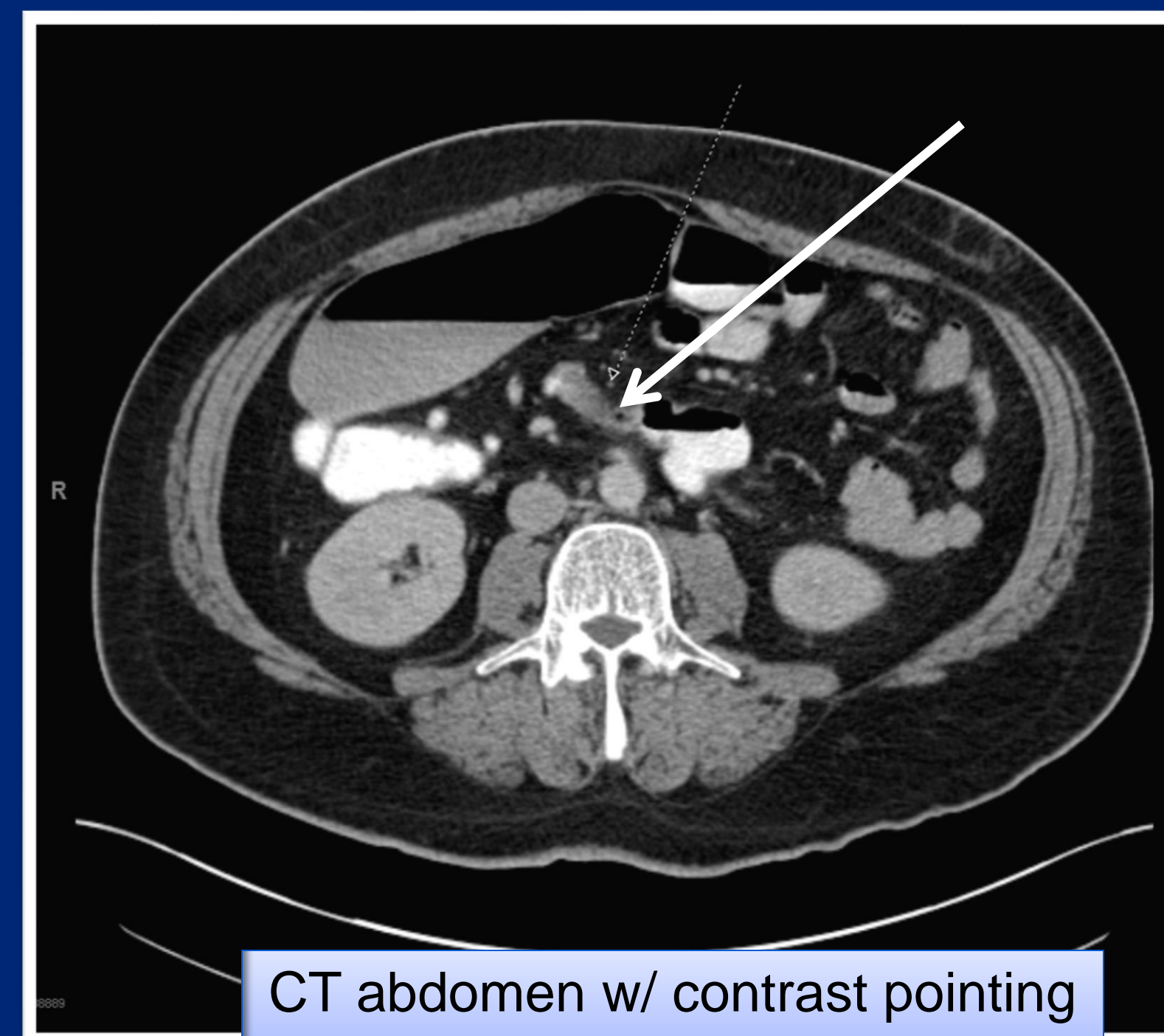
Course of Illness



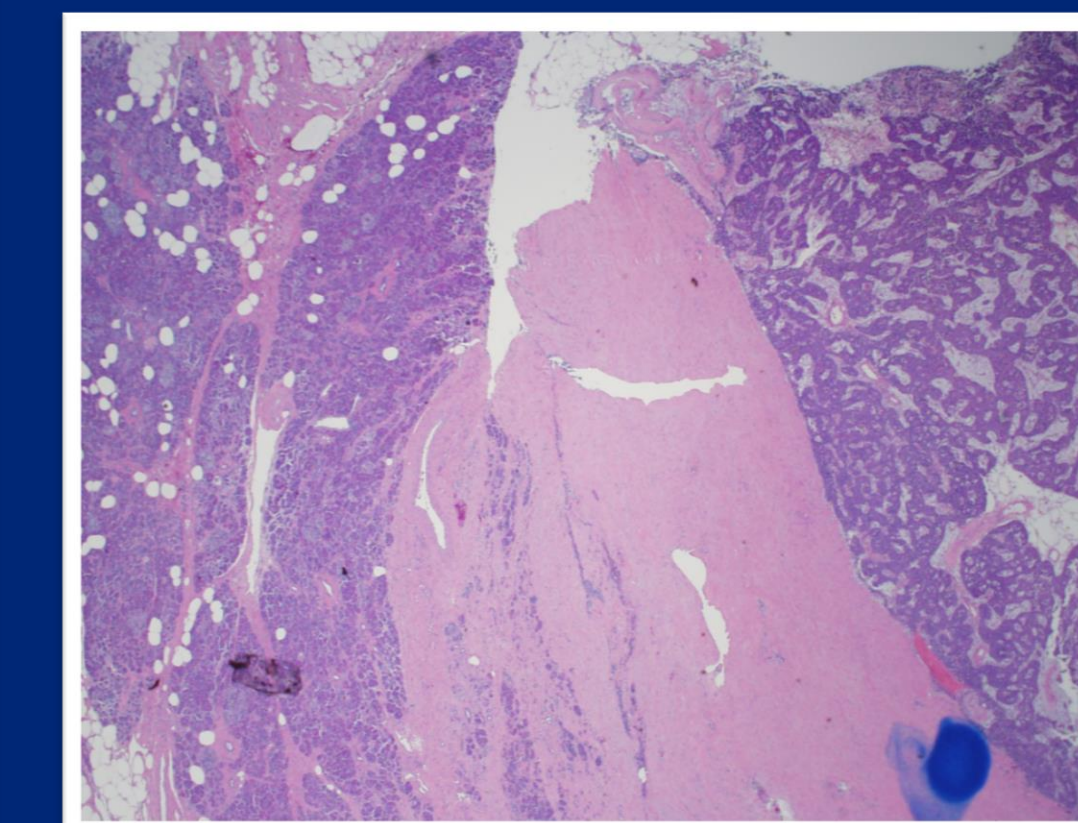
Diagnosis & Treatment

VIP diagnosis:

- Secretory diarrhea (osmotic gap <50 mOsm/kg)
- R/O other etiologies (infectious, factitious, etc.)
- VIP level >75 pg/mL
- Imaging (CT w/ contrast or MRI)
- If inconclusive endoscopic US or Somatostatin receptor scintigraphy



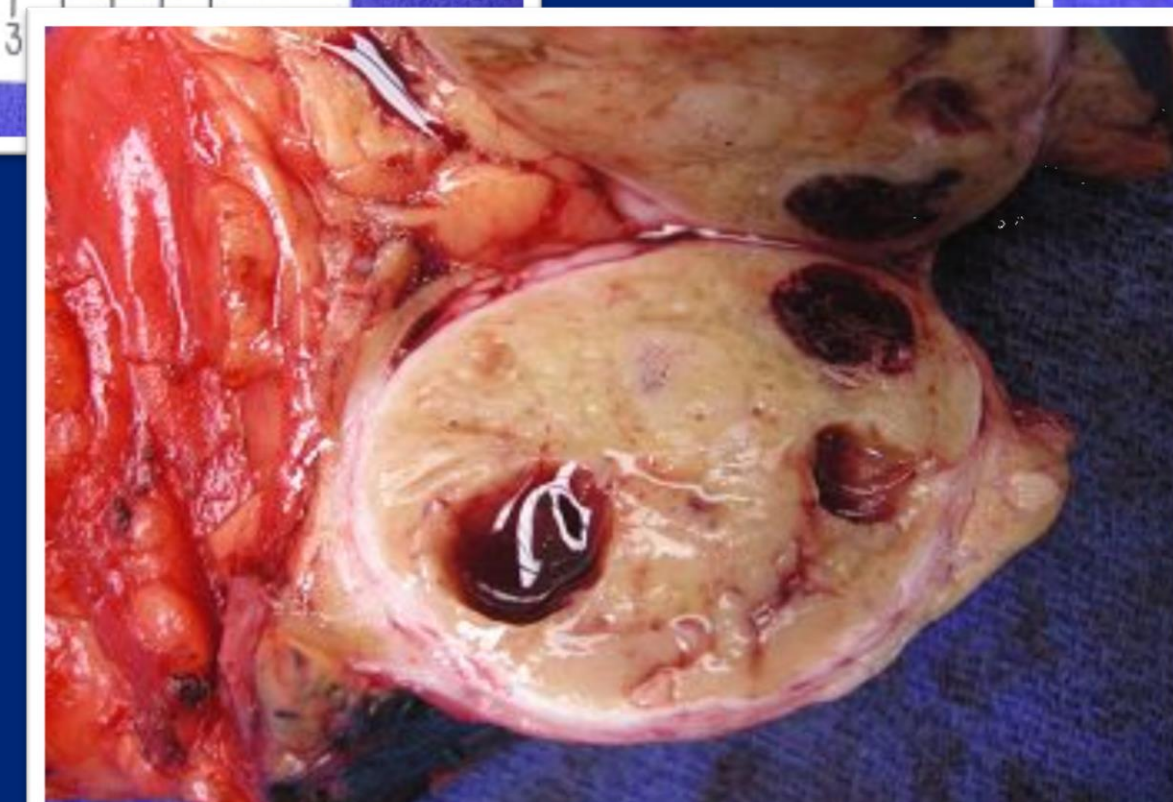
CT abdomen w/ contrast pointing to Pancreatic Mass.



Immunohistochemical stains for synaptophysin and chromogranin are positive in the neuroendocrine tumor.

Treatment

- Replete fluids / electrolytes
- Octreotide (drip vs. Q 8 hours subcutaneous vs. depot every month)
- If no metastasis → resection (curative)
- If refractory → interferon alfa



Distal pancreas with a 6.5 cm intermediate grade neuroendocrine tumor.

Take Home Points

- Importance of continued workup and diagnostic evaluation in a patient who presents with profound diarrhea without evident etiology.
- Diagnostic workup includes clinical presentation, laboratory testing including electrolytes and VIP level, imaging studies, histologic and immunohistochemistry findings.
- Histologic findings support clinical history and include: nested/trabecular arrangement of small/medium cells, finely granular eosinophilic cytoplasm, central round/oval nuclei, stippled chromatin

Acknowledgements

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References