

Malignant Gastrointestinal Neuroectodermal Tumor of the Small Intestine, a Rare Cause of Iron Deficiency Anemia.

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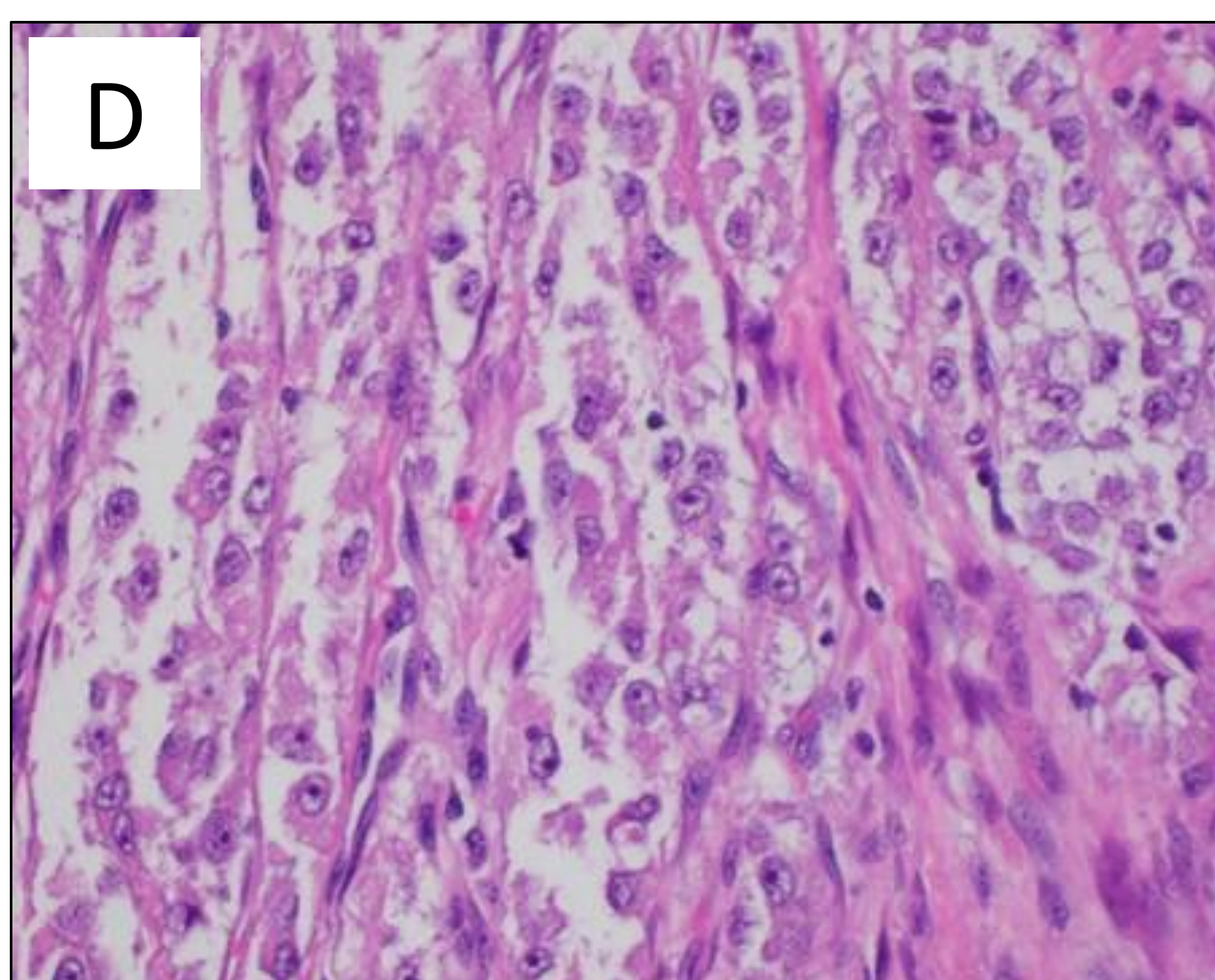
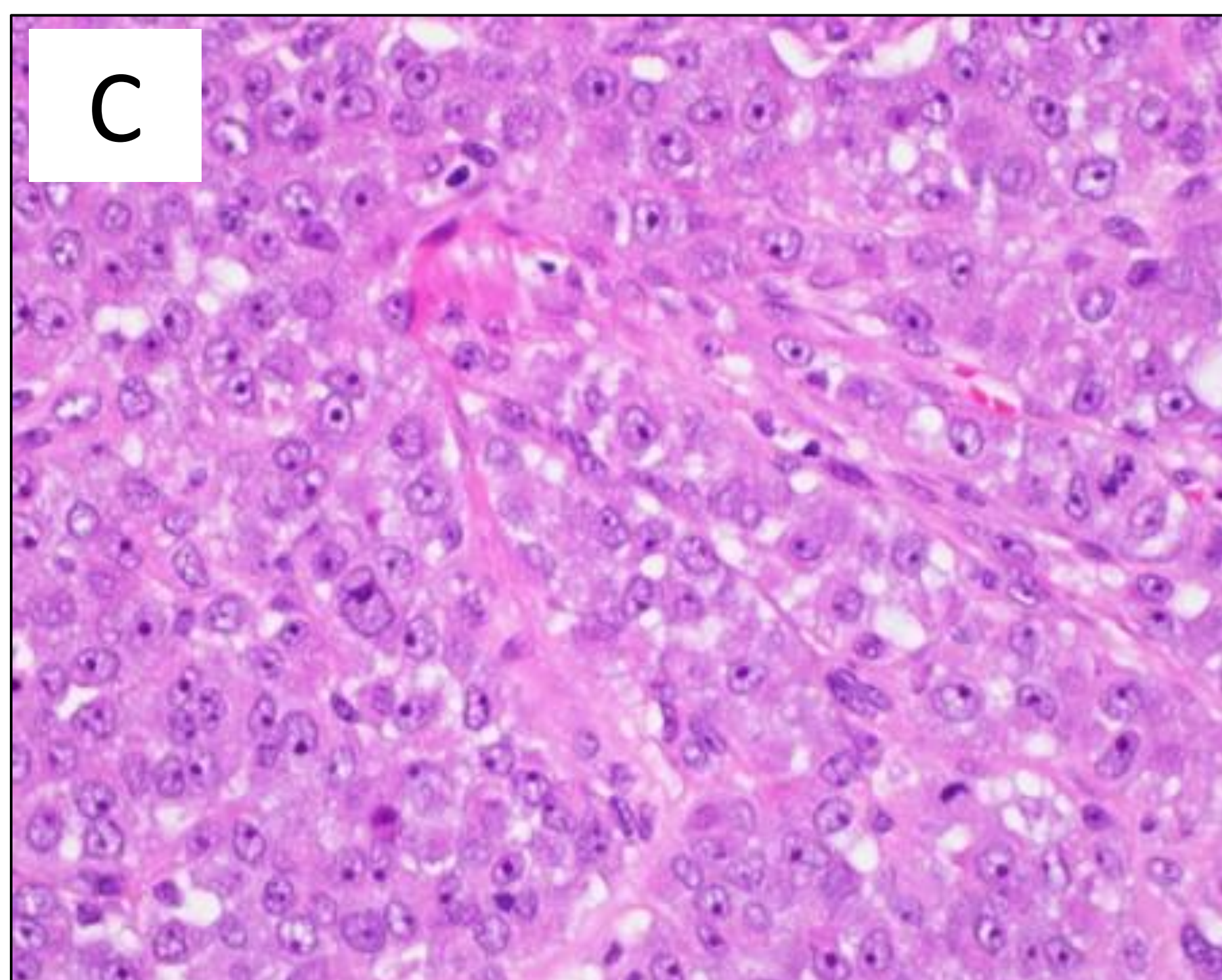
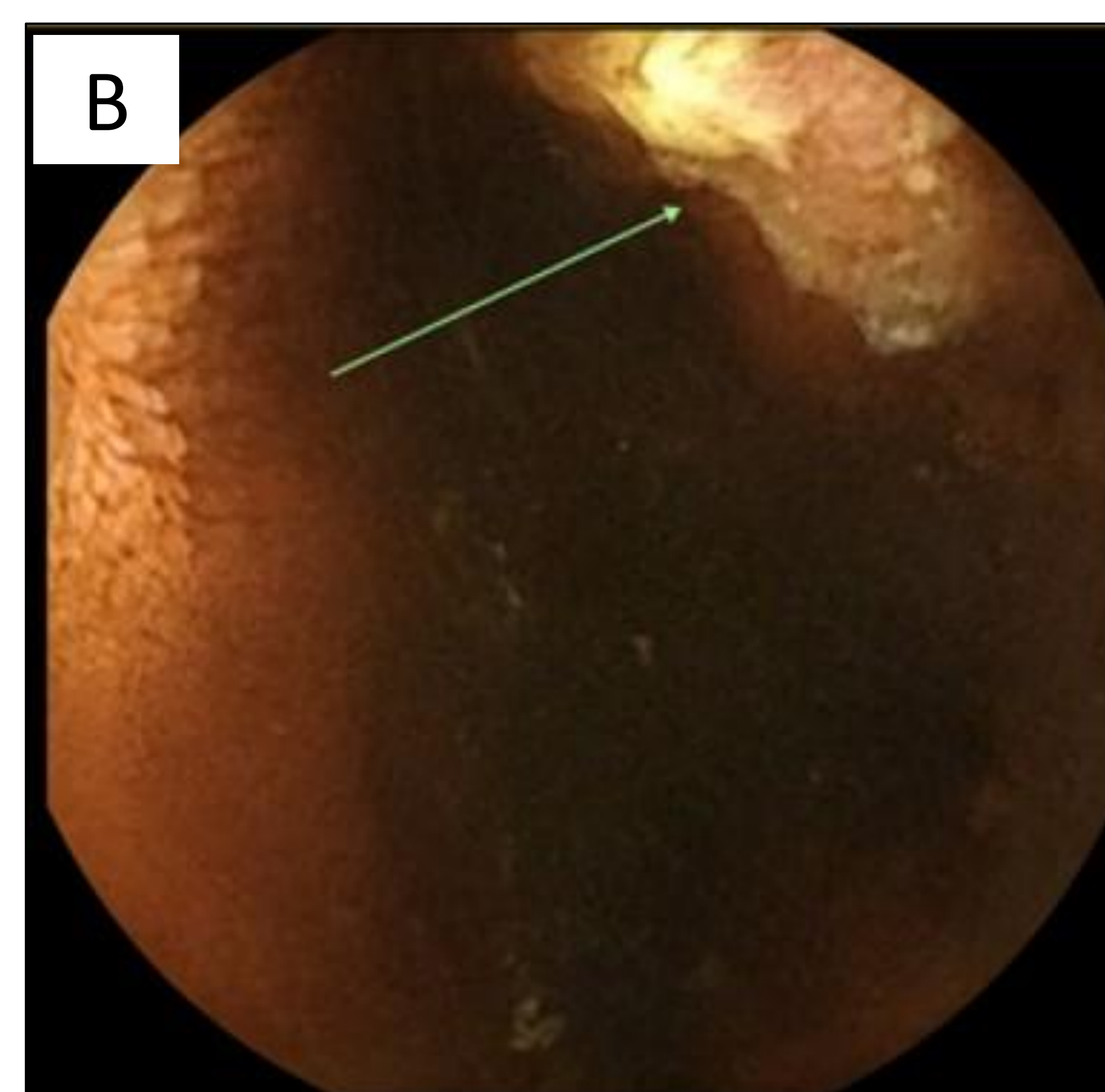
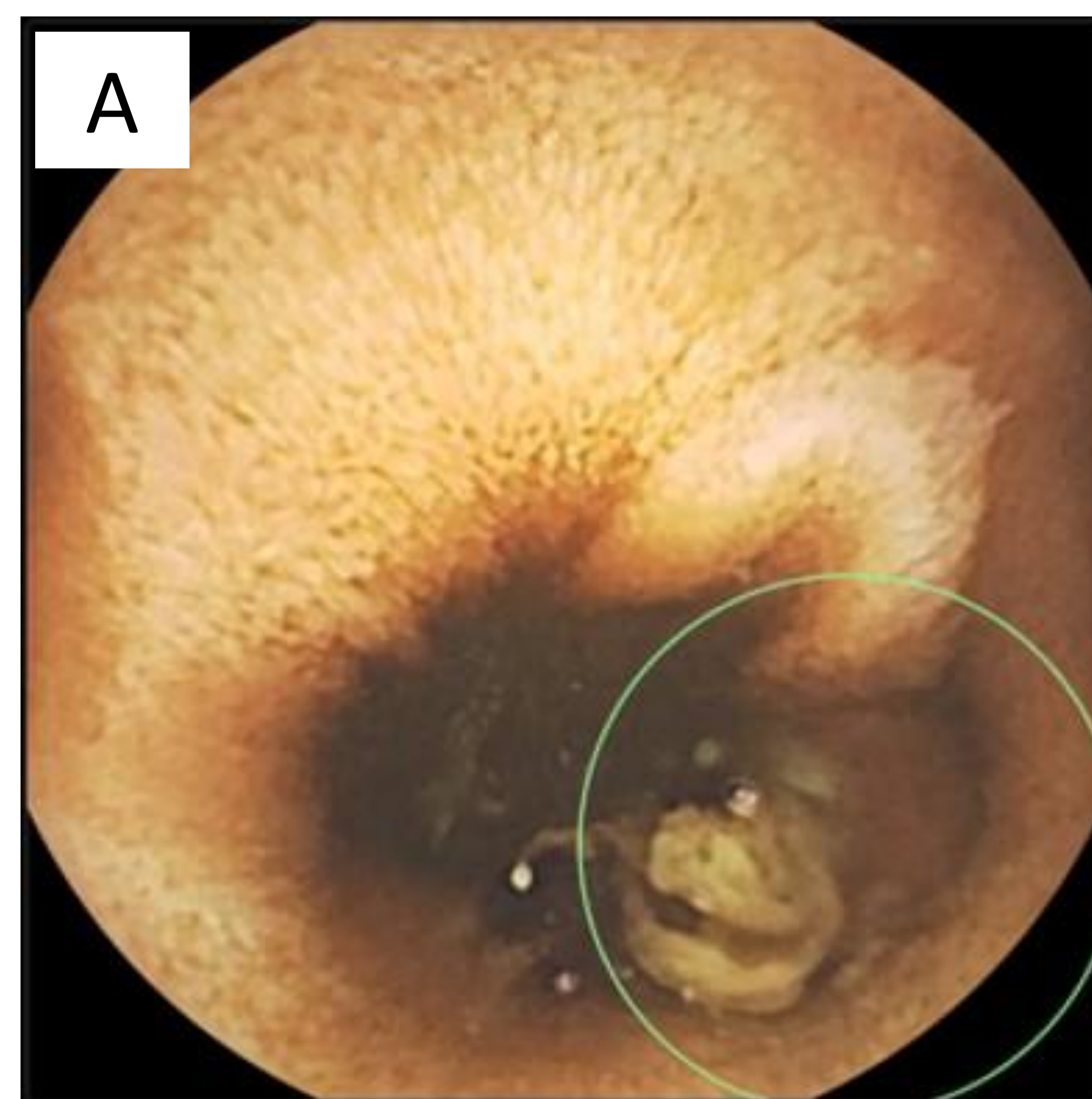
Introduction

- Gastrointestinal neuroectodermal tumors (GNET) are a type of high-grade sarcoma of the GI tract
- They share features with clear cell carcinoma of tendon sheaths and aponeuroses
- Typically, patients present with clinical signs and symptoms of small bowel obstruction, intussusception or abdominal mass
- These tumors can arise in the stomach, colon, as well as metastases to liver or regional lymph nodes
- We present a case of ileal GNET of an asymptomatic patient with Iron-deficiency Anemia (IDA) diagnosed using video capsule endoscopy (VCE)

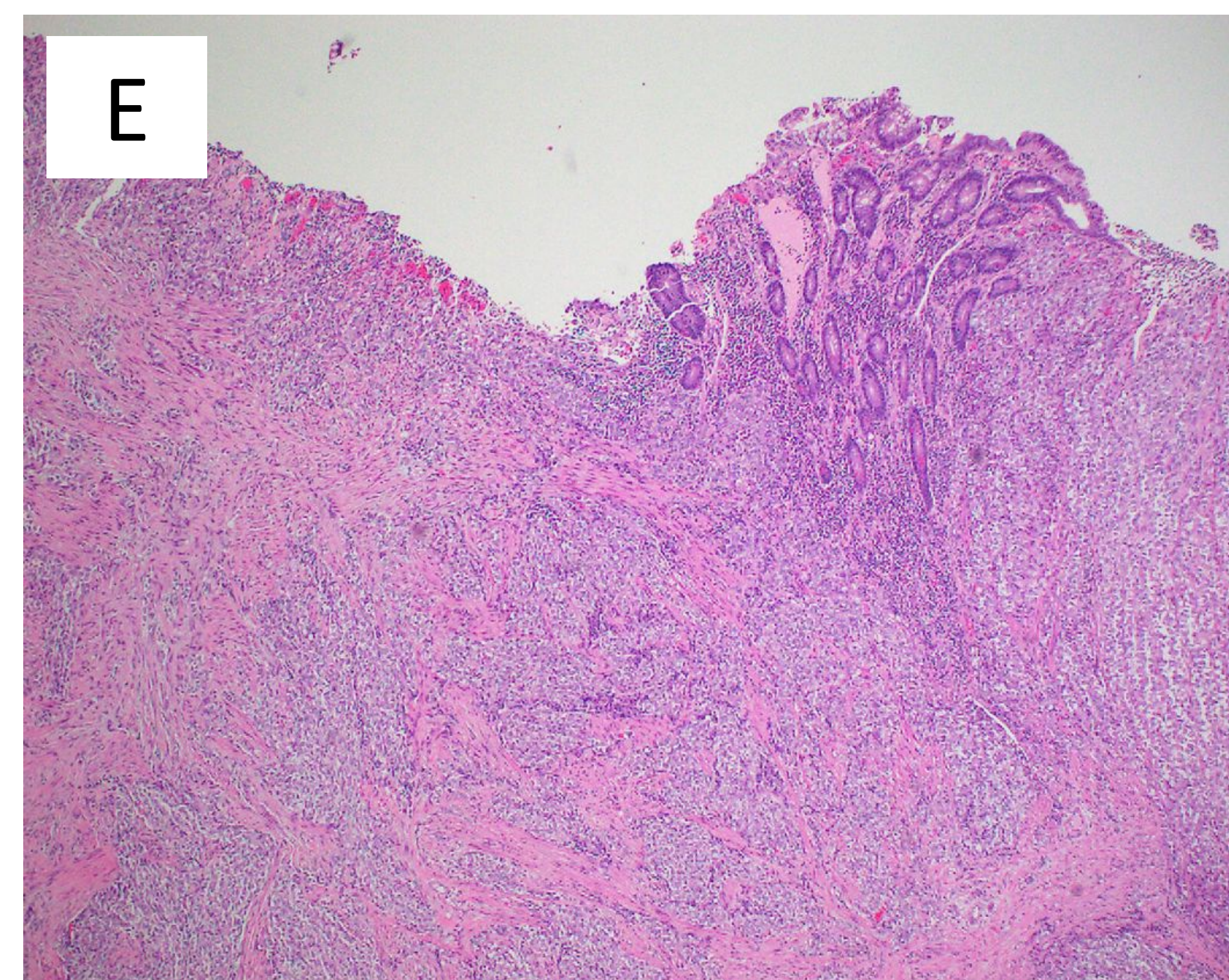
Case Presentation

- 58yo Hispanic male with PMH of GERD and obesity presented with chronic microcytic anemia
- Hemoglobin 8.6 g/dl, MCV 62.2, iron sat. 2.6%, transferrin 307 mg/dl, ferritin < 5 ng/ml, reticulocyte index 1.34; normal folate, vitamin B12 and TSH
- He was taking oral ferrous sulfate and vitamin C for IDA. He reported heartburn but denied melena, hematochezia, hematemesis, weight loss and had an unremarkable colonoscopy 4 years prior
- EGD with mild erosive esophagitis and mild chronic active gastritis with negative biopsies. Colonoscopy was unremarkable with optimal bowel preparation.
- VCE revealed an ulcerated mass in the ileal lumen. CT of the abdomen noted asymmetric wall thickening with enhancement of the small bowel
- Laparoscopy-assisted enteroscopy with rendezvous technique allowed direct visualization of a 3 cm proximal ileum mass requiring open segmental resection with primary anastomosis
- Pathology suggested malignant GNET or clear cell sarcoma-like tumor of the GI tract. IHC positive for neural crest markers (S100, SOX10) and Vimentin; negative for CD117, DOG1, HMB45, MART1, MAA, chromogranin, and synaptophysin. FISH positive for EWSR1 (22q12) rearrangement
- Clear surgical margins without regional lymph node spread; due to lack of standard guidelines it was recommended to surveil with imaging every three months without adjuvant radiation or chemotherapy

Figures



- A - Luminal view of small bowel tumor on VCE
 B - Close up view of small bowel tumor ulceration on VCE
 C - Histopathology slide showing epithelioid cells with eosinophilic cytoplasm, pleomorphic nuclei with prominent nucleoli and vesicular nucleoli. Scattered mitotic figures identified.
 D - Focal areas of clear cell change
 E - GNET involvement of the bowel wall with malignant cells forming sheets and nests. Tumor cells infiltrating the lamina propria and with mucosal ulceration



Discussion

- Differential diagnosis for GNET include Ewing's sarcoma tumors of the GI tract, Metastatic melanoma, Gastrointestinal stromal tumors, Leiomyosarcoma
- GNET typically present at a median age of 33 years old
- Typically arise from muscularis propria of ileum
- Surgical removal is preferred mode of treatment; chemotherapy and radiation have not been effective
- Median survival of 10 months

Conclusions

This case highlights the importance of VCE for small bowel assessment after negative bidirectional endoscopy to identify the etiology of IDA per American Gastroenterology Association guidelines. In this case, an ulcerated GNET was the etiology of IDA persisting despite iron supplementation.

References

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