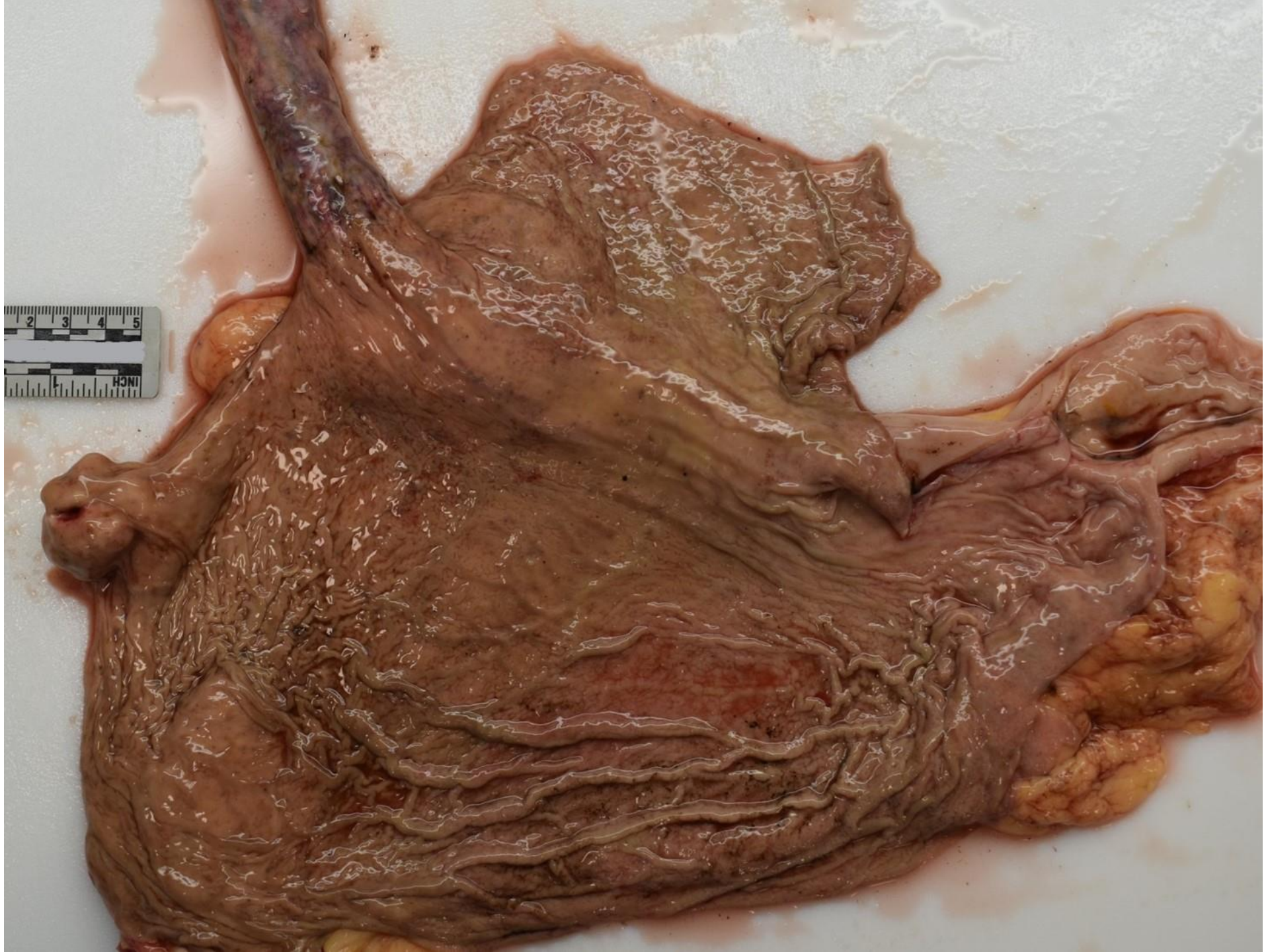
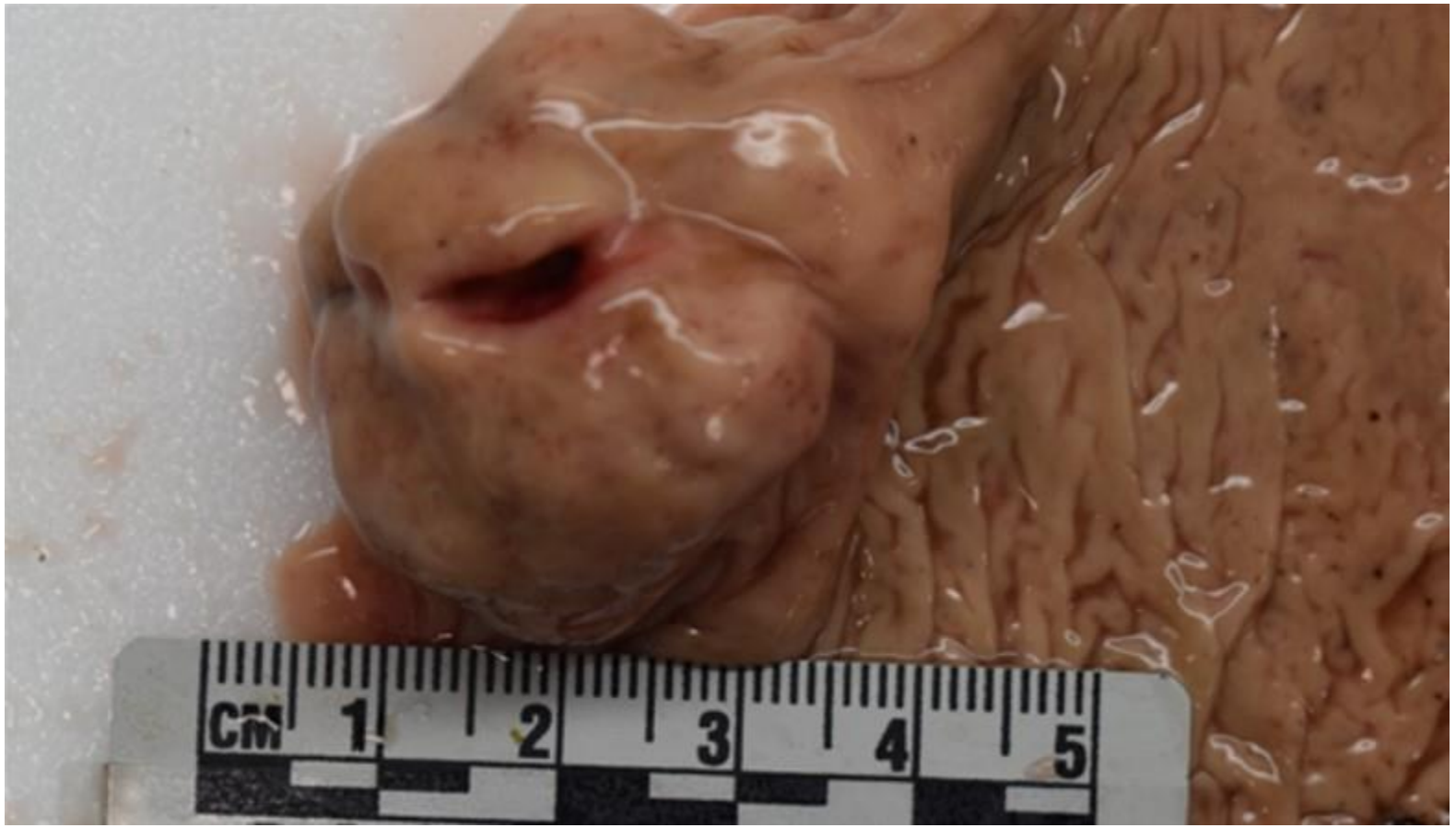
Case #98

NAME Educational Activities Committee

Case provided by:

Sarah Thomas (KCU College of Osteopathic Medicine) and Dr. Christine James (Johnson County Medical Examiner's Office in Olathe, Kansas)


2
3
4
5
INCH


CM
1
2
3
4
5

1. A 58 year-old man with a history of chronic alcoholism was witnessed by to “vomit blood” and collapse to the floor. At autopsy a mass was identified in the stomach. Microscopic examination revealed a partially calcified spindle-cell lesion that stained positively for CD34, CD117, and S100. What is the diagnosis?

- ◯ Gastrointestinal stromal tumor (GIST)
- ◯ Epithelioid hemangioendothelioma (EHE)
- ◯ Congenital anomaly of the stomach
- ◯ Inflamed Leiomyoma
- ◯ Gastric desmoid fibromatosis

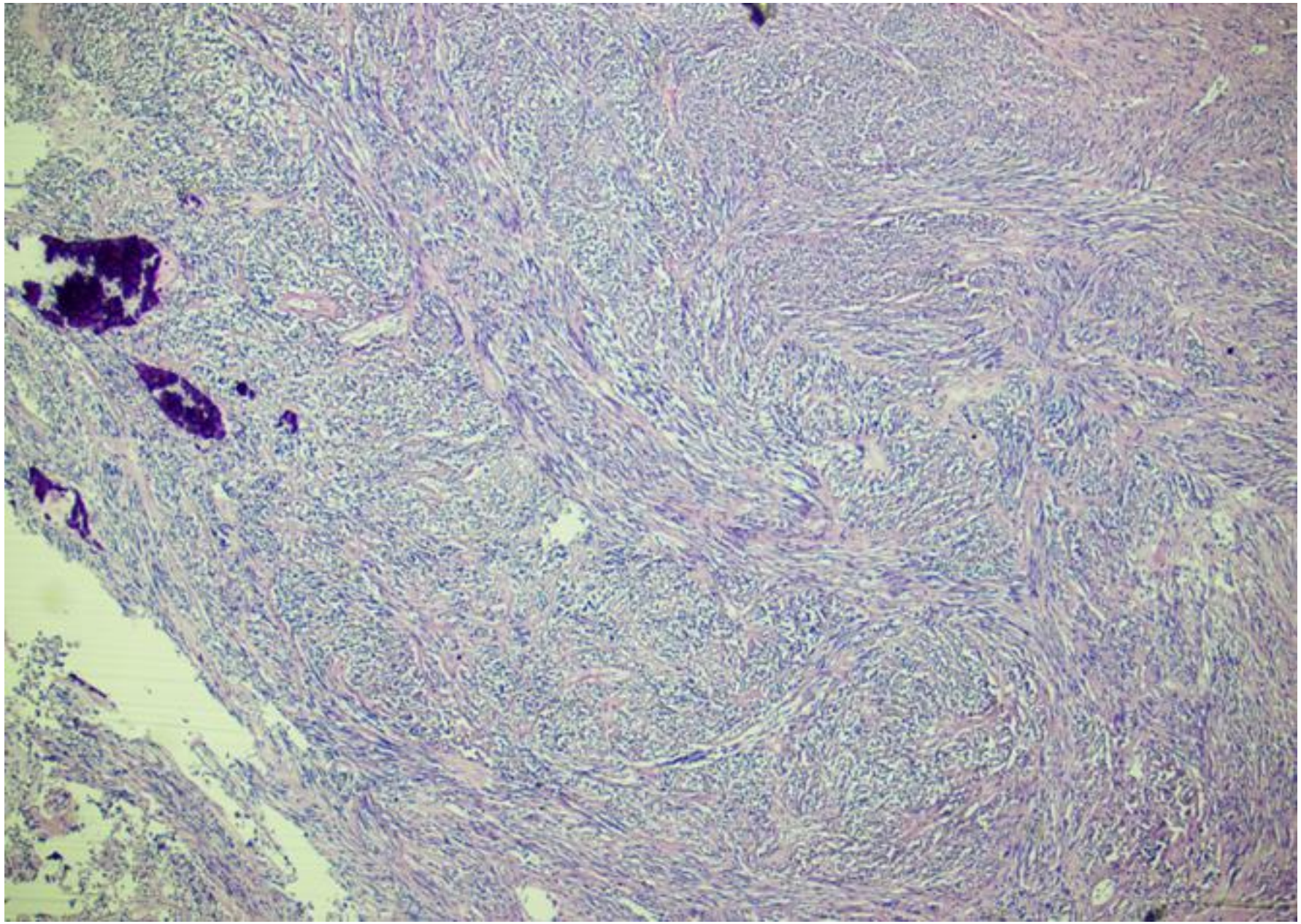
Answer...

A. Gastrointestinal stromal tumor (CORRECT ANSWER, 84.17% of responses)

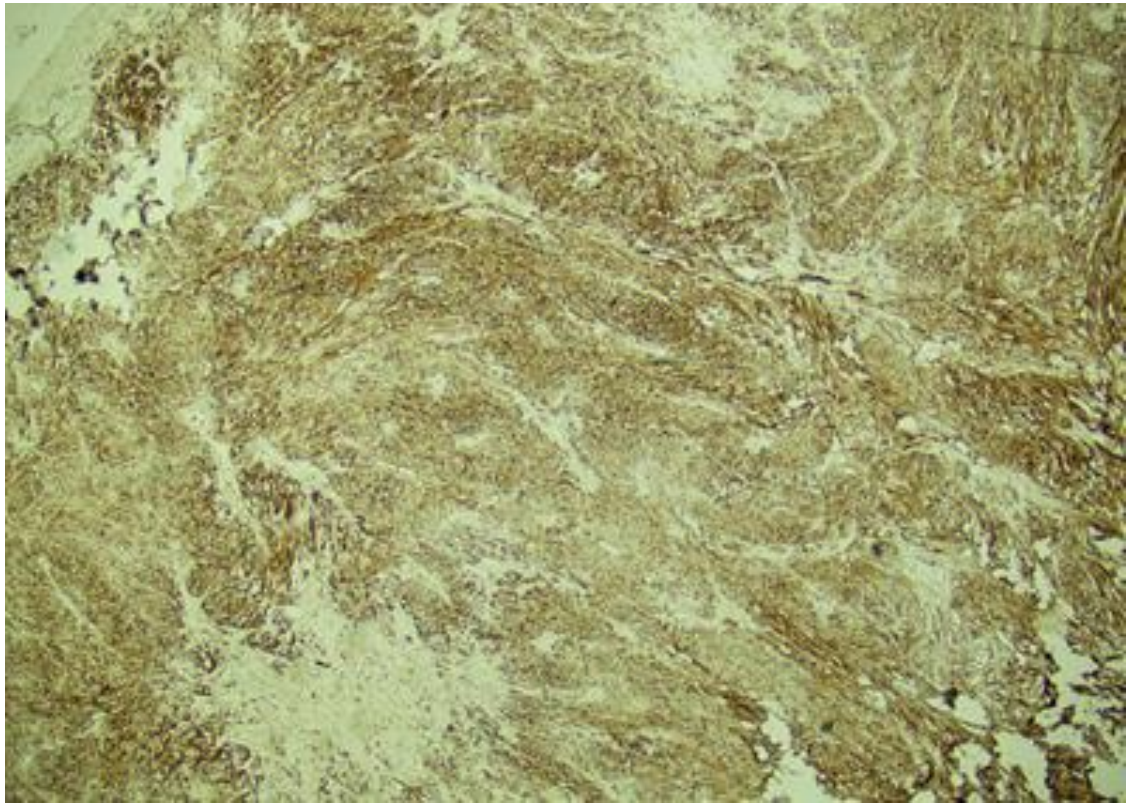
In addition to the information provided, the decedent was jaundiced, with scattered bruises and icteric sclerae. The peritoneal cavity contained 1700 mL of ascites and the liver was cirrhotic. Of note, there were no esophageal varices, and the 3.5 cm mass identified in the stomach was thought to be the source of his bleeding. Histology confirmed that the mass was a gastrointestinal stromal tumor (GIST).

GISTs are often benign and are the most common mesenchymal tumors of the GI tract. Sixty percent of GISTs occur in the stomach, and the median age of diagnosis is in the 60s. The most common presenting symptom is GI bleeding, such as melena or hematochezia. Though they can have several histologic patterns, approximately 70% are spindle cell type. The most common immunohistochemical markers are KIT, anoctamin-1 (also known as DOG1), CD117 and CD34. S100 is uncommon, and expression correlates with a poorer prognosis.

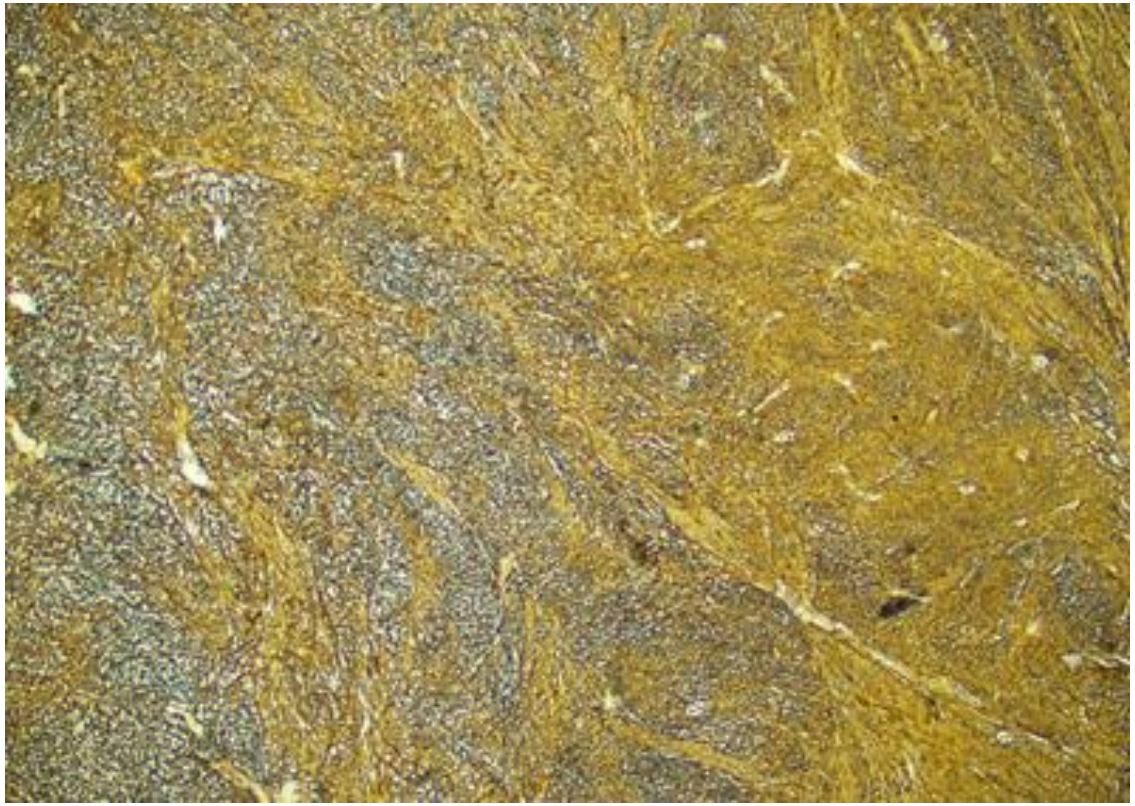
The decedent's cause of death was certified as upper gastrointestinal hemorrhage due to gastrointestinal stromal tumor (GIST) of the stomach. Chronic alcoholism was listed as a contributory factor, and the manner was certified as natural.



Spindle-cell morphology with scattered calcifications.



CD34



CD117

Other responses…

B. Epithelioid hemangioendothelioma (EHE)

EHE is a very rare vascular tumor or anomaly arising from the cells lining the blood vessels. EHE can arise anywhere in the body. An origin from a blood vessel can be demonstrated in roughly 50% of cases. It can present with symptoms of GI bleeding, but its treatment is different from GISTs. EHE typically has positive endothelial differentiation markers: CD31, ERG, CD34 and FLI-1.

C. Congenital anomaly of the stomach

While anomalies can be present in the stomach and GI tract, the histologic and immunohistochemical features of this lesion were most consistent with a GIST.

D. Inflamed leiomyoma

Leiomyomas are rarely found in the stomach, and they typically go unnoticed unless they grow large enough to impact nearby structures or if they become ulcerated and cause epigastric pain and/or bleeding. Though also spindle-cell lesions histologically, leiomyomas are usually positive for smooth muscle actin (SMA) and desmin, and typically negative for CD117 (c-kit protein) and CD34.

E. Gastric desmoid fibromatosis

Gastric desmoid fibromatosis is an extremely rare lesion. Typically found intramurally, these usually follow an expansile growth pattern that mimics GIST. Desmoid fibromatosis is typically locally aggressive and infiltrative, with approximately 50% of presentations in the abdominal wall and 40% within extra-abdominal soft tissues. Only about 8% affect the mesentery or even more rarely, arise from the small bowel mimicking GIST. The most common immunohistochemical markers are β-catenin and focal cytoplasmic positivity for SMA.

REFERENCES

1. Burch J, Ahmad I. Gastrointestinal Stromal Cancer. [Updated 2022 Sep 26]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK554541/
2. Sarlomo-Rikala M, Kovatich AJ, Barusevicius A, Miettinen M. CD117: a sensitive marker for gastrointestinal stromal tumors that is more specific than CD34. Mod Pathol. 1998 Aug;11(8):728-34. PMID: 9720500.
3. Stacchiotti S, Miah AB, Frezza AM, et al. Epithelioid hemangioendothelioma, an ultra-rare cancer: a consensus paper from the community of experts. ESMO Open. 2021;6(3):100170. doi:10.1016/j.esmoop.2021.100170
4. Nigam, Neha MD; Chalhoub, Walid MD; Abdelaziz, Dalia MD; Haddad, Nadim MD, FACG. Gastric Leiomyoma: A Case Report: 756. American Journal of Gastroenterology 108():p S226, October 2013.
5. Lee JC, Curtis D, Williamson JB, Ligato S. Gastric Desmoid Fibromatosis - Report of a Rare Mimic of Gastrointestinal Stromal Tumor. Cureus. 2021;13(11):e19614. Published 2021 Nov 15. doi:10.7759/cureus.19614