

**Case History:**

A 49-year-old male with a past medical history of GERD, chronic hepatitis B and C, COPD, smoking, and polysubstance dependence, presented for evaluation of worsening heartburn and dysphagia to liquids with regurgitation. CT abdomen/pelvis demonstrated an irregular gastric mass measuring 2.5 cm (Figure 1), and upper GI endoscopy showed a 2.5 cm subepithelial mass in the gastric cardia, 2 cm distal to the gastroesophageal junction (Figure 2). The mass was stable in size and prior biopsies had been nondiagnostic. Repeat biopsy was performed (Figures 3-5).

Figure 1. CT abdomen/pelvis imaging demonstrating gastric mass (arrow).

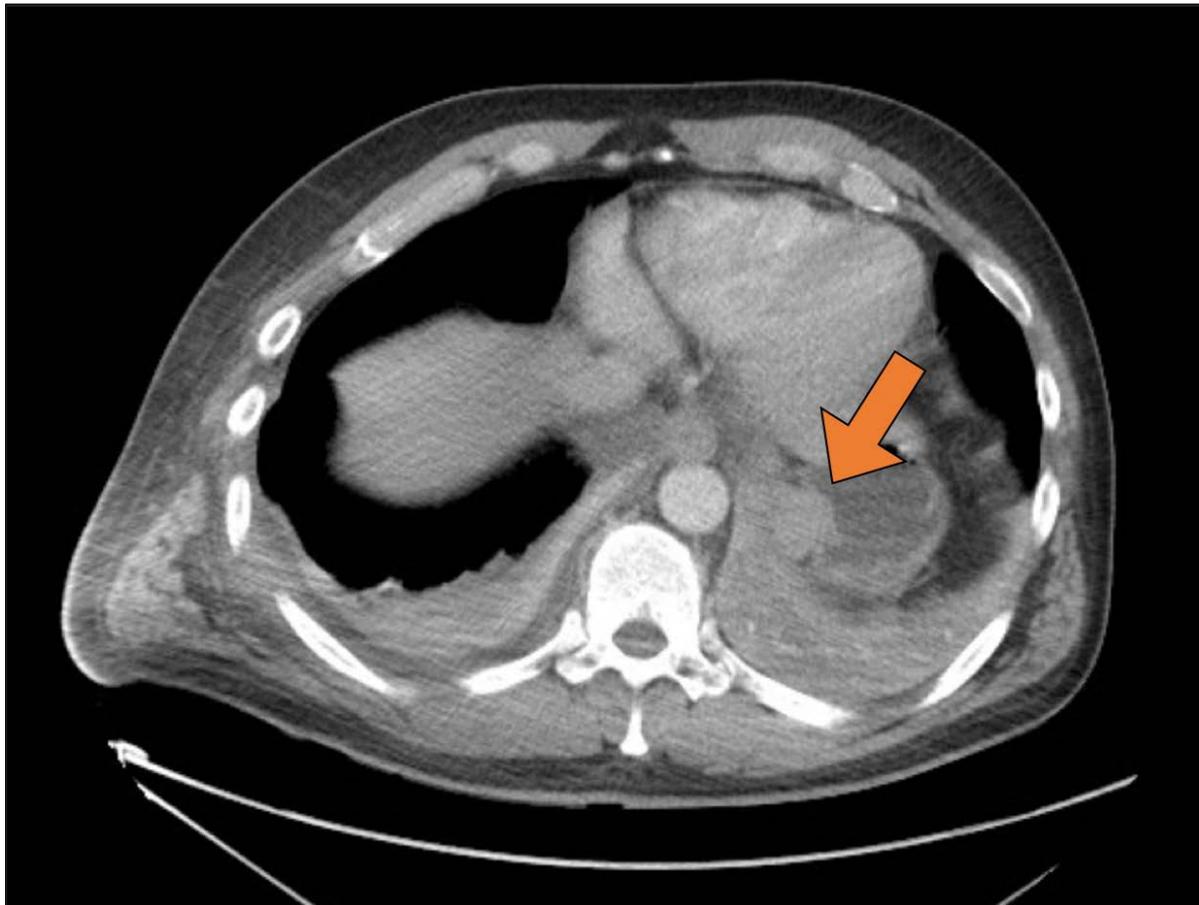


Figure 2. Endoscopic appearance of the subepithelial gastric mass.



Figure 3. Core biopsy, low power view.

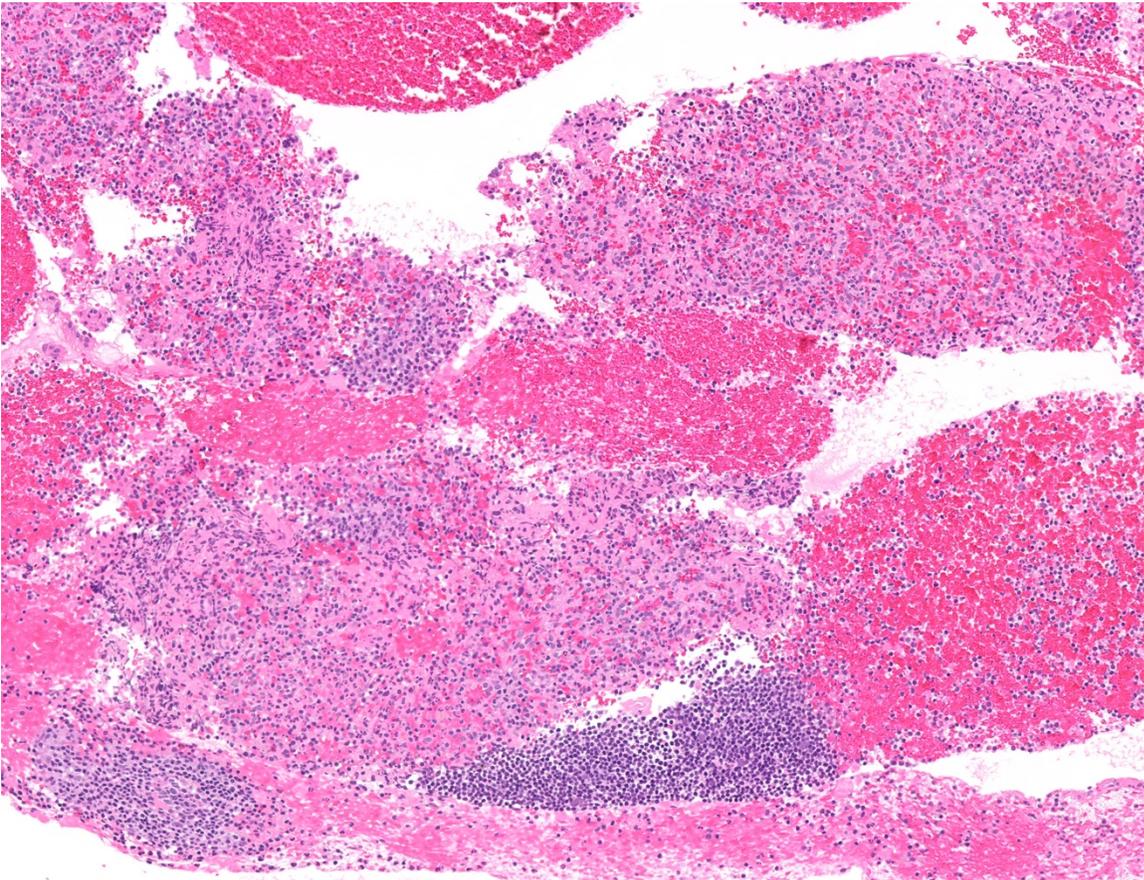


Figure 4. Core biopsy, medium power views.

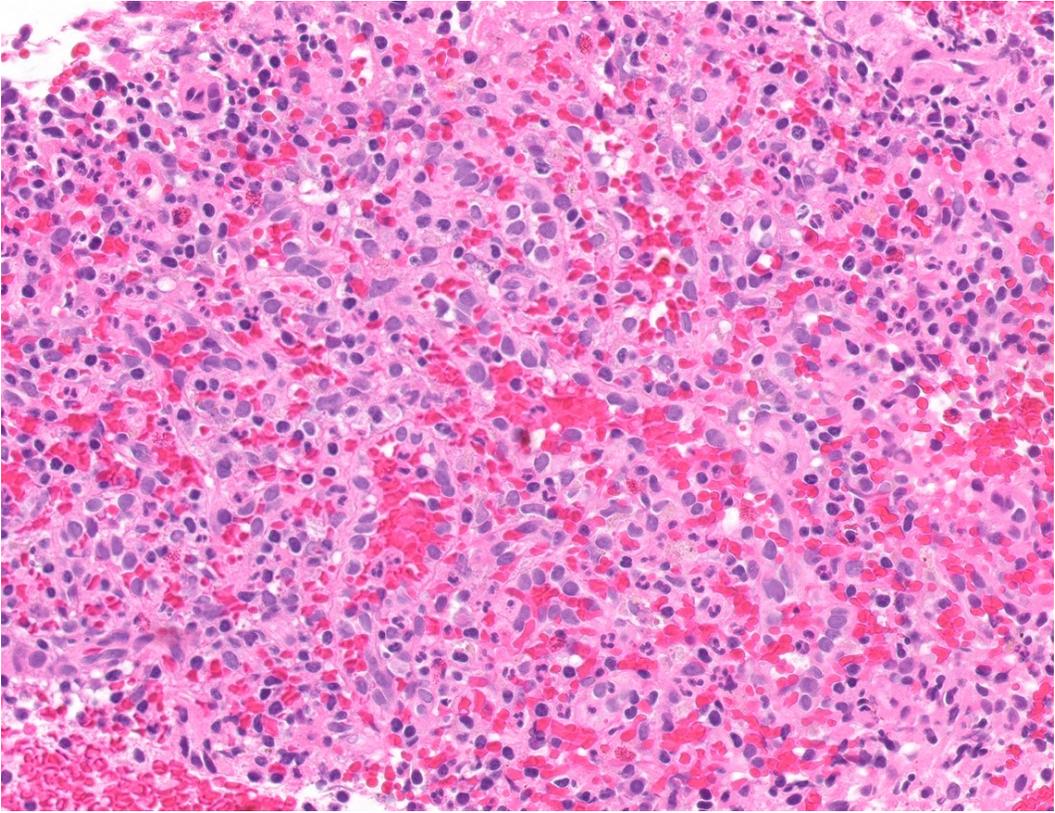
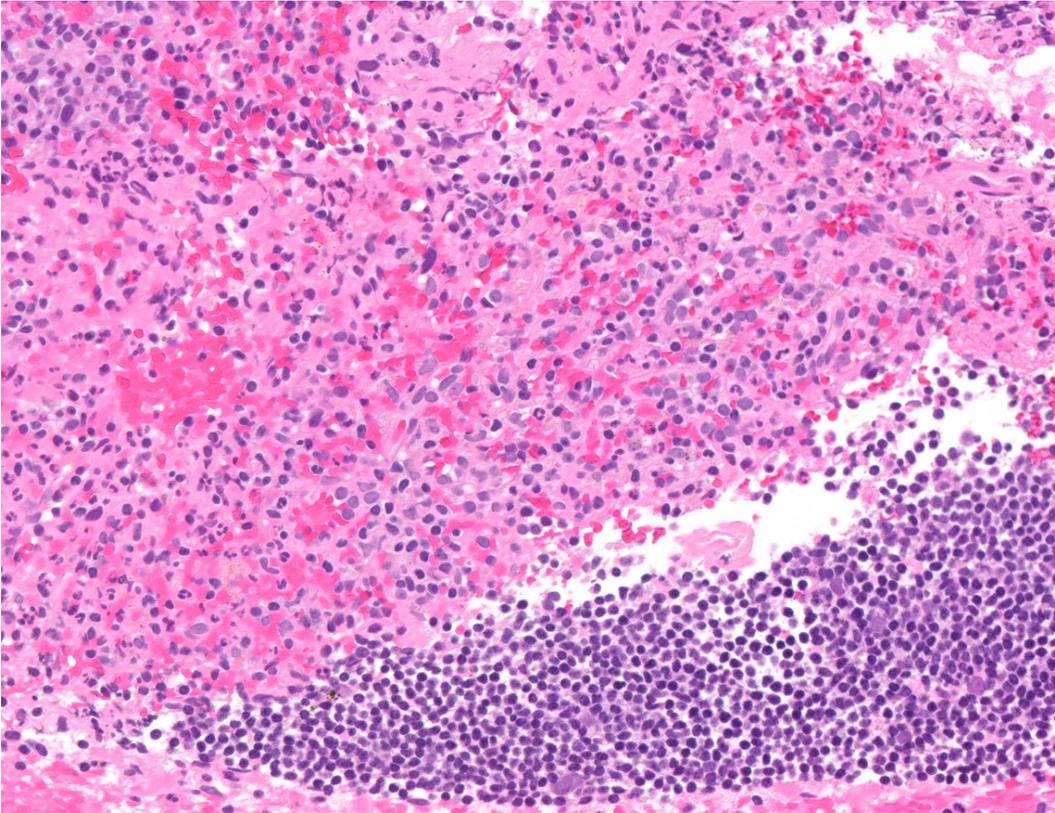
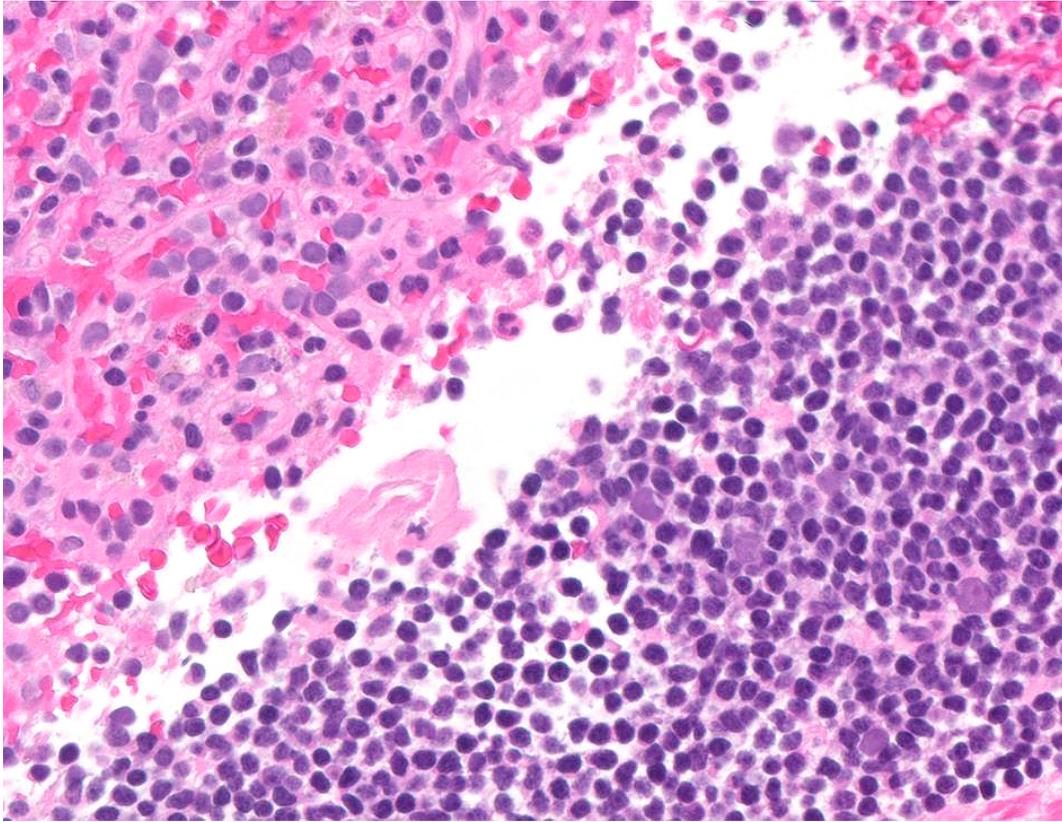


Figure 5. Core biopsy, high power view.



**What is your diagnosis?**

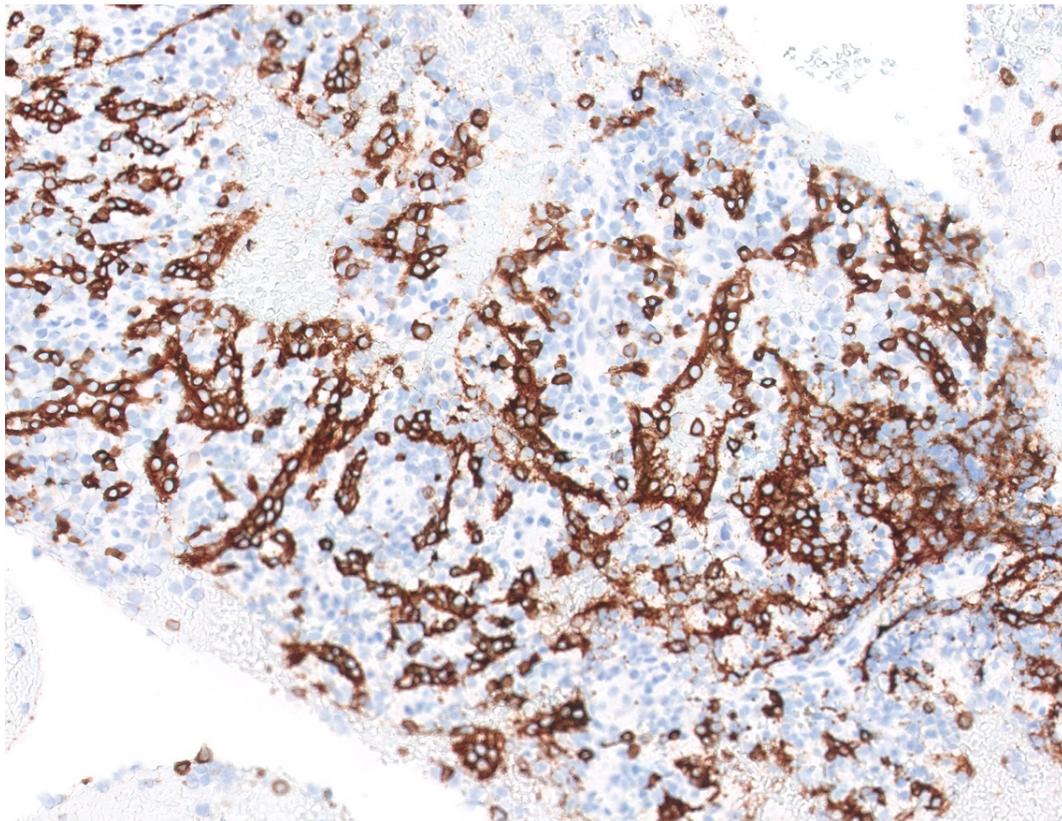
- A) Gastrointestinal stromal tumor (GIST)
- B) Glomus tumor
- C) Schwannoma
- D) Splenosis
- E) Lobular capillary hemangioma

**Correct Answer:**

D) Splenosis

The biopsy shows fragments of vascular and lymphoid tissue with areas containing lymphoid aggregates (consistent with white pulp) and areas with numerous vessels with a lymphoid background (consistent with red pulp). A CD8 immunostain highlights the splenic littoral cells lining the vascular spaces (Figure 6). The findings are consistent with splenosis. Upon investigation, the patient had a remote history of traumatic splenectomy following a motor vehicle accident.

Figure 6. CD8 immunostain highlighting the littoral cells.



## Discussion:

**Splenosis** is the heterotopic auto-transplantation of splenic tissue following disruption of the spleen's capsule by traumatic rupture (67% of cases) or iatrogenic splenic injury.<sup>1,2</sup> It is most commonly seen in the abdominal and pelvic cavities, particularly on the serosal surfaces of the small intestine and mesentery, greater omentum, parietal peritoneum, and diaphragm.<sup>3</sup> Splenosis can also involve solid organs, most commonly the liver, but also extra-abdominal organs such as the lungs.<sup>1,4</sup> It is hypothesized that the mechanism is direct implantation of splenic tissue from the rupture of the spleen, but splenosis in some sites such as lung may occur via the hematogenous route.<sup>2</sup>

There have been fewer than 20 reported cases of gastric splenosis, all of which involved the gastric fundus except one case of the gastric cardia.<sup>4</sup> Involvement of the serosa and/or muscularis propria is typical, but rare cases have involved submucosa. Gastric splenosis is usually asymptomatic and is found incidentally during endoscopy or surgery, but occasionally can present with upper gastrointestinal (GI) bleeding.<sup>1,3</sup> The mean age of presentation is 44 years old (range 17-68 years old), with an average post-splenectomy interval period of 14.5 years (range 4-38 years). The mean size of a splenic implant is 2.3 cm (range 1.1-5 cm). Most cases of gastric splenosis cases are initially suspected to be gastrointestinal stromal tumors (GISTs) prior to histopathologic examination.

Histologically, splenosis is composed of red and white pulp components of the normal spleen. The sinusoids of the red pulp are lined by a distinctive cell type called the "splenic littoral cell," which has a characteristic dual endothelial and histiocytic immunophenotype. Notably, these cells are positive for CD8, as well as CD31, ERG, WT1, Factor VIII, and CD68; they are negative for CD34, distinguishing these vascular structures from those of a vascular tumor such as hemangioma.<sup>5</sup>

Resection of splenosis is not necessary unless the patient is symptomatic from intestinal obstruction or GI bleeding, or if malignancy cannot be ruled out on a limited small biopsy specimen.

**GIST** is the most common mesenchymal tumor of the GI tract, with the stomach being the most common site (60% of cases). As such, GIST is high on the differential diagnosis of a subepithelial gastric mass. GISTs are typically composed of lobules of uniform spindle or epithelioid cells, but are not generally associated with prominent vessels or lymphoid stroma. CD117 (c-kit) and DOG1 are highly sensitive and specific immunohistochemical markers for the diagnosis of GISTs<sup>6</sup>, and should be performed to confirm the diagnosis.

**Glomus tumors** are mesenchymal neoplasms composed of modified smooth muscle cells that most commonly involve the skin, but rarely can involve the GI tract, almost exclusively in the stomach.<sup>7,8</sup> Glomus tumors of the stomach show a female predominance and typically present with GI bleeding and/or ulceration.<sup>8</sup> Microscopically, the tumors are well-circumscribed and composed of nodules of uniform, small, round-to-polygonal cells with round nuclei and delicate, homogenous chromatin. These cells can resemble lymphocytes, such as those seen in splenosis, but are positive for smooth muscle actin (SMA) and calponin, and negative for lymphoid markers by immunohistochemistry. In addition, glomus tumor cells are also characteristically organized around vessels; however, unlike the sinusoidal vascular spaces of the splenic red pulp, those found in glomus tumors are prominent, dilated, thin-walled hemangiopericytoma-like vessels.<sup>8</sup>

**Schwannomas** of the GI tract almost always arise in the muscularis propria and classically have peripheral lymphoid cuffs that are readily identified at low magnification.<sup>9,10</sup> The tumor is composed of spindle cells but can have focal epithelioid areas, and a small biopsy specimen with mesenchymal and lymphoid areas, such as in this case, raises the possibility of a schwannoma. Immunohistochemically, the tumor cells are positive for S100 and SOX10.

**Lobular capillary hemangiomas** are benign vascular proliferations that uncommonly occur in the GI tract and resemble their counterparts at other locations. There have been around 10 case reports of gastric lobular capillary hemangiomas in the literature; most patients presented with upper GI bleeding or melena and were found to have a pedunculated polypoid lesion in the stomach.<sup>11</sup> Microscopically, there is a lobular arrangement of closely-packed small vessels that is often associated with overlying erosion or ulceration. Thus, the vessels tend to be lined by reactive, plump endothelial cells that morphologically resemble the splenic littoral cells. However, these cells are positive for all vascular markers including CD34 and are negative for CD8. Inflamed granulation tissue may be seen overlying a lobular capillary hemangioma, but a dense lymphoid infiltrate is not typically seen.

## References:

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## Contributing authors:

M. Lisa Zhang, MD

PGY-4 in Anatomic and Clinical Pathology

Joseph Misdraji, MD

Associate Professor of Pathology

Department of Pathology, Massachusetts General Hospital, Boston, MA