

Introduction

Gastrointestinal stromal tumors (GISTs) make up the more common of the two groups of mesenchymal GI neoplasms, typically of the subepithelium. GISTs are unique as they are thought to arise from the pacemaker cells of the GI tract, the interstitial cells of Cajal, that give them CD34 positivity and are typically associated with mutation of KIT gene, identified via CD117 positivity. GISTs typically occur in the stomach (40-60%) or jejunum/ileum (25-30%) but can occur anywhere in the gastrointestinal tract, including mediastinum and accessory structures like mesentery, peritoneum or omentum.

Case Description

A 62-year-old male with a past medical history of type 2 diabetes mellitus presented to the emergency department for abdominal pain. He had one day of epigastric pain radiating to his back, worse with oral intake. He endorsed a 10-pound unintentional weight loss over the previous two months.

Computed Tomography (CT) of the abdomen with intravenous (IV) contrast demonstrated inflammation in the porta hepatis with accompanying porta lymphadenopathy and a large paraesophageal, retrocrural mass.

Esophagogastroduodenoscopy (EGD) with endoscopic ultrasound (EUS) was performed revealing 5.6 cm paraesophageal mass with central anechoic degeneration and a 1.8 cm fourth layer gastric cardia subepithelial lesion, both suspicious for GIST. Both were biopsied and were CD34 and CD117 positive, consistent with GISTs. The patient was later discharged home with GI follow but was lost to follow up.

Discussion

Although GISTs are the most common type of stromal tumor in the GI tract, they only make up approximately 1% of primary GI malignancies. GISTs have an incidence of 0.68 cases per 100,000 population. Mediastinal GISTs are more unique, with only twelve cases reported. Furthermore, multiple synchronous GISTs have only been reported in three other cases when not associated with GIST syndromes.

CT is the imaging modality of choice for GISTs. GISTs usually appear as smooth, solid masses easily seen with CT scan. EGD is especially useful for diagnosing GISTs if located in the esophagus, stomach or duodenum appearing as smooth, submucosal masses with possible central ulceration and/or protrusion into the lumen. On EUS, GISTs appear hypoechoic, homogenous with clear margins arising most commonly from the fourth layer (muscularis propria) and less likely from the second layer (muscularis mucosa).

GIST management often consists of surgery and/or imatinib depending on the characteristics of the GIST.

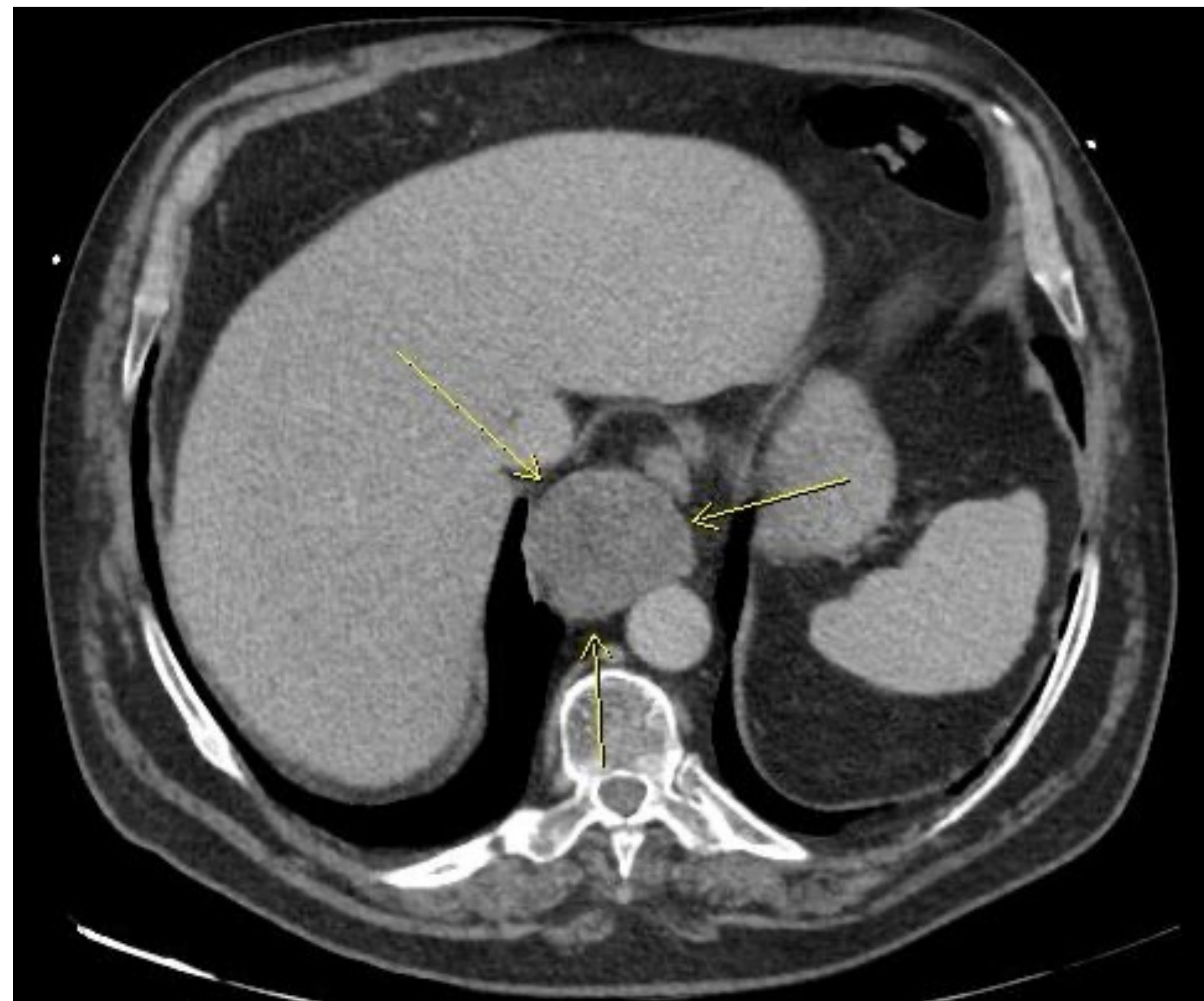


Figure 1: Transverse CT View of Paraesophageal GIST



Figure 2: Sagittal CT View of Paraesophageal GIST

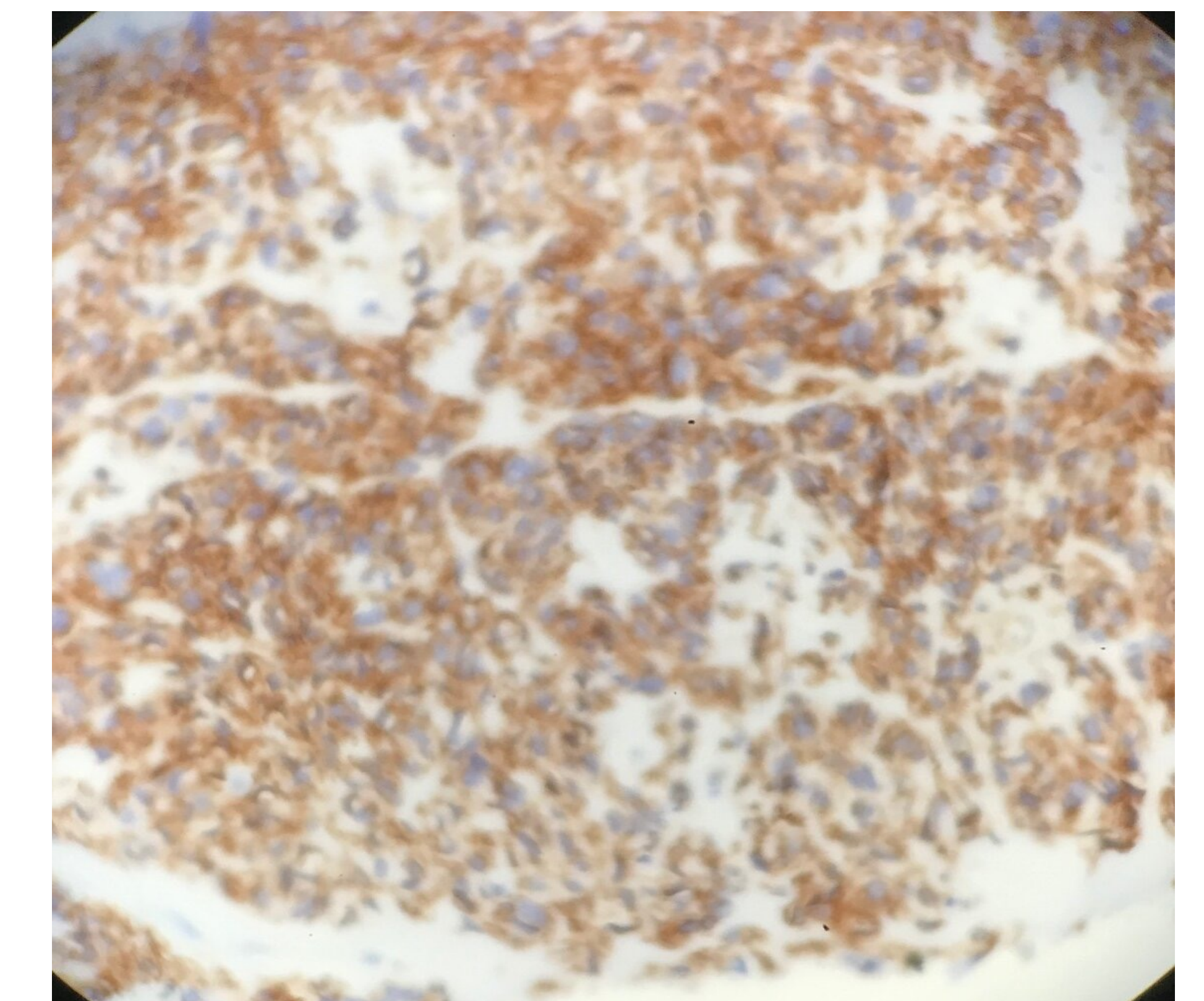
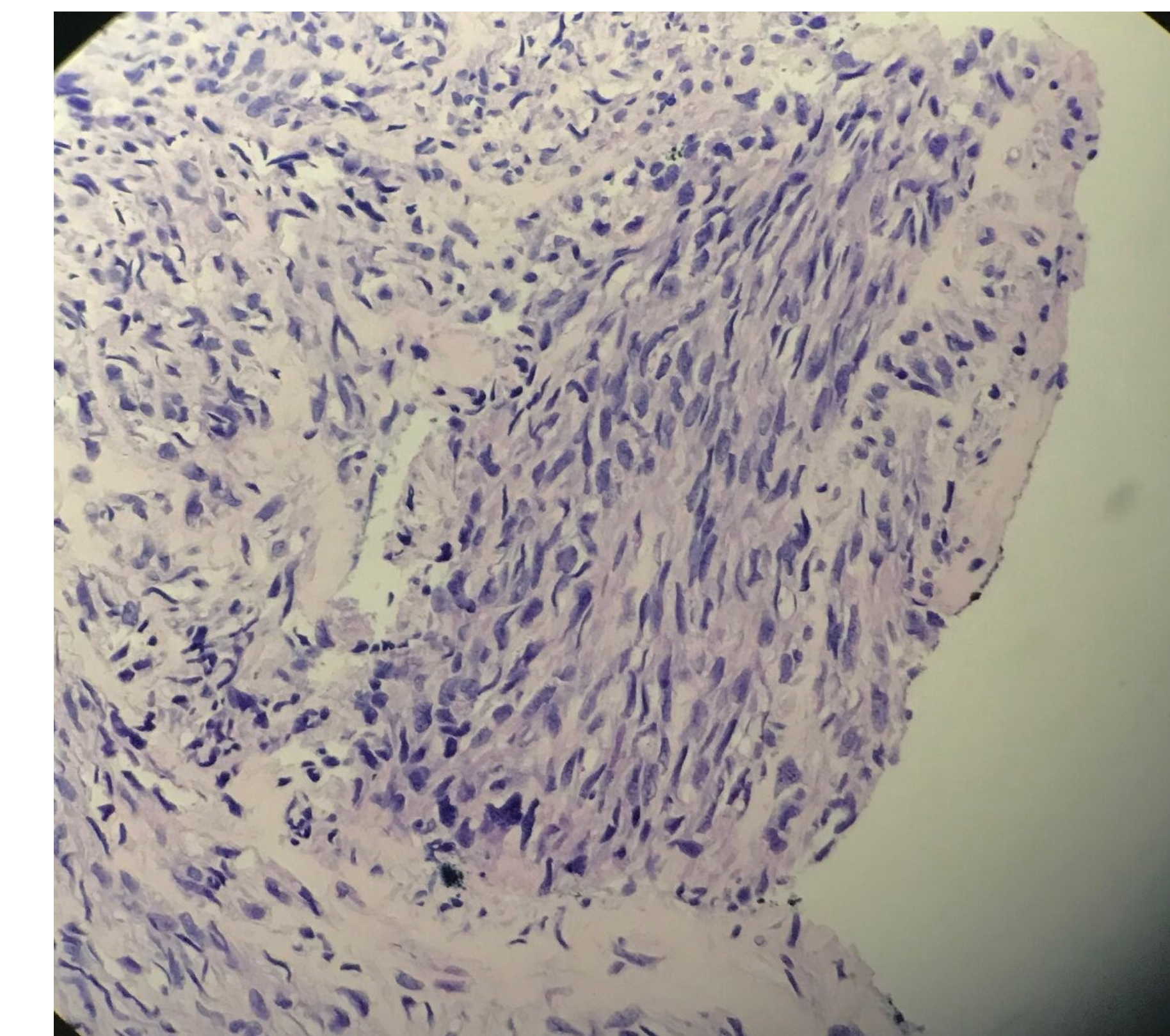


Figure 3 & 4: H&E and DOG1 Stain Confirming GIST

Contact

Brandon Wiggins, DO, MPH
 Ascension Genesys Hospital
 Email: Brandon.wiggins@ascension.org
 Phone: 248 227 7896