

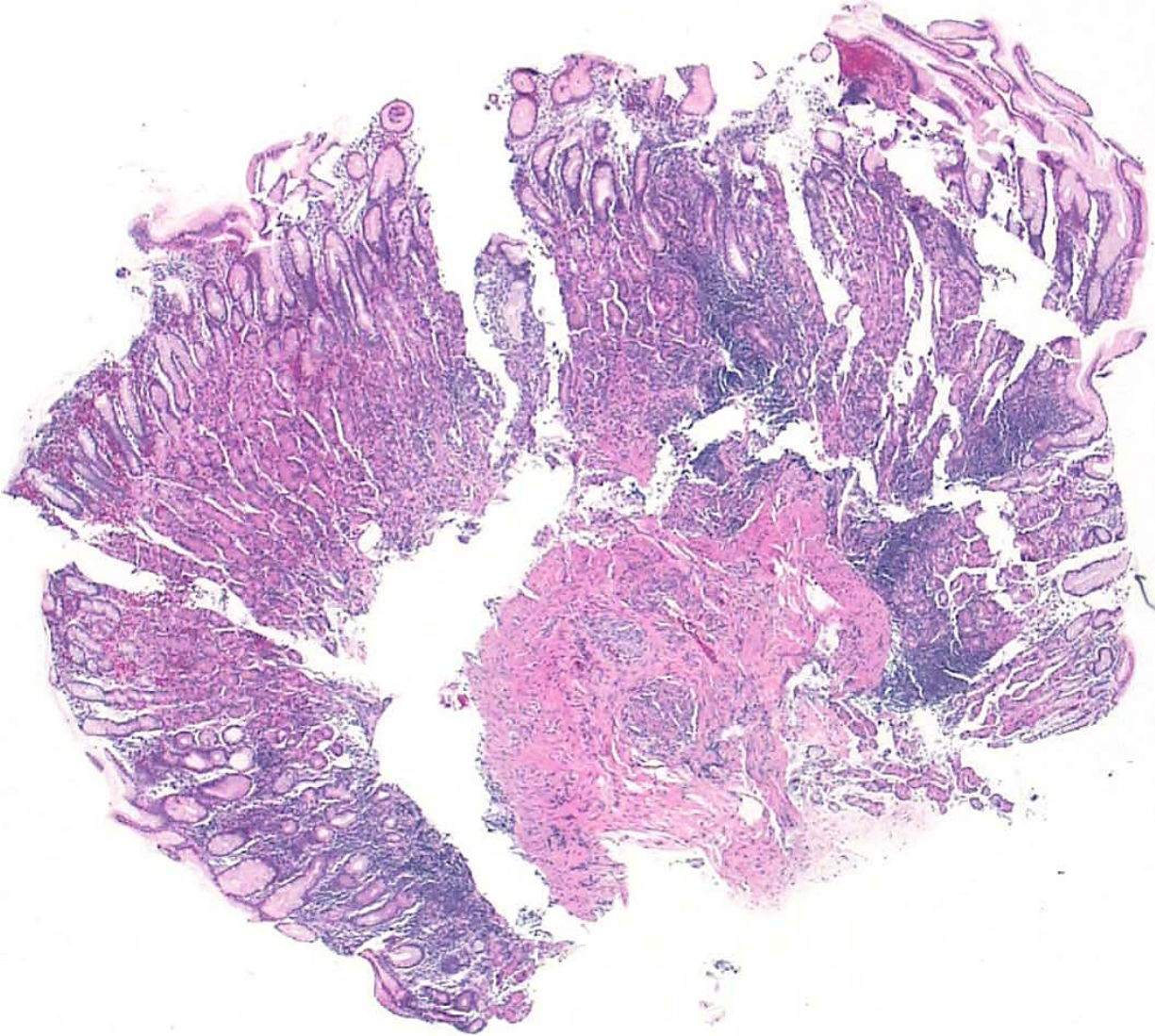
Clinical History

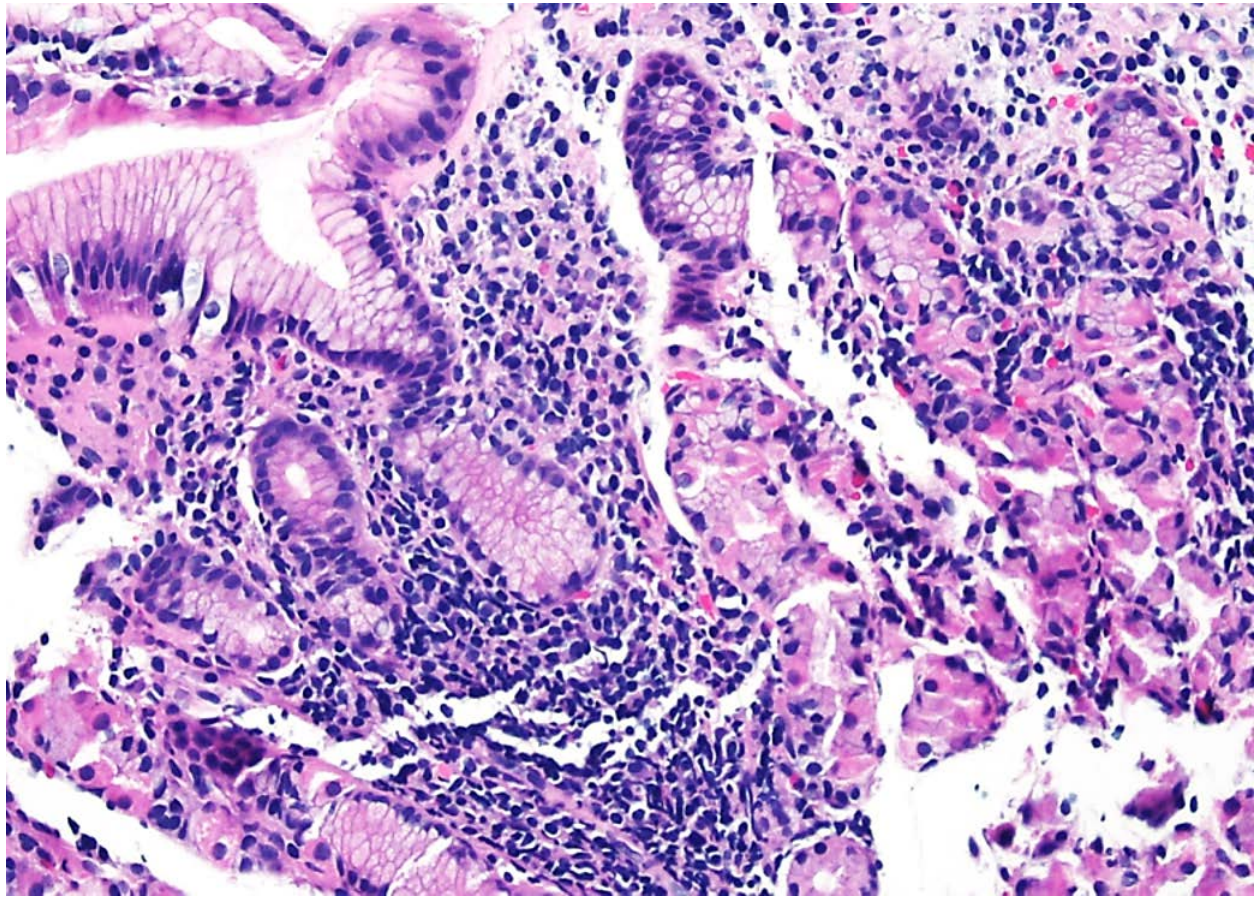
A 27-year-old male with no previous history presented for iron deficiency anemia. Upper GI endoscopy showed striped, mildly erythematous mucosa without bleeding in the gastric body and a single erythematous 2 to 3 mm papule with no stigmata of recent bleeding in the gastric fundus.

Figure 1.



Figure 2. H&E (low and high power)





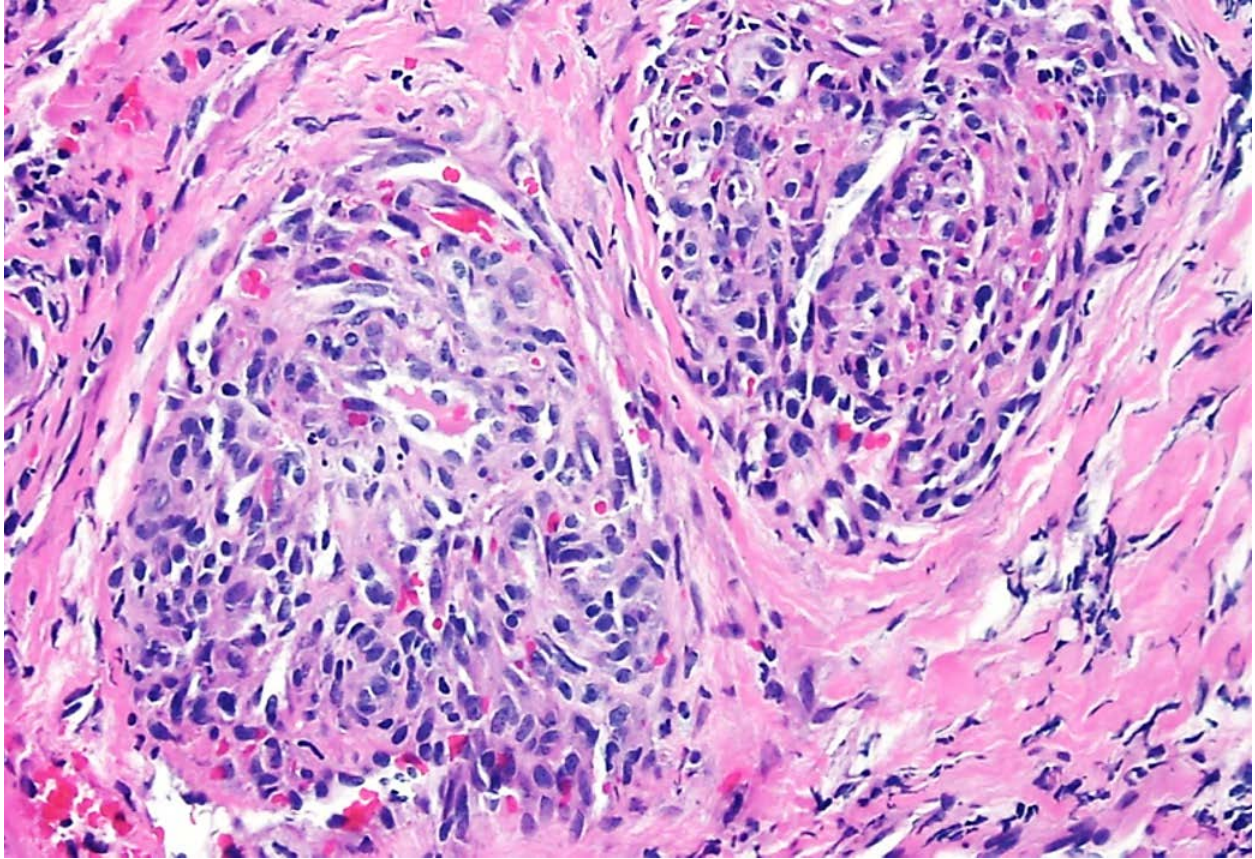


Figure 3. HHV-8 immunohistochemistry

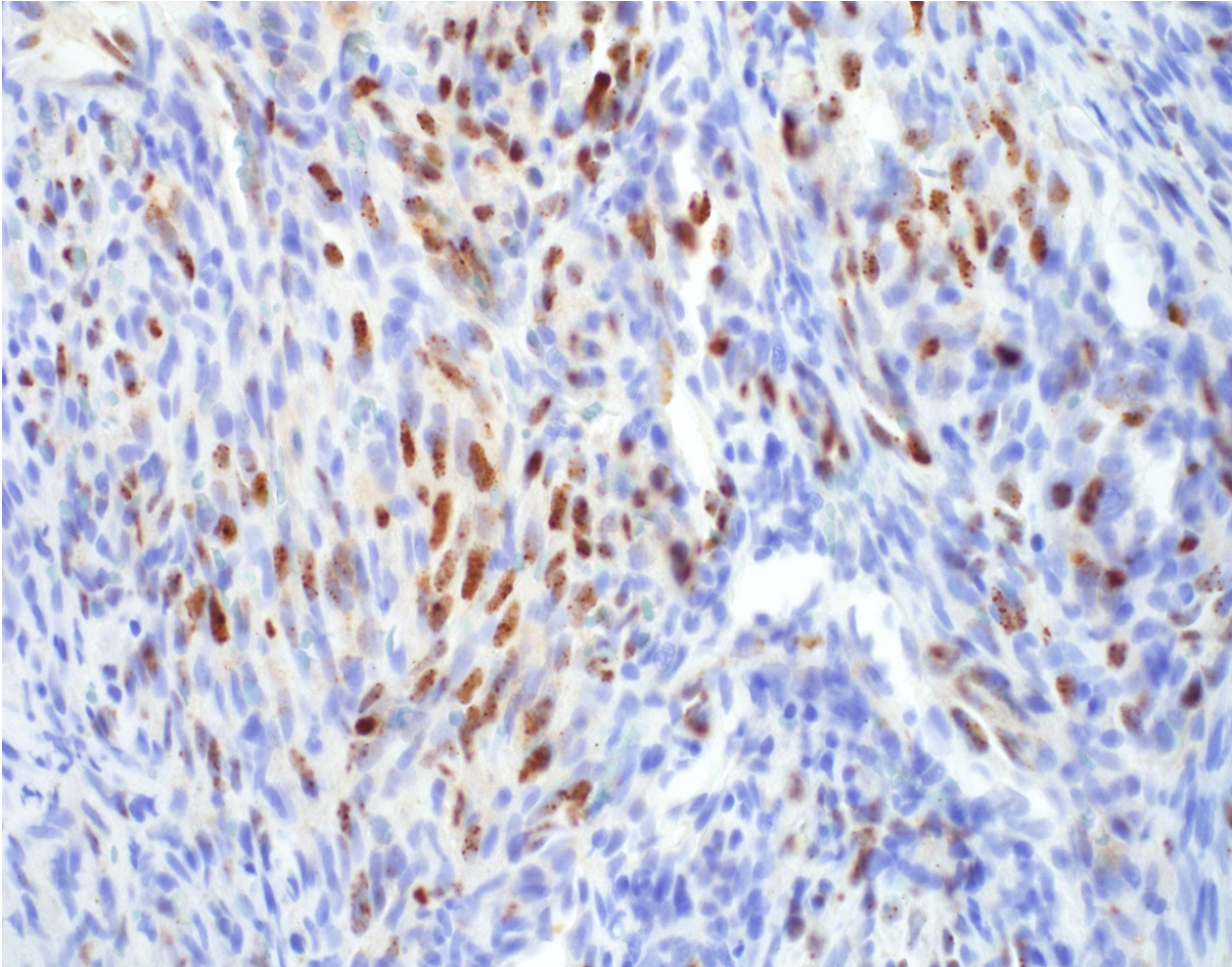
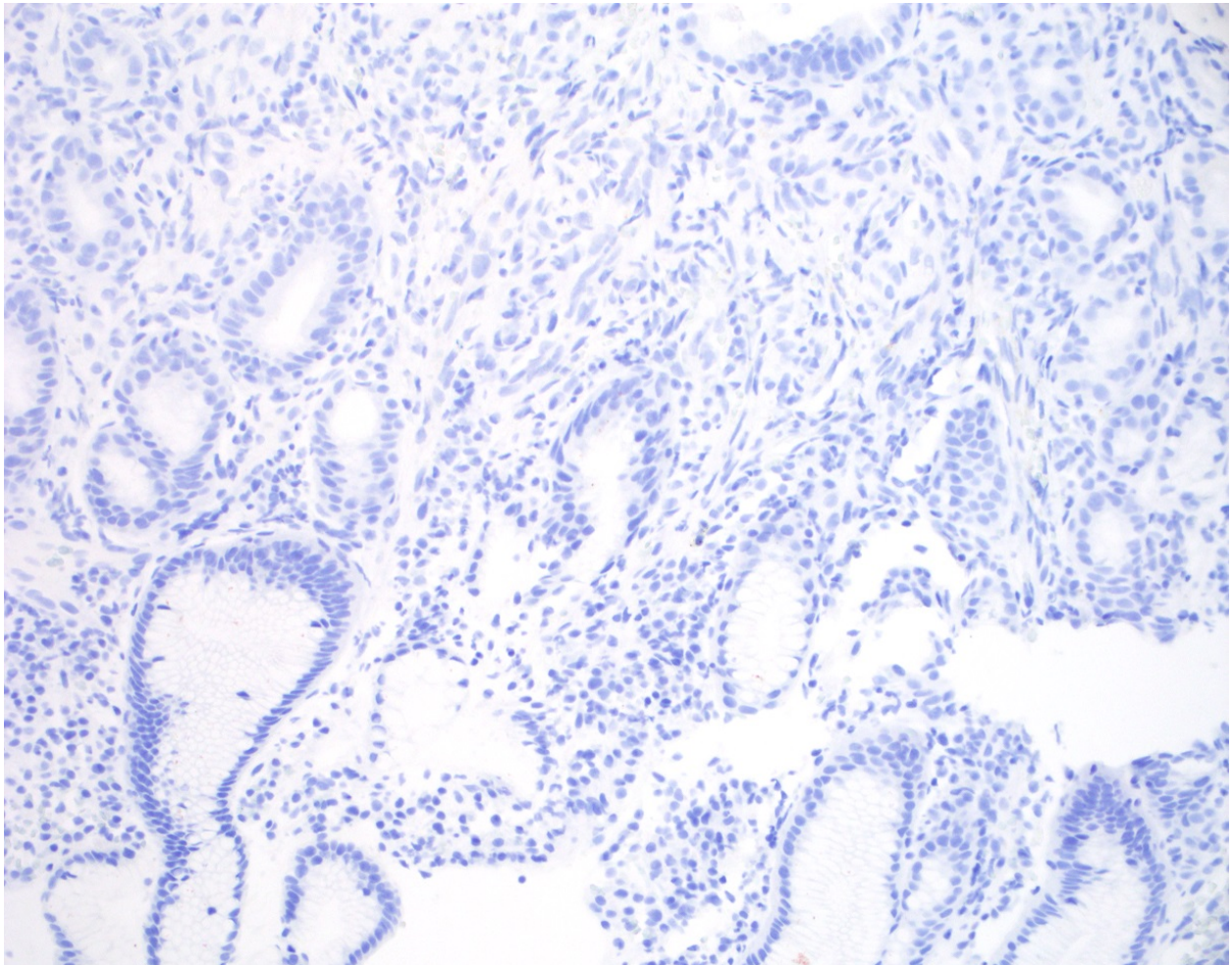


Figure 4. H. pylori immunohistochemistry



What is your diagnosis?

- A) Gastrointestinal stromal tumor
- B) Glomus tumor
- C) Spindle cell hemangioma
- D) Well-differentiated neuroendocrine tumor
- E) Kaposi sarcoma

Correct answer is:

E) Kaposi sarcoma

Discussion:

What is Kaposi's sarcoma?

Kaposi sarcoma (KS) is an uncommon vascular lesion caused by human herpes virus-8 (HHV-8). KS primarily affects patients with HIV/AIDS, as well as those who are otherwise immunosuppressed; endemic forms occur in Africa and the Mediterranean. The extent of the disease could vary from minimal mucocutaneous disease to multifocal systemic involvement, including the gastrointestinal tract.

Endoscopic and histologic features of KS

At endoscopy, early lesions look like mucosal erythematous changes; and more advanced lesions can present as a plaque/nodule, sometimes with central ulceration. Microscopically, the lesions are located primarily in the submucosa and consist of spindle cells and vascular-appearing endothelial-lined spaces. The spindle cells may show a wide range of nuclear pleomorphism; mitoses or apoptosis can be seen. Red blood cells fill some of the spaces between the spindle cells, which can be very subtle in some cases such as this one. Intracytoplasmic hyaline globules can be seen as well. The lamina propria can be filled with lymphoplasmacytic infiltrate, mimicking *H. pylori*-associated gastritis.

Immunohistochemical profile of KS

An immunostain for HHV8 is very helpful in confirming the diagnosis, particularly for equivocal cases. Vascular markers such as ERG, CD34, CD31, or von Willebrand factor may also be helpful but are not nearly as specific for Kaposi's as HHV8.

Differential diagnosis of KS

A) Gastrointestinal stromal tumor (GIST) can have spindle cell morphology, but GISTs do not show slit-like vascular channels, and immunostains for DOG-1 and CD117 (c-kit) are positive in GISTs. Of note, Kaposi's sarcoma can stain weakly for CD117 (c-kit) as well.

B) Glomus tumor commonly displays branching capillary-sized vessels lined by endothelial cells, which are surrounded by collars of uniform glomus cells in a hyalinized or myxoid stroma. Spindling of the stromal cells sometimes can be seen. Tumor cells are round with indistinct borders and sharply punched out nuclei in amphophilic to eosinophilic cytoplasm. Smooth muscle actin or muscle specific actin stains highlight glomus cells, and collagen type IV staining clearly illustrates the vascular pattern.

C) Spindle cell hemangioma shows spindled endothelial cells with slit-like channels but also has epithelioid cells. It usually lacks plasma cells and hemorrhage. HHV8 is negative.

D) Well-differentiated neuroendocrine tumors (NETs) can show a spindle cell morphology, but NETs typically have salt-and-pepper nuclei and are positive for neuroendocrine markers such as synaptophysin and chromogranin.

Clinical Outcome

Most patients are initially asymptomatic; however, KS with visceral involvement can ultimately be associated with poor prognosis. If the KS is associated with HIV or other immunosuppressive state, then the underlying disorder should be addressed (e.g., antiretroviral therapy for HIV). Ultimately, treatment is often palliative, and the presence or absence of symptoms determines the treatment regimen. The treatment options include antiretroviral therapy, chemotherapy, radiation therapy, or combination therapy.

Since a serologic test for HIV had been done on our patient after the diagnosis of Kaposi's sarcoma and was positive, the patient was subsequently treated with antiretroviral therapy.

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References

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