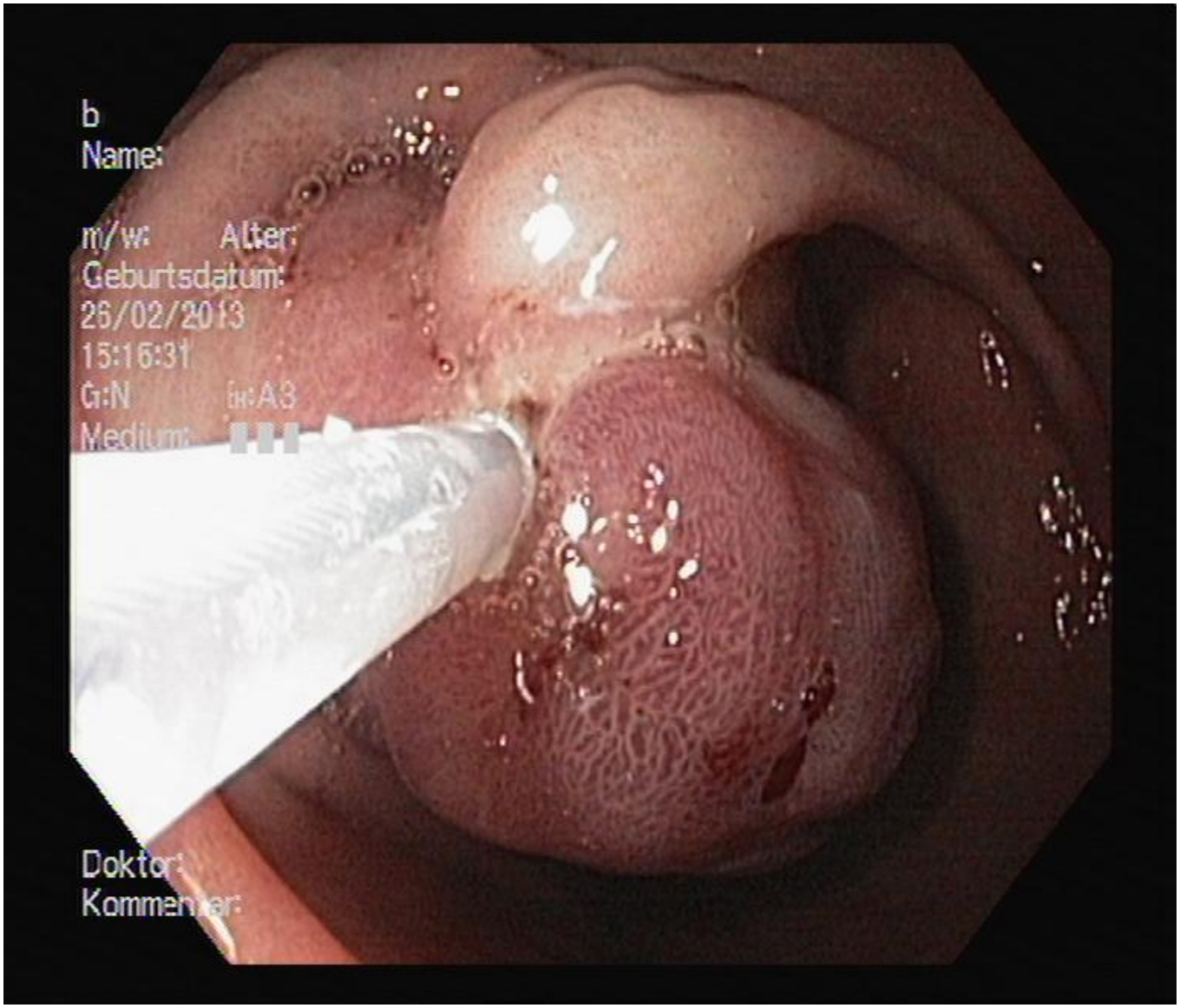
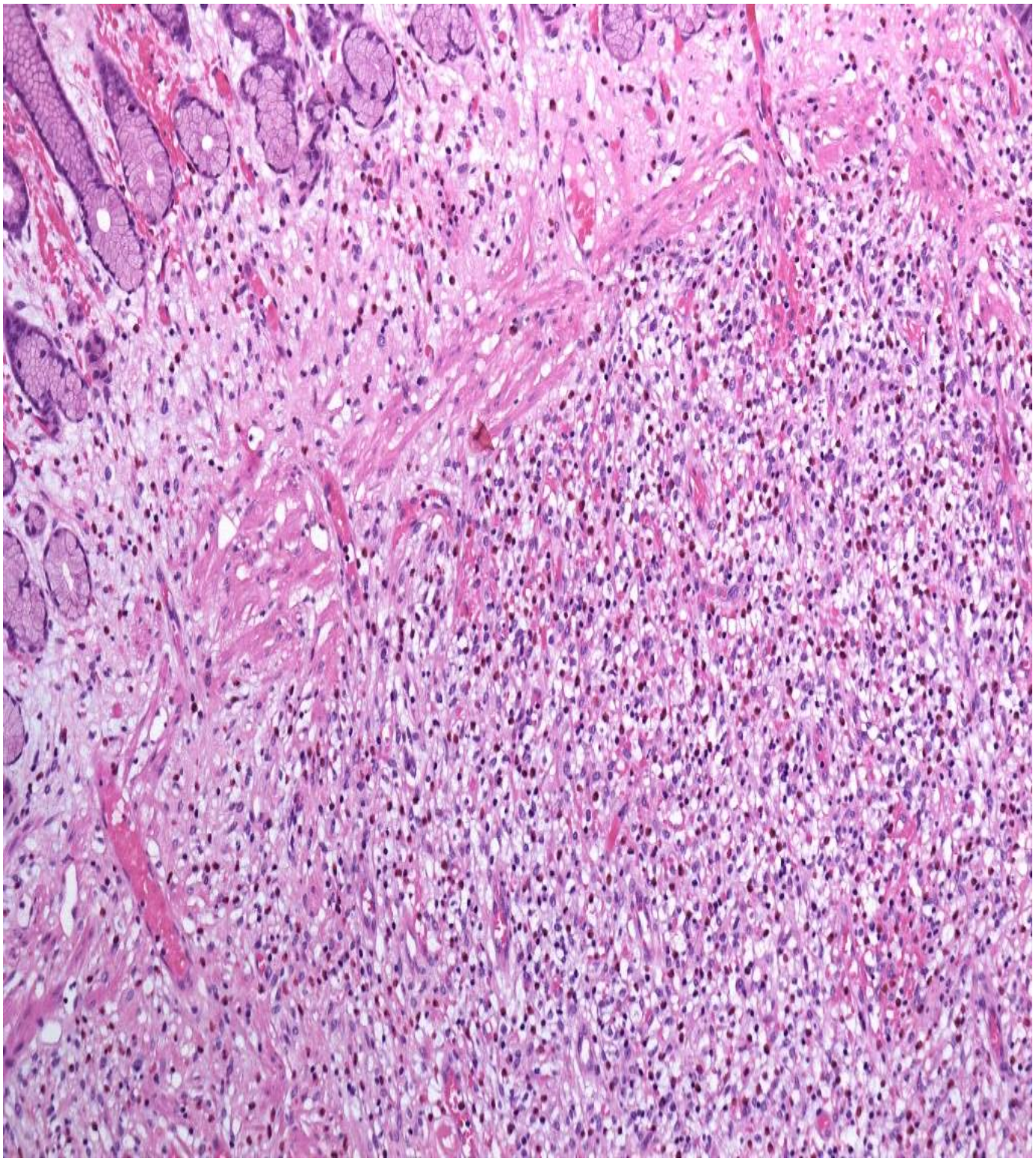
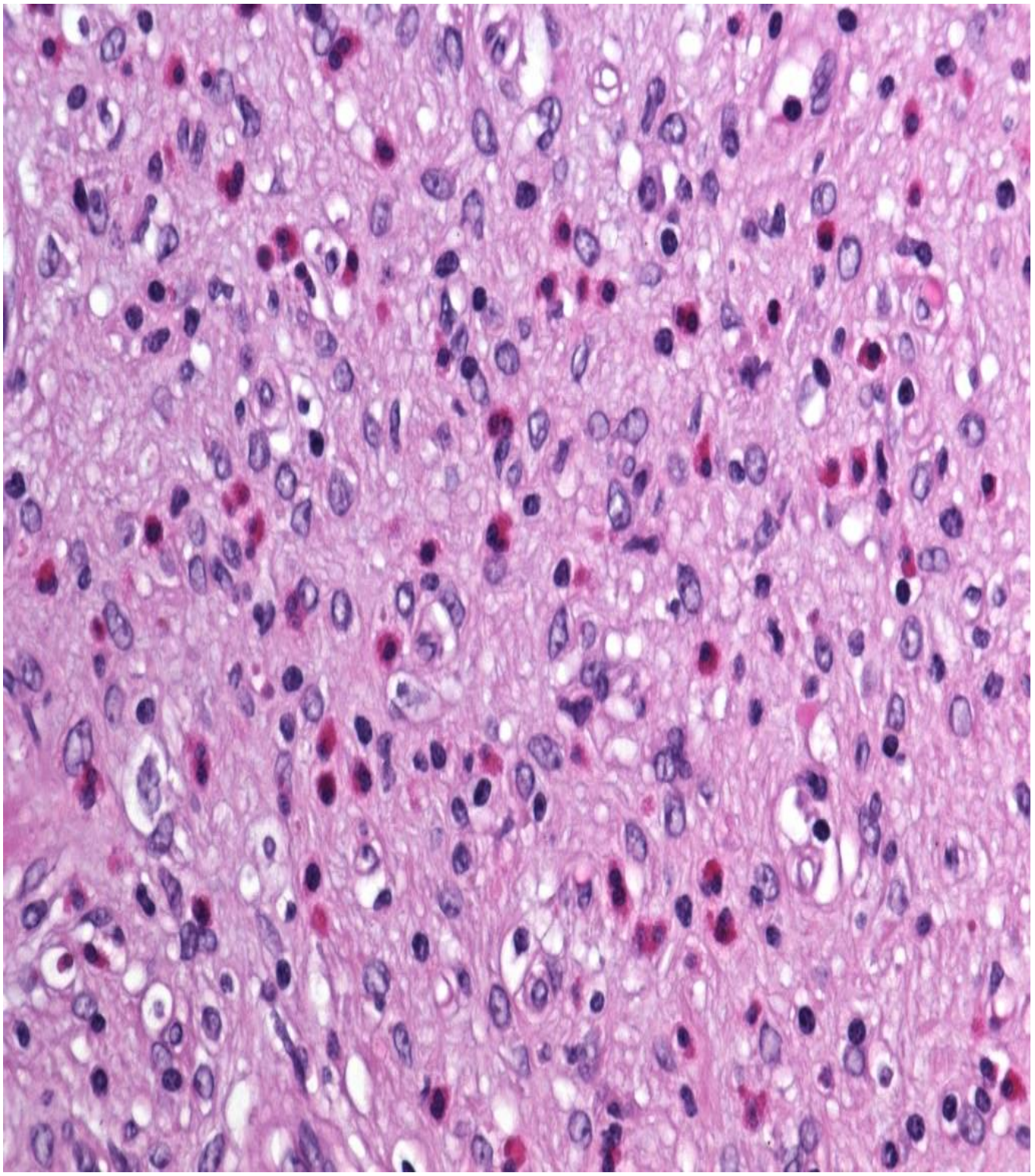
July 2013

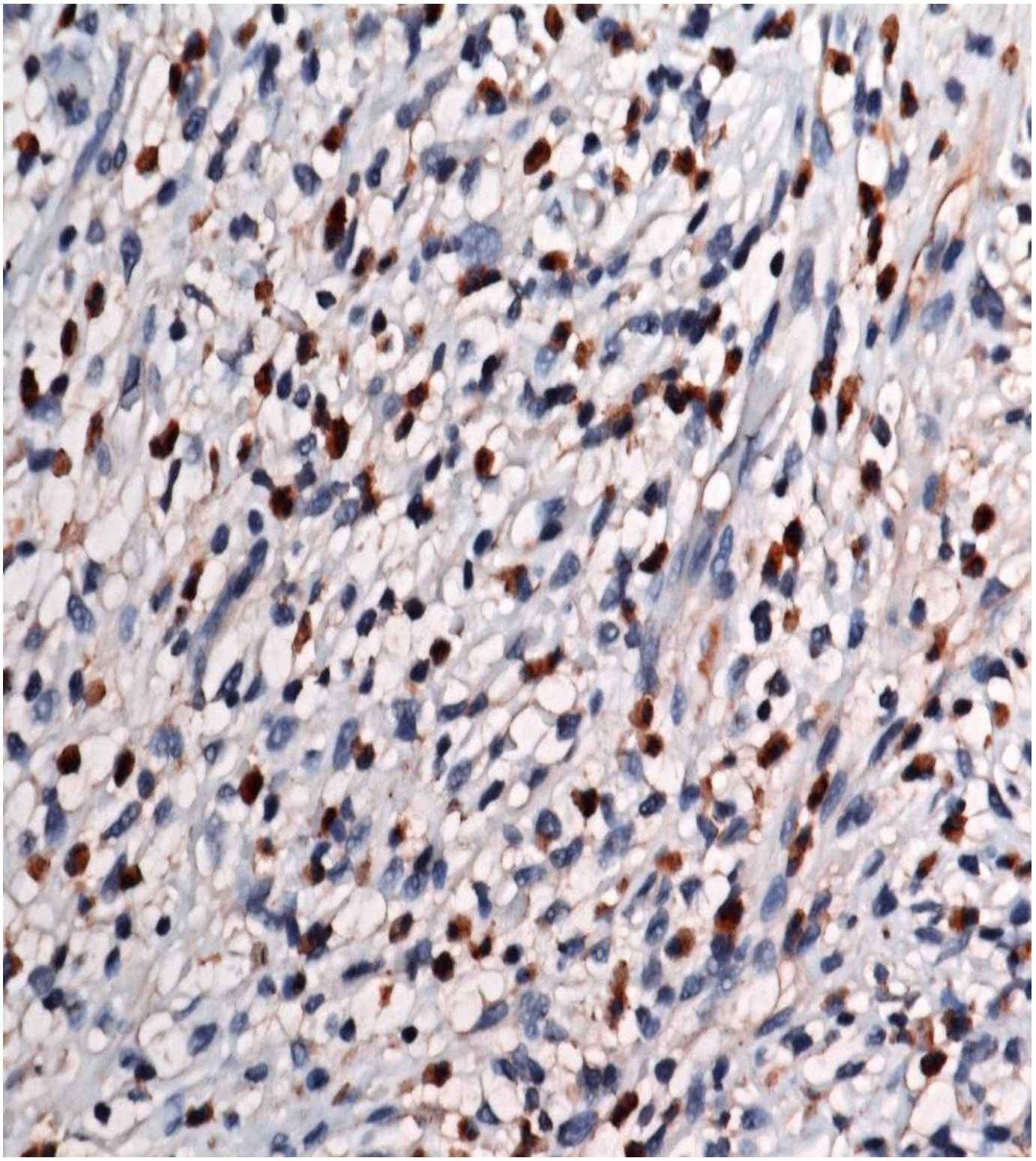
Irregular polyp in the prepyloric antrum of a 53-year-old female.

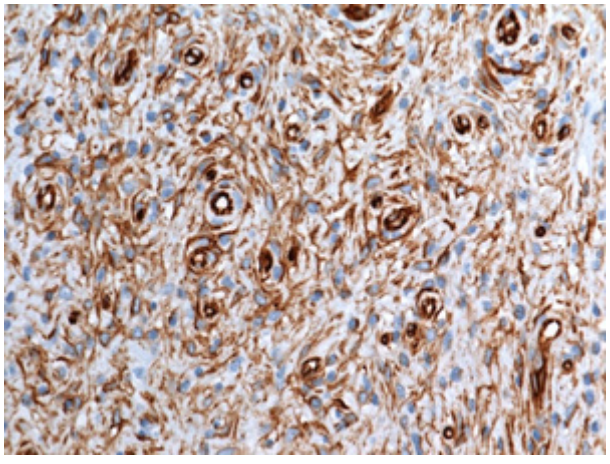
What is your diagnosis?

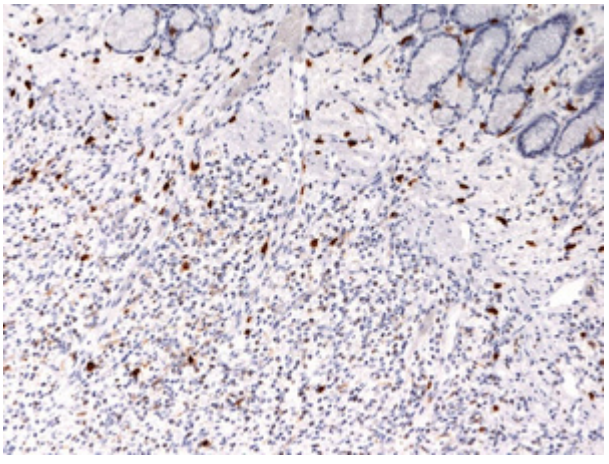












Diagnosis

Inflammatory fibroid polyp (Vanek’s tumour).

Comment

The polypectomy specimen shows a submucosal lesion of bland spindle cells with prominent vasculature which is densely infiltrated by numerous eosinophils (Panels B and C) which are positive for EPX, an antibody recognizing eosinophil peroxidase (Panel D). The lesion is strongly positive for CD34, indicating an “onion-skin” arrangement of cells around vessels (Panel E). The tumour cells are constantly negative for CD117 (KIT) which, however, stains intralesional mast cells (Panel F).

Inflammatory fibroid polyps are rare benign lesions that may occur throughout the whole gastrointestinal tract. Most often they are seen in the stomach, particularly the antrum. Large polyps have been reported to cause anaemia or gastric outlet obstruction. In the small bowel, they may lead to intussusception. Upon histology, inflammatory fibroid polyps show a proliferation of loose spindle cells arranged in short fascicles or whorled structures, commonly in an “onion-skin” arrangement around the abundant vessels. Usually large amounts of eosinophils are seen, but lesions with sparse numbers of eosinophils have also been described. In this case, the positive reaction with CD34 as well as the CD117 positivity of frequent intratumoral mast cells may be a source of confusion with gastrointestestibal stromal tumours (GISTs).

On the molecular level, inflammatory fibroid polyps are driven by activating mutations in the platelet-derived growth factor receptor alpha (PDGFRA) gene in a localization-specific pattern: exon 12 mutations predominate in the small intestine, while exon 18 mutations occur frequently in the stomach (also in our case, not shown). KIT mutations have not been detected.

For further reading

- Greenson JK. Gastrointestinal stromal tumors and other mesenchymal lesions of the gut. Mod Pathol. 2003;16:366-75.
- Huss S, Wardelmann E, Goltz D, et al. Activating PDGFRA mutations in inflammatory fibroid polyps occur in exons 12, 14 and 18 and are associated with tumour localization. Histopathology. 2012;61:59-68.
- Liu TC, Lin MT, Montgomery EA, et al. Inflammatory fibroid polyps of the gastrointestinal tract: spectrum of clinical, morphologic, and immunohistochemistry features. Am J Surg Pathol. 2013;37:586-92.

Presented by

Dr. Cord Langner, Graz, and Dr. Franz Siebert, St. Veit/Glan, Austria