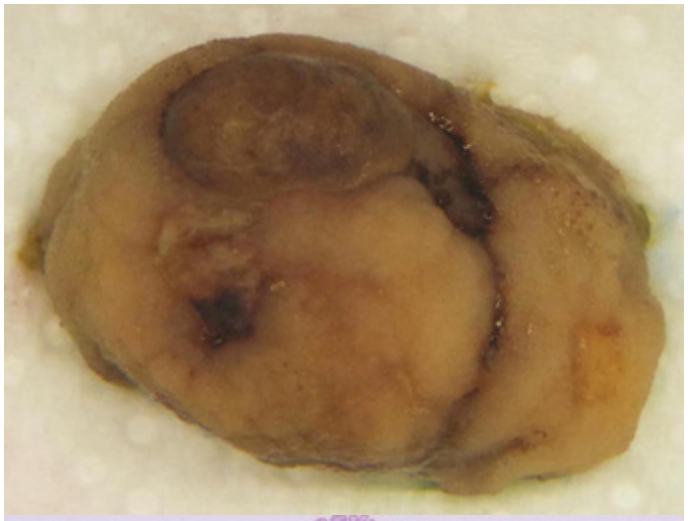
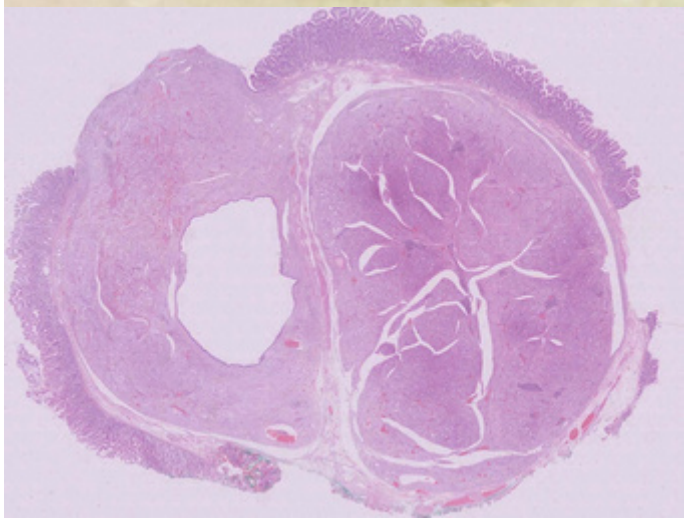
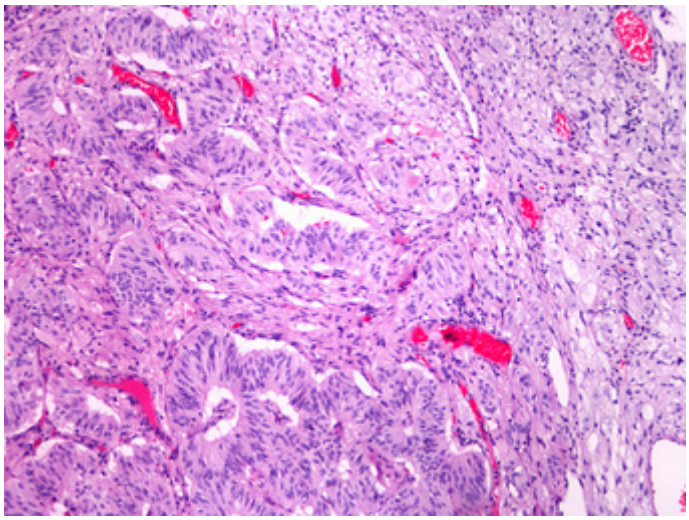
September 2021

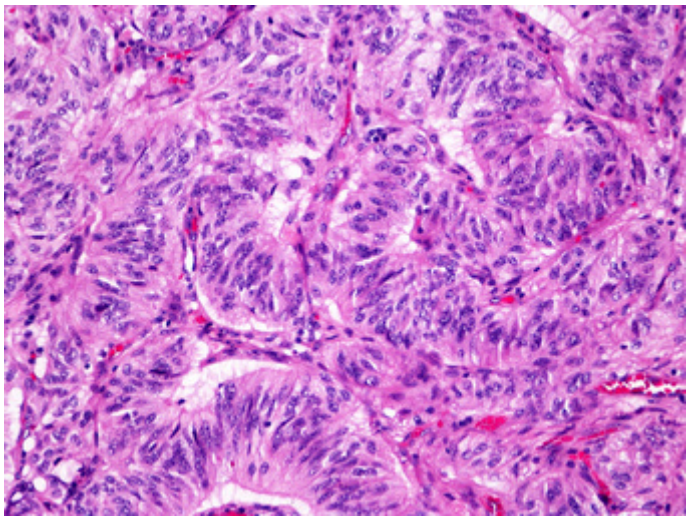
A 67-year-old male with a bleeding tumor of the duodenum

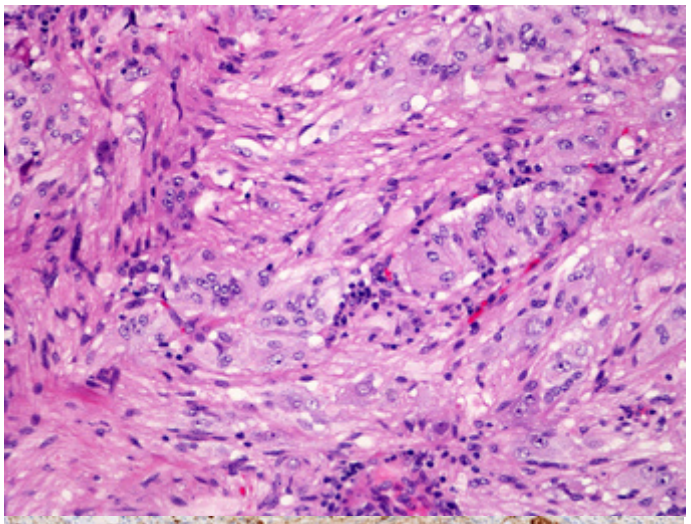
What is your diagnosis?

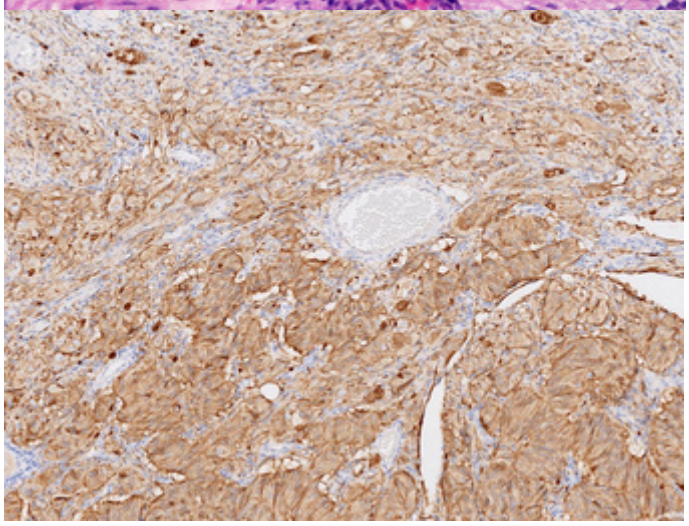


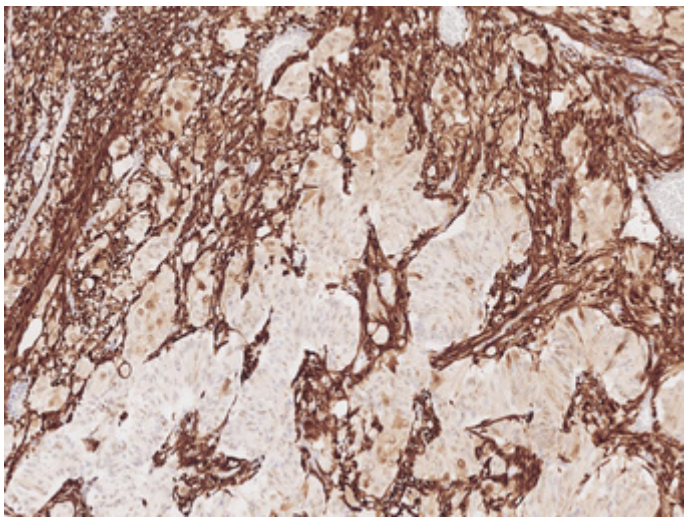


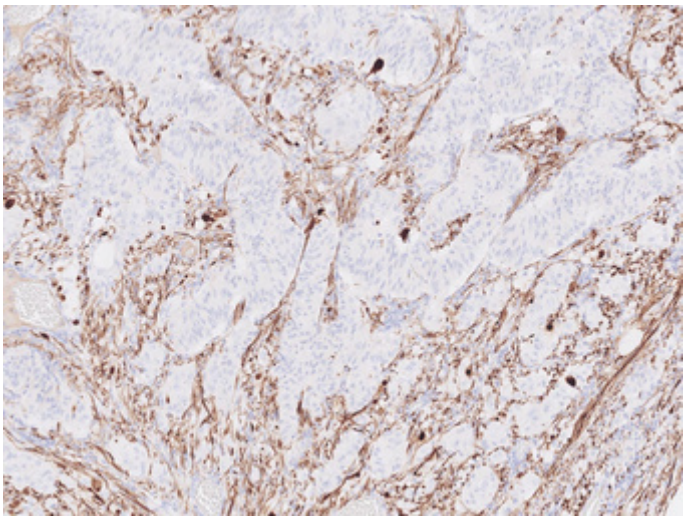












Diagnosis:

Gangliocytic paraganglioma.

Comment:

The tumor was excised. It measured 2.5 x 1.5 cm and was located in the duodenal wall, covered by intact mucosa, macroscopically resembling gastrointestinal stromal tumor (GIST) (Panel A). Histopathology showed a tumor in the submucosa and muscularis propria of the duodenal wall (Panel B), composed of a mixture of neuroendocrine tumor corresponding to NET G1, ganglion cells with the characteristic prominent nucleoli and abundant cytoplasm, and spindle-shaped Schwann cells (Panels C-E), with no atypia or necrosis and with a low proliferation activity (Ki67 index approx. 2%). The NET component stained immunohistochemically for neuroendocrine markers chromogranin and synaptophysin (Panel F) and pancreatic polypeptide, while the ganglion and Schwann cells were S100-positive (Panel G) and NF-positive (Panel H); these two markers were negative in the NET component. The tumor did not express CD117.

Gangliocytic paraganglioma (GP) is a rare tumor, composed of a mixture of neuroendocrine cells, ganglion cells and Schwann cells. It usually arises in adults, mostly in duodenum, but rare cases have been described in the jejunum, stomach and other locations. GP may present with gastrointestinal bleeding, abdominal pain, duodenal obstruction, pancreatitis and/or anemia. In most patients, it is characterised by indolent behavior, and excision of the tumor is curative. However, metastases to the lymph nodes and liver have been reported in approximately 10% of cases, and there is a single report of a tumor-associated death.

Differential diagnosis includes GIST, nerve sheath tumors, ganglioneuroma and NET. Rarely, it may mimick malignant tumors, for example clear cell sarcoma-like tumors and carcinoma. On the basis of morphology and immunohistochemistry, it is mostly not difficult to make the diagnosis of GP, if all components are present. If not, it can be misdiagnosed as NET, particularly in small endoscopic biopsies. It has been recommended to use pancreatic polypeptide and progesterone receptors which is usually positive in GP but not in NET. This distinction is important as NETs can behave more aggressively than GP.

For further reading:

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- Mahmoud S, Salami M, Salman H. A rare serious case of retroperitoneal paraganglioma misdiagnosed as duodenal gastrointestinal stromal tumor: a case report. BMC Surg. 2020; 20(1):49.
- Okubo Y, Yoshioka E, Suzuki M, et al. Diagnosis, pathological findings, and clinical management of gangliocytic paraganglioma: a systematic review. Front Oncol. 2018; 8:291.
- Okubo Y. Gangliocytic paraganglioma: an overview and future perspective. World J Clin Oncol. 2019; 10(9):300-2.

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