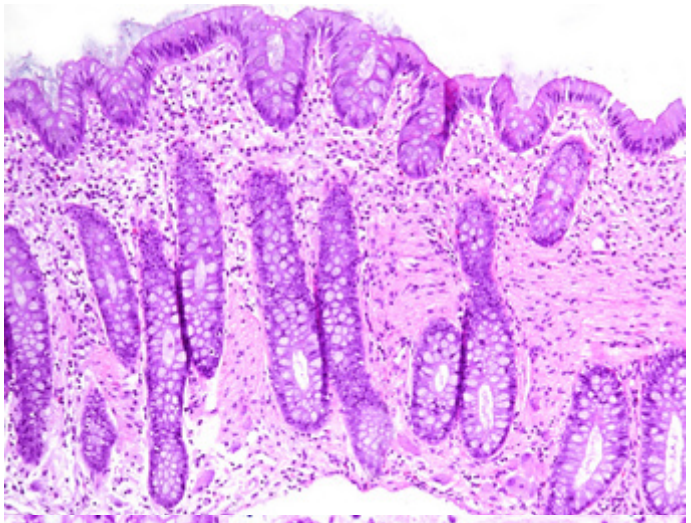
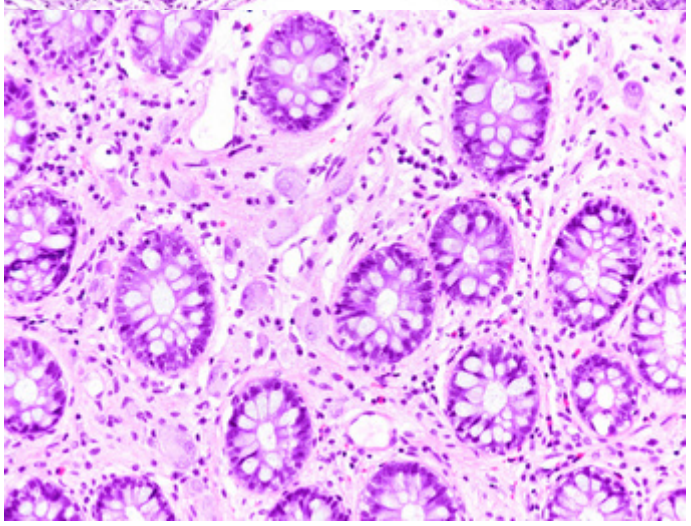
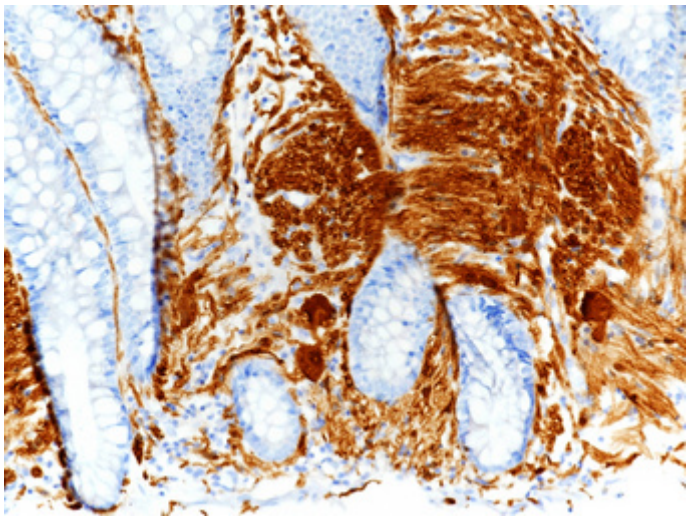
April 2017

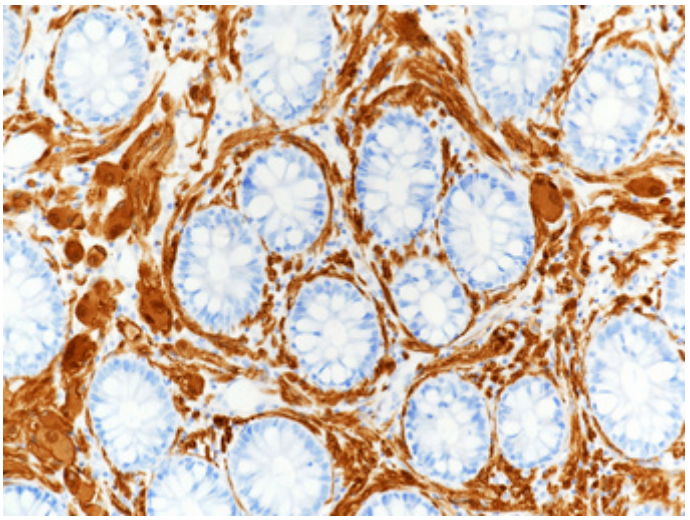
Solitary polyp in the sigmoid colon of a 61-year-old female.

What is your diagnosis?









Diagnosis:

Solitary polypoid ganglioneuroma.

Comment:

The lamina propria is occupied by a schwannian stromal proliferation with clusters of ganglion cells (panels A-B). Immunohistochemistry reveals a diffuse S100 protein expression in the Schwann and ganglion cells (panels C-D), prompting diagnosis of ganglioneuroma.

Although ganglioneuromas can occur at any level of the tubular gastrointestinal tract, they are most commonly situated in the colorectum. Lesions can be categorized into three distinct subtypes: solitary polypoid ganglioneuromas, ganglioneuromatous polyposis, and diffuse ganglioneuromatosis. Patients with solitary polypoid ganglioneuromas are usually asymptomatic and do not have an increased risk of systemic manifestations, the polyps are usually incidentally found, sessile/pedunculated and measure only a few millimeters. Ganglioneuromatous polyposis, and diffuse ganglioneuromatosis can be associated with a variety of disorders e.g. Cowden syndrome, juvenile polyposis syndrome, neurofibromatosis type 1, multiple endocrine neoplasia syndrome.

Differential diagnosis includes mucosal perineurioma, mucosal Schwann cell hamartoma, schwannoma, and gangliocytic paraganglioma.

Perineuriomas are characterized by concentric, whorled spindle cell proliferation around crypts, and are usually associated with an epithelial hyperplasia and serrated features. Using immunohistochemistry perieuriomas are negative with S100 protein and SMA, but express claudin-1, collagen IV, GLUT-1 and weakly EMA. Schwann cell hamartomas are ill-defined, bland spindle cell lesions that are characteristically restricted to the mucosal layer. They consist of S100 positive Schwann cells, without the ganglion cell component characteristic of ganglioneuromas. Gastrointestinal schwannomas and gangliocytic paragangliomas are usually well circumscribed and submucosal. In contrast to their CNS counterparts gastrointestinal schwannomas are usually surrounded by a rim of lymphocytic infiltration, do not show the typical Antoni A and Antoni B regions, and also lack a true capsule. Gangliocytic paragangliomas have areas identical to ganglioneuromas, but they also have a second component in keeping with the morphology of a paraganglioma.

In some instances ganglion cells can also be found in the lamina propria of IBD patients as a consequence of chronic mucosal injury, but these cases invariably lack the schwannian stromal background.

For further reading:

- Srinivasan R, Mayle JE. Polypoid ganglioneuroma of colon. Dig Dis Sci. 1998; 43: 908-9.

- Chan OT, Haghighi P. Hamartomatous polyps of the colon: ganglioneuromatous, stromal, and lipomatous. Arch Pathol Lab Med. 2006; 130: 1561-6.
- De Leon MP. What clinicians wish to know about benign colorectal polyps: an operative classification. Pathol Res Pract. 2014; 210: 645-8.

Presented by:

Dr. Bence Kővári, Szeged, Hungary, and Dr. Cord Langner, Graz, Austria.