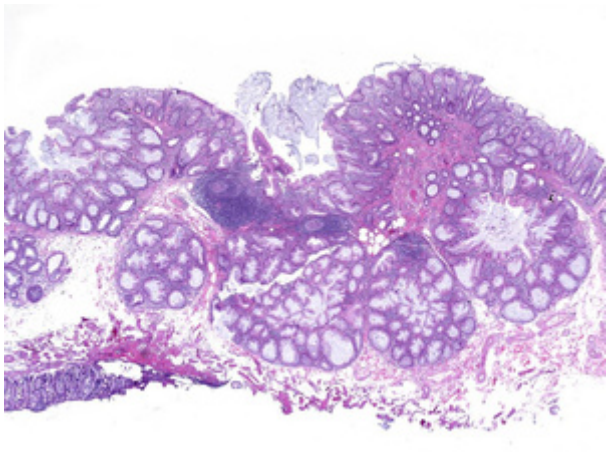
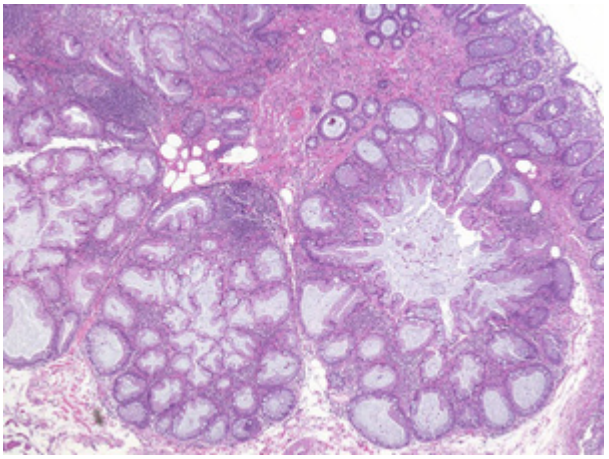
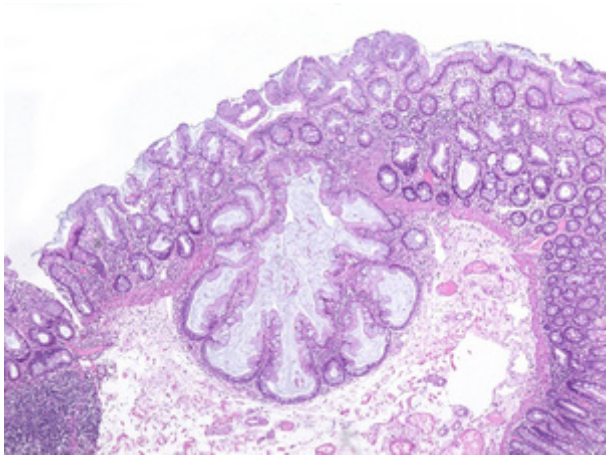
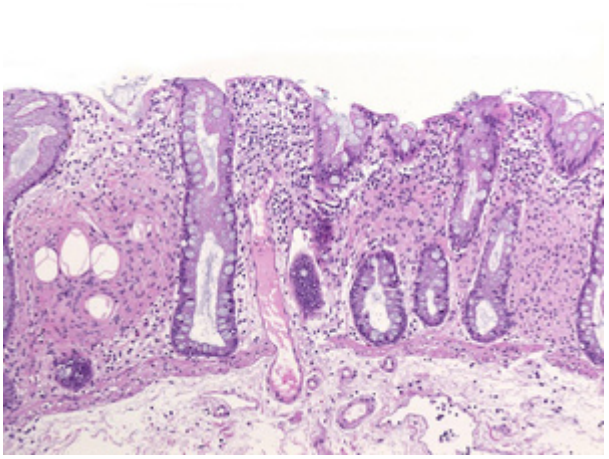
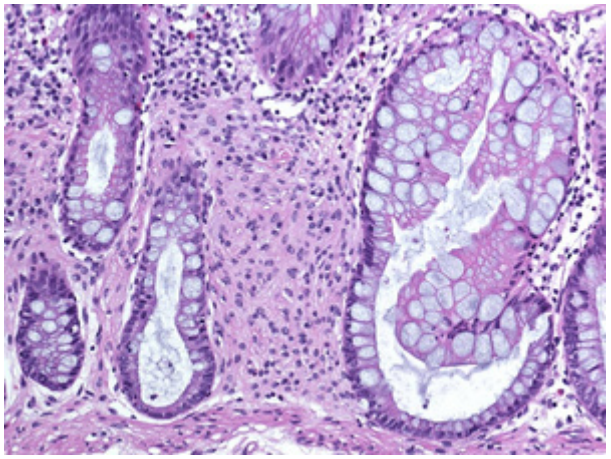
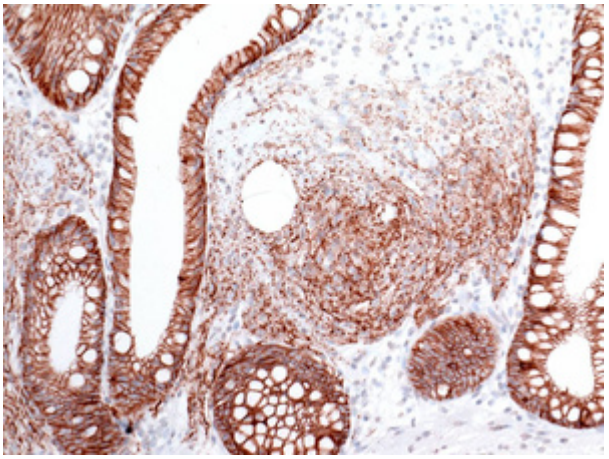
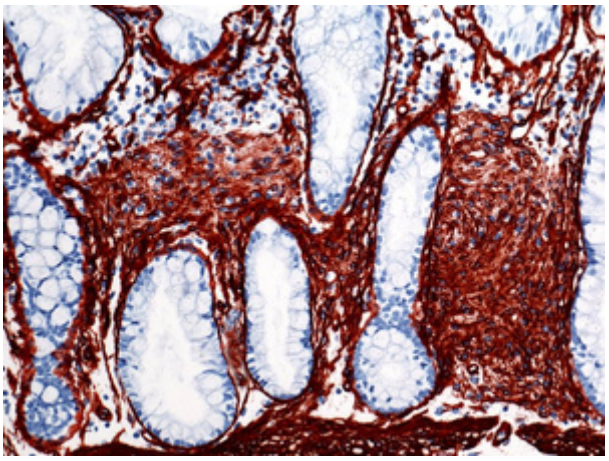
August 2014

Sessile serrated adenoma with perineurial-like stromal proliferation (fibroblastic polyp).

What is your diagnosis?



Diagnosis

Sessile serrated adenoma with perineurial-like stromal proliferation (fibroblastic polyp).

Comment

Different magnifications (Panels A-C) show a sessile serrated adenoma with characteristic “L-” and “T-shaped” crypts and marked epithelial misplacement (herniation through the muscularis mucosae), but no cytological dysplasia. The lateral portion of the lesion (Panel D) is characterized by an unusual stromal component with bland and monomorphic spindle cells. On high power (Panel E), these elements have eosinophilic cytoplasm and indistinct cell borders. Upon immunohistochemistry, the spindle cells are strongly and diffusely positive for perineural markers such as Claudin-1 (Panel F) and GLUT-1, and they are also positive for Collagen IV (Panel G) and EMA.

Fibroblastic polyps (or hyperplastic polyps with perineurial-like/perineurioma-like stroma) represent about 0.2 % of all colonic polyps and occur most commonly in women, with a mean age of 55 years. They are usually small lesions (2 to 9 mm), located in the rectosigmoid colon, mostly detected as an incidental finding.

These mucosal polyps consist of two distinct and closely merged components: a glandular-epithelial one, characterized by serrated appearance without dysplasia and a stromal one, characterized by bands of monotonous spindle cells, with pale eosinophilic indistinct cytoplasm, oval nuclei with vescicular chromatin and inconspicuous nucleoli, in the absence of nuclear atypia, mitoses or necrosis. The epithelial component shows the morphology of typical hyperplastic polyps or, rarely, sessile serrated adenomas. The stromal sheets displace the colonic epithelium and surround the crypts in an onion-skin like pattern. The stromal component shows an expansive-type growth pattern and, in some cases, a distinct separation to the superficial epithelium by a thin layer of lamina propria is evident.

The perineurial-like stromal cells are positive for perineurial markers such as Claudin-1, GLUT-1, Collagen IV, and EMA. They are also positive for Vimentin, sometimes for CD 34 and negative for SMA, Protein S100, Desmin and CD117. Differential diagnosis includes mucosal prolapse-associated polyps, schwannomas, fibromatosis, lamina propria fibrosis, GISTs, inflammatory fibroid polyps (Vanek tumors), juvenile-type hamartomatous polyps, inflammatory myoglandular polyps, leiomyomas.

The pathogenesis of the lesion is largely unknown. It is of note that the stromal and the epithelial components are closely related. At first it was suggested that the serrated epithelium could be a reactive phenomenon related to the stromal proliferation. Recently, however, BRAF mutations typical for serrated polyps, i.e. hyperplastic polyps and sessile serrated adenomas were reported, indicating a neoplastic nature of the epithelial component. Interestingly, BRAF mutations were present only in perineuriomas with crypt serration, but not in perineuriomas without crypt serration. To turn the coin around: Is the perineurial-like stromal

proliferation the reactive (non-neoplastic) phenomenon related to the epithelial proliferation or is the stromal component neoplastic in nature, i.e. the independently proliferating component of a true mixed or hybrid tumour? Until now, this question has not been sufficiently answered.

For further reading

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- Pai RK, Mojtahed A, Rouse RV, Soetikno RM, Kaltenbach T, Ma L, Arber DA, Plesec TP, Goldblum JR, Pai RK. Histologic and molecular analyses of colonic perineurial-like proliferations in serrated polyps: perineurial-like stromal proliferations are seen in sessile serrated adenomas. Am J Surg Pathol. 2011;35:1373-80.
- Groisman GM, Hershkovitz D, Vieth M, Sabo E. Colonic perineuriomas with and without crypt serration: a comparative study. Am J Surg Pathol. 2013;37:745-51. Presented by: Dr. Francesca Sarocchi, Genova, Italy, and Dr. Cord Langner, Graz, Austria.

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