Perineurioma of the sigmoid colon

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DESCRIPTION

A 24-year-old man presented to the surgical outpatient clinic with a 1-year history of intermittent, painless, fresh rectal bleeding associated with defaecation. The blood was noticeable as streaks on the toilet paper. His bowel habit remained otherwise unchanged. He reported no weight loss or family history of bowel cancer. His medical history was unremarkable.

On examination, there were no clinical features of anaemia, jaundice, cyanosis, clubbing, oedema or cervical lymphadenopathy. Proctoscopy revealed an internal haemorrhoid at the 7 o'clock position. Blood test results included a haemoglobin level of 158 g/L, mean corpuscular volume of 87.0 fL and mean corpuscular haemoglobin concentration of 331 g/L, all of which were within normal limits.

On flexible sigmoidoscopy, a 3 mm polyp within the sigmoid colon was identified. This was a well-circumscribed, flat elevated lesion (Paris classification type 0–IIa) (figure 1). Its surface features on narrow band imaging were not typical of an adenomatous or hyperplastic polyp. Given its unusual appearance, the polyp was removed.

Histology showed it to be a bland mesenchymal lesion of the colonic wall. The lamina propria appeared to be expanded by a vaguely nodular proliferation of spindle cells percolating between distorted non-serrated crypts (figure 2A). There was no cytological atypia, mitotic activity or necrosis. Very small blood vessels were scattered throughout. There was no perivascular condensation or eosinophilia to suggest an inflammatory fibroid polyp. Immunohistochemical staining showed widespread although weak positivity for epithelial membrane antigen (figure 2B). DOG1, CD34, CD117, smooth muscle actin, desmin, S100 and cytokeratin AE1/3 were all negative. The features were consistent with a perineurioma.

Colonic perineuriomas, or fibroblastic polyps, are rare benign mucosal lesions comprising a proliferation of bland spindle cells expressing markers of perineurial differentiation.[1] They are usually found in the rectum and sigmoid colon, although they have been reported proximal to the splenic flexure.[2] With an estimated incidence of 0.1%–1.46% of all colonic polyps, they tend to be discovered incidentally in patients undergoing routine screening colonoscopy.

Endoscopically, they appear as solitary, sessile, well-circumscribed mucosal lesions and range in size from 0.2 cm to 1.5 cm. Characteristic histological features include spindle cell proliferation within the lamina propria which often separates and distorts the colonic crypt architecture.

Two variants are recognised: those with and those without accompanying serrated epithelium; the former may superficially resemble hyperplastic polyps or sessile serrated lesions. The pathogenesis of the perineurial-type stroma is poorly understood. There is increasing evidence to suggest that the epithelial component of serrated fibroblastic polyps harbours BRAF mutations which may trigger epithelial–mesenchymal interactions, causing fibroblast transdifferentiation and proliferation. The development of the less common non-serrated variant, of which this case is an example, has yet to be determined.[3] It is possible that the two variants represent entirely separate biological entities.

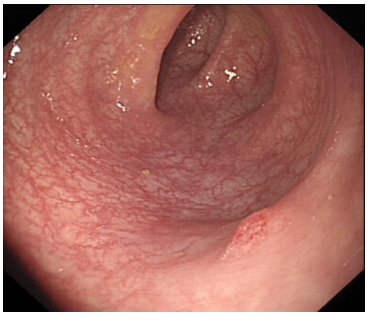

Figure 1 Flexible sigmoidoscopy. Gross appearance of the polyp under white light.

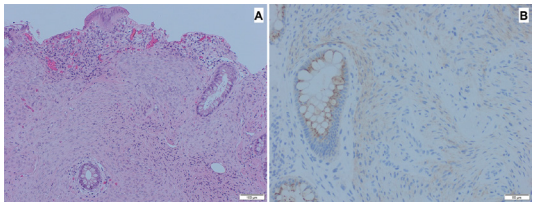

Figure 2 Photomicrograph. (A) Nodular spindle cell lesion percolating between distorted but non-serrated colonic crypts (H&E stain, ×100 magnification). (B) Weak staining in the spindle cells (epithelial membrane antigen stain, ×200 magnification).

Learning points

- Colonic perineuromas are rare benign mucosal lesions; however, their characterisation is important in determining colonoscopic surveillance.
- Decisions about follow-up surveillance colonoscopy should be based on the nature and size of the accompanying serrated epithelial component (if present), according to existing guidelines for serrated colorectal polyps in general.
- There are no reported cases of recurrence or metastases.

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To our knowledge, there have been no reported cases of recurrence or metastases of colonic perineuriomas. However, the neoplastic nature of the serrated variant means that they should be classified based on the criteria used for serrated colorectal polyps in general, making postpolypectomy surveillance in some cases appropriate.[3]

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