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Case Reports

Colonic Ganglioneuroma

Report of a Case in a Patient with Neurofibromatosis, Multiple Colonic Adenomas and Adenocarcinoma

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Summary. A 73 year old woman with cutanous neurofibromatosis developed colonic carcinoma. The resected colon also contained multiple tubular adenomas and a polypoid ganglioneuroma. Multiple neurofibromas were seen during the operation over the serosal surface of the small intestine. Other cases of colonic ganglioneuromas and of combined neurogenic and epithelial colonic tumours are reviewed.

Key words: Colon – Ganglioneuromas – Adenomas – Adenocarcinoma – Neurofibromatosis

Introduction

Ganglioneuromas of the peripheral nervous system are benign tumours composed of nerve fibres and mature ganglion cells (Harkin and Reed 1969). Few cases have been described in the alimentary tract. In 1957 Dahl et al. reported a case from the duodenum and reviewed 11 previously recorded cases from the stomach (2 cases), ileum (2 cases) appendix (5 cases) and ileum and large intestine (2 cases). Since then, the English literature searched contained another duodenal (Gemer and Feuchtwanger 1966) and six more large intestinal ganglioneuromas (Gherardi 1961; Gleason et al. 1962; Donelly et al. 1969; Raszkowski and Hufner 1971; Brodey and Hoover 1974; Bibro et al. 1980). We here report a further case of colonic ganglioneuroma which was present in a patient with cutaneous and intestinal neurofibromatosis (NF). The patient had concomitant colonic multiple adenomas and adenocarcinoma.

Case Report

A 73 year old white woman with NF presented with three weeks' history of abdominal distention, colicky pain, vomiting and alternating diarrhoea and constipation. There was no relevant past history and no family history of large bowel neoplasms or NF.

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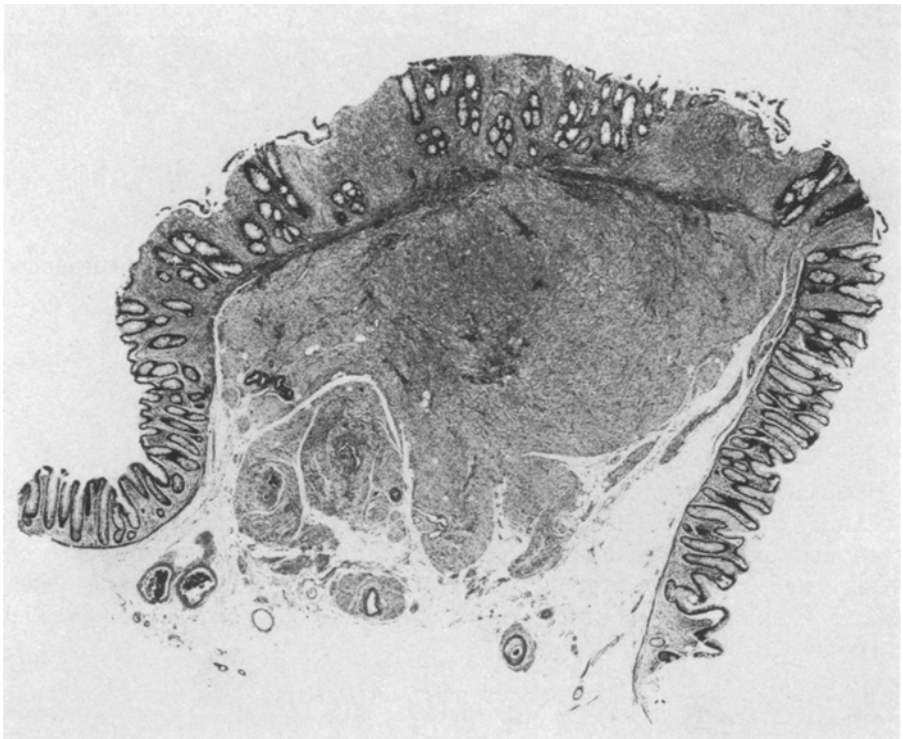

Fig. 1. Colonic ganglioneuroma. The lesion affects the mucosa and submucosa. Note atrophy of glands and mucosal ulceration. H&E. ×9

On examination the patient had abdominal distention and numerous skin neurofibromas and café-au-lait patches. The skin tumours ranged in size between 0.5 and 1.5 cm and were present on the face, arms, abdomen, back and legs. Abdominal X-ray confirmed the presence of dilated loops of small and large bowel. A barium enema showed an irregular stricture at the splenic flexure.

At laparotomy a constricting carcinoma was found at the splenic flexure with proximal gross dilatation of the colon and caecum. No metastases were present but several nodules, having the same appearance and consistency as the cutaneous neurofibromas, were present on the serosal surface of the small bowel. An extended right hemicolectomy was performed. The skin and small intestinal tumours were not biopsied.

Histopathology

The resected specimen was 65 cm long and consisted of stump of terminal ileum, caecum, appendix, ascending and transverse colon. The tumour was 3.0 cm from the distal plane of resection, had an ulcerated surface and measured 3.5 cm in maximum dimension. It encircled the bowel lumen completely and infiltrated the whole thickness of its wall. One sessile and three pedunculated polyps were present in the dilated colon 34, 32, 27 and 6 cm proximal to the main tumour. The largest polyp, at 34 cm, was 3.0 cm long and up to 0.5 cm in diameter.

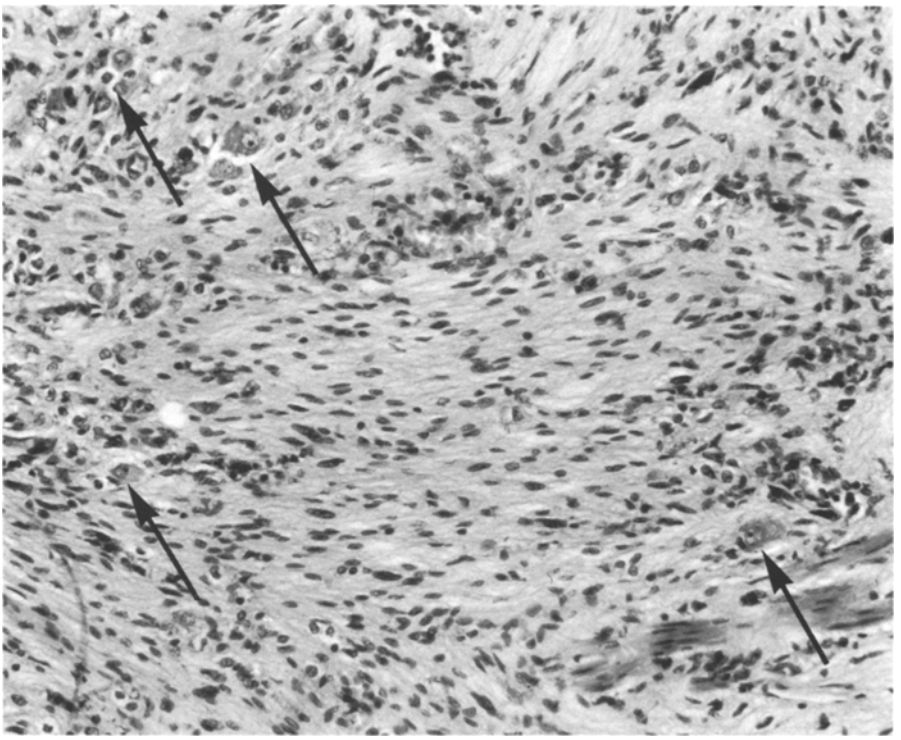

Fig. 2. Colonic ganglioneuroma. High power view to show the parallel bundles of spindle shaped cells and fibres, and scattered ganglion cells (arrows). H&E. ×160

Histologically, the main tumour was a moderately differentiated adenocarcinoma with a rich fibrous tissue stroma. It infiltrated the adjacent pericolic fibrofatty tissue but all the 14 lymph nodes dissected from the specimen were free of tumour.

The pedunculated polyps were tubular adenomas composed of long stalks formed of normal colonic mucosa and submucosa, with the adenomatous changes present only at the tips of the lesions. The adenomatous glands showed moderate degrees of dysplasia and a few Paneth cells were present in one lesion.

The sessile polyp was present in the dilated part of the colon 32 cm proximal to the carcinoma and measured 1.0 cm in maximum dimension. Histologically the lesion was well defined but not encapsulated, and was present in the mucosa and submucosa (Fig. 1). It consisted of compact interlacing bundles of spindle-shaped cells and fibres with interspersed ganglion cells (Fig. 2). The latter were large ovoid cells with abundant cytoplasm and large ovoid vesicular nuclei, each with a single nucleolus. These cells were scattered singly and in groups in the mucosa and submucosa. The glands in the affected areas were atrophied, few in number and widely separated from each other. The overlying surface epithelium was ulcerated in some areas. The muscularis mucosae was mostly

intact except for small gaps through which bundles of tumour fibres were seen extending from one side to the other (Fig. 1). These appearances are consistent with the diagnosis of ganglioneuroma (Harkin and Reed 1969).

Sections from the bowel wall distal to the tumour and from the small intestine were normal.

Random sections from the dilated colon showed marked thinning out of the wall. This was mostly due to atrophy of the muscularis propria. In some areas the muscle layer was completely replaced by dense fibrous tissue reaction which extended into the submucosa. Ganglion cells were absent in the fibrotic areas but present outside them.

Discussion

Patients with NF may develop intestinal neurogenic tumours. These are usually in the form of neurofibromas (Schmincke 1956; Grodsky 1957; Levy and Khatib 1960; Wilson and Anderson 1960; Ghrist 1963; Staple et al. 1964; Raszkowski and Hufner 1971; Case records of the Massachusetts General Hospital 1974) which may show evidence of malignant change (Levy and Khatib 1960; Ghrist 1963). In few patients with NF, ganglioneuromas have been reported in the small intestine and appendix (Schmincke 1956; Dahl et al. 1957; Ghrist 1963). Ours seems to be the first case of a ganglioneuroma of the colon to be reported in a patient with NF.

Harkin and Reed (1969) in their discussion of ganglioneuromas mention that they are occasionally found in the subcutaneous tissues of patients with NF. Perhaps the intestinal tract should also be considered a possible, though rare, site of ganglioneuromas in these patients. Bearing in mind that they may also occur in the intestinal tract as isolated lesions in the absence of NF.

The concomitent presence of NF and colonic epithelial tumours in our patient is probably the result of a chance association; as both conditions are relatively common. However, it may be worth mentioning here that our case brings to five the number of patients in whom both colonic epithelial and peripheral neurogenic tumours were reported. Levy and Khatib (1960) and Raskin and Dodd (1976) reported two patients with NF who had concurrent epithelial tumours. These were an adenomatous polyp and juvenile polyposis respectively. Bibro et al. (1980) reported the only other case in which a colonic ganglioneuroma was associated with colonic carcinoma, and Donnelly et al. (1969) reported a case of combined ganglioneuromas and juvenile polyposis. Neither of these patients had cutaneous NF.

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Note Added in Proof

A further case of colonic adenocarcinoma associated with diffuse ganglioneuromatosis of colon has been reported after submitting this manuscript.

Snover DC, Weigent CE, Sumner HW (1981) Diffuse mucosal ganglioneuromatosis of the colon associated with adenocarcinoma. Am J Clin Pathol 75:225–229