Inflammatory Fibroid Polyp of the Caecum in a Patient with Neurofibromatosis
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Abstract
Introduction: Inflammatory fibroid polyp of the large intestine is uncommon. To our knowledge, this condition has not been reported in a patient with neurofibromatosis. Clinical Picture: In this report, a 66-year-old woman with neurofibromatosis was found to have a large polyp in the caecum. Treatment: Right hemicolectomy was performed because of the size of the polyp. Outcome: Pathological examination showed that the polyp was an inflammatory fibroid polyp. Conclusion: Clinicians should be aware that inflammatory fibroid polyps could be one of the many manifestations of neurofibromatosis in the gastrointestinal tract.

Adenomatous polyps had also been resected from the sigmoid colon and splenic flexure. A small bowel series showed no other polyps in the small bowel. A right hemicolectomy was performed through a small transverse incision because of the considerable anaesthetic risks. The opened specimen revealed a polyp 3.5 cm in diameter (Fig. 2). There were no postoperative complications and the patient was discharged well 7 days later. She was well and without recurrence 1 year after the operation.

The patient was also diagnosed to have von Recklinghausen disease (neurofibromatosis) from numerous skin neurofibromata and cafe au lait spots. Six neurofibromata have been excised between the years 1980 and 1986. One of her 3 daughters also has the disease. Warfarin was prescribed since cerebral emboli caused a grand mal epileptic fit 9 years ago. A total hysterectomy and bilateral salpingo-oophorectomy had also been performed for menorrhagia.

Histological examination of the polyps revealed fibroblast-like spindle cells intermingled with large numbers of mixed inflammatory cells (Fig. 3). There were also numerous eosinophils and marked vascularity. Some of the

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vessels also exhibited thick hyalinised walls. The spindle cells were negative for neural marker (S-100) and smooth muscle marker (desmin). These features were consistent with an inflammatory fibroid polyp.

Discussion

Helwig and Ranier introduced the term “inflammatory fibroid polyp” in 1953. The aetiology of inflammatory fibroid polyp remains elusive. It is thought to be a result of chronic irritation and inflammation and may represent a peculiar form of granulation tissue formation.

The disease has no sex predilection and peak prevalence was in the 5th and 6th decades of life. Lesions are almost invariably solitary and occur most commonly in the stomach. In the present case, the lesion was found in a person of slightly older age of 66 and was located in an uncommon site, the caecum. On the other hand, the appearance, the size and the microscopic features of the polyp fit well with the descriptions in the literature. On macroscopic examination, specimens appear as sessile or polyloid lesions localised to the bowel wall and protrude into the lumen. The size varies from 2 to 4 cm. It often has a homogeneous appearance akin to a leiomyoma. Microscopic analysis shows abundant fibroblasts arranged in whorls around blood vessels with prominent inflammatory cells, particularly eosinophils.

Inflammatory fibroid polyps can present in many different ways including pain, bleeding, anaemia, persistent polyp intussusception and intestinal obstruction. Obstruction tends to be the commonest presentation. Also, inflammatory fibroid polyp can mimic carcinoma macroscopically and thus surgery is usually performed to remove the lesion. In our patient, the polyp was detected on follow-up colonoscopy for anaemia and suspected colonic cancer.

Von Recklinghausen’s disease is an autosomal dominant hereditary condition that may affect the gastrointestinal tract in approximately 25% of cases. Such gastrointestinal involvement occurs in 3 main forms: a) hyperplasia of the submucosal and myenteric nerve plexuses and mucosal ganglioneuromatosis, leading to disordered bowel motility; b) gastrointestinal stromal tumours with different degrees of neural and smooth muscle differentiation; and c) distinctive carcinoid tumour of the periampullary region which may be associated with phaeochromocytoma. There have been no case reports of neurofibromatosis with inflammatory fibroid polyps. We postulate that the inflammatory fibroid polyp may be a result of repeated chronic inflammation arising from disordered bowel motility found in neurofibromatosis.
In conclusion, inflammatory fibroid polyp of the colon is uncommon. This is, to our knowledge, the first time this lesion has been reported in a patient with neurofibromatosis. Thus, in a patient with neurofibromatosis, clinicians should be aware that inflammatory fibroid polyp could be one of the many manifestations in the gastrointestinal tract. In our patient, inflammatory fibroid polyp can mimic carcinoma on endoscopy and surgery was performed to remove the lesion.

REFERENCES