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Case Report

Solitary polypoid ganglioneuroma in the sigmoid colon: a rare finding during colonoscopy

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ABSTRACT

Ganglioneuromas (GN) are rare, benign tumours of the autonomic nervous system which are seldom encountered in the gastrointestinal tract. They may occur as solitary lesions or more commonly as multiple lesions, also known as ganglioneuromatosis. Endoscopically they have no identifiable phenotypic characteristics and therefore diagnosis can only be confirmed through histological analysis. Microscopically they are composed of ganglion cells, nerve fibres and Schwann cells and show S-100 protein immunoreactivity. Clinically, there are no specific symptoms eluding towards the diagnosis of solitary ganglioneuromas of the gastrointestinal tract with most patients remaining asymptomatic. Due to their rarity, no guidelines current exists for solitary colonic GNs, however consensus exists that endoscopic resection is curative with no evidence of recurrence following total excision. This case gives an account of a solitary colonic ganglioneuroma in the sigmoid colon encountered during a screening colonoscopy in a 47-year-old male.

Keywords: Ganglioneuroma, Polypoid ganglioneuroma, Sigmoid colon, Colonoscopy, S100 tumour

INTRODUCTION

Ganglioneuroma's (GN) are benign neuroectodermal tumours which originate from undifferentiated neural crest cells including ganglion cells, nerves fibres and glial cells.[1,2] They can occur anywhere along the sympathetic chain, but have a predilection for the head, neck and adrenal glands, and only very rarely present in the gastrointestinal tract.[2,3] Shekita et al categorised gastrointestinal ganglioneuromas into three morphological subtypes: polypoid ganglioneuromas, diffuse ganglioneuromatosis and ganglioneuromatous polyposis.[1-3] Polypoid ganglioneuromas which are mostly solitary, have no association with genetic syndromes such as juvenile polyposis, neurofibromatosis 1 or multiple endocrine neoplasia IIB syndrome (MEN-IIB), and are only very rarely encountered.[2] Solitary ganglioneuromas of the gastrointestinal tract are usually asymptomatic however may present with symptoms including abdominal pain, constipation, ileus, obstruction or weight loss.[2,3] They are frequently discovered incidentally during routine endoscopy, surgery or autopsy.[4]

CASE REPORT

A 47-year-old Caucasian male with an unremarkable past medical history underwent an elective colonoscopy for per rectal bleeding and anaemia. He had intermittent diarrhoea occurring approximately once a fortnight and a small quantity of per rectal bleeding. He denied a history of abdominal pain or unintentional weight loss. There was no personal or family history of colorectal malignancy or genetic syndromes such as neurofibromatosis 1, Cowden syndrome or MEN-IIB syndrome. The colonoscopy revealed fourth degree haemorrhoids, diverticular disease and a 20 mm flat unusual appearing polyp in the distal sigmoid colon just above the rectosigmoid junction. The polyp was tattooed

and resected using hot snare (see Figure 1-3). Histopathological evaluation of the polyp revealed a benign ganglioneuroma. The neural proliferation extended around nerves and vessels of the submucosa and extended to deep diathermied margins. Spindle cell and ganglion cell proliferation was seen and the lesion demonstrated S100 immunoreactivity.

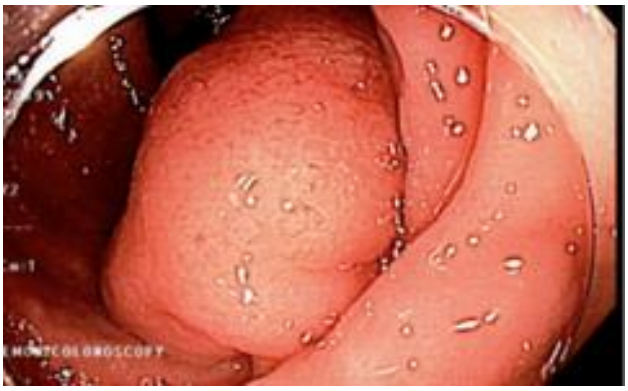

Figure 1: Endoscopic photograph of the 20 mm distal sigmoid polyp.

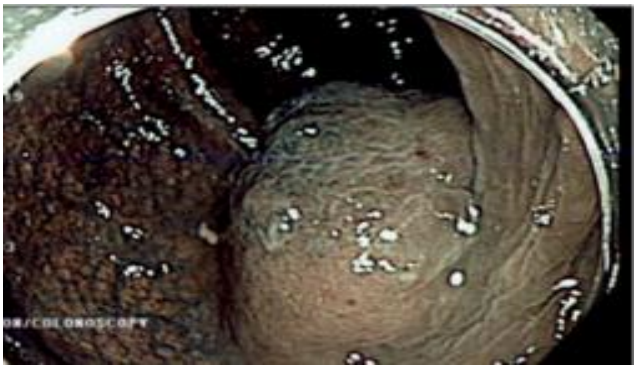

Figure 2: Endoscopic photograph of the 20 mm distal sigmoid polyp under narrow-band imaging.

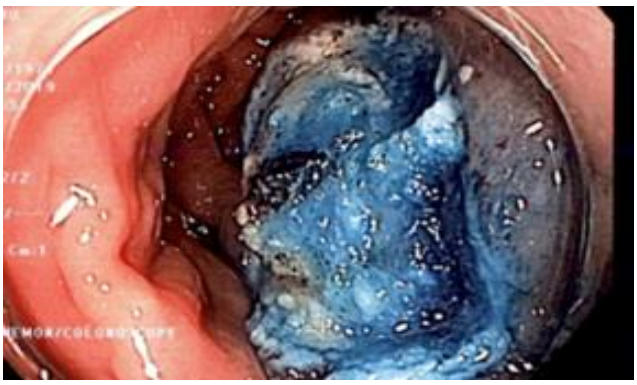

Figure 3: Endoscopic photograph of the 20 mm distal sigmoid polyp after tattoo.

DISCUSSION

Ganglioneuromas (GNs) are benign hamartomatous tumours of neural crest origin which arise from sympathetic or peripheral nerves.[2,5] These tumours may occur anywhere along the paravertebral sympathetic plexus but are most commonly found in the retroperitoneal, mediastinal or cervical regions.[6] GNs of the gastrointestinal tract are extremely rare and are infrequently found in the colon.[6]

Gastrointestinal ganglioneuromas occur in two settings: as solitary isolated lesions (polypoid ganglioneuromas) and syndromically as multiple lesions that either produce multiple exophytic polyps (ganglioneuromatous polyposis) or poorly demarcated transmural proliferations (diffuse ganglioneuromatosis).[2,4,7] In solitary examples, polypoid ganglioneuromas are generally small (<2 cm), sessile or pedunculated polyps that endoscopically are difficult to distinguish from hyperplastic polyps, adenomas or juvenile polyps.[1-3,8] Ganglioneuromatous polyposis are multiple (usually more than 20), sessile or pedunculated and display greater variability than sporadic ones, ranging in size from 1 to 2.2 cm.[2,3] Diffuse ganglioneuromatosis (DG) presents as disseminated, nodular lesions often up to 17cm in size that frequently extend transmurally to involve the myenteric plexus.[2,3,7] They can be poorly demarcated and distort the architecture of the surrounding tissue.[7] Most cases of DG are associated with Von Recklinghausen's disease (VRD) or MEN-IIB.[7,8]

In solitary ganglioneuromas, there is no known gender predominance, and peak incidence is in the 4th to 6th decades of life, however lesions have been detected in adults from ages 20 to 90 years old.[2,5,7] Reported cases of solitary gastrointestinal GNs often involve the colon and rectum, and less commonly the appendix and ileum.[9] Solitary GN are usually asymptomatic.[3,6] They can however lead to thickening of the bowel wall, strictures or submucosal nodularities and therefore patients may present with gastrointestinal bleeding, abdominal pain, constipation, obstruction, or weight loss.[3,6]

Endoscopically, solitary intestinal ganglioneuromas have no discerning phenotypic characteristics therefore definitive diagnosis requires biopsy.[2] Microscopic examination reveals the lamina propria expanded by an infiltrate of spindle cells and ganglion cells.[10] During immunohistochemistry, stains for S100 protein and neuron-specific enolase confirm the presence of spindle cells and ganglion cells respectively.[11]

Management of gastrointestinal GNs depends on the patient's history and clinical presentation as well as the size, location and subtype. Treatment for solitary colonic GNs detected during colonoscopy usually involves endoscopic polypectomy.[2,7] There are no current guidelines for solitary ganglioneuromas however most clinicians agree that endoscopic resection is curative and surveillance colonoscopy is not necessary following resection given the benign nature of the lesions which seldom recur.[2,7] Contrarily, patients with syndromic

ganglioneuromas require careful surveillance based on their specific syndromes.[7]

Prognosis for patients with solitary colonic GNs is usually excellent and without complications as evidenced by a study of 28 patients with polypoid ganglioneuroma which found that after an average follow-up period of 8 years, none of the patients developed MEN-IIB, Von Recklinghausen disease or subsequent complications.[2,6]

CONCLUSION

In conclusion, solitary polypoid ganglioneuromas in the colon are rare and benign neoplasms with endoscopic resemblance to sessile polyps frequently encountered in clinical practice. They can be safely excised endoscopically, with most clinicians agreeing that surveillance colonoscopy or screening for genetic syndromes is not required given their benign nature. If diffuse ganglioneuromatosis is seen however, syndromic causes must be considered such as neurofibromatosis type 1, MEN-IIb or Cowden syndrome.

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