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## Single Case

# Colonic Ganglioneuroma: A Rare Finding during Colorectal Cancer Screening

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## Keywords

Ganglioneuroma · Autonomic nervous system · Ascending colon · Screening colonoscopy · S100 tumor

## Abstract

Ganglioneuromas are very rare clinical entities, and their occurrence in the large bowel lays further emphasis on their rarity. Ganglioneuromas are benign tumors of undifferentiated neural crest cells. Their clinical presentation is mostly asymptomatic, and if any symptoms are present at all, they are usually nonspecific, with excellent prognosis. We report an asymptomatic, 65-year-old male with a solitary ascending colonic polyp found on screening colonoscopy. Histology revealed benign polypoid spindle-cell proliferation as well as S100 reactivity, consistent with ganglioneuroma. We report on the clinical presentation and discuss the origin, epidemiology, treatment, and management of this lesion.

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## Introduction

Ganglioneuromas are rare and benign tumors of the sympathetic nervous system composed of ganglion cells, nerve fibers, and glial cells, which originate from neural crest cells [1]. Solitary ganglioneuromas should be distinguished from ganglioneuromatous polyposis and diffuse ganglioneuromatosis. They are not associated with genetic syndromes such as neurofibromatosis 1, multiple endocrine neoplasia 2B syndrome, and juvenile polyposis. Clinically, patients are either asymptomatic or present with vague symptoms of abdominal pain, constipation, weight loss, bleeding, and signs of bowel obstruction [2]. Though there are no current guidelines, most clinicians agree that surveillance colonoscopy is not necessary following endoscopic resection. Herein, we describe a case of a 65-year-old male with an ascending colonic polyp found on screening colonoscopy, which proved to be a ganglioneuroma following histopathology.

## Case Presentation

A 65-year-old male with a medical history of chronic obstructive pulmonary disease with partial right-lung resection 16 years ago, hypertension, diabetes mellitus type 2, and gastroesophageal reflux disease presented for colonoscopy for colorectal cancer screening. Our patient, who is a nonsmoker, did not report any active complaints or symptoms. Physical examination was unremarkable. The heart rate was 101/min, and he had a blood pressure of 158/83 mm Hg, a respiratory rate of 20/min, an oxygen saturation of 95% on room air, and a temperature of 98.4°F. Laboratory findings showed a white cell count of  $10 \times 10^9/L$ , hemoglobin of 13.5 g/dL, hematocrit of 41%, and platelets of  $359 \times 10^9/L$ .

During screening colonoscopy, 2 nonbleeding polyps were identified in the ascending colon, measuring 6 mm each. Both polyps were removed by cold forceps excisional polypectomy. Following histopathology, hematoxylin and eosin staining showed spindle-cell proliferation, and immunohistochemistry demonstrated S100 immunoreactivity, consistent with ganglioneuroma (Fig. 1, Fig. 2, Fig. 3).

## Discussion

Ganglioneuromas are benign hamartomatous tumors of the autonomic nervous system [1]. As a subtype of tumors of undifferentiated neuroblastic origin, ganglioneuromas are composed of ganglion cells, nerve fibers, and supportive glial cells [3, 4]. Ganglioneuromas are uncommon worldwide and can occur at any autonomic location. However, a review of published cases shows that ganglioneuromas have a predilection for the head, neck, and/or adrenal glands [2]. Furthermore, ganglioneuromas bear no predilection for gender and have been detected among adults in their 3rd–10th decades of life with peak incidence between the 4th and 6th decades [5, 6].

Gastrointestinal ganglioneuromas are morphologically classified into 3 categories, namely polypoid ganglioneuromas, ganglioneuromatous polyposis, and diffuse ganglioneuromatosis [7]. Polypoid ganglioneuromas are mostly solitary, small (measuring <2 cm), and may be sessile or pedunculated polyps [6]. Ganglioneuromatous polyposis are multiple, mostly more than 20, ranging from 1 mm to 2.2 cm in size, and can also be sessile or pedunculated. Some may be filiform [6]. Ganglioneuromas classically occupy the lamina propria

but can extend into the submucosa. Diffuse ganglioneuromatosis is nodular and diffuse, up to 17 cm in size, and often extends transmurally to involve the myenteric plexus [8].

Ganglioneuromatous polyposis and diffuse ganglioneuromatosis are associated with genetic syndromes such as neurofibromatosis 1, multiple endocrine neoplasia 2B syndrome, and juvenile polyposis. However, solitary ganglioneuromas are not associated with systemic and familial syndromes [9]. Patients with solitary ganglioneuromas are usually asymptomatic and are detected on routine colorectal cancer screening. However, some patients may present with vague symptoms of abdominal pain, constipation, weight loss, hematochezia, and signs of obstruction [2]. While hematochezia due to a solitary ganglioneuroma is very rare, a review of the literature shows that bleeding is highly associated with this tumor [2, 10, 11]. However, the majority of patients is asymptomatic or presents with chronic diarrhea [12].

Endoscopically, intestinal ganglioneuromas have no discerning phenotypic characteristics. They are definitively diagnosed by biopsy followed by histology, which demonstrates immunoreactivity to S100 with comma-shaped nuclei mixed with aggregates of ganglion cells. Ganglioneuromas can also be positive for vimentin, glial fibrillary acidic protein, and neuron-specific enolase. Hematoxylin and eosin staining usually shows a high proliferative index of spindle cells with a collection of ganglion cells [6].

Gastrointestinal ganglioneuromas are usually treated endoscopically and completely excised. Currently, no guideline exists on the management of solitary ganglioneuromas or recommendations for surveillance colonoscopy. However, most authors agree that repeat colonoscopy is not necessary due to the benign nature of the lesion, which tends not to recur [6, 13, 14]. Furthermore, prognosis is usually excellent and without complications. Evidently, a study of 28 patients with solitary ganglioneuromas found that after an average follow-up time of 8 years, none of the patients developed von Recklinghausen disease, multiple tumor syndromes, or subsequent complications [7].

In conclusion, gastrointestinal ganglioneuromas are rare and benign hamartomatous tumors that originate from the autonomic nervous system. Bearing no predilection for gender, the average incidental age is 50 years with excellent patient prognosis. Solitary ganglioneuromas are not associated with any systemic or genetic conditions and tend not to recur. As a result, they can be safely excised endoscopically without complications. Our patient was similarly treated with a favorable clinical outcome.

#### Statement of Ethics

Informed consent was obtained from the patient.

#### Disclosure Statement

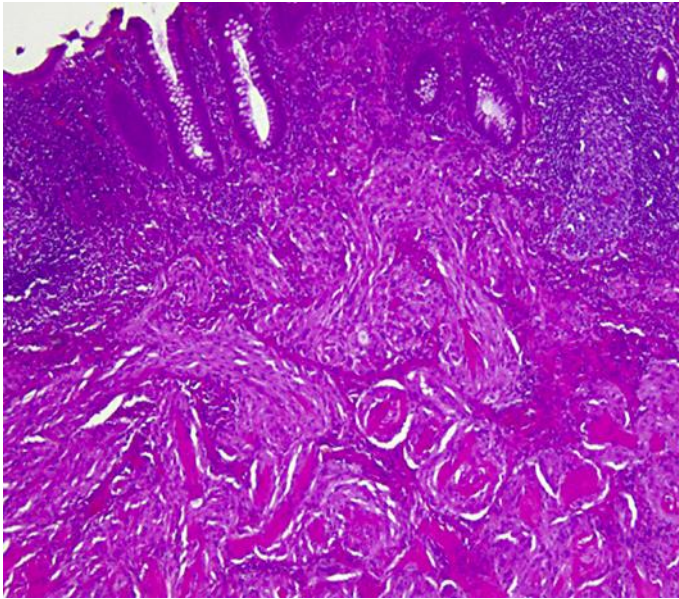
The authors have no conflicts of interest or financial relationships to disclose.

### Author Contribution

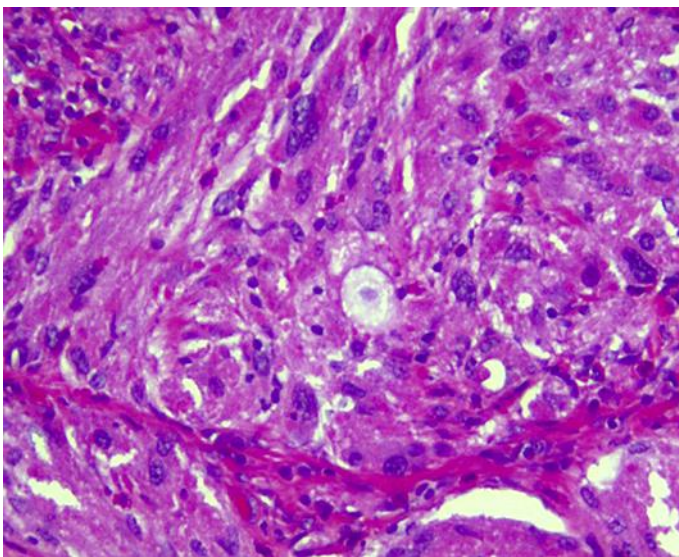
E.O., M.O., D.R., and M.R. were responsible for the conception, design, and the drafting of the article. E.O., M.O., D.R., M.R., T.H., and P.X. critically revised the article for important intellectual content. E.O., M.O., D.R., M.R., T.H., and P.X. gave final approval to the article.

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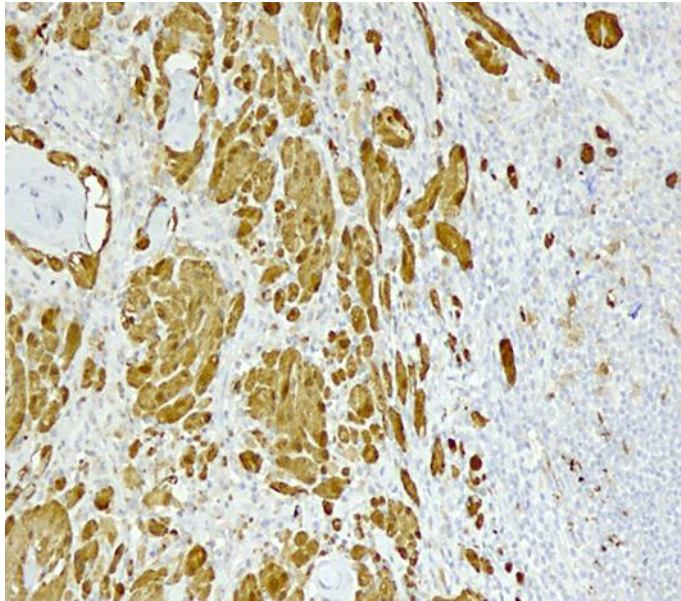
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**Fig. 1.** Hematoxylin and eosin staining showing ganglion and stromal cells. Magnification,  $\times 40$ .



**Fig. 2.** Hematoxylin and eosin staining revealing nodular expansion of neural bundle and ganglion cells. Magnification,  $\times 100$ .



**Fig. 3.** Immunohistochemical staining demonstrating S100 immunoreactivity. Magnification,  $\times 40$ .