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(CASE REPORT)



## A 80-year-old man with intestinal ganglioneuroma: A rare case

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### Abstract

Ganglioneuroma (GN) is a rare benign neurogenic tumor, usually asymptomatic, which arises along the sympathetic ganglion chains. It accounts for more than 0.1%-0.5% of all tumors of the nervous system. Ganglioneuromas most frequently occurs the abdomen, however these tumors can grow anywhere sympathetic nervous tissue is found. Other common locations include the adrenal gland, paraspinal retroperitoneum, posterior mediastinum, head and neck. However, examples arising in the gastrointestinal tract are extremely uncommon. The main therapy for this lesion is the complete surgical excision. In this article we report a case of 80-year-old man with ganglioneuroma localized in a polypoid lesion of the sigmoid bowel.

**Keywords:** Ganglioneuroma (GN); Rare tumor; Sympathetic nervous system; Sigmoid bowel

### 1. Introduction

Ganglioneuromas (GN) are unusual hamartomatous tumors of the sympathetic nervous system composed of ganglion cells, nerve fibers and glial cells, which originated from neural crest cells[1][2].

The first authenticated case was described by Lorezt in 1870. There are no known risk factors for ganglioneuromas and they are not associated with genetic syndromes such as neurofibromatosis 1, multiple endocrine neoplasia 2B syndrome and juvenile polipolys [3][4]. They are rarely seen in the gastrointestinal tract where they can present either as a solitary lesion or as multiple polyps in the colon and/or terminal ileum [5]. GN commonly occurs in the mediastinum (41,5%) and retroperitoneum (37,5%) [6].

Clinically, patients are either asymptomatic or present with vague symptoms of abdominal pain, constipation, weight loss, bleeding and signs of bowel obstruction [7]. These tumors may produce hormones, which can cause diarrhea, an enlarged clitoris (in females), high blood pressure, increased body hair, and sweating [8].

Histologically, ganglioneuroma is characterized by the presence of mature ganglion cellular elements, immunoreactive for S-100 and for enolase-neuron-specific (NSE) and devoid of aspects of malignancy, nerve fibers, blood vessels. There are no current guidelines about therapeutic management. Most clinicians agree that surveillance colonoscopy is not necessary following endoscopic resection.

When the lesion occurs, it is removed by surgical resection; GN can be diagnosed through physical examination or, moreover, through histological sample.

Herein, we describe a case of 80-year-old male with a sigmoid polyp found on colonoscopy, which is histopathologically compatible with ganglioneuroma.

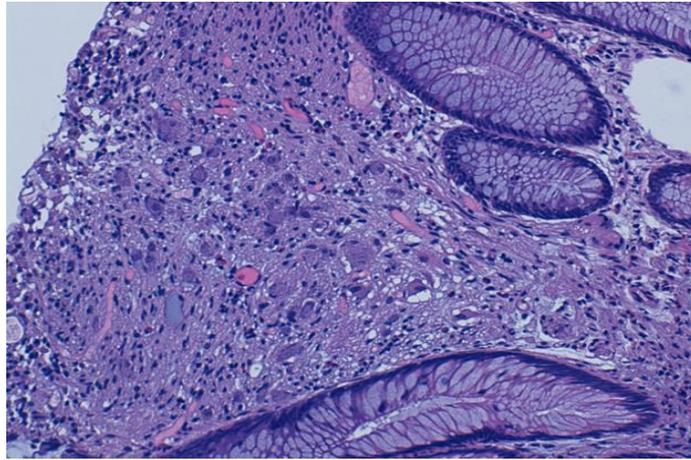
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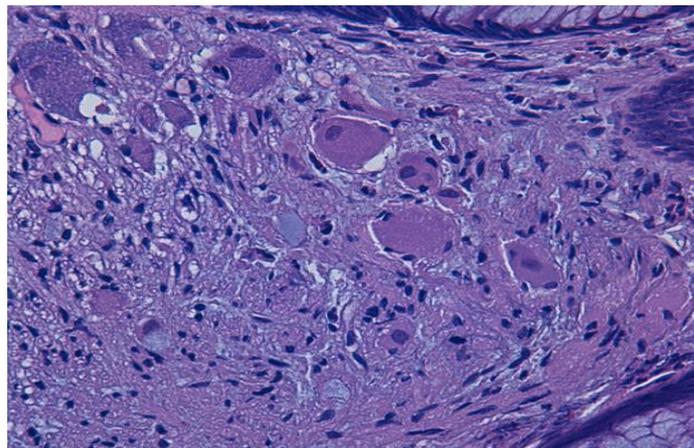
## 2. Case report

In May 2019, an 80-year-old man showed up at a private clinic showing an intense abdominal pains at the level of the left iliac pit. For this reason, a screening colonoscopy was carried out, up to the cecum.

It revealed that the visceral lumen is fully elongated and very convoluted in the sigma, where there are numerous diverticula even off the intestinal collar, and it is clogged with faecal material; in the sigma a sessile polyp was detected and removed during colonoscopy. The sample was formalin fixed and paraffin embedded and finally microscopically observed on routine slides in the Surgical Pathology Service at Casale Monferrato Hospital. Histological picture highlighted fragment of colic mucosa characterized by thinning of the glandular amount for the intra-mucosal proliferation of interstitial fused and giant cells with abundant cytoplasm and no mitotic activity (figures 1,2) suggestive for ganglion origin as immune-reactive for S100 protein (figure 3).

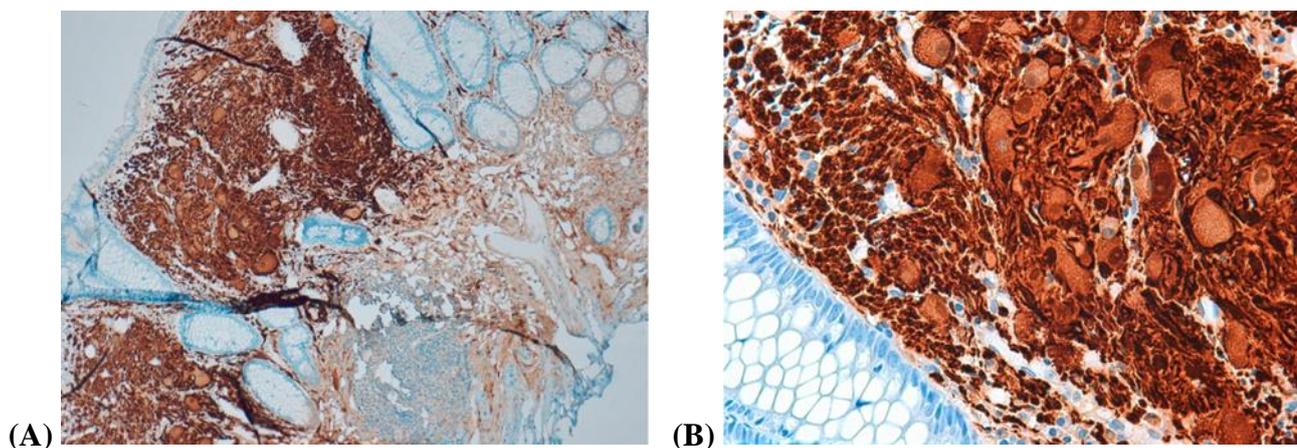


**Figure 1** Hematoxylin and eosin staining showing ganglion and stromal cells (Magnification 20x).



**Figure 2** Hematoxylin and eosin staining showing proliferation of ganglion cells and benign spindle-shaped Schwann cells (Magnification 40X).

Endoscopically, intestinal ganglioneuromas have no peculiar morphological characteristics; they are definitively diagnosed by biopsy followed by histology which demonstrates immunoreactivity for S100 protein in cells with comma-shaped nuclei mixed with aggregates of ganglion cells. Ganglioneuromas can also be positive for Vimentin, glial fibrillary acidic protein (GFAP) and NSE [9]. Histologically, ganglioneuroma is mainly composed of ganglion cells, mucus matrix, nerve fibers and Schwann cells and the first two of them are its pathognomonic components [10].



**Figure 3** (A) and (B) Immunohistochemical stain of the bowel mucosa with ganglioneuroma demonstrating S100 immunoreactivity. Magnification: (A) 10x and (B) 40x.

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### 3. Discussion

Ganglioneuroma (GN) is a rare benign tumor, derived from autonomic nervous system. These tumors are rarely found in the gastrointestinal tract as most frequently occur in head, neck, or adrenal glands. Retroperitoneal GNs account for 37.5% of all GNs and about 0.72%-1.6% of primary retroperitoneal tumors. Bearing no predilection for gender, the average incidental age is 50 years with excellent clinical outcome.

They can be divided into three different morphological subtypes, namely polypoid GNs, ganglioneuromatous polyposis, and diffuse ganglioneuromatosis [11].

Typically polypoid GN is the most common form and can be found as sessile or peduncolated bowel polyps, <2 cm in size, and sited in the lamina propria but sometimes extending into the sub-mucosa. Solitary ganglioneuromas are not associated with any systemic or genetic conditions and tend not to recur.

Clinical presentation can be asymptomatic, but depending on the size, location, and degree of involvement of the affected segment; patients can present with abdominal pain, weight loss, obstruction, and bleeding.

Most commonly, the lesions are encountered incidentally during colonoscopy as polyps, as in the present case and the excision is curative. Diagnosis is usually made on histological and immunochemical examination of the removed specimen through S100 protein immunoreactivity of the ganglion cells and nerve fibers.

No association between GNs and bowel carcinoma has been found yet [12]. Surgical excision is the best treatment for GN, and postoperative adjuvant radiotherapy and chemotherapy are unnecessary.

However, patients with GN still need long term radiological follow-up [13].

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### 4. Conclusion

Ganglioneuromas is an uncommon tumor which can only be diagnosed through histopathologic examination. Histology of the lesion shows multiple spindle cells on hematoxylin and eosin stain, with positivity for S100 protein and NSE on immunohistochemical staining. The main treatment for this lesion is complete surgical excision; nevertheless, patients without complications after tumor removal still need regular follow-up.

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### Compliance with ethical standards

#### *Disclosure of conflict of interest*

Authors declare that there are no conflicts of interest in connection with this paper, and the material described is not under publication or consideration for publication elsewhere.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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