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Colonic Ganglioneuroma: A Rare Incidental Finding

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INTRODUCTION

Ganglioneuromas are rare and benign tumors of neural crest origin that are comprised of ganglion cells arising from nerves.[1] These tumors are characterized by overgrowth of nerve ganglion cells, nerve fibers, and supporting cells in the gastrointestinal tract. They can be classified based on the extent of involvement in the gastrointestinal (GI) tract and are divided into three subgroups: polypoid ganglioneuroma, ganglioneuromatous polyposis, and diffuse ganglioneuromatosis. These three subclasses are differentiated based on their association with other pathologies or syndromes and the extent of their involvement in the GI tract.

Solitary polypoid ganglioneuroma has no association with other pathologies or syndromes.[2,3] On the other hand, ganglioneuromatous polyposis and diffuse ganglioneuromatosis are associated with various syndromes including multiple endocrine neoplasia 2B syndrome, neurofibromatosis type 1, juvenile polyposis, nonfamilial adenomatous polyposis, tuberous sclerosis, Cowden's syndrome, and Ruvalcaba-Myhre-Smith syndrome.[4] Clinically, patients are usually asymptomatic. In some cases, however, they may present with abdominal pain, constipation, bleeding, weight loss or show signs of bowel obstruction. Herein, a case of a 61-year-old male is presented, who after undergoing a routine colonoscopy, was found to have an incidental polyp that was characterized by the pathology lab as ganglioneuroma.

CASE REPORT

A case of 61-year-old male with past medical history of hypertension and type 2 diabetes mellitus presented to an outpatient endoscopy clinic for a routine colonoscopy screening. The patient had stable vital signs with a blood pressure of 110/72 mmHg, oxygen saturation of 98% on room air, and heart rate of 102 bpm. Physical exam was unremarkable. His social history was significant for being a former smoker.

During screening, colonoscopy showed a 6 mm polyp in the ascending colon, 5 mm polyp in the sigmoid colon, and small hyperplastic polyps in the rectum. A single-piece polypectomy was performed using a cold snare. Following histopathology, staining and analysis with hematoxylin and eosin and S100 immunostaining, results were consistent with ganglioneuroma.

DISCUSSION

Ganglioneuromas are rare, benign tumors characterized by overgrowth of nerve ganglion cells, nerve fibers, and supporting cells in the gastrointestinal tract.[1] Clinically, patients usually are asymptomatic. In some cases, however, they may present with abdominal pain, constipation, bleeding, weight loss, or show signs of bowel obstruction, especially in the case of ganglioneuromatosis where there is an association with multiple endocrine neoplasia IIB. Solitary ganglioneuromas have no association with genetic syndromes and patients are asymptomatic in most of the cases. As in our case, those patients are diagnosed incidentally during routine colon cancer screening.[5,6] Grossly, polypoid isolated ganglioneuromas may resemble juvenile polyposis or adenomas and are usually 1-2 cm in size. They can be found at any section in the colon and classically occupy the lamina propria. Therefore, their diagnosis cannot be done endoscopically and thus require histopathological analysis. A biopsy stained in hematoxylin and eosin would show spindle and ganglion cells (Figures 1, 2 and 3). It would demonstrate immunoreactivity to S100 highlighting both ganglion cells and neuronal components (Figure 4).

Given their association with various syndromes, findings of such tumors should prompt clinicians to investigate the presence of other symptoms and signs further. In the presence of high clinical suspicions to any of the forementioned syndromes, genetic testing may be done for a confirmed diagnosis to the patient and proximal relatives.

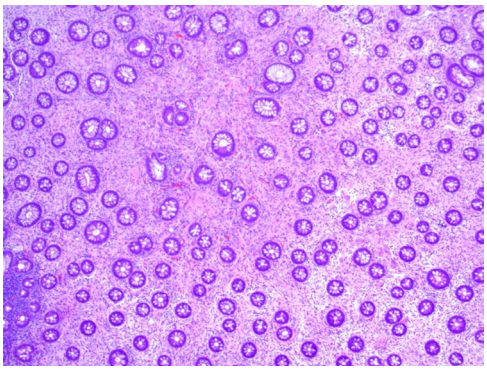

Figure 1. Low power view (40x) of polypoid colonic mucosa with lamina propria expansion.

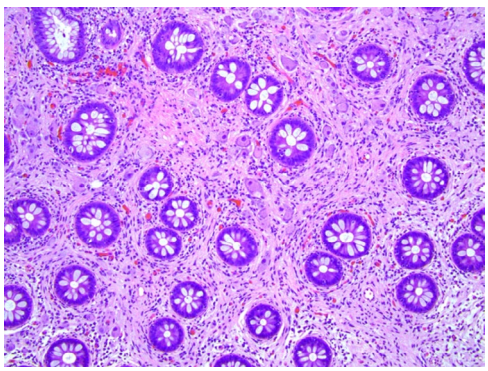

Figure 2. Intermediate power view (100x) showing bland spindle cells intermixed with plump epithelioid cells in between colonic glands.

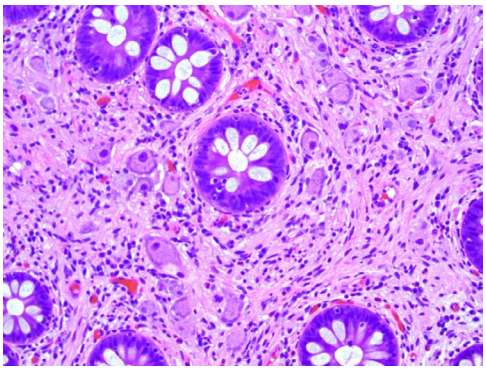

Figure 3. High power view (200x) shows ganglion cells (Plump cells with abundant amphophilic cytoplasm, oval eccentric nuclei, and prominent nucleoli) with admixed neuronal spindle cells with wavy nuclei.

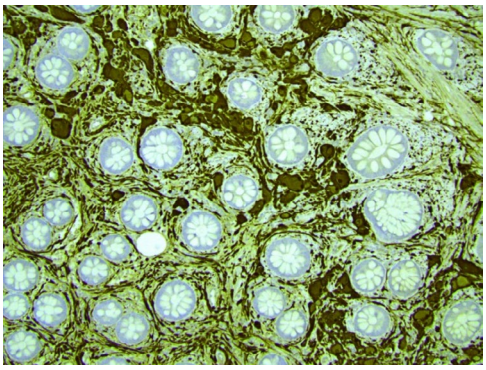

Figure 4. S100 immunostaining highlighting both the ganglion cells and the neuronal components (brown color).

Treatment of ganglioneuromas is mainly through endoscopic excision of the polyp.[5] Following excision, symptomatic treatment, and management of the associated syndromes (if present), there is no need for any routine surveillance colonoscopy other than the one used for the colorectal cancer screening. These polyps are benign, thus there is no added benefit from repeating colonoscopy.

REFERENCES

[1] Zabaglo M, Dreyer MA. Neuroma. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing, 2022.
[2] Chan OTM, Haghighi P. Hamartomatous polyps of the colon: Ganglioneuromatous, stromal, and lipomatous. Arch Pathol Lab Med 2006; 130(10): 1561-1566. PMID: 17090203.
[3] Rubin BP, Hornick JL. Mesenchymal Tumors of the Gastrointestinal Tract. In: JLHornick (Ed). Practical Soft Tissue Pathology: A Diagnostic Approach. Second Edition. Philadelphia: Elsevier, 2019, pp. 459-498. ISBN 9780323497145.
[4] Abraham G, Prakash SR. Solitary colonic ganglioneuroma: A rare incidental finding of hematochezia. Case Rep Gastrointest Med 2015; 2015:794985. PMID: 26075113.
[5] Baiomi A, Abbas H, Niazi M, Remotti H, Daniel M, Balar B. Colonic ganglioneuroma: A rare lesion with extremely different presentations and outcomes in two patients. Gastroenterology Res 2021; 14(3):194-198. PMID: 34267836.
[6] Voltaggio L, Montgomery EA. Gastrointestinal tract spindle cell lesions--just like real estate, it's all about location. Mod Pathol 2015; 28(Suppl 1):S47-S66. PMID: 25560599.

Keywords: ganglioneuroma, endoscopy, S100 proteins, colonoscopy, gastrointestinal tract