Case Report

A case of ganglioneuroma masquerading as hyperplastic colonic polyp from a histopathologist perspective

Nayak Mamita, Panda Sasmita

Department of Oncopathology, Acharya Harihar Post Graduate Institute of Cancer, Cuttack, Odisha, India

Abstract

Ganglioneuromas are uncommon benign neuroectodermal tumors of undifferentiated neural crest cells characterised by hyperplasia of ganglion cells ,nerve fibres and supporting cells.They are found in diverse sites ,most common site being in adrenal glands and head and neck region whereas the gastrointestinal tract being a very unusual site .Though solitary as well as multiple ganglioneuromas occur in colorectal site, still it is an unusual site. Usually solitary ganglioneuromas are asymptomatic .We herein report a case of colonic ganglioneuroma diagnosed histopathologically and by immunohictochemistry, in a 42 year old male who presented with a hyperplastic polyp on colonoscopy complaining of abdominal pain.

Keywords: Colon, ganglioneuroma, histopathology, polyp, S100

Address for correspondence: Dr. Mamita Nayak, Department of Oncopathology, Acharya Harihar Post Graduate Institute of Cancer, Cuttack, Odisha, India.
E-mail: nayak_mamita@yahoo.com

Submitted: 06-Oct-2021 **Accepted:** 03-Nov-2021 **Published:** 22-Dec-2021

INTRODUCTION

Ganglioneuromas (GNs) are tumors of the sympathetic nervous system consisting of ganglion cells, nerve fibers, and glial cells. Hence, these tumors usually grow along the distribution of sympathetic nervous tissue. Ganglioneuroma (GN) commonly occurs in the posterior mediastinum (60%–80%) and abdominal cavity (10%–15%), most commonly localized to the retroperitoneum and the adrenals.[1] However, 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.[2] Solitary ones are usually incidental findings unrelated to any genetic syndromes whereas the reverse is true for multiple polyps. Patients with solitary GNs are usually asymptomatic or present with vague symptoms of abdominal fullness, pain, rectal bleeding, weight loss, occasional times with intussusception, or with hematochezia.[1,2] Histologically characterized by the presence of mature ganglion cells admixed with spindle cells and immunohistochemically by expression of S100 and neuron-specific enolase. Definitive diagnosis of GN can only be made through histopathological examination.[3]

We describe a case of incidental GN in a 42-year-old male who underwent colonoscopy, complaining of abdominal pain with the colonoscopic diagnosis of the hyperplastic polyp.

CASE REPORT

A 42-year-old male presented to the outpatient department with abdominal pain persistent for 2 months. His bowel habits were not altered, no history of rectal bleeding, no abdominal distension, no weight loss, and no drug history. There were no other relevant clinical complaints.

Access this article online	
Quick Response Code:	Website: www.ijcpc.org
	DOI: 10.4103/ijcpc.ijcpc_10_21

How to cite this article: Mamita N, Sasmita P. A case of ganglioneuroma masquerading as hyperplastic colonic polyp from a histopathologist perspective. Int J Clinicopathol Correl 2021;5:88-90.

Personal history was unremarkable with no history of diabetes, hypertension, or any chronic disease. History did not reveal any significant findings. Family history was quite normal without any clue hinting toward any genetic syndromes. Physical examination was unremarkable with a normal heart rate of 98 beats/min and blood pressure of 120/90 mm Hg. Laboratory findings revealed a hemoglobin level of 11.2 gm/dl, white blood cell count of 12,000 cells/cumm, and a normal platelet count of 402×10^9/L. His occult blood test was negative. Abdominal ultrasonography was normal. Colonoscopy revealed a nonbleeding polyp in the ascending colon measuring around 7 mm and colonoscopically was diagnosed to be a hyperplastic polyp. Excisional polypectomy was done and sent for histopathology. Microscopically, the sections showed normal-appearing colonic epithelium and gland crypts lined by goblet cells with the lamina propria composed of ganglion cells admixed with haphazardly arranged spindle cells featuring nuclear buckling and collagenous stroma [Figure 1a and b]. There was no significant atypia or mitotic figures. Immunohistochemical staining was positive for S100 [Figure 2a and b] and negative for SMA [Figure 2c]. Based on these findings, a definitive diagnosis of GN was given. The patient is in regular follow-up and is asymptomatic post 6 months of polypectomy.

DISCUSSION

Ganglioneuromas are hamartomatous tumor of the autonomic nervous system.[4] They consist of ganglion cells, nerve fibers, and Schwann sheath elements.[5] They can occur at any anatomic site, with the commonly affected sites being the adrenal, head, and neck. Gastrointestinal site is one of the uncommon sites of predilection.

Gender wise there is no specific predilection. With regard to age, there is a peak incidence between the 4th and 6th decades.[1,6,7] Clinically, their presentation is usually asymptomatic and nonspecific in form of abdominal pain, constipation, weight loss, bleeding, chronic diarrhea, signs of bowel obstruction, or rarely hematochezia or intussusceptions.[1,3,8] These tumors may produce hormones, which can cause diarrhea, an enlarged clitoris (in females), high blood pressure, increased body hair, and sweating.[9]

Gastrointestinal GNs are morphologically three types, namely, (1) Polypoid ganglioneuromas, (2) Ganglioneuromatous polyposis, and (3) Diffuse ganglioneuromatosis. Polypoid GNs are solitary with size, usually <2 cm masquerading as juvenile polyp, adenomas, or hyperplastic polyps, whereas the ganglioneuromatous poylposis is multiple (20–40) with size varying from 1 mm to 2.2 cm. In either of the entities, they can be either sessile or pedunculated. Diffuse ganglioneuromatosis is nodular and diffuse with the size varying up to 17 cm in size.[7]

Solitary GNs are not related to any hereditary or genetic syndromes, the other two categories are associated with genetic syndromes such as Von Recklinghausen's disease (neurofibromatosis 1), multiple neuroendocrine neoplasia 2B syndrome, and juvenile polyposis. Ganglioneuromatous polyposis is also associated with Cowden's disease and Ruvalcaba-Myhre-Smith syndrome.[10]

Solitary GNs are mostly incidentally detected on colonoscopy as polyp. Colonoscopically, there are no typical features which can elucidate it as GN. Ganglion cells are normally not seen in gastrointestinal mucosa.[5] However, in solitary GNs, histology reveals mature ganglion cells which are large cells with eccentrically placed nuclei and abundant eosinophilic cytoplasm admixed in a stroma of haphazardly arranged spindle cells with many of them having wavy/buckled nuclei (both spindle and Schwann cells) in the lamina propria. However, sometimes, they can extend into the submucosa. Usually, these lesions show positive immunostaining to S100

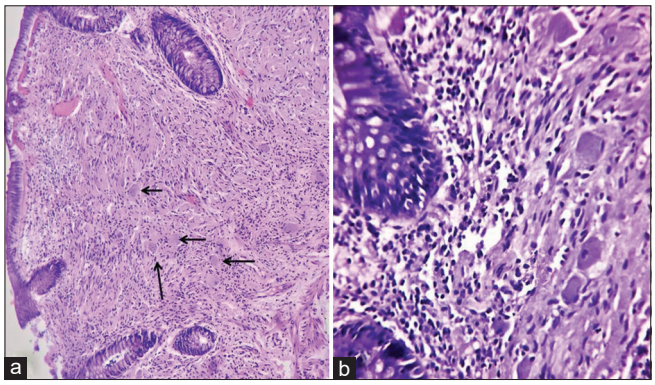

Figure 1: (a) Ganglioneuroma showing ganglion cells (marked by arrows) and spindle cells (H and E, ×200). (b) Mature ganglion cells and proliferated spindle cells with wavy nuclei (H and E, ×400)

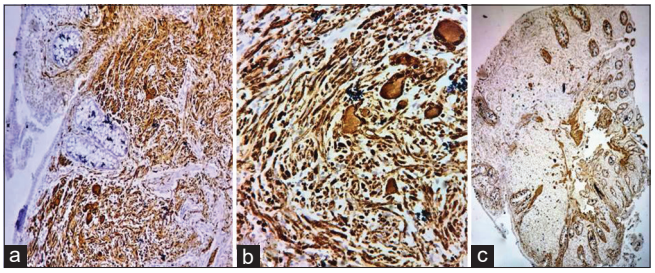

Figure 2: (a) Ganglion cells and spindle cells are positive for S100 (×200). (b) Strong positivity of ganglion and spindle cells for S100 (×400). (c) Spindle cells are negative for SMA (×100)

protein, which is helpful in confirming the neural origin of the lesion. Definitive diagnosis of GN can only be made through histopathological examination aided by immunohistochemistry with S100. Ganglioneuromatous polyposis which is multiple polyps is characterized by mature ganglion cells in the mucosa and submucosa. The diffuse ganglioneuromatosis have hyperplasia of nerve fibers and ganglion cells leading to band-like enlargement of the submucosal plexus and myenteric nerve plexus.[5]

GNs are treated by endoscopic complete excision. There are no established guidelines or recommendations for follow-up/surveillance colonoscopy in these cases. Being benign in nature with no tendency for recurrence, the prognosis is usually excellent without any recurrence, subsequent complications, or predilection toward malignancy or development of any multiple tumor syndromes.[4,10] Hence, does not necessitate the requirement for long-term clinical follow-up in these cases. Our case is without any clinical complaint post 6 months clinical follow-up.

Thus, gastrointestinal GNs are unusual tumors developing from the undifferentiated neural crest cells. Solitary GNs being one of them are commonly asymptomatic clinically and are incidentally identified colonoscopically as a polyp. These polyps must be excised endoscopically and must be subjected to the histopathological examination which is the only mode of definitive diagnosis. The accurate diagnosis will help in excluding other causes, alleviate unnecessary anxiety of any subsequent cancer, subsequent complications, will eliminate the need for surveillance colonoscopy, and omit the need for familial screening or genetic counseling. Although there is not much evidence, still physicians do not recommend the need for surveillance colonoscopy following endoscopic resection.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ofori E, Ona M, Ramai D, Huang T, Xiao P, Reddy M. Colonic ganglioneuroma: A rare finding during colorectal cancer screening. Case Rep Gastroenterol 2017;11:434-9.
2. Abraham G, Prakash SR. Solitary colonic ganglioneuroma: A rare incidental finding of hematochezia. Case Rep Gastrointest Med 2015;2015:794985.
3. Nehme C, Ghazaleh S, Patel D, Hasan S, Nawras A. Incidental ascending colon ganglioneuroma in the setting of hematochezia. Cureus 2020;12:e9447.
4. Chan OT, Haghighi P. Hamartomatous polyps of the colon: Ganglioneuromatous, stromal, and lipomatous. Arch Pathol Lab Med 2006;130:1561-6.
5. Katherine K, Javier R, Michael D, Sushil A. Isolated ganglioneuroma of the colon. Am J Gastroenterol 2010;105:S343-4.
6. Bakker JR, Haber MM, Garcia FU. Gastrointestinal neurofibromatosis: An unusual cause of gastric outlet obstruction. Am Surg 2005;71:100-5.
7. Voltaggio L, Montgomery EA. Gastrointestinal tract spindle cell lesions – Just like real estate, it's all about location. Mod Pathol 2015;28 Suppl 1:S47-66.
8. Soccorso G, Puls F, Richards C, Pringle H, Nour S. A ganglioneuroma of the sigmoid colon presenting as leading point of intussusception in a child: A case report. J Pediatr Surg 2009;44:e17-20.
9. Spinelli C, Rossi L, Barbetta A, Ugolini C, Strambi S. Incidental ganglioneuromas: A presentation of 14 surgical cases and literature review. J Endocrinol Invest 2015;38:547-54.
10. Ledwidge SF, Moorghen M, Longman RJ, Thomas MG. Adult transmural intestinal ganglioneuromatosis is not always associated with multiple endocrine neoplasia or neurofibromatosis: A case report. J Clin Pathol 2007;60:222-3.