The Gang's All Here: A Rare Case of Polypoid Ganglioneuroma of the Colon

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Introduction

Ganglioneuromas are rare benign tumors characterized by proliferation of nerve ganglion cells, nerve fibers and the supporting cells of the nervous system. These tumors are usually found in the mediastinum, retroperitoneum or adrenal glands. Rarely they occur in the gastrointestinal tract. Due to paucity of large-scale studies, there is currently a lack of guidelines regarding best practice management for intestinal ganglioneuromas. Here, we report a rare case of an isolated cecal ganglioneuroma.

Case Report

- A 63-year-old female with no pertinent past medical history presented for a routine screening colonoscopy.
- She had normal vital signs & no abnormal laboratory findings.
- During the colonoscopy, cold forceps polypectomy was performed to retrieve a total of 3 polyps.
- Hematoxylin and eosin staining of the cecal polyp revealed evidence of spindle cell proliferation accompanied by large cells with prominent nucleoli, consistent with ganglion cells (table 1).
- The immunostaining of the cecal polyp was reactive for the S100 protein but was negative for markers of stromal tissue (table 1).
- The concurrence of these findings was consistent with a diagnosis of polypoid ganglioneuroma.

Table 1: Histopathological & immunostaining characteristics of the polyps

Polyps retrieved	- Two 3 mm polyps from ascending colon - One 2 mm polyp from cecum
Hematoxylin & eosin staining of cecal polyp	- Spindle cell proliferation - Large cells with prominent nucleoli - Consistent with ganglion cells
Immunostaining of cecal polyp	- S100 + - CD34-, CD117-, SMA-, Desmin-, EMA-

Table 2: Classification of intestinal ganglioneuromas (modified from reference 1)

1. **Polypoid ganglioneuroma**	- Solitary & small (< 2 cm) - Sessile or pedunculated
2. **Ganglioneuromatous polyposis (GP)**	- Numerous (often 20 - 40) - Sessile or pedunculated - May be associated with genetic syndromes
3. **Diffuse ganglioneuromatosis (DG)**	- can range from 1-17cm in size - present as disseminated, nodular, intramural or transmural lesions - May be associated with genetic syndromes

Discussion

- The pathophysiology of these tumors is not well understood but may involve disordered replication of normal tissue cells.
- Gastrointestinal ganglioneuromas have been classified by Shekitka et al into 3 groups as shown in table 2.[1]
- Clinical features depend on location & size but can include abdominal pain, bleeding, constipation or obstruction.
- Few reports have reported an association with colorectal cancer.[2]
- There are no standard treatment guidelines for these tumors but similar to other cases reported in the literature, the tumor in our case was excised safely endoscopically.
- Currently there is a lack of guidelines for screening & surveillance endoscopies in patients with solitary ganglioneuromas.
- The US Multi-Society Task Force recommends genetic testing in any person with 2 or more lifetime hamartomatous polyps or a family history of such polyps.[3]
- Overall, the prognosis of solitary ganglioneuromas is good.

Conclusion

Even though there are no standard treatment guidelines, endoscopic polypectomy can be curative in cases of solitary ganglioneuromas.

We believe that awareness & proper counselling will translate into better care of patients with such tumors.

References

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