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Case Report

Colonic Hypoganglionosis in an Adult Female: A Case Report

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Abstract Hypoganglionosis is a rare cause of intestinal obstruction characterized by a reduced number of ganglionic cells in the colon. Patients with this condition may present with constipation and acute intestinal obstruction. Based on the segment of the intestine affected, it can be classified into type one, in which the pathology is confined to a segment of the colon, and type two, in which the hypoganglionosis is widespread but less severe. In children, the condition can be isolated or associated with Hirschsprung's disease. Hypoganglionosis is extremely rare in adults and is not easy to diagnose. We present this 39-year-old female patient who presented with symptoms of acute intestinal obstruction and was initially managed medically. Still, she ultimately underwent a total colectomy, and the diagnosis of colonic hypoganglionosis was made on histopathological evaluation. Her recovery was uneventful, and she has been symptom-free.

Keywords: Hypoganglionosis, Constipation, Abdominal distension, Colectomy

Introduction

Hypoganglionosis is a rare disease that affects 5% of cases of neuronal intestinal malformation.[1] The clinical features of hypoganglionosis are like Hirschsprung's disease, where they present with chronic constipation or as acute pseudo-intestinal obstruction. Intestinal hypoganglionosis is divided into two subtypes: isolated hypoganglionosis and the Hirschsprung-associated form. Isolated hypoganglionosis is a distinct and rare disorder that has been proposed as a distinct entity, with two further subtypes: congenital isolated hypoganglionosis (CIHG) and acquired isolated hypoganglionosis (AIHG). In isolated hypoganglionosis, the submucosal nerve plexus is normal, while a significant reduction in the myenteric ganglia is noted. In congenital hypoganglionosis, the number and size of ganglion cells are reduced at birth, and although their size increases with time, their number does not change. Acquired hypoganglionosis often occurs as a late-onset finding and is characterized by degeneration of ganglion cells and findings of gliosis on histopathological study.[2,3]

We report a case of hypoganglionosis in an adult woman who experienced turbulent years of worsening abdominal symptoms before receiving a definitive diagnosis. Her clinical journey highlights the importance of considering rare neuromuscular disorders in patients

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with stubborn constipation and underscores the role of full-thickness biopsies in confirming the diagnosis.

Clinical Presentation

A 39-year-old Chinese lady presented to the emergency department with complaints of progressive abdominal distension and intermittent abdominal pain for the last 3 days.

History of presenting illness

The patient reported progressive abdominal distension and intermittent episodic abdominal pain over the last 3 days. The pain was generalized, crampy, non-radiating, and partially relieved by passing flatus. She reported no passage of stools for 4 to 5 days but had been able to pass flatus. She denied nausea, vomiting, or fever.

She described a long-standing history of constipation since the age of 25, requiring laxatives or enemas 2–3 times per week. Her bowel habits had progressively worsened, and she passed stools with the help of these agents once a week. She had tried various dietary modifications and other forms of medications with limited relief. She gave a history of weight loss as reduced food intake due to her constipation. There were no previous surgeries or chronic illnesses. There was no history of weight loss, gastrointestinal bleeding, recent travel, or antibiotic use. She had no previous surgeries or known chronic illnesses.

Medical/Surgical history

The past medical history was significant in that during this period she had multiple CT scans and colonoscopies, but no definite diagnosis was made. An MRI defecography done in 2022 revealed moderate rectal prolapse and large rectoceles. A rectal mucosal intersectional colonic study done in the same year suggested colonic inertia. The patient had no other known comorbidities. Or any surgeries done. She had used over-the-counter laxatives and enemas frequently for her symptoms and has no known drug allergies on record.

Family history

Family history was insignificant, as no one in her family suffered from a similar problem, a history of Hirschsprung's disease. No history of GI malignancies or Inflammatory bowel disease was present.

Social

She is a non-smoker, though she takes an occasional glass of alcohol. She is an accountant by profession. Her social life has been rather constricted due to her prevailing problems.

Clinical examination

On physical examination, she was mildly distressed due to her distension, but there was no pallor, and her hydration was adequate. Her vital signs were BP 118/80, PR 90/mt, temperature was within normal limits, and SpO2 98% on room air.

On examination of the abdomen, it was soft, distended, and non-tender or guarded. There was a non-tender palpable mass occupying the entire left flank. The abdomen was tympanic, and the bowel sounds were normal. Digital rectal examination showed an empty rectum with no feces or masses felt.

Investigations

Blood investigations, which included a full blood count, electrolytes, thyroid function, and serum calcium levels, were normal. A CT scan of the abdomen revealed a megacolon extending to the cecum, with greater dilation in the left descending colon (10 cm) and a large fecaloma (Figure 1).

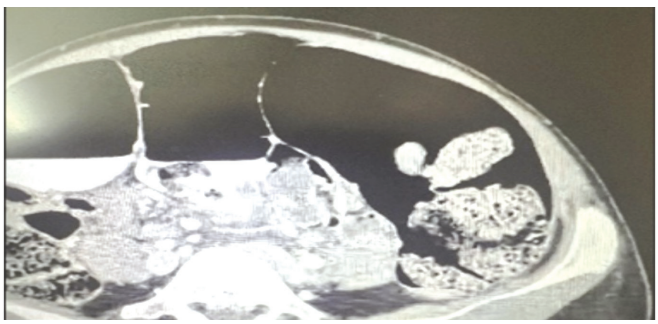

Figure 1 Computerized tomography of the abdomen, showing the dilated transverse colon

Management

Conservative management began with intravenous fluids, analgesics, and antibiotics. Although the therapy was continued, it was eventually discontinued due to a lack of improvement. It was then decided to proceed with surgical intervention, specifically an open laparotomy. Intraoperatively, as shown in the CT scan, there was a megacolon extending to the cecum, with more pronounced dilation over the left descending colon. The sigmoid colon was highly redundant. The colon was impacted with feces (Figures 2 and 3). The transition point was located at the upper rectum. Other organs appeared

normal. A total colectomy was performed, followed by a stapled side-to-end ileo-anal anastomosis and a covering loop ileostomy. Post-operative recovery was uneventful, and she was discharged.

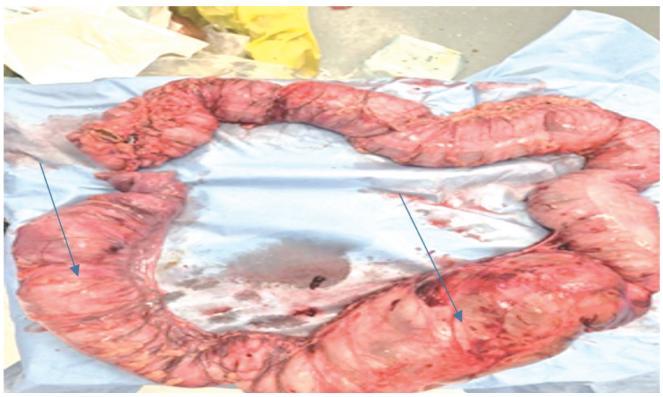

Figure 2 Specimen from a total colectomy (the arrow indicates the dilated segment)

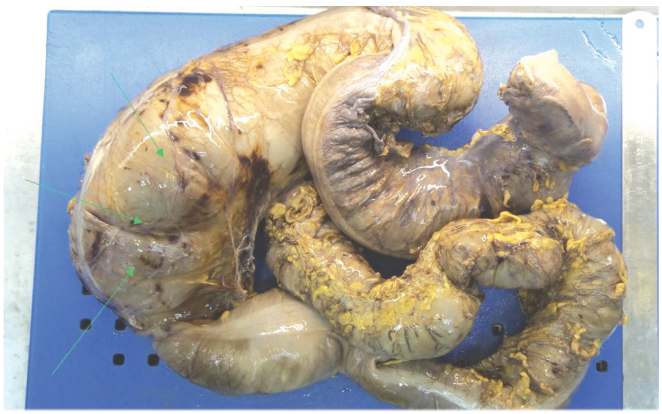

Figure 3 The macroscopic specimen of the resected total colectomy specimen (the green arrows indicate the dilated segment)

Histopathology

The histopathological reports (Histological slides, Figure 4) showed morphological features of hypoganglionosis. The mucosal layer shows ganglions, but the number of ganglion cells appears to be reduced and appears from the cecum to the sigmoid colon. The submucosal layer is normal.

Follow-up

On follow-up, she was cheerful and coping well with the stoma. There was significant relief of abdominal symptoms with the stoma functioning well. She has also shown a gain in weight compared to her admission. Closure of the stoma is planned in the coming months with improvement in her function.

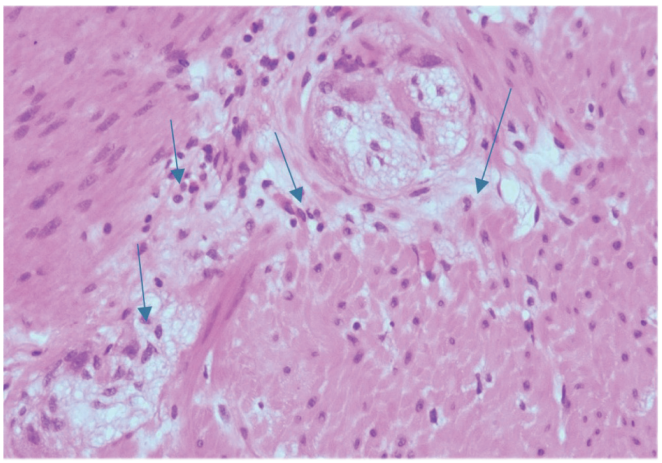

Figure 4 The histological examination of the specimen highlights the features of hypoganglionosis with a reduced number of ganglionic cells. (The blue arrow indicates the reduced number of ganglionic cells)

Discussion

Intestinal hypoganglionosis is a disorder of intestinal innervation that is commonly seen in children. It is one of the rarest gastrointestinal neuromuscular disorders. It affects the submucosal and myenteric neural plexus of the rectal wall and involves a specific length of the intestine just above the rectum.[4] Hypoganglionosis is characterized by a reduced number of ganglia, with fewer than one ganglion per 10 mm or an average of fewer than 2 neurons per ganglion. The diagnosis is usually confirmed through histological examination of the full-thickness specimen, including two-thirds of the resected segment's circumference. Hematoxylin and eosin staining are used to verify the diagnosis of hypoganglionosis.[5,6] It can be classified into type one, which is focal with a transitional zone, and type two, which is diffuse without a transitional zone. The transitional zone in type one often shows fewer, or no ganglion cells compared to the proximal dilated segment of the colon.[7]

The clinical presentation of intestinal hypoganglionosis involves long-term constipation and may include acute intestinal obstruction, abdominal distension, and ongoing constipation. Gas retention in the abdomen can cause pain. Common imaging tests, such as abdominal x-ray and CT scans, often show signs of intestinal blockage.[6] A significantly enlarged proximal colon segment with a transitional zone and a narrow distal segment on CT in a patient with chronic, unresponsive constipation typically indicates hypoganglionosis. Barium enema images usually support these findings to confirm the diagnosis.[8]

The management of hypoganglionosis involves surgical resection, which may include either a segmental resection of the colon or a subtotal colectomy. The most common complications observed are frequent and loose stools, especially in patients who have undergone a subtotal colectomy, while other complications, such as anastomotic leaks and enterocutaneous fistulas, are noted as rare. The choice of surgical procedure depends on the patient's presentation and the affected segment of hypoganglionosis.[9–11] This case report highlights the importance of the role of hypoganglionosis in chronic constipation in adults. After performing radiological investigations, performing serial biopsies may be beneficial in these patients; however, these have to be full-thickness biopsies, which may hinder their use in preoperative investigations of these patients.[12,13]

Conclusion

Hypoganglionosis of the colon is a rare cause of intestinal obstruction in adults, and it is crucial to establish the diagnosis for proper management and to prevent complications. These patients should be monitored for recurrent constipation to avoid issues like acute intestinal obstruction.

Conflicts of Interest

There is no conflict of interest.

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