

Isolated Adult Hypoganglionosis Presenting as Sigmoid Phytobezoar: A Case Report

Bulabai Karpagam¹, S Vinayagam², Jai Kishore³, Justine Samuel³

¹Associate Professor, Department of Radiology, SRM Medical College Hospital and Research Institute, Kancheepuram, Tamil Nadu, India, ²Professor, Department of Radiology, SRM Medical College Hospital and Research Institute, Kancheepuram, Tamil Nadu, India, ³Resident, Department of Radiology, SRM Medical College Hospital and Research Institute, Kancheepuram, Tamil Nadu, India

Abstract

Bezoars are the mass found trapped in the gastrointestinal system. They can also be termed as Gastrolith. The word "bezoar" is derived from the Persian pād-zahr, which means "antidote." Trichobezoars and phytobezoars are the most frequent and usual form in the stomach and can pass into the small bowel where they occasionally cause obstruction. Most bezoars are found in patients with a history of previous gastric surgery. Here, we report an interesting rare case of isolated adult hypoganglionosis presented as sigmoid phytobezoar causing chronic progressive abdominal pain with distension. Computed tomography showed the air mottled intraluminal mass with dilated sigmoid colon and rectosigmoid focal smooth narrowing.

Key words: Bezoar radiology, Focal hypoganglionosis, Large bowel obstruction, Sigmoid colon phytobezoar

INTRODUCTION

Isolated hypoganglionosis is a rare cause of intestinal innervation defects, can present as enterocolitis or poor bowel function, in infancy or childhood. We report a case of hypoganglionosis with an unusual presentation of sigmoid phytobezoar in a 57-year-old male patient. Bezoars usually form in the stomach and can pass into the small bowel where they can cause gastric or small bowel obstruction (SBO). Only few cases of colonic bezoar have been reported in the literature and were noted in post-operative stricture and psychiatric patients according to Agha *et al.*¹ We report a case of hypoganglionosis of rectosigmoid presenting as phytobezoar. Large bowel obstruction due to colonic bezoar is a diagnostic challenge and may cause life threatening complications. For colonic bezoar computerized tomography (CT) seems to be the main diagnostic modality, also for detecting the etiology as in our case report and the associated complications.

CASE REPORT

A 57-year-old male patient had progressively increasing abdominal pain and constipation for 2 weeks. He had a history of chronic constipation since childhood, for which he was taking high fiber diet and was treated with laxatives. Physical examination revealed distended abdomen with a hard mass in the lower abdomen. A CT scan demonstrated gross dilatation of the sigmoid colon of 15 cm diameter with the air mottled intraluminal mass. There was no enhancing solid component on intravenous (IV) contrast study and no flow of oral contrast distal to the lesion. Per-rectal contrast revealed suspicious focal rectosigmoid smooth narrowing for 5 cm, beyond which colonoscopy could not be passed. Patient was initially managed conservatively, as symptoms progressed was subjected for laparotomy, sigmoidectomy with a side-to-side colorectal anastomosis was performed. Histopathology of the resected specimen showed occasional ganglion cells and hypertrophied nerve bundles in the muscle layers, suggesting hypoganglionosis corresponding to the site of narrowing demonstrated on CT.

DISCUSSION

Isolated hypoganglionosis is a rare cause of intestinal innervation defects. It accounts for 5% of all intestinal

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Corresponding Author: Dr. Bulabai Karpagam, Department of Radiology, SRM Medical College Hospital and Research Institute, Kancheepuram, Tamil Nadu, India. Phone: +91-9894777929. E-mail: karaso1987@yahoo.com

neuronal malformations, characterized by sparse and small mesenteric ganglia, with hypertrophy of muscularis mucosa. Isolated hypoganglionosis can present as enterocolitis or poor bowel function, in infancy or childhood and can be isolated or associated with Hirschsprung's disease. Only 92 cases of isolated hypoganglionosis were reported from 1978 to 2009 according to Qadir *et al.*² Isolated hypoganglionosis predisposing to phytobezoar is even more rare, and this case report describes the atypical presentation of the disease. Patients usually present with severe acute and chronic constipation, pseudo-obstruction or enterocolitis. Until date, there is a similar case of isolated hypoganglionosis of sigmoid colon has been reported in the literature,² but presented as sigmoid volulus.

Bezoars consist of ingested foreign materials that accumulate within the gastrointestinal tract and are classified according to the materials of which they are composed. Approximately, 2-4% of SBOs are caused by bezoars and decreases as goes distally. In addition, presentation with features of acute surgical abdomen is extremely rare, accounting for only 1% of the patients. A bezoar is a concretion of indigestible material found in the gastrointestinal tract, which usually forms in the stomach and passes into the small bowel, where it can cause SBO. It can be classified into one of the four major types: trichobezoar, pharmacobezoar, lactobezoar, and phytobezoar. Phytobezoars are the most common, and are composed of vegetable matter such as celery, pumpkin, grape skin, prune and persimmons and it contains large amount of non-digestible fibers such as cellulose, hemicellulose, lignin and fruit tannins.³⁻⁵

Radiological investigations being the initial modality of choice of investigation for patients with suspected bowel obstruction. Findings on radiographs can suggest the presence of bezoars, barium, and CT studies can confirm it.³ The most extensive series published in this literature by Ripollés *et al.*⁶ has emphasized on CT and no other study has yet contrasted the usefulness of the different imaging techniques in application to bezoars.

Abdominal radiography demonstrated a distended sigmoid colon and rectum with dissipated feces (Figure 1). CT is much more sensitive and specific, and published series have shown the focal well-circumscribed air-mottled intraluminal mass, as noted in our case also. CT may be considered the imaging technique of choice for confirming the diagnosis of gastrointestinal bezoars. CT aids to establish the site of obstruction and to determine the cause and detect the existence of additional intestinal or gastric bezoars. A CT scan demonstrated gross dilatation of the sigmoid colon of 15 cm diameter with the air mottled intraluminal

mass (Figures 2a and b). There was no enhancing solid component or bowel wall enhancement on IV contrast study. And there was no distal flow of oral contrast to the rectum. Per-rectal contrast, revealed suspicious focal rectosigmoid smooth narrowing for 5 cm, demonstrated on reconstructed CT images (Figure 3), and also beyond which colonoscopy also could not be passed.

The method of management of bezoar removal depends on the site of impaction, the size, nature, and



Figure 1: X-ray abdomen shows distended sigmoid with dissipated faeces

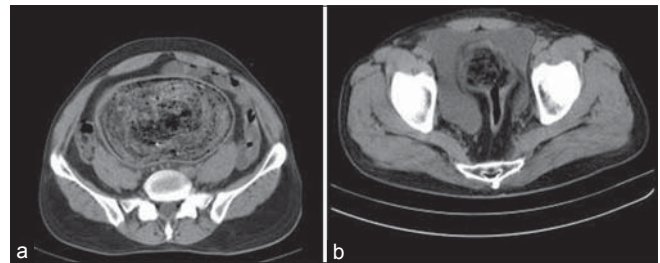


Figure 2: (a and b) Computerized tomography scan demonstrated gross dilatation of the sigmoid colon of 15 cm diameter with air mottled intraluminal mass

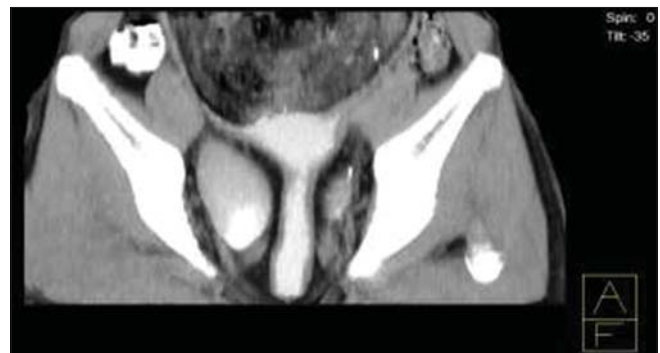


Figure 3: Computerized tomography reconstructed images shows smooth narrowing of rectosigmoid, without abnormal wall thickening and proximally dilated sigmoid colon with mottled appearance



Figure 4: (a and b) Surgical specimen shows bezoar with fecaloma causing dilated sigmoid colon

complications. Moreover surgical management is reserved for those presenting with life-threatening complications such as sigmoid volvulus, hemochezia or peritonitis⁷ and in cases when conservative management by digital evacuation, enemas and colonoscopy fails.

Our patient was initially managed conservatively, as symptoms progressed, and CT findings favored colonic bezoar, he was subjected for laparotomy, sigmoidectomy with a side-to-side colorectal anastomosis was performed. Operative confirmation of distal sigmoid focal narrowing (Figure 4a and b) indicated hypoganglionosis by histopathology and immunochemistry. Underlying narrowed segment predisposing to phytobezoar can be explained due to long-term laxatives and high fiber diet.

CONCLUSIONS

Isolated hypoganglionosis is a rare disease with clinical and epidemiological features similar to Hirschsprung's disease, but the age at diagnosis is higher. High degree of suspicion in cases of chronic constipation needs evaluation and CT may reveal the established features of phytobezoar and the predisposing factors. A definitive diagnosis requires histopathological analysis of full-thickness intestinal biopsies. And the treatment should be tailored according to the extent of hypoganglionosis.

REFERENCES

1. Agha FP, Nostrant TT, Fiddian-Green RG. "Giant colonic bezoar:" A medication bezoar due to psyllium seed husks. *Am J Gastroenterol* 1984;79:319-21.
2. Qadir I, Salick MM, Barakzai A, Zafar H. Isolated adult hypoganglionosis presenting as sigmoid volvulus: A case report. *J Med Case Rep* 2011 8;5:445.
3. Ali WA, Gondal ZI, Yammahi AA, Hushki SF, Badri F, ElTayeb YH. A case of small bowel obstruction due to phytobezoars. *J Surg Case Rep* 2013;2013. pii: rjt046.
4. Arvind B, Sachin K, Mathews VV, Telisinghe PU. Colonic obstruction secondary to phytobezoar. *Brunei Int Med J* 2010;6:100-4.
5. Bala M, Appelbaum L, Almogy G. Unexpected cause of large bowel obstruction: colonic bezoar. *Isr Med Assoc J* 2008;10:829-30.
6. Ripollés T, García-Aguayo J, Martínez MJ, Gil P. Gastrointestinal bezoars: Sonographic and CT characteristics. *AJR Am J Roentgenol* 2001;177:65-9.
7. Wang LT, Hsiao CW. Clinical spectrum and treatment of bezoars in adults: Experience of 20 cases in a single institute. *J Soc Colon Rectal Surg Taiwan* 2008;19:9-15.

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