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Acute large bowel pseudo-obstruction due to atrophic visceral myopathy: A case report

Sean M. Wrenn^{a,*}, Charles S. Parsons^{a,c}, Michelle Yang^b, Ajai K. Malhotra^a^a University of Vermont Medical Center, Department of Surgery, 111 Colchester Avenue, Burlington, VT, 05401, United States^b University of Vermont Medical Center, Department of Pathology, 111 Colchester Avenue, Burlington, VT, 05401, United States^c Beth Israel Deaconess Medical Center, Department of Surgery, 330 Brookline Avenue, Boston, MA, 02215, United States

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ABSTRACT

PURPOSE: Atrophic visceral myopathy is a pathological diagnosis characterized by atrophy of the smooth muscle layers of the viscera with intact ganglia. Rarely, it can present acutely as an intestinal pseudo-obstruction. We describe a rare case report and explore how this diagnosis can be distinguished from other forms of intestinal obstruction.

CASE DESCRIPTION: A 60-year-old male with a past medical history of hypothyroidism presented to the emergency department with a two-day history of worsening abdominal distention and pain associated with nausea and vomiting. Upon evaluation patient was found to have tachycardia, with abdominal distention and localized tenderness with peritonitis. Computed tomography demonstrated large bowel obstruction, likely caused by sigmoid volvulus. The patient underwent emergent laparotomy. Intraoperatively, the entire colon was found to be extremely dilated and redundant. With a working diagnosis of recurrent sigmoid volvulus causing intermittent large bowel obstruction, a sigmoid colectomy and primary anastomosis was performed. Pathology revealed atrophic visceral myopathy, with an extremely thin colonic wall and atrophic circumferential and longitudinal muscularis propria without inflammation or fibrosis. The ganglion cells and myenteric plexus were unaffected. Post-operatively, the patient developed prolonged ileus requiring nasogastric decompression and parenteral nutrition. The ileus resolved with pro-kinetic agents, and patient was discharged home on post-operative day fifteen.

CONCLUSIONS: Atrophic visceral neuropathy is a rare cause of intestinal pseudo-obstruction. While often presenting with chronic obstruction in younger populations, we present a rare late-onset acute presentation that may have been secondary to underlying hypothyroidism.

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1. Introduction

The loss of intrinsic smooth muscle and neuronal activity within the small and large intestines impairs motility and has previously been demonstrated to be a potential cause of pseudo-obstruction of the bowel. Pseudo-obstruction has previously been defined as signs and symptoms of intestinal obstruction in the absence of a true lumen-occluding mechanical obstruction [1]. It can be differentiated from true obstruction via radiography with enteric contrast or via endoscopy.

Atrophic visceral myopathy, a relatively rare diagnosis, typically presents with chronic pseudo-obstruction symptoms. We present a case of a previously asymptomatic 60 year-old male who developed

severe acute onset pseudo-obstruction. The case is presented in accordance with the SCARE guidelines for surgical case reports [2]. Prior to publication the patient provided written consent for the de-identified presentation of his clinical course.

2. Case presentation

A 60 year-old male with a past medical history of hypothyroidism (on daily levothyroxine therapy) and prior inguinal hernia repair (14 years prior) presented to our emergency department with complaints of acute onset abdominal pain, nausea, vomiting, obstipation, and abdominal distention of two days duration. On presentation, patient had tachycardia (heart rate 140 beats per minute), a temperature 37.6 °C, with regular blood pressure (136/77 mmHg) and oxygen saturation (SpO₂ 95% on room air). His abdominal examination was significant for distention, diffuse tenderness to palpation (worse on the left side of abdomen), and tympany. There was no noted abdominal guarding or rebound tenderness. Laboratory values drawn at the time were significant for an

* Corresponding author at: UVM Department of Surgery, 111 Colchester Avenue, Burlington, VT, 05401, United States.

E-mail addresses: Sean.Wrenn@uvmhealth.org (S.M. Wrenn), chasparsons@gmail.com (C.S. Parsons), Michelle.Yang2@uvmhealth.org (M. Yang), Ajai.Malhotra@uvmhealth.org (A.K. Malhotra).

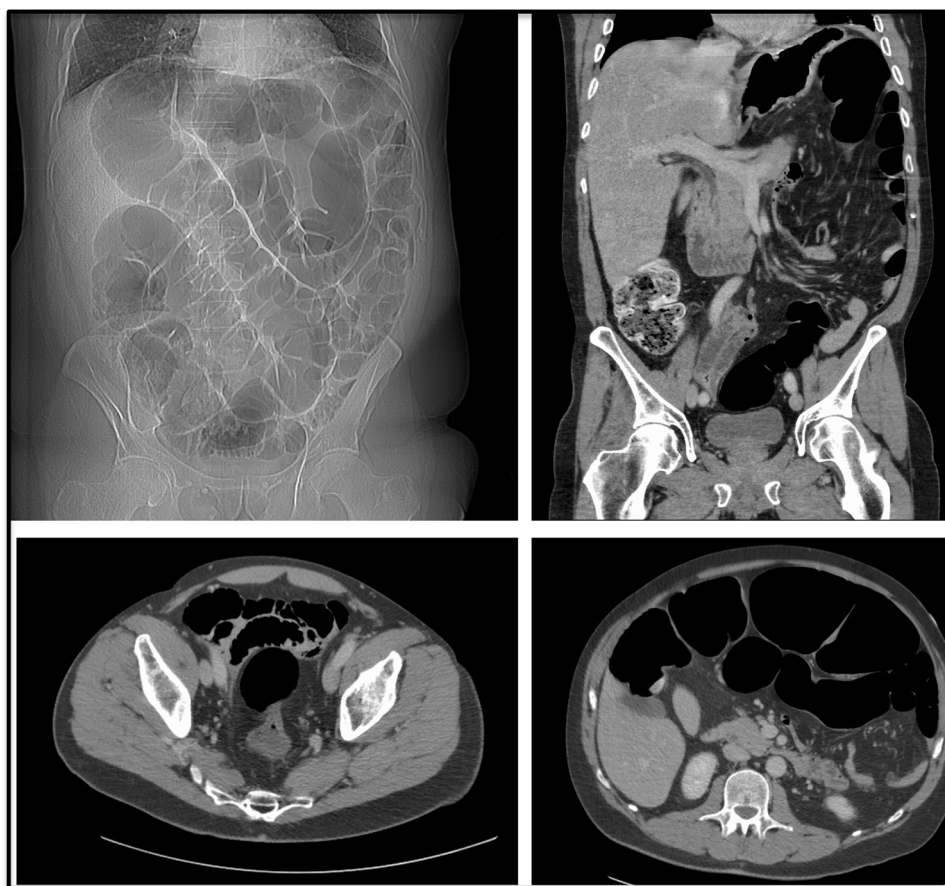


Fig. 1. Initial computed tomography (CT) abdomen and pelvis scan findings on patient presentation.

elevated white blood count of $12.53 \times 10^3/\text{cm}^2$ (89.0% neutrophils). Serum lactate was 1.7 mmol/L. A stat computed tomography (CT) scan of the abdomen and pelvis with intravenous contrast was suggestive of an acute obstruction of the sigmoid colon (Fig. 1). The imaging however did not reveal a clear transition point, was atypical for sigmoid volvulus, and there was no radiological evidence of ischemia. A rectal enema was recommended to further visualize a potential transition point, however due to the patient's worsening clinical picture, emergent operative exploration was chosen. The patient was consented and taken to the operating room for exploratory laparotomy.

A laparotomy incision was utilized in the lower midline abdomen. Upon exposure, an extremely large colon was noted (including ascending, transverse, descending, and sigmoid colon) (Fig. 2). The sigmoid mesocolon was elongated with a narrow base. No evidence of internal herniation, focal ischemic segment, or adhesive band was identified. The small bowel was run from the ligament of Treitz to the ileocecal valve, and all appeared healthy and well perfused. Based on the findings and the extremely dilated and redundant sigmoid colon, and the risk of possible intermittent sigmoid volvulus, the decision was made to perform a sigmoid colectomy with primary anastomosis. This was performed with a linear cutting stapler in a side-to-side fashion to two sized-matched regions of large bowel without tension. No diverting ostomy was created due to the satisfactory appearance of the anastomosis and negative intraoperative leak testing.

Upon evaluation by surgical pathology, the segment of resected colon (95 cm in length, 12 cm in circumference) was noted to have extreme thinning of the bowel wall throughout. Microscopically, the thin wall showed atrophic internal circumferential and exter-



Fig. 2. Intraoperative findings and gross appearance of colon.

nal longitudinal layers of the muscularis propria, with the most severe atrophy and loss of muscle fibers identified in the external longitudinal muscle layer. Ganglion cells and the myenteric plexus appeared normal in unaffected wall, and no fibrosis was noted (Fig. 3).

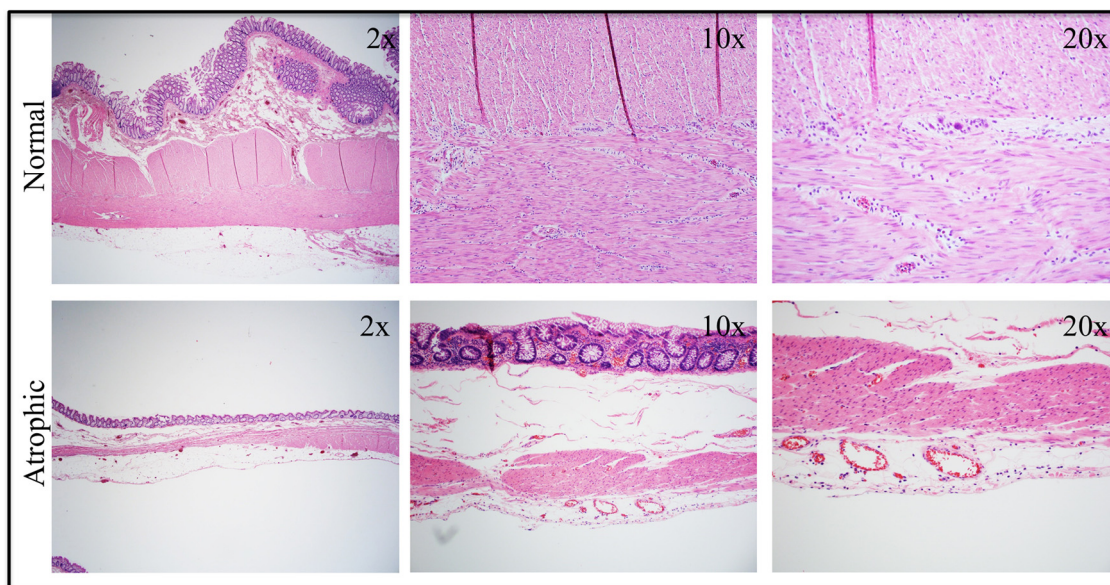


Fig. 3. Hematoxylin and Eosin (H&E) stain histopathological slides representing normal colon and atrophic visceral myopathy at 2×, 10×, and 20× magnification.

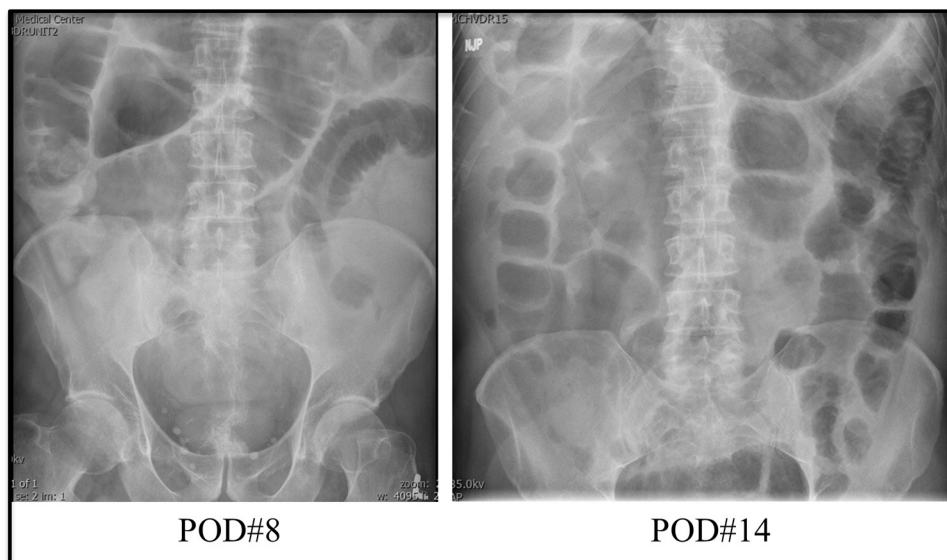


Fig. 4. Select abdominal plain films representing continued diffuse dilation of bowel in the postoperative period consistent with adynamic ileus.

Postoperatively, the patient was admitted to the general floor. He had a prolonged postoperative course with slow return of bowel function, requiring parenteral nutrition support until postoperative day fourteen (See Fig. 4). By discharge, he was tolerating a regular diet with regular flatus and multiple bowel movements.

Patient had a previously undergone screening colonoscopy one and a half years prior, with no remarkable findings (Supplemental Image 1).

Of note, patient’s levothyroxine was increased in dosage during his hospital course due to an elevated TSH (see Fig. 5). Upon further workup patient’s free T4 was found to be below the lower limit of normal range (0.7 ng/dL), consistent with clinical hypothyroidism. His repeat laboratory values at his follow-up appointment showed a normalization of his TSH.

At his one-month follow-up appointment, patient was recovering appropriately with return of normal gastrointestinal function (tolerating a regular diet with regular bowel movements). His wound was well healed and he was reportedly ready to return to

work. Patient also had follow-up arranged with an endocrinologist and gastroenterologist.

3. Discussion

Though rarely reported, the presentation of pseudo-obstruction from colonic neuropathy and myopathy has been established for 40 years [1]. Faulk et al. reported an apparently autosomal dominant visceral myopathy characterized by thinning of the longitudinal muscle as found on biopsy.

Potential causes of visceral myopathy include both familial and hereditary forms, megacystis microcolon, and scleroderma. It is also commonly associated with chronic constipation [3]. Smooth muscle motor function is vital to the various colonic processes and functions, such as transit, storage, absorption of nutrients, salts, water, stool shaping, and excretion, though the underlying mechanisms remain poorly understood [4]. This case is unusual as it presents a profound myopathy that presented in an acute man-

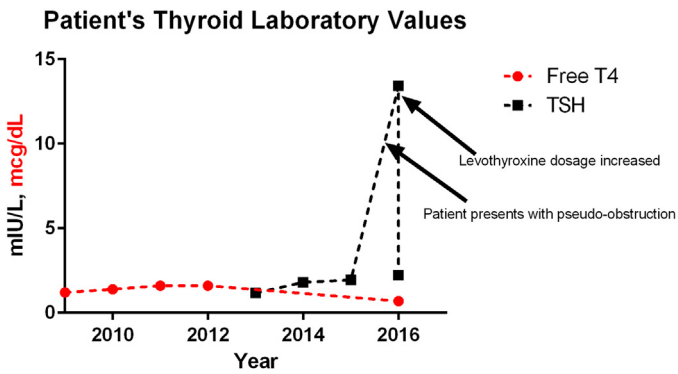


Fig. 5. Chart demonstrating patient’s acutely elevated serum thyroid stimulating hormone (TSH) and decreased serum free T4 at the time of intestinal pseudo-obstruction.

ner (with minimal to no prior symptoms), rather than in a chronic or pediatric presentation.

Perhaps the most common form of pseudo-obstruction is the acute variant, colloquially known as Ogilvie’s Syndrome. Ogilvie’s Syndrome [5] is now attributed to a variety of heterogeneous and idiopathic causes. Some of the most common causal factors include recent surgery, prolonged bed-rest, narcotic pain medication, and electrolyte abnormalities [6].

One rare potential cause of Ogilvie Syndrome is clinical hypothyroidism [7,8]. Visceral effects of thyroid derangement are previously well described, though pseudo-obstruction is a much more rare manifestation. Severe hypothyroidism may present mildly with atrophic gastritis, constipation, ileus, bloating, and flatulence, or more severely with pseudo-obstruction or megacolon [9]. This “myxedema ileus” is exceedingly rare, and can be potentially reversed with administration of exogenous thyroid hormone [10]. As our patient’s thyroid levels were below normal limits and consistent with potential clinical hypothyroidism, it is unclear if

this was a contributing or precipitating factor for his pathology. Of note, our patient did not report other more common manifestations of clinical hypothyroidism such as weight gain, fatigue, hair loss, edema, cold intolerance, or hoarseness [11].

Chronic intestinal pseudo-obstruction has been divided into multiple categories, including neuropathies, myopathies, and “mesenchymopathies” (characterized by disruption of the interstitial cells of Cajal) [6]. The classic myopathies frequently manifest histologically with fibrosis within the replaced muscle layers, which was notably absent from our patient [6]. The alimentary tract is usually diffusely involved, with symptoms often present in esophagus, stomach, and small bowel as well [12].

Large bowel obstruction can be subcategorized into mechanical (true) obstruction, and pseudo (false) obstruction. Pseudo-obstruction can be further categorized into acute and chronic, as well as primary and secondary forms. The differential diagnosis of large bowel obstruction is diverse and can make accurate diagnosis complex (See Fig. 6). Radiological examination, particularly studies with enteric contrast can assist in differentiation true luminal obstruction from pseudo-obstruction. Had our patient not showed signs of clinical decompensation necessitating urgent operative intervention, it was planned to perform a contrast enema study to rule out a pseudo-obstruction. While laparotomy and colonic resection provided tissue biopsy for the ultimate diagnosis, it is of uncertain clinical benefit in this population and should be avoided if possible. Unfortunately, this patient was not able to undergo pre-operative contrast enema, and did not have rectal contrast in his initial CT scan, as this would have allowed clear diagnostic differentiation between true mechanical obstruction and pseudo-obstruction. Colonoscopy was another potentially useful diagnostic and therapeutic tool that was not utilized due to the patient’s unstable clinical status.

While metoclopramide appeared to be of therapeutic benefit to the patient to aid in the resolution of his postoperative ileus, the drug’s mechanism of action (motilin receptor agonism) is not an

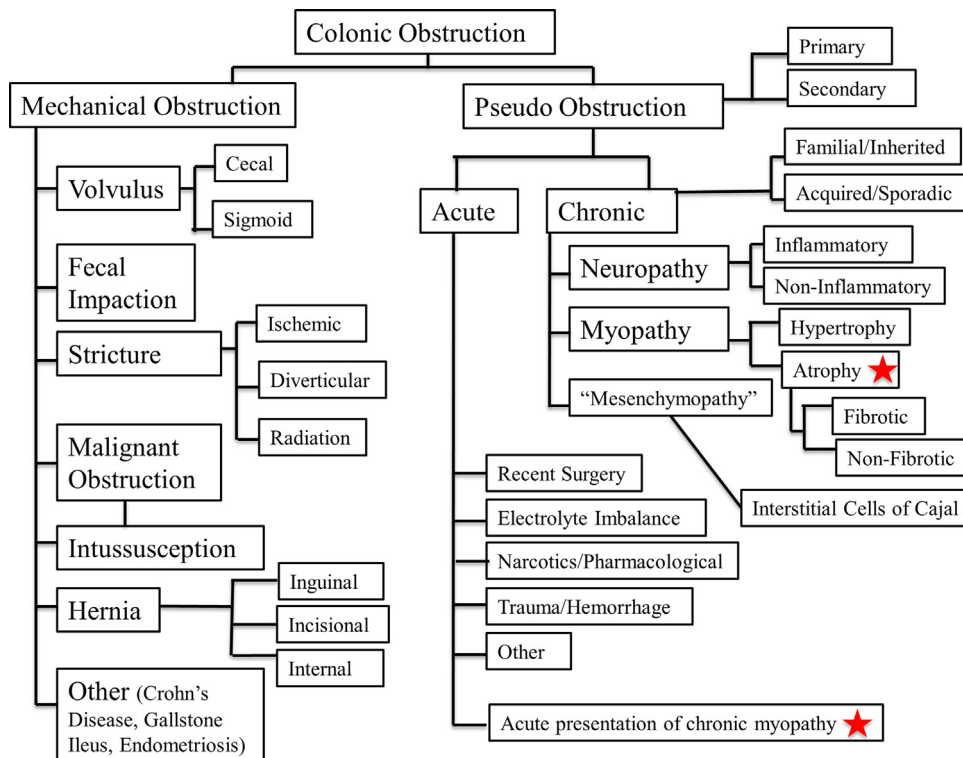


Fig. 6. Schematic representing the differential diagnosis of large bowel obstruction.

effective treatment of colonic pseudo-obstruction. It is therefore reasonable to assume that the patient's ileus involved both foregut and hindgut dysmotility (and perhaps foregut myopathy as well).

4. Patient perspective

The patient states gratitude for his care, and particularly for not receiving an ostomy, which he feels would have had a negative impact on his quality of life. He wishes to stress that the initiation of the metoclopramide caused significant relief in his postoperative ileus symptoms, and greatly facilitated his postsurgical recovery and discharge home. He endorses a slight increase in his frequency of bowel movements that has not significantly impacted him.

5. Conclusion

The presentation of acute colonic obstruction presents a diagnostic challenge, with a variety of complex etiologies and contributing factors. Radiography (particularly contrast enema), endoscopy, and manometry can help distinguish potential diagnoses, however patients presenting with hemodynamic instability may be diagnosed after emergent surgical intervention. Atrophic visceral myopathy is a rare cause of intestinal pseudo-obstruction, and has usually been reported as a chronic phenomenon. When more common risk factors are not present, it is imperative that the clinician consider more rare causes of pseudo-obstruction including hypothyroidism.

Conflicts of interest

The authors report no relevant conflict of interest related to this study.

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Ethical approval

Patient provided permission for the publication of this clinical case report. IRB approval was not otherwise obtained per the policy of our institution.

Consent

Verbal and written consent from the patient for the case report was provided and attached. All identifying details have been removed from the manuscript.

Author contributions

SW wrote the manuscript, CP critically reviewed and edited the manuscript, Michelle Yang provided the histopathological diagno-

sis, pathology images, and assisted with editing and reviewing the manuscript, AM provided final critical review of the manuscript and oversaw/devised the study.

Registration of research studies

Researchregistry1753 (UIN).

Guarantor

Sean Wrenn, MD is the guarantor for the work.

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at <http://dx.doi.org/10.1016/j.ijscr.2017.02.040>.

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