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Ankush Kalra, MD
Thomas Jefferson University, ankush.kalra@jefferson.edu

Anthony Dimarino, MD
Thomas Jefferson University, anthony.dimarino@jefferson.edu

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Chronic Idiopathic Intestinal Pseudo-Obstruction: A Working Diagnosis

Ankush Kalra, MD and Anthony J DiMarino, MD

INTRODUCTION

Chronic intestinal pseudo-obstruction (CIP) is a rare and disabling motility syndrome, yet one that demands an extensive review of digestive motility and peristaltic pathophysiology. Primarily a disorder of the small intestine, CIP was first described by Dudley and colleagues in 1958; it is defined by severe signs and symptoms of intestinal obstruction (abdominal pain and distention, nausea, vomiting, and constipation), in addition to radiographic evidence of dilated bowel in the absence of a true, mechanical obstruction. Symptoms are often slowly progressive and diagnosis requires the presence of symptoms for at least six months.[1] A 2013 national survey in Japan estimated the prevalence of CIP at 0.8 to 1.0 per 100,000, with an incidence rate of 0.21 to 0.24 per 100,000. In the same survey, the mean age at diagnosis is 63.1 years for males and 59.2 for females.[2] CIP encompasses an extensive differential diagnosis, a complex, multidisciplinary work-up, and a vast array of potential treatment options based in intricate pathophysiology.

CASE PRESENTATION:

RC is a 75-year-old male who presented with recurrent small bowel obstructions (SBOs) between August and September 2014. He has no chronic medical conditions, and his past medical history is significant only for a community-acquired pneumonia and pleural empyema at age 50. His surgical history is significant only for a right inguinal hernia repair at age 7. Initially, his symptoms began in April 2014 and were mild, limited to constipation relieved with over the counter laxatives. After two brief admissions for SBOs that resolved with nasogastric tube decompression, RC presented on September 9, 2014 with a distended, tympanitic abdomen with absence of bowel sounds and minimal tenderness to palpation. A computed tomography (CT) scan demonstrated multiple dilated loops of small bowel with a transition point in the proximal ileum. A nasogastric tube was again placed but the obstruction persisted clinically and on repeat X-Rays. During an exploratory laparoscopy on September 15, 2014, the right colon and entire small bowel were palpated. No transition zone or small bowel abnormality was found and the peritoneal surfaces of all abdominal organs appeared normal. Ultimately, an ileocecectomy was performed and RC underwent a thorough diagnostic workup.

DIFFERENTIAL DIAGNOSIS:

While CIP is a rare and elusive diagnosis, it is not one of exclusion. It is a clinical diagnosis typically confirmed by endoscopic or radiologic exclusion of a mechanical obstruction. In the case of RC, the lack of mechanical obstruction was confirmed by manual palpation of the entire small bowel and colon. The rest of the workup focused on the etiology of CIP, which may be either idiopathic or secondary. Secondary CIP was ruled out with lab testing for collagen vascular diseases, hypothyroidism, diabetes, porphyria, and celiac disease, as well as a lack of any iatrogenic factors. A thorough paraneoplastic work-up was unremarkable; Ho, Yu, and neuronal nuclear antibodies were negative, CT scan of the thorax ruled out thymoma, thyroid ultrasound revealed benign nodules, and testicular ultrasound demonstrated no masses. The ileocecectomy specimen demonstrated small bowel mucosa with prominent reactive lymphoid hyperplasia. While nonspecific, this pathology is most consistent with an inflammatory neuropathy.

OUTCOME/FOLLOW-UP:

An exploratory laparotomy performed on September 24, 2014 demonstrated significantly dilated small bowel at the ligament of Treitz. There was no evidence of mechanical obstruction such as mass, adhesions, stricture, kinking, or intussusception. Palpation of the colon was normal minus significant transverse colon dilation. Ileocecectomy was performed and a gastrostomy tube was also placed for venting. With continued symptoms and radiographic pseudo-obstruction after ileocecectomy, RC was started on total parenteral nutrition (TPN). After failure of stool softeners, laxatives, and prokinetic lubiprostone, treatment with prokinetic linaclotide was initiated. Moderate success

was achieved with the addition of somatostatin analog octreotide and antibiotic rifaximin, as RC began to move his bowels. Acetylcholinesterase inhibitor neostigmine was also administered with successful movement of the bowels within less than five minutes, further supporting a neuropathic etiology. Ultimately, RC was discharged on October 14, 2014 with linaclotide, octreotide, polyethylene glycol, and TPN. Significant progress was made over the ensuing months, as he began to tolerate oral intake, discontinued TPN, and had his medications tapered. Currently, RC only takes linaclotide twice weekly and maintains a gastrostomy tube for intermittent venting. He has returned to his normal lifestyle and has daily, formed bowel movements without symptomatic or radiographic evidence of bowel obstruction.

DISCUSSION:

Reflecting on the case of RC, it can be said that the true mechanism of his return to health remains uncertain. CIP is classified using three histological categories: neuropathies (either inflammatory or degenerative), myopathies (smooth muscle fibrosis), and mesenchymopathies (dysfunction of the pacemaker interstitial cells of Cajal). Inflammatory neuropathies are the most common cause of CIP, defined by myenteric plexus ganglionitis and encompassing etiologies such as paraneoplastic syndromes, infections, and connective tissue disorders.[1] Interestingly, most case reports describe CIP as the first presentation of another disease, most commonly small cell lung cancers, lupus, and scleroderma.[3] While the appropriate screening tests were done for these conditions, one 2012 case report describes CIP as the initial manifestation of an atypical, seronegative systemic sclerosis.[4] Octreotide has been used with much success in CIP secondary to scleroderma. Gastrointestinal transit studies have demonstrated that, while octreotide has been proven to slow intestinal transit by inhibiting intermittent, low-amplitude contractions, it enhances short burst, high amplitude contractions.[5] A 1991 study demonstrated that scleroderma patients with an inability to generate migrating motor complexes were able to produce 3.6 complexes every three hours after administration of 100 micrograms of octreotide.[6] The stasis associated with CIP is believed to generate a cycle of small intestinal bacterial overgrowth (SIBO) that leads to mucosal inflammation and further dysmotility[1], and this might explain why RC developed transverse colon dilation later on in his course. While rifaximin and other antibiotic regimens are often used to treat SIBO, it has been demonstrated that 50 micrograms of octreotide every evening for three weeks will reduce breath hydrogen excretion, a marker of SIBO, from 25 to 4 parts per million.[6] Therefore, the SIBO that is both a contributor to and consequence of CIP may be successfully treated by targeting small intestinal motility. As CIP generally has a chronic, relapsing course and overall poor prognosis, it is important to regularly monitor these patients and remain vigilant about pursuing further workup and therapies with any change in clinical status.

KEY POINTS:

The case of RC is a complicated one defined by uncertainty and a multitude of diagnostic and therapeutic interventions. While he is currently symptom free and maintained only on linaclotide twice weekly, history tells us that this case may soon be revisited and a more certain diagnosis obtained. While there is a need for physicians to become knowledgeable about CIP, the medical community must continue to inquire about targeted therapies. It is essential that these stories be shared. The pathophysiological basis of treatments used in these cases continues to be a key focus of research.

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