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Case Report

CHRONIC PSEUDO-OBSTRUCTION OF THE SIGMOID COLON: A CASE REPORT

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SUMMARY – Chronic intestinal pseudo-obstruction (CIPO) is a rare syndrome characterized by signs of intestinal obstruction lasting for 6 months or more, in the absence of a definitive cause of obstruction. We report a case of CIPO in a 49-year-old female patient with a 6-month history of ongoing irregular bowel movements, manifested as constipation and diarrhea accompanied by abdominal pain and bloated feeling. Contrast-enhanced abdominal computed tomography and magnetic resonance enterography revealed focal thickening of a segment of the lienal flexure and intermittent areas of wider and narrower caliber along the sigmoid colon. No signs of a definitive cause of obstruction were found, but evidence for dolichosigma was revealed, which was later confirmed with colonoscopy. Due to persisting symptoms, the patient agreed to elective resection of the sigmoid colon. Following the procedure, symptoms regressed with a significant improvement in the quality of life. The patient has been regularly monitored in an outpatient setting and reports absence of the symptoms since the procedure. Pathophysiology of the resected section revealed more prominent lymphatic tissue, follicular arrangement, and reactively altered germinal centers, which can suggest CIPO.

Key words: *Chronic intestinal pseudo-obstruction; Dolichosigma; Constipation; Abdominal pain; Abdominal computed tomography; Abdominal magnetic resonance enterography*

Introduction

Intestinal pseudo-obstruction was first reported by Dudley *et al*. in 1950. It is a rare syndrome of disordered peristalsis, characterized by symptoms of intestinal obstruction but lacking any cause of mechanical obstruction[1,2]. In 1978, Christensen started using the term chronic intestinal pseudo-obstruction (CIPO) to describe cases of intestinal pseudo-obstruction that persisted for more than 6 months. Due to its rarity and lack of symptom specificity, it is commonly misdiagnosed, leading to a large proportion of advanced and severe cases[3-5]. CIPO is very rare, which makes estimating the prevalence and incidence of the syndrome very difficult. Estimates suggest an incidence of 1 *per* 40000 live births, while data from the American Pseudo-Obstruction and Hirschsprung's Disease Society

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estimate that there are 100 infants born with intestinal pseudo-obstruction every year in the United States[6,7].

Chronic intestinal pseudo-obstruction mostly occurs in children and can be divided into three groups based on the etiology, which are congenital, acquired, and idiopathic. Congenital CIPOs generally seem to have a genetic component and may involve autosomal dominant, autosomal-recessive, and sex-chromosome-related inheritance. Acquired types are more common in adults and are most often secondary manifestations of systemic neurological, endocrinologic, connective tissue diseases, and malignancy. Idiopathic cases, as the name suggests, have no definitive cause[8,9].

Further division can be made for the congenital and acquired types of CIPO based on histopathologic parameters. It is typically separated into the following three groups: neuropathies, myopathies, and mesenchymal anomalies. In the case of intestinal neuropathies, the main histologic feature is neurodegeneration and dysplasia of the enteric nervous system. Intestinal myopathies are characterized by diffuse abnormalities in the muscular layering and supernumerary intestinal muscle coat. Intestinal mesenchymopathies involve abnormalities of interstitial Cajal cells and collagenous tissues of the muscularis propria[10-14].

Case Report

Chief complaints

A 49-year-old patient presented to the Emergency Department, Sestre milosrdnice University Hospital Center, complaining of constipation accompanied by pain under the left costal arch.

History of present illness

Patient history-taking revealed 6-month ongoing irregular bowel movements, involving constipation and diarrhea accompanied by abdominal pain, cramps, and bloated feeling.

History of past illness

The patient had undergone a surgical procedure due to carcinoma, which consisted of right adnexectomy and bilateral salpingectomy.

Physical examination

Upon arrival, the patient was in severe pain but alert, communicative, mobile, eupneic, and with signs of normal peripheral blood circulation. Blood pressure was 140/90 mm Hg, oxygen saturation was 98%, and body temperature was 37.5 °C. The abdomen was painful to touch in the periumbilical region, without signs of peritoneal inflammation. The liver and spleen were not palpable. The rest of physical examination showed no signs of pathology.

Imagining examination

Preliminary gastroenterological evaluation using plain abdominal x-ray and multi-slice computed tomography (MSCT) did not reveal any evidence for pathology or obstruction, only signs of dolichosigma. Further gastroenterological evaluation was carried out using contrast-enhanced abdominal CT and magnetic resonance (MR) enterography. Contrast-enhanced CT revealed colonic dilatation with formed gas-liquid levels, affecting a region stretching from the cecum to the splenic flexure, with a maximum diameter of 74 mm at the transition from the caecum to the ascending colon. Other segments were less dilated, with the transverse colon reaching a maximum diameter of 63 mm and the descending segment having a maximum diameter of 40 mm. Evidence for focal thickening of a short segment of the lienal flexure was found, along with intermittent areas of wider and narrower diameter along the dolichosigma, with a maximum diameter of up to 47 mm. No definitive cause of mechanical ileus was revealed (Fig. 1A). MR enterography confirmed the CT findings, and visualization of the transitional zone was achieved in the elongated double-curved splenic flexure region, showing only discrete haustral thickening (Fig. 1B). The use of peroral contrast was contraindicated due to suspicion of mechanical ileus, thus colonoscopy was performed to confirm the absence of mechanical obstruction. No signs of mechanical obstruction were found, but colonoscopy confirmed the findings of dolichosigma.

Laboratory examination

Blood biochemistry showed no signs of electrolyte disbalance, making 'mimickers' an unlikely cause of the patient's symptoms. Microbiological sampling turned up negative. Further evaluation by a multidisciplinary team excluded autoimmune, endocrinologic, and neurological causes of the colonic pathology that had been revealed by gastroenterological evaluation.

Definitive diagnosis was CIPO.

Treatment

After a few months, despite the use of conservative therapy with spasmolytics and fiber-rich enteral nutrition, symptoms continued to persist. The patient's con-

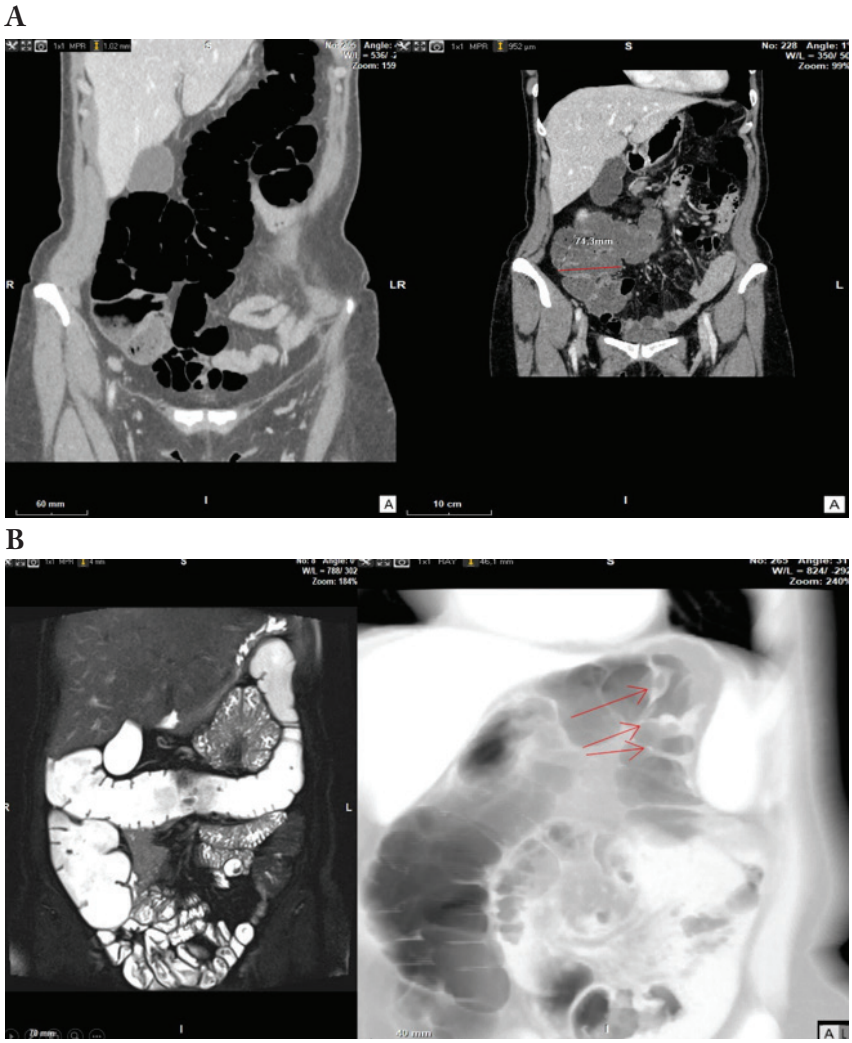

Fig. 1. Imaging findings: (A) contrast-enhanced computed tomography (CT) revealed colonic dilatation from the cecum to the splenic flexure, with no signs of a distal obstructing lesion; (B) magnetic resonance enterography confirmed the contrast-enhanced CT findings of colonic dilatation and dolichosigma. The transitional zone showed only discrete haustral thickening in the region of the elongated double curved splenic flexure (red arrows).

dition was further complicated by an episode of bacteriemia due to chronic constipation, accompanied by systemic inflammation, which was successfully treated with antibiotic treatment. Due to the patient's history of adnexectomy and salpingectomy, a multidisciplinary team suggested laparoscopic exploration for adhesions and elective resection of the sigmoid colon in the absence of any significant findings. The patient agreed to proceed with the recommended procedure. During the procedure, we found no evidence for adhesions, and the decision was made to proceed with resection of the sigmoid colon.

Outcome and follow-up

Following the procedure, the patient's symptoms regressed, with significant improvement in the quality of life. The patient has been regularly monitored in an outpatient setting and reports absence of symptoms for over 1 year since the procedure. Histopathology of the resected colonic tissue revealed more prominent lymphatic tissue, follicular arrangement, and reactively altered germinal centers. More specifically, moderate signs of focal chronic inflammatory infiltrates were found in the lamina propria, as well as edema of the submucosa and dilated vascular areas engorged with

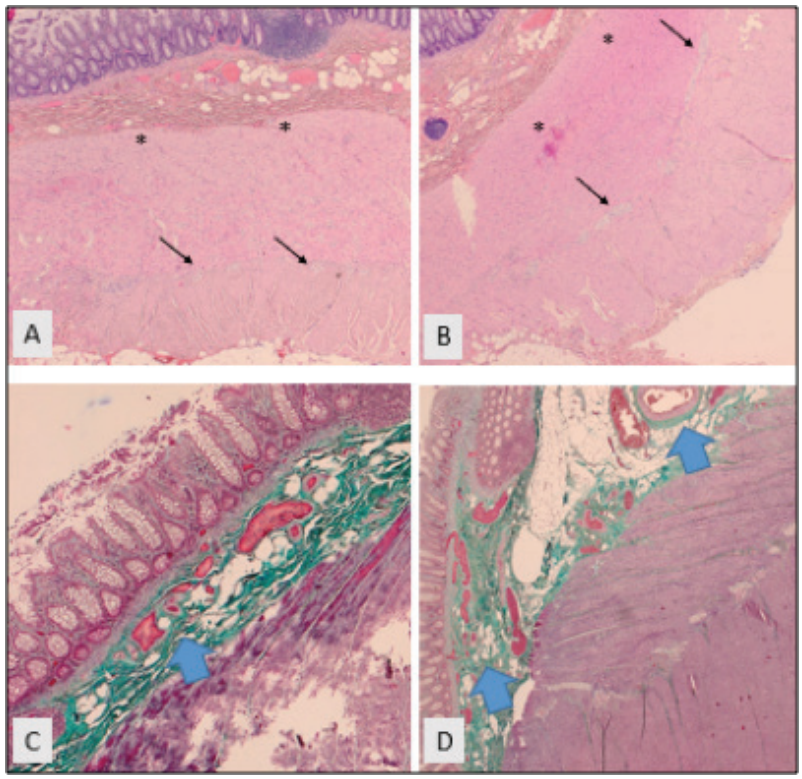

Fig. 2. Histopathologic findings: (A, B) the Meissner plexus was absent from the submucosa (*), whereas the Auerbach's myenteric plexus was preserved (black arrows); (C, D) evidence for submucosal layer proliferation and perivascular fibrosis (blue arrows).

blood. Muscularis propria showed evidence for hyperplasia and absence of the Meissner plexus, which is usually located in the submucosa between the circular muscle and the mucosa. The Auerbach's myenteric plexus, located between the longitudinal and circular muscle layers, was persevered, unlike the Meissner plexus (Fig. 2A and B). Further inspection revealed submucosal layer proliferation and perivascular fibrosis resulting from the CIPO (Fig. 2C and D). These findings are most consistent with the neuropathic type of CIPO.

Discussion

Our case is a presentation of a patient with an anatomic variant of dolichosigma that resulted in a clinical picture of subileus. Preliminary evaluation in the Emergency Room was insufficient to ascertain the cause of the patient's complaints. Full gastroenterological evaluation was necessary to elucidate the cause of the patient's constipation and exclude any mechanical obstructions. Use of peroral contrast is warranted for excluding intestinal mechanical obstructions, but the risk of contrast retention and fecal stone formation is a cause for concern, as they can aggravate the obstruction. The method has been largely replaced with the use of high-resolution CT and MR imaging[15-17]. For this reason, our primary evaluation consisted of MR enterography and contrast-enhanced CT using only an intravenous contrast medium. Conditions such as toxic megacolon and adynamic ileus are referred to as 'mimickers' because they can present with signs of large bowel obstruction, without any mechanical cause. Since these conditions are associated with electrolyte imbalances, blood biochemistry must be analyzed to exclude them as a possible cause[18]. Once definitive causes of mechanical obstruction and possible 'mimickers' are excluded, a diagnosis of CIPO is established based on the clinical manifestations, in the presence of extensive intestinal dilatation and multiple gas-liquid levels[9]. As this was the case in our patient,

the diagnosis was established, and we proceeded with conventional treatment. After a few months of no improvement, a decision was made to proceed with elective surgical removal of the dolichosigmoid segment of the colon.

Literature suggests a more conservative approach to the treatment of CIPO, minimizing surgical interventions due to concerns for the development of postoperative inhibition of peristalsis and induction of intestinal failure[9,19,20,22,23]. While this approach may be best suited for most cases, it ignores the subjective nature of the patient's ailments and pain. Our patient had periods of up to 7 days without passing any bowel content, which caused severe pain and distress. The condition often reoccurs even after successful conservative treatment and sometimes requires substantial surgical intervention following the first recurrences, such as subtotal or total colectomy[17]. Unwarranted postponement of abdominal surgical interventions can be risky if the patient's condition worsens to the point of requiring emergency surgery, which holds a greater risk of complications and has a significantly worse outcome[21]. We suggest that in some cases, elective surgery is a better option as it addresses the patient's symptoms more swiftly, reduces the risk of complications (such as the incidence of bacteriemia in our patient), and possibly helps avoid more excessive surgical interventions in the future.

Conclusion

In our patient's case, the symptoms were severe, pronounced, and long-lasting. We conclude that in the case of CIPO, the progression of symptoms should be recognized in time, and elective surgical treatment should be considered earlier when there is no response to conventional conservative therapies.

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Sažetak

KRONIČNA PSEUDO-OPSTRUKCIJA SIGMOIDNOG KOLONA – PRIKAZ SLUČAJA

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Kronična intestinalna pseudo-opstrukcija (KIPO) je vrlo rijedak sindrom obilježen znakovima intestinalne opstrukcije koji traju 6 mjeseci ili duže, a u odsutnosti definitivnog jasnog uzroka. U našem prikazu slučaja radi se o 49-godišnjoj bolesnici koja je više od 6 mjeseci imala stalne nepravilne stolice koje su se manifestirale izmjenom proljeva i konstipacije, a bile su praćene nadutošću i bolovima u trbuhu. Učinjenom obradom koja je uključivala kontrastnu kompjutoriziranu tomografiju i magnetsku enterografiju kod bolesnice je utvrđeno žarišno zadebljanje segmenta lienalne fleksure i isprekidana područja šireg i užeg kalibra duž sigmoidnog kolona. Nije nađen jasni uzrok opstrukcije, ali je otkrivena anatomska varijanta dolihosigme koja je kasnije potvrđena i kolonoskopijom. Zbog vrlo izraženih dugotrajnih simptoma koji nisu regredirali na konzervativne metode liječenja bolesnica je pristala na preporučenu elektivnu resekciju sigmoidnog kolona. Nakon zahvata simptomi su se povukli uz značajno poboljšanje kvalitete života, a bolesnica se redovito prati u gastroenterološkoj ambulanti. Patohistološki nalaz reseciranog dijela sigmoidnog kolona otkrio je istaknutije limfno tkivo, folikularni raspored i reaktivno promijenjene germinativne centre, što ide u prilog dijagnoze KIPO-a.

Ključne riječi: *Kronična intestinalna pseudo-opstrukcija; Dolihosigma; Konstipacija; Bol u trbuhu; CT abdomena; MR abdomena; Elektivna resekcija sigmoidnog kolona*