

Type I Congenital Colonic Pouch Resection With Terminal Ileostomy in a 43-year-old Woman. Case Report

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Research Article

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Abstract

INTRODUCTION: Congenital colonic pouch syndrome is an extremely rare condition, particularly in Western countries. In this condition, the colon is replaced with a partially or completely abnormal pouch, connected to the genitourinary tract by a fistula or may end in a blind pouch. As it is a congenital condition, the cases reported are found in the neonatal population, so its detection in adulthood is extremely rare.

CASE PRESENTATION: We present the case of a 43-year-old female, who was admitted to the general surgery service with suspected fistulizing CROHN's disease. She attended with a two-month clinical picture, presenting a painful ulcer in the left perianal and gluteal region, accompanied by discharge of purulent material and feces through the vaginal introitus. Laboratory tests (CAT, colonoscopy and MRI) did not present conclusive information. An exploratory laparotomy was performed, finding a congenital type I colonic pouch, so it was resected, and a terminal ileostomy was performed.

CONCLUSIONS: The present case aimed to report our experience with a pathology that has not been reported in this particular age group in our country. Imaging studies before surgery were unable to diagnose the pathology, so the exploratory laparotomy was decisive for the treatment.

LEVEL OF EVIDENCE: I

Introduction

Congenital colonic pouch (CCP) is a rare variant of ano-rectal malformations (ARM), endemic from northern India, where about 92.2% of reported cases originate (1). In this condition, the colon is replaced with a partially or completely abnormal pouch connected to the genitourinary tract by a fistula (colovesical). CCP has been included, as a rare variant, in the Krickenbeck classification of ARM (2). There are several anatomical classifications of the congenital colonic pouch (3), but basically four types are considered based on the extension of the involved colon described by Narasimharao et al. (4). In type I the normal colon is completely absent and the opening of the ileum enters directly into the colonic pouch; in type II the cecum and a segment of normal colon are present proximal to the colonic pouch; in type III the normal proximal colon extends at least to hepatic flexure, but does not reach the descending colon; in type IV the descending colon is almost normal and only the terminal portion, the rectum and a variable length of sigmoid, are included within the colonic pouch. The CP, especially in newborns, is thin-walled and distended at the expense of meconium and flatulence. However, with a wide fistula or in older children, it may be smaller and thick-walled. In Type I/II CCP, the proximal ileum or colon usually enters the pouch from right to left in a low position, close to the fistula (5). Subtypes I and II are more common in women. In most cases, the colonic pouch is reported to open into the vagina or into a persistent cloaca.

The aim of this study is to present the clinical case of a congenital colonic pouch type I, diagnosed in a 45-year-old woman who was surgically treated with a terminal ileostomy.

Case Presentation

The patient is a 43-year-old female resident of Nuevo León, Mexico, who was admitted to the general surgery department with suspected fistulizing CROHN disease. The patient denied any significant heredofamilial history. Within her personal pathological history, she was diagnosed with chronic nonspecific ulcerative colitis (CNUC) at the age of 17. Subsequently, at the age of 24 she was diagnosed with CROHN disease managed with prednisone and mesalazine and two years later biological management (infliximab) was added. At the age of 31, the patient experienced multiple episodes of urinary tract infection, and a diagnosis of rectal/vesical fistula was made. Four years later, after continuing with recurrent urinary tract infections, the patient developed pyoderma gangrenosum in the pelvic limbs, treated with thalidomide, pemecrolimus and colchicine. During 2018, CROHN disease reactivates, and she also debuts with type 2 diabetes that was managed with insulin glargine. The patient provided reports of two examinations performed two years before her admission to our service, where the following data of interest are reported: 1) Magnetic Resonance Imaging (MRI): Rectum with partial distension and presence of a marked decrease in the thickness of the posterior wall, which also shows an irregular surface suggestive of ulcerated lesion; 2) Colonoscopy: A fistulous orifice with small bowel mucosa, enteric fistula was observed (Image No.1).

Regarding the current condition, the patient came to the surgery service, referred from the gastroenterology, with a two-month clinical picture. It was characterized by a painful ulcer in the left perianal and gluteal region, with a discharge of purulent material and stool through the vaginal introitus. Vaginal examination revealed the presence of transvaginal discharge of fecal matter, scanty and foul-smelling. At the time of rectal examination, abundant fecal matter was detected in the rectal ampulla. The rectal sphincter was normotonic and no fistulous tracts or masses were palpated. The following results were obtained: 1) Colonoscopy: enterovaginal and enterocolonic fistula, with two fistulous orifices and ulcer covering 80% of the lumen; 2) CT of the abdomen and pelvis: apparent surgical changes corresponding to colectomy, ileal-colonic anastomosis, discrete thickening of inflammatory aspect at rectal level. Anomaly of the lumbosacral transition type Castellvi IIa (Supplements 1 and 2). The CAT report, which mentions surgical changes corresponding to colectomy, is noteworthy, given that the patient had no history of previous surgery. Therefore, given the ambiguity of the presumptive presurgical diagnosis (rectal/vaginal fistula), the patient was scheduled for an exploratory laparotomy.

Surgical Technique

Under general anesthesia, asepsis and antisepsis, a supra and infra umbilical incision was made in the midline. Using diathermy, the incision was performed in planes until entering the abdominal cavity, multiple firm adhesions were identified, omentum-wall, omentum-colon (colonic pouch), the ileocecal valve and the cecum were identified, which was communicating with the rectum (Image No.2). Dissection of the blind pouch Toldt's fascia and the mesocolon were performed. The inferior mesenteric artery and vein were identified, dissected, ligated and cut with 2 - 0 silk. A mesorectal window was then made and resected with an 80 mm linear stapler at the level of the sacral promontory. A section was made in the

terminal ileum 20 cm from the ileocecal valve and the specimen was extracted (Image No.3). The ileostomy was externalized in the right flank, fixed to the skin with 3 – 0 vicryl, hemostasis was performed, and closure by planes, vicryl 1 aponeurosis, 3 – 0 vicryl subcutaneous cellular tissue, 3 – 0 nylon skin, and 3 – 0 nylon skin was performed.

The trans-surgical findings are detailed as follows: Multiple omentum-abdominal wall adhesions were found, omentum-colon (Congenital colonic pouch) Zulkhe III, IV, V. Congenital colonic pouch type 1, ileocecal valve with junction to the cecum which ends in a blind pouch, where it joins with the sigmoid colon, presenting a dilatation of approximately 20–25 cm, in addition, the absence of the cecal appendix was identified (Image No.4).

Discussion And Conclusions

CCP is an unusual anomaly in which a pouch-like dilatation with a variable degree of shortening of the colon is associated with an ARM. The pouch usually ends in a fistulous communication with the genitourinary tract. The M: F ratio ranges from 2.25:1 to 7:1 (5). In this study, we present a case of CCP in a female patient. The clinical presentation is usually in the neonatal period with abdominal distension and the presence of an anorectal malformation; there may be fecaluria in 50% of cases in male patients due to communication of the urinary tract with the colonic pouch (6). Our patient has a clinical history of gastrointestinal pathology of several years of evolution, especially linked to CROHN syndrome, so the presence of a colonic pouch had never been suspected. It was not until she was 43 years old that we found it as part of an important finding, after an exploratory laparotomy. This is especially important to consider the possibility that a pathology that is normally expressed and resolved in the neonatal stage, may be present even in adulthood. In addition, it may be masked by other conditions that make its diagnosis and resolution difficult. In a retrospective study by Singh et al., where all cases of colonic pouch, managed over a period of 5 years, were recorded, the age of presentation ranged from 1 to 15 days after birth (7). Of 64 cases reviewed, coloplasty was performed on 34 occasions, with excellent cosmetic and functional results after stoma closure.

In our country the presence of this type of malformation is highly unusual, which makes its diagnostic suspicion very difficult; therefore, an adequate pre-surgical anamnesis can help to plan its approach. However, there is always the possibility that the diagnosis cannot be made until surgical exploration. It should be recalled that our patient was carefully examined with the appropriate laboratory and laboratory studies; nonetheless, the reports obtained were confusing and some even suggested the existence of sequelae of a previous, non-existent surgery. Gallegos et al., present the clinical case of a 2-day-old female whose diagnosis (Congenital colonic pouch) was made until the time of laparotomy (8). Gallegos mentions that, in most cases, radiography may be sufficient to establish the diagnosis; however, in the case they present, none of the studies performed helped to establish the diagnosis with certainty. Timely diagnosis, in addition to offering adequate treatment, allows avoiding the multiplicity of approaches aimed at other pathologies, as was the case of our patient who was 43 years old, with treatments focused on treating a fistula secondary to CROHN's disease.

List Of Abreviations

ARM

ano-rectal malformations

CCP

Congenital colonic pouch

CNUC

Chronic Nonspecific Ulcerative Colitis

CT

Computed Axial Tomography

MRI

Magnetic Resonance Imaging

Declarations

- Ethics approval and consent to participate: Autograph consent was obtained from the patient for the presentation and publication of her case.
- Consent for publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.
- A copy of the written consent is available for review by the Editor-in-Chief of this journal.
- Availability of data and materials: All data provided are available in the medical file located within the "Hospital Valentín Gomez Farías del Instituto de Seguridad y Servicios Sociales de los Trabajadores del Estado", in Zapopan Jalisco, Mexico.
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- Authors' contributions: CGJ, OMJ, EPR, SGM, TSQL were involved in the patient's care. All authors have contributed substantially to collecting data, revising the article, and given necessary intellectual inputs. All authors read and approved the final manuscript.
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Figures



Figure 1

Colonoscopy (Entero-enteric fistula)



Figure 2

Congenital colonic pouch resection type I + ileostomy ends

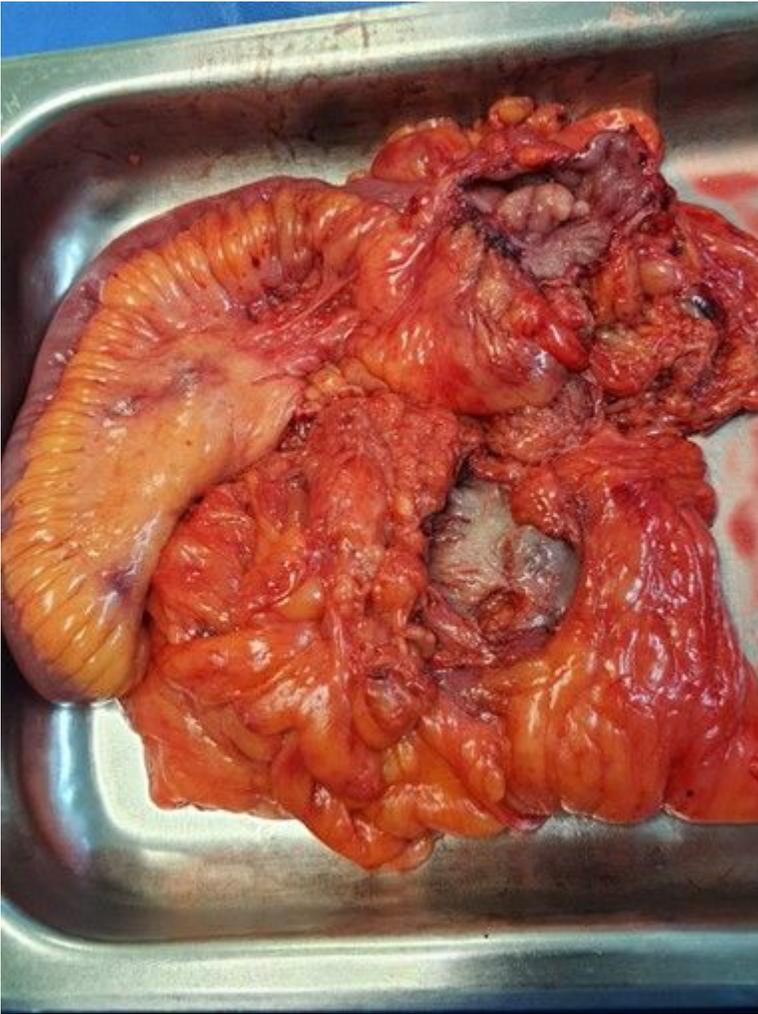


Figure 3

Extraction of the piece, identifying the ileocecal valve with union to the cecum which ends in a blind pouch



Figure 4

Congenital colonic pouch type 1

Supplementary Files

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