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Case Report



Total Proctocolectomy Resulting from Toxic Megacolon in A Young Patient with Ulcerative Rectocolitis: Systematic Review and Case Report

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Abstract

Introduction: Ulcerative colitis is a chronic, relapsing inflammatory bowel disease, generally beginning between 20 and 40 years old, with an etiology that is still under debate.

Methods: based on the PRISMA method, articles related to "toxic megacolon; ulcerative colitis" published in the last 5 years were identified, as well as comparing them with a case of a young patient with a rare presentation of toxic megacolon requiring total proctocolectomy.

Case report: we report a rare case of ulcerative colitis without previous signs, culminating in perforated toxic megacolon requiring total proctocolectomy, with good postoperative results, describing the actions taken by the general surgery team at a reference hospital.

Discussion: Total proctocolectomy with anal anastomosis with ileal pouch represents the standard approach for patients with refractory UC or complications, including toxic megacolon. The exact moment to perform surgery in patients with moderate to severe UC still remains a challenge, but several factors must be considered and medication alternatives must be offered to patients without risk factors for complications.

Conclusion: it is necessary to individualize the treatment plan and the same patient may require multiple modalities to achieve remission or improvement in quality of life.

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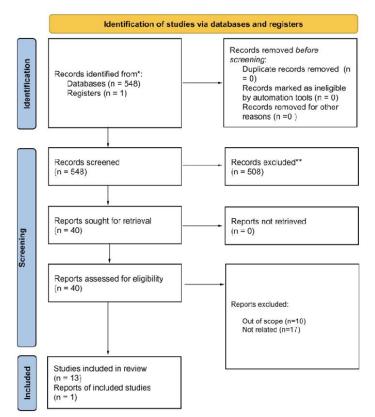
Keywords: Perforated megacolon; Total proctocolectomy; Toxic megacolon; Ulcerative colitis

Introduction

Ulcerative Colitis (UC) is an idiopathic chronic inflammatory bowel disease of the colon that generates continuous inflammation of the mucosa, being characterized by a relapsing and remitting pathology. The most common symptoms include bloody diarrhea with tenesmus and rectal urgency. The onset of the disease is mainly observed between 20 and 40 years of age [1,2]. Although the etiology is still under debate, more evidence is emerging regarding underlying autoimmune components. In this article, we discuss ulcerative colitis in its clinical aspects and treatment, as well as its association with toxic megacolon, using recent literature and comparing it with a rare case described in a young patient in a reference service.

Methods

This study was based on the methodological procedures described in Preferred Report Items for Systematic Reviews and Metaanalysis (PRISMA) of the experimental type to identify, select and critically evaluate research already published on the exposed topic. The review was carried out on original articles available in the PubMed databases, and were selected using the terms: "toxic megacolon; ulcerative colitis", which were determined based on previous studies on the topic. The selection criteria were studies, available in full, published in the last 5 years. Articles that were not related to the topic studied or the research question were excluded.



Case Report

Woman, 22 years old, white, with no relevant personal or family history, was admitted to the emergency room in a reference service reporting persistent diarrhea for 3 weeks, having been hospitalized 9 days ago with the medical clinic team to investigate the condition. However, she escaped from the hospital after 3 days of hospitalization, aware of the risks. Although, the patient returned to the service one month after the incident, reporting a worsening of her pain and progressive abdominal distension, associated with massive watery diarrhea, and no longer presenting with hematochezia. Upon examination of the patient, she was in poor general condition, dehydrated and discolored, with a distended abdomen, diffusely decreased bowel sounds, hypertympanic percussion and intense diffuse pain on deep palpation, but without signs of peritonitis. The tests requested in the emergency room showed marked inflammatory tests (CRP 355.5), significant anemia (Hb 7.4; Ht 22.1), absence of leukocytosis but with an increase in rods, and a slight change in the coagulogram (INR 1.38). A total abdominal tomography with contrast was also performed, which had as changes a marked liquid and gaseous distension of the colic frame, with the formation of levels, noting foci of parietal pneumatosis in the cecum and proximal ascending colon, in addition to mild distension of the distal ileum with air-fluid levels and a small amount of free liquid in the pelvic excavation (Figure 1).

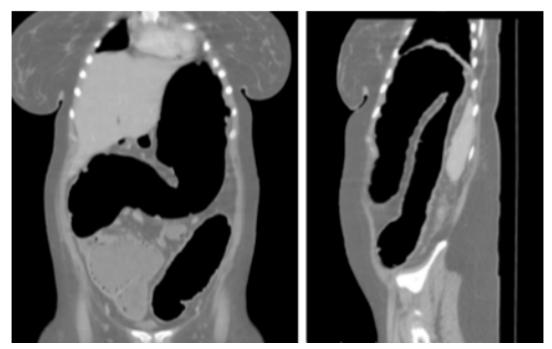


Figure 1: Abdominopelvic computed tomography with contrast, showing liquid and gaseous distension of the colic frame, with formation of levels, noting foci of parietal pneumatosis in the cecum and proximal ascending colon, in addition to distension of the distal ileum with air-fluid levels and a small amount of free fluid in the pelvic excavation.

Due to the condition and laboratory and imaging tests, the patient was hospitalized by the general surgery team with a surgical plan for exploratory laparotomy, due to suspicion of toxic megacolon. She was kept fasting, with an open nasogastric tube in drainage, 2 bags of plasma were transfused, 2 bags of packed red blood cells, and ciprofloxacin and metronidazole were prescribed as antibiotic therapy, and vitamin K. During the preparation time for surgery, the patient's condition worsened, presenting on examination with signs of peritonitis and increased abdominal distension. The patient was therefore taken to the surgical center where a median xiphopubic laparotomy was performed, opening the planes in the same direction. An inventory of the cavity was carried out where multiple perforation points were observed throughout the colon, a large amount of feces in the abdominal cavity. It was then decided to perform a total Proctocolectomy, with the terminal ileum being stapled with the aid of a 75 mm linear stapler, and subsequently the told line was sectioned, detaching the right colon, hepatic angle, left colon and splenic angle. The transverse mesocolon omentum was then separated, with access to the retroperitoneum, ligation and section of the inferior mesenteric vein and inferior mesenteric artery close to the origin of the vessels. The ileocolic and middle colic arteries were also ligated and sectioned, with subsequent excision of the colon, completing excision of the low rectum with the aid of Contour. The cavity was then washed with 0.9% saline solution, and a circular incision was made on the right flank

to create and mature a terminal ileostomy. A silicone drain was placed in the topography of the rectal stump and externalization on the left flank. After reviewing hemostasis, plane closure was performed and the specimen was sent to the anatomopathologist.

The report of the anatomopathological examination of a surgical specimen composed of a segment of small intestine, ileocecal valve, cecal appendix and segment of large intestine was subsequently released, confirming the hypothesis of toxic megacolon, showing a picture of extensively ulcerated chronic inflammation, as an etiology Ulcerative colitis (Figure 2).



Figure 2: Product of total proctocolectomy affected by ulcerative colitis after resection for toxic megacolon.

After the surgical procedure, the patient was sent to the ICU, presenting hemodynamic stability due to vasoactive drugs (VAD), but still in serious condition. Subsequently, the patient evolved with clinical and hemodynamic stability without the use of DVA and sedation, reporting improvement in abdominal pain, with a functioning ileostomy, being discharged to the infirmary ward.

Results

Reference	Ago/sex	Predisposing factors	Clinic	Entrance path	Treatment	surgery and outcome
Luciano; et al, 2023	22-year-old, fernale	History of ulcerative colitis at the age of 5.	abdominal pain nausea bilious emesis tachycardic, mildly hypertensive diffuse abdominal tenderness mild distention toxic megacolon	abdominal pain, nausea and bilious emesis. Time of symptom onset not described.	Not described	urgent exploratory laparotomy, total abdominal colectomy with end-ileostomy; discharged after 7 days
Rohman; et al, 2024	12 years old, female	Not described	Abdominal pain Poor appetite Diarrhea Weight loss Tachycardia Microcytis anemia Hypoolbumiernia Abdominal tenderness Dilated transverse colon Pneumonia Basal hung nodules Basal Bowel perforation	Four-week abdominal pain, poor appetile, diarrhea and weight loss history	Not described	Subtotal colectomy and ileostomy, discharged
Hayashi; et al, 2023	59-year-old, male	Not described	Ulcerative lesion Dilatistico of the transverse colon Pneuronia Covid-19 Liver dysfunction Jaundice anemia (hth=7.9g/dL)	Complain of abdominal pain and bloody stools. Time of symptom onset not described	Initiated on day 1 with prednisolone (80 mg/day) Initiated on day 4 tacrolimus (4 mg/day)	Subtotal colectomy and ileostomy, <u>retai</u> mucous fistala, discharged after 77 days post operation. Rectal resection and ileoreetal anastomosis eight months post-operation
Apostolov; et al, 2020	24-year-old, male	Not described	Extreme frequent evacuation Tachyc ardic Chronic colitis Dilatation of the transverse colon C. difficile infection	2 week history of lose and bloody bowel actions, abdominal discomfort	Not described	Colectomy, discharged after 2 weeks
Morosanu; et al, 2022	42-year-old, female	psoriasis in Ixekizumab therapy; Smoker	diarthea abdominal pain recial bloeding fever anemin (Hb=10.9g/dL) electrolytes imbalances hypoalbuminemia toxic megacolon	abdominal pain and diarrhea for 7 days (after Ixekizumab administration)	Initiated on day 3 with infliximab (5 mg/kg)	total colectomy with ileostomy and rectum preservation; discharged after 7 days
Garate, et al, 2021	22-year-old, female	Not described	diarthea bernatochezia addominal pain weight loss anemia (Hb=8.1g/dL) C. difficile infection toxic megacolon	diarthea, hematochezia and intense abdominal pain for 2 month	Initiated on day 4 with Hydrocortiso ne (300 mg/day) IV; on 8 day, initiated infliximab (5 mg/kg)	no surgery required;
Pomerantz, et al, 2021	39-year-old, male	five-year history of moderate ulcerative colitis	C. diff infection Cheneic diarrhea nauxen vomiting bematochozia rectal perforation	abdominal and rectal pain for one week	Not described	urgent exploratory laparotomy, total abdominal colectomy and end ileostomy due; No secondary follow-up surgery was required

Table 1: Compares the reports found in the literature [3-9]. These are studies with 1 or more patients described, comparing them based on common factors, such as gender, age at onset of symptoms, clinic and treatment, as well as prognosis.

It is worth highlighting the scarce literature on patients suffering from UC after total proctocolectomy, as well as the similarity with the case presented in this article. In fact, the most important relationship seems to be found in the pattern of age and sex, since UC apparently affects a greater number of female patients, with 57.1% compared to males with 42.8%. Furthermore, it is inferred

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that younger women (under 30 years of age) are at greater risk of complications and the need for surgery, accounting for 42.8% of cases, while young men account for only 14.2%. However, as men age, their risk of UC increases to 28.5%, while women remain at 14.2%. Furthermore, factors such as earlier onset of symptoms culminated in more serious complications, requiring urgent surgery, such as a case similar to the one reported in this article, of a young woman with toxic megacolon. Regarding treatment, 71.4% of patients underwent total proctocolectomy and 100% had a good prognosis, with subsequent discharge and follow-up. However, no possible complications such as infections, wound dehiscence or ICU admission were reported.

Discussion

Epidemiologically, some risk factors have been described, such as age, genetics, microbiota and diet. UC has a bimodal age distribution with a peak incidence in the second or third decades of life, followed by a second peak between 50-80 years of age. Around 8-14% of patients have a family history of inflammatory bowel disease and some markers such as the TNFSF15 (TL1A) locus, which increases the risk of UC with more serious complications, have been identified. It is assumed that the development of IBD is an immunological response to food. The Western-style diet is associated with an increased risk of developing UC, as well as increased intake of total fat, animal fat, and polyunsaturated fatty acids. Several studies point to dysbiosis of the intestinal microbiota in IBD, as this altered composition of commensal bacterial populations leads to dysregulation of the immune response to bacterial antigens [1]. Clinically, UC presents with bloody diarrhea, rectal urgency, tenesmus and varying degrees of abdominal pain, which is relieved after defecation. Typically, symptoms have intermittent attacks that can become more frequent and occasionally severe, which require hospitalization. The diagnosis depends on the aforementioned clinical features associated with histological findings on endoscopy and biopsy. Recent studies suggest that patients at higher risk of relapse are younger, with extensive disease, previous relapses, and fecal calprotectin greater than 190 ug/g on repeat tests [1]. Risk factors associated with increased disease extent include obesity, history of appendectomy, and severe disease activity. Over the years, many scoring systems have been developed to assess disease activity, such as the modified Riley score, the Gupta index, and the Geboes scoring system [10].

A feared complication in UC is toxic megacolon, in which the local inflammatory process can disrupt the neuromuscular function of the colon, leading to its dilation and subsequent perforation. TM is characterized by dilation of the colon (\geq 5-6 cm), along with signs of systemic toxicity, and is often associated with infectious colitis,

which can lead to mortality if not treated [5]. The goals of treatment focus on improving quality of life, remission and minimizing the risk of cancer. The basic approach is based on severity, distribution, age of onset, duration of the disease, evolution, frequency of relapse, previous medication, medication side effects and extraintestinal manifestations. Severe acute ulcerative colitis is defined as six or more bloody bowel movements per day, associated with other systemic signs and symptoms. The therapeutic approach includes timely diagnosis and exclusion of enteric infections, with methylprednisolone 60 mg for 24 hours normally used as initial therapy. For patients who do not respond to corticosteroid therapy, we should consider colectomy in 3 to 5 days, as delays in surgery can significantly increase mortality. Other important points include fluid and electrolyte replacement, flexible sigmoidoscopy to rule out infections, stool cultures, nutritional support and DVT prophylaxis. The most commonly performed surgery is restorative proctocolectomy with ileal pouch-anal anastomosis [1,11].

Total proctocolectomy with anal anastomosis with ileal pouch represents the standard approach for patients with refractory UC or complications, including toxic megacolon. Studies show that early and late complications, mainly infections, can occur in more than 20% of patients, while long-term complications occur in less than 10% of patients [2]. However, proctocolectomy is associated with a significant improvement in patients' quality of life. The exact moment to perform surgery in patients with moderate to severe UC still remains a challenge, but several factors must be considered and medication alternatives must be offered to patients without risk factors for complications. The impact of the disease on the patient's quality of life, predictive factors of negative evolution and risk of side effects with medications must be taken into account. When both therapeutic options are valid, discussion with a multidisciplinary team is essential, balancing potential risks and benefits. The patient always needs to be well informed about the steps necessary to obtain a fully functional pouch, potential limitations and complications postoperatively and after restoration of intestinal flow [2,12].

Conclusion

Predicting the prognosis of patients with UC is very difficult, requiring individualization of the treatment plan and the same patient may require multiple modalities to achieve remission or improvement in quality of life. In summary, this is a rare presentation of ulcerative colitis in a young patient, without comorbidities or previous history, with rapid progression to perforated toxic megacolon. Fortunately, timely total proctocolectomy allowed for greater survival for the patient in question, as well as a better quality of life, even without adjuvant treatments.

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