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Toxic Megacolon in A Patient with Ulcerative Colitis and A History of Hirschprung's Disease Case Report

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1. Introduction

Hirschsprung's disease (HD) is the most common cause of functional intestinal obstruction in paediatric patients. It is characterized by contiguous intestinal ganglionitis of the myenteric plexuses of Auerbach and the submucosal plexus of Meissner, extending proximally from the rectum in variable degrees, with a histological transition zone from ganglionic to a ganglionic occurring before a normally innervated area [1].The rectosigmoid form is the classic presentation (75-80%); long-segment colon disease (10-15%) or total colon ganglionitis with ileal involvement (5-7%) are uncommon [2].Clinically, it most frequently presents as intestinal obstruction of the colon, or in newborns with a history of failure to evacuate meconium within 24-48 hours of birth; other clinical manifestations include abdominal distension, non-bilious vomiting, and constipation [3]. Ulcerative colitis (UC) is a chronic disease of the colon mucosa that results from the interaction of genetic and environmental factors [4].Characterized by mucosal inflammation that begins distally and likely extends proximally, eventually involving the entire colon. It exhibits a bimodal distribution with respect to its incidence, with an initial peak between 20 and 30 years of age and a second peak between 50 and 80 years of age, affecting men and women equally; It has shown an increase in incidence in recent years [5]. The clinical course is unpredictable and is characterized by episodes of remission and relapses or exacerbations. The clinical presentation includes bloody diarrhoea, increased bowel frequency, abdominal pain, fatigue, and faecal incontinence [6].

2. Clinical Case

A 23-year-old male, originally from and residing in the state of Hidalgo, with no significant family history. He had a history of constipation since he was four months old. He underwent a Lynn biopsy, which revealed no ganglion cells in the various sections of intestine sent for pathology, and was found to have Hirschsprung disease.

Surgical history: Diverting colostomy due to a ganglionic megacolon (Hirschsprung's disease) on 12/00/1999. Duhamel's descent was performed on 6/24/2000, with histopathological report showing absence of ganglion cells in the myenteric and submucosal nerve plexuses, with a single nerve level of eleven in total. Colostomy closure on 10/11/2000, with no apparent complications.The illness began on 08/03/2022 with intense hypogastric pain, abdominal distension and the presence of rectal bleeding, in addition to reporting weight loss of 11 kg in two weeks. He was evaluated in a private setting where a colonoscopy was performed on 08/28/2022, showing a rectum with ulcerated mucosa, with a 5x4 cm ulcer, in addition to loss of haustra, concluding acute proctitis due to probable ulcerative colitis (Image 1). Histopathological report on 07/31/2022 reporting abundant inflammatory infiltrate composed of polymorphonuclear cells, lymphocytes and plasma cells, identifying data of mild epithelial regeneration, with abundant areas of hemorrhage and necrosis, concluding ulcerative colitis with data of activity (Image 2).During hospital staypersists with approximately eight evacuations of haematochezia characteristics daily, with transfusion requirements, iscompleted mint diagnostic protocol with C-reactive protein (PCR), viral panel, TORCH and AB toxins, with negative results. Adjusting treatment with steroid at a rate of hydrocortisone 100 mg daily, with a PCR result of 154 despite said management. HO index is performed (Table 2) with a result of 6 points, with a probability greater than 85% of not responding to medical treatment. In addition, according to Oxford criteria (Table 3) with 85% probability of meriting surgical management due to number of bowel movements and lack of response to treatment. Presenting an unfavourable evolution meeting clinical, biochemical and radiographic criteria for toxic megacolon, as an acute complication of UC. Therefore, surgical management is decided, being a patient a candidate for total proctectomy plus terminal ileostomy and placement of a left JJ catheter.The patient was taken to the operating room on 10/6/2022 under balanced general anaesthesia. The patient underwent a suprainfraumbilical incision, revealing multiple loop-to-loop, loop-wall, and loop-rectum adhesions with their release. A megacolon was found with maximum dilation in the transverse region, as well as interloop enema. Told's fascia was released, dissecting until the mesentery layer of the distal ileum was separated, followed by separation of the transverse colon from the greater omentum; continuing with the release of the rectum in the presacral plane, transection, and closure of the same, achieving dissection and release of the ascending colon, and extracting the surgical specimen (Image 6). A Noble maneuver was performed, with subsequent exteriorization of the matured terminal ileostomy using the Brooke technique. Total blood loss was 600 cc.

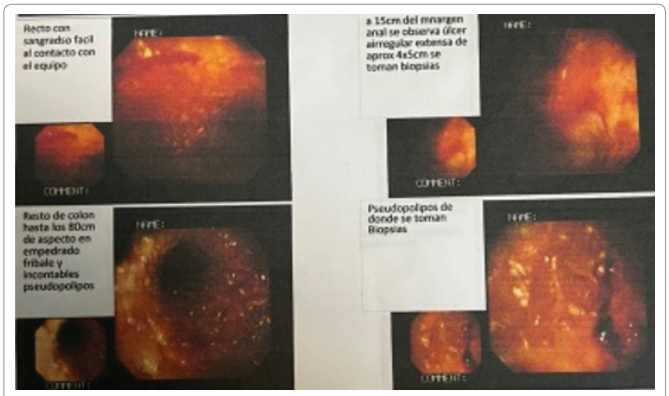

Image 1: Rectum with bleeding feasy to touch, with an extensive irregular ulcer measuring 4x3 cm at 15 cm from the anal margin.

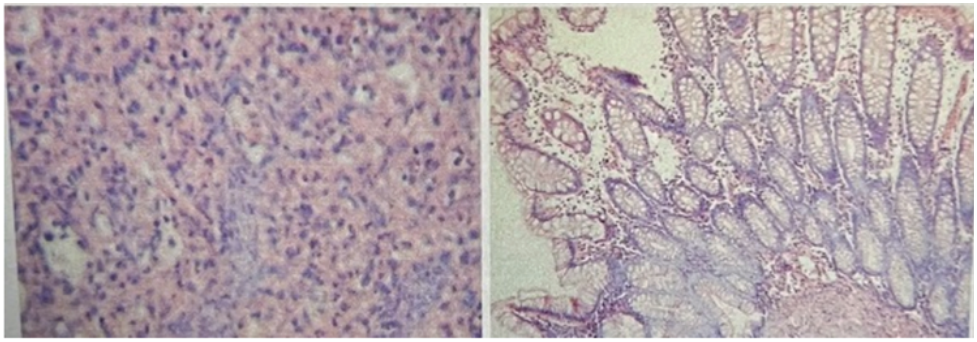

Image 2: Sections stained with hematoxylin and eosin, observing a solution of continuity of the mucosa and the thickness.

Table 1: ClassiMontreal establishment. Basedontheextensionofthe CU.

Termino	Distribucion	Descripcion
E1	Proctitis	Afectacion limitada al recto (esto es, la extension proximal de la unión rectosigmoidea)
E2	Colitis izquierda	Afectacidnlimitada a la portion del colon distal a la flexuraesplenica (analoga a «colitis distaU)
E3	Extensa	La afectacion se extiende hasta la flexuraesplenica, incluye pancolitis

Table 2: Adjustingtreatmentwithsteroid at a rateofhydrocortisone 100 mg daily, with a PCR resultof 154 despitesaidmanagement. HO indexisperformed.

Evaluaciondia 3-5	Puntajc
n de deposicionesen 24 h	
<4	0
4-6	1
7-9	2
>9	4
Dilatacion de colon: Presente	4
Hipoalbuminemia< 3,0 g/dL	1

Table 3: Travis (Oxford) index to estimate risk of corticosteroid refractoriness and needforcolectomy.

Evaluation dia 3-5	Riesgo
> 8 deposiciones en 24 h	Valor predictivopositivo de necesidad de colectomia: 85%
3-8 deposicionesen 24 h + proteina C reactiva> 45 mg/L	

Image 3: Abdominal X-ray, showing dilation of the ascending and transverse colon.

3. Discussion

The history of HD in the previously described case is known as short-segment disease, affecting the rectum and a small area of the sigmoid colon. It is the most common type, with a prevalence of up to 80%. It is diagnosed postnatally and is less severe. Infants younger than 6 months should be evaluated with high levels of suspicion: symptoms of intestinal obstruction (biliary vomiting, abdominal distension, failure to evacuate), failure to evacuate meconium within 48 hours of birth, history of constipation or family history of HD, Trisomy [21].Rectal biopsy is the gold standard for confirming the diagnosis, with the sample taken at least 2 cm posterior to the dentate line to avoid the physiological aganglionic zone. However, it should be preceded by non-invasive tests to support the diagnosis, such as colon enema and/or anorectal manometry. The diagnosis is established by the absence of ganglion cells in the rectal biopsy, including the presence of nerve fibre hypertrophy, increased acetylcholinesterase activity, and decreased or absent calretinin-immunoreactive fibres in the lamina propria.

The primary objective is surgical resolution of the pathology. The goal is to respect the affected segment of the rectum and colon, lowering the intestine with normal lymph node levels below the anastomosis, and preserving the function of the internal anal sphincter.Regarding the presentation of UC in this clinical case, it is important to highlight the increasing prevalence of this disease in patients of this age group. This is also important to stage severity based on clinical presentation and biochemical parameters according to the modified TLW criteria (Table 4), in order to assess in-hospital treatment with intravenous steroids, as well as the risk of progression to a second line of treatment, including the risk of colectomy [7].Using endoscopic studies, such as the Montreal classification for assessing disease spread, as part of the diagnostic and staging protocol for disease activity, taking into account the Mayo score (Table 5), which is more frequently used due to its simplicity and applicability; however, the UCEIS (Ulcerative Colitis Endoscopic Index of Severity) is the most valid score for assessing severity [8].SomeIndices or criteria such as those of Travis (Oxford) and Ho have been developed to identify patients who, on the third day of intravenous steroid treatment, are at greater risk of failure and require second-line therapy or surgical management. The use of both criteria is proposed to identify a more unfavourable outcome in the management of UC, taking into account the number of bowel movements, CRP levels, colonic dilation, and albumin levels among their variables [9].The mega colorn toxic is a total or segmental colonic dilation that occurs in the context of systemic toxicity, mostly considered a complication of inflammatory bowel disease type UC, to a lesser extent it can occur in patients with Crohn's disease (CD), or

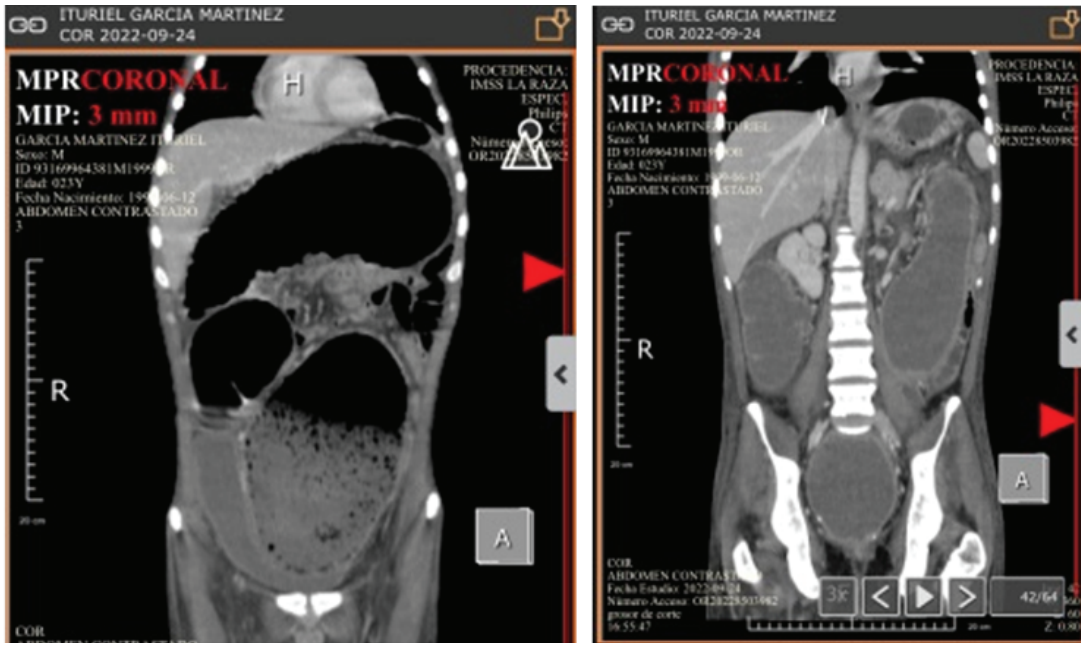

Image 4 and 5: Partially distended stomach, with diffuse increase in colonic dimensions, ascending colon of 7 cm, transverse colon of 10 cm and descending colon of 14 cm, mucosa with enhancement upon passage of contrast, dilation of the left renal pelvis of 30 mm and ipsilateral ureter of 19 mm.

any inflammatory or infectious condition process of the colon such as infection by Chloridoids and Cytomegalovirus, hence the importance of ruling out these pathologies. Being the way in which up to 5-10% of patients with UC debut, being caused by certain mechanisms such as the increase in the production of nitric oxide (NO) synthase by macrophages and smooth muscle cells, which inhibits intestinal tone favoring dilation; in addition to the typical mucosal inflammation, damage to the myenteric plexus of the colon has been related; as well as the use of Sulfasalazine and 5-ASA which can precipitate this pathology and must be restarted until it is resolved [10].The diagnosis of this presentation was based on patients with abdominal distension and diarrhoea, combined with clinical findings of inflammatory response and evidence of colonic dilatation, using the following criteria broadly:

1. Radiographic evidence of colonic dilation (greater than 6 cm).
2. Plus at least 3 of the following: fever greater than 38°C, heart rate greater than 120 bpm, leukocytosis greater than 10,500 with neutrophilia, anemia.
3. Plus at least 1 of the following: dehydration, altered alertness, fluid and electrolyte disturbances, hypotension.

The following cases are indicative of surgical management:

l. Emergency: perforation, massive hemorrhage, multiple organ dysfunction syndrome (MODS).
2. Urgentcia: severe acute colitis (5-7 days after failure of medical therapy), toxic megacolon.
3. Elective: failure of medical therapy, stricture formation, extraintestinal manifestations, dysplasia or cancer.

Surgical management the surgical management of UC represents a challenge. Treatment failure is the most common indication in chronic patients. However, this depends on the staging of the disease and the patient's condition. Up to 34% of patients in this group require colectomy. The optimal procedure is total colectomy with Brooke ileostomy in most cases, taking into account the duration or permanence of the stoma. It presents an overall mortality rate of less than 1%, with a morbidity rate of around 30%. Both open and laparoscopic approaches are appropriate in the emergency setting; an open approach is preferred in cases of perforation, in order to achieve damage control [11,12].

Image 6: Product of total proctolectomy secondary to toxic megacolon.

4. Conclusion

UC requires multidisciplinary management due to its complexity and variety of presentations, with the goal of achieving the best functional response and minimizing the need for a stoma as much as possible. The complexity of the clinical case presented demonstrates the combination of two pathologies that directly affect the colon, adding to the diagnostic and therapeutic challenges they entail.

Table 4: ModifiedTrueLove-Wittscriteria. ESR = Erythrocytesedimentationrate.

Pantuación	3 puntos	2 puntos	I punto
Numero de deposiciones	>6	4-6	<4
Sangre en las deposiciones	++++++	+	-
Hemoglobina 2/1 Hombre	<10	10-14	> 14
Mujer	< 10	10-12	>12
Albumina (g/1)	<30	30-32	>32
Fiebre ("C)	>38	37-38	<37
Taquicardia	>100	80-100	<80
VSG	>30	15-30	<15
Leucocitos (N 1.000)	>13	10-13	< 10
Potasio	<3	3-3.8	>3.8
Inactive: <11	Brote lwave: 11-15	Brote moderado: 16-21	Brote grave: 22-27

Table 5: Mayo score. Grades of endoscopic findings.

GRADO	HALLAZGOS ENDOSCÓPICOS
0	Normal O enfermedod inactiva
1	Enfermedad leve: eritema, disminución del patrón vascular, leve friabilidad.
2	Enfermedad moderada: eritema acentuado, ausencia del patrón vascular, friabilidad, erosiones.
3	Enfermedad grave: sangrado espontáneo, ulceración.

References

1. Das K, Mohanty S. Hirschsprung disease - current diagnosis and management. Indian Journal of Pediatrics. 2017; 84(8): 618-623.
2. Esperanza Lopez M, Singer J, Hoppin A.Congenital aganglionic megacolon (Hirschprung disease). 2022.
3. Kale A, Badwaik A, DhulseP, Maurya A, Kurian B.A Case Report on 2 years Child: Hirschprung's Disease. Journal of Pharmaceutical Research International. 2021; 558-564.
4. Gallo G, Kotze PG, Spinelli A. Surgery in ulcerative colitis: When? How?, Best Practice & Research Clinical Gastroenterology. 2018.
5. Yamamoto-Furusho JK, Gutierrez-Grobe Y, Lopez-Gomez JG.The Mexican Consensus Group on Idiopathic Chronic Ulcerative Colitis. Journal of Gastroenterology of Mexico (English Edition). 2018; 83(2): 144-167.
6. FotedarS. Toxic megacolon: A rare but lethal complication of ulcerative colitis. The Journal of the Association of Physicians of India. 2022; 70(11): 88-88.
7. SegalJP, LeBlanc JF, Hart AL. Ulcerative colitis: an update. Clinical Medicine (London, England). 2021; 21(2): 135-139.
8. Coimbra R, Celentano V, Wani I. WSES-AAST guidelines: management of inflammatory bowel disease in the emergency setting. World Journal of Emergency Surgery. 2021; 16(1): 23.
9. Rubin DT, Ananthakrishnan AN, Siegel CA, Sauer BG. ACG clinical guideline: Ulcerative colitis in adults: Ulcerative colitis in adults. The American Journal of Gastroenterology. 2019; 114(3): 384-413.
10. Iborra M, Fernández-de la Varga M, BeltránB. Diagnostic and therapeutic protocol for severe acute forms of ulcerative colitis. Medicine. 2020; 13(11): 635-641.
11. Feuerstein JD, Moss, AC, Farraye FA. Ulcerative colitis. Mayo Clinic Proceedings. Mayo Clinic. 2019; 94(7): 1357-1373.
12. Nakamura H, Lim T, PuriP. Inflammatory bowel disease in patients with Hirschsprung's disease: a systematic review and meta-analysis. Pediatric Surgery International. 2018; 34(2): 149-154.