SURGERY INSIGHTS

MIDITERRANEAN JOURNALS

Case Report

DOI: 10.55085/si.2022.601

Idiopathic Localized Dilation of Ileum

Alberto Robles Méndez Hernández[1], Roberto Jauregui Brechu[1], Oscar Alejandro Mora Torres[1], Andrew M. Sorsby Vargas[1], Gabriel Isaac Castro Enríquez[1], Erick Mauricio Aceves Rodríguez[1], Sandra Berenice Somarriba Domínguez[2]

[1] General Surgery Department, Hospital Angeles Metropolitano, Mexico city, Mexico.
[2] Department of Internal Medicine, Hospital Angeles Metropolitano, Mexico city, Mexico.

Received: 24 Nov 2021
Revised: 28 Avr 2022
Accepted: 13 May 2022
Published: 27 Jun 2022

Academic Editor:
Jaume Tur-Martínez

Correspondence: Alberto Robles Méndez Hernández. General Surgery Department, Hospital Angeles Metropolitano, Mexico city, Mexico.
Email: albertormh01@gmail.com

Cite this article as: Hernández ARM, Brechu RJ, Torres OAM, Vargas AMS, Enríquez GIC, Rodríguez EMA, Domínguez SBS. Idiopathic Localized Dilation of Ileum. Surg Insights. 2022;1:601. [https://doi.org/10.55085/si.2022.601]

Authors' contributions
The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript, and provided approval for this final revised version.

Acknowledgements
We want to thank the pathology service of our hospital, over all Dr Víctor Hugo Méndez Cano for helping us with the diagnosis.

Funding
The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests
None declared.

Patient consent for publication
Obtained.

ABSTRACT

Introduction: Idiopathic localized dilation of the ileum (ILDI) is rare in adults, debuting with nonspecific symptomatology.
Case report: a 36-year-old male patient with no past surgical history with intense abdominal pain and acute abdomen upon physical examination and CT imaging evidence of intestinal occlusion. Initial laparoscopic approach, a 30 cm terminal ileum mass with multiple inflammatory mesenteric nodes was found and resected. Post-surgically, further laboratory evaluations failed to justify any other diagnosis rather than ILDI, excluding histopathologic and laboratory compatible with abdominal cocoon syndrome. The patient was discharged without complications and no recurrence of ILDI.
Conclusions: Idiopathic localized dilation of the ileum is a rare pathology specially on adults, in children is often associated to other mid-gut congenital anomalies, gastrointestinal blood loss, with or without chronic anemia, and small-bowel obstruction, ILDI should be considered as a exclusion differential diagnose of unrelated intestinal obstruction.

Keywords: Gastrointestinal, Surgery, Acute, Abdomen.

INTRODUCTION

Idiopathic localized dilation of the ileum (ILDI), also called segmental dilation of the small bowel, (1) ileal dysgenesis, (2) segmental megaileum, (3, 4) and giant Meckel´s diverticulum refers to a rare, small bowel lesion of unclear etiology. Studies suggest a congenital origin as it has been associated with other mid-gut congenital anomalies. It is characterized as a well-defined aperistaltic dilated segment of the ileum with abrupt proximal and distal transition to normal bowel, most presented in neonates. No association between ILDI and abnormal ganglia innervation has been found. Clinical manifestations include abdominal pain, intermittent bowel obstruction and occult intestinal bleeding. (5-7) Only 19 cases of ILDI in adults have been described since 1955. (4, 8-14).

CASE REPORT

A 36-year-old male patient with medical history includes bipolar disorder on treatment with Lithium Carbonate and no surgical history, 16 hours onset of symptoms with abdominal mesogastrium pain associated nausea and vomiting on 3 occasions, at physical examination distended acute abdomen is found. His hemoglobin was 17.7g/L, leucocyte was 16.7 ×109/L with neutrophilia of 93%. The CT findings were a transition zone in the right lower quadrant, with no evident observable masses (Figure 1).
A diagnostic laparoscopic is performed and firm loop-loop adhesions on terminal ileum and 30 cm small intestine dilatation, inguinal region was normal, ileocecal valve and appendix preserved, rest of the abdominal cavity without findings, conversion to open surgery was performed. The ileal dilatation was resected (Figures 2 and 3) and ileo-ileal end-to-end anastomosis in two layers war made for reconstruction.

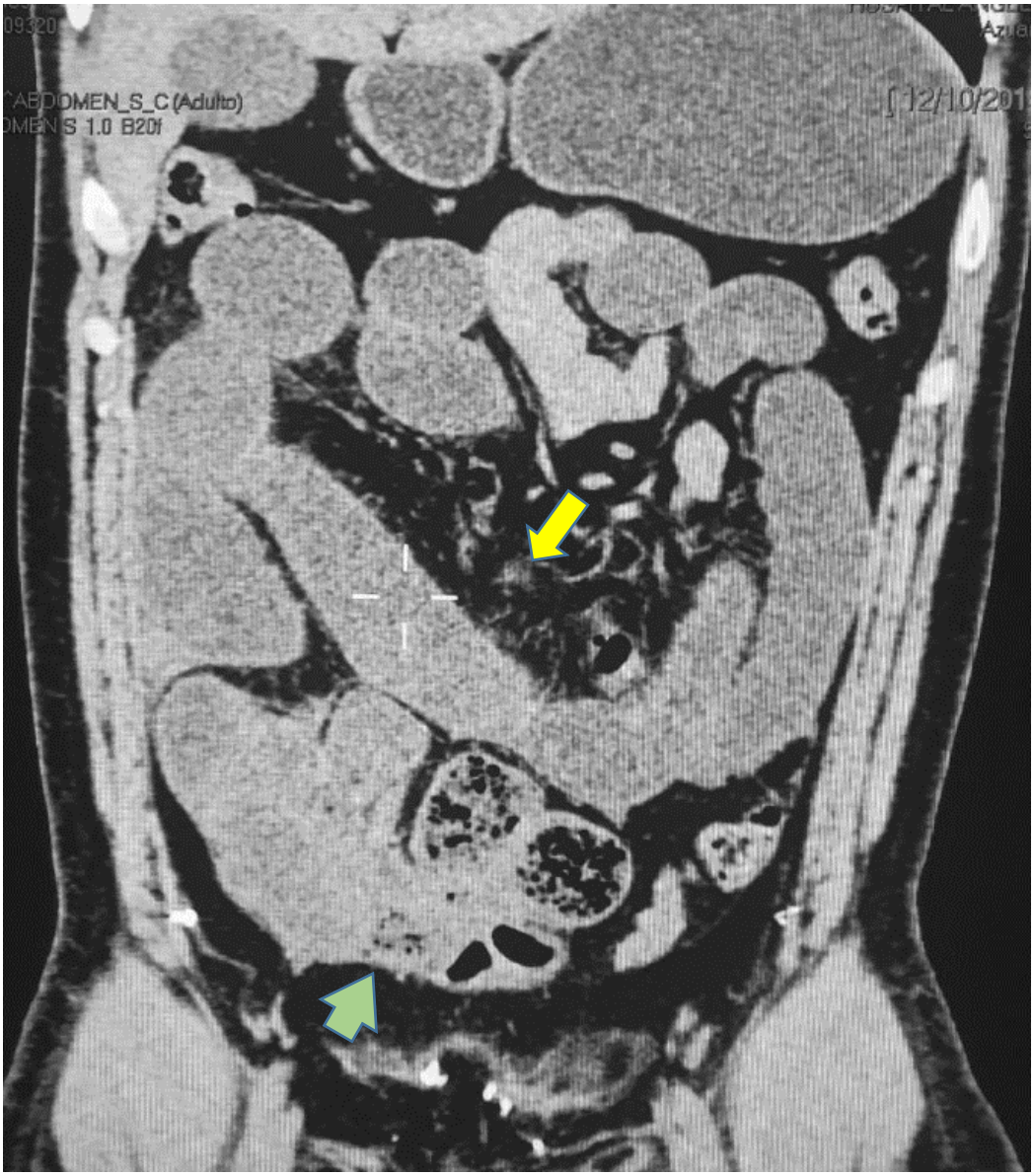

Figure 1. Coronal section of computed axial tomography image in which multiple loops of small intestine with hydro-air levels and inter-edema are observed (green arrow) with striation of intestinal fat (yellow arrow).

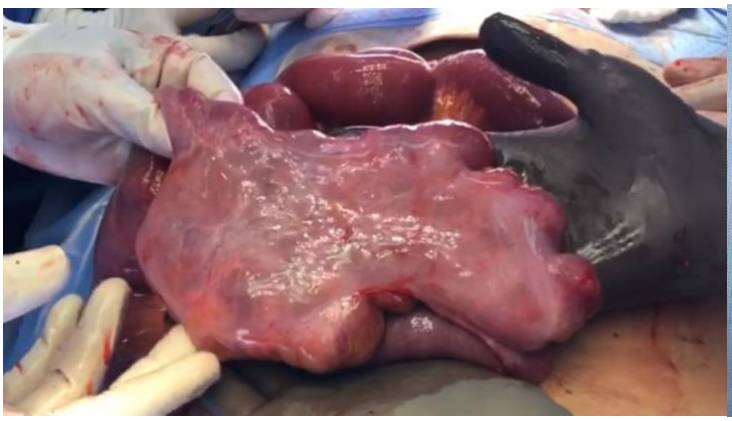

Figure 2. Dilated ileum during surgery. As no dissection plane was observed with suspicion of malignancy, no peritoneal sac was dissected.

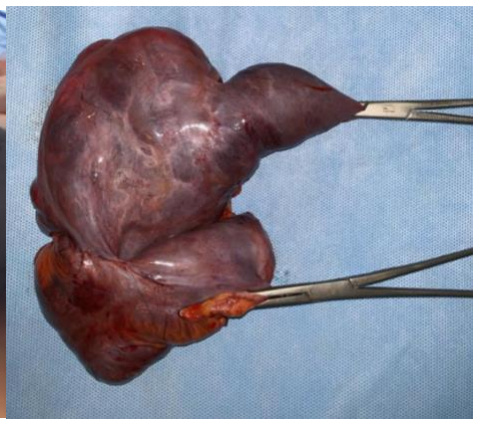

Figure 3. Macroscopic view of the resected dilated ileum.

Following the surgical procedure we approached differential etiology with: IgA, IgG, IgM, IgE, complement C3, C4, C5, antigenic and functional determination of C1 inhibitor and urinary porphobilinogen values were reported normal; HIV and hepatitis serology, ANA, C-ANCAS, P-ANCAS, anti-Saccharomyces Cerevisiae antibodies, anti-DNA antibodies, anti-Actin-F antibodies, anti-acetylcholine receptor antibodies, anti-CCP antibodies, anti-SSA

antibodies, anti-SSB antibodies, anti-SM antibodies, anti-Musk antibodies, anti-LA antibodies, anti-Ro/SSA antibodies and LE cells were negative.
Histopathological analysis of the resected dilated segment revealed minimal changes in the mucosa (image 4.A), vascular congestion along with interstitial edema in submucosa (Figure 4.A) and chronic inflammatory infiltrate along with fibroblastic proliferation in subserosa. (Figure 4.B, 4.C, 4.D). Negative staining for amyloid protein, IgG4 and tuberculosis.
The patient was discharged within 5 days, semestral follow-up was performed during 3 years without recurrent disease.

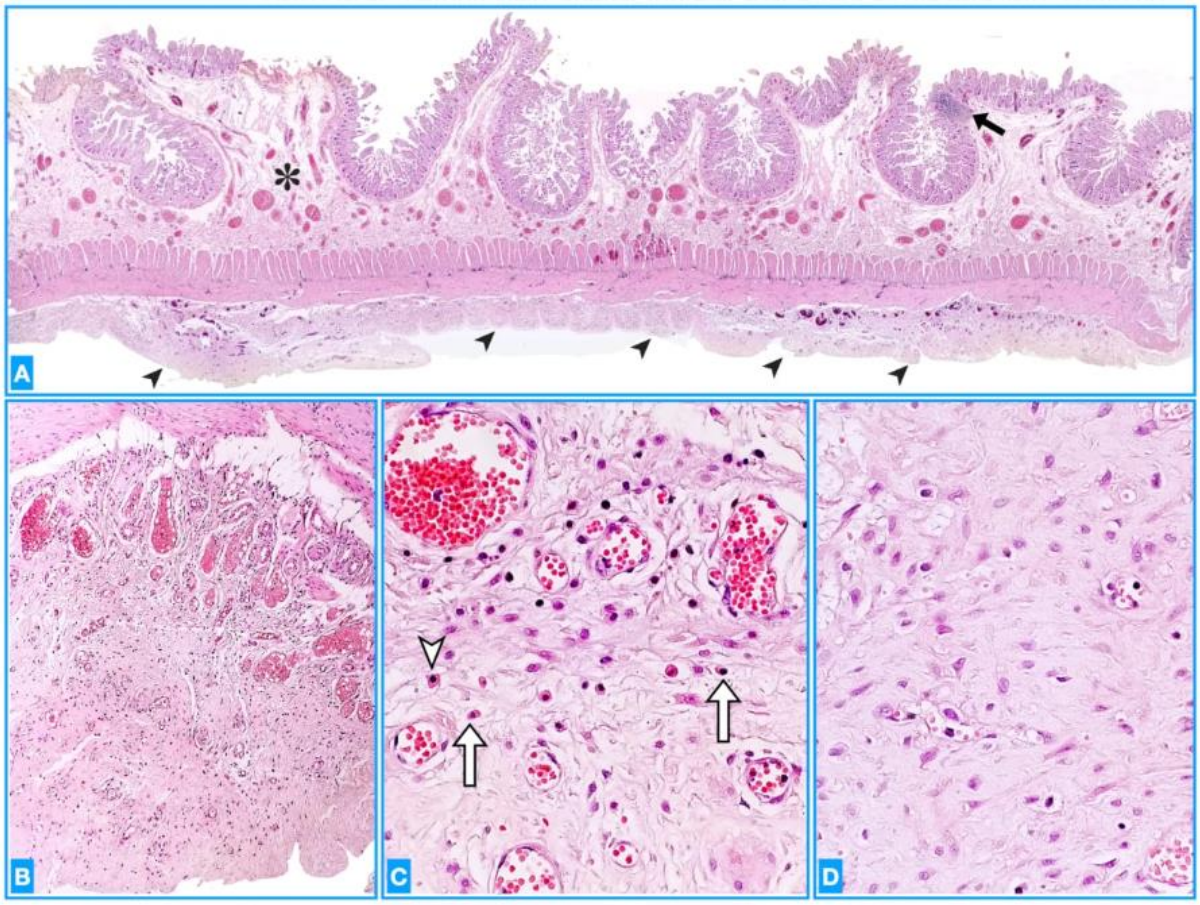

Figure 4. Resected tissue analysis (**A**, 10 x ‘) shows mucosa with minimal changes and preserved lymphoid aggregates (black arrow), submucosa with vascular congestion and interstitial edema (asterisk) and subserosa with thickened appearance (black arrowheads). Under greater augmentation (**B**, 200 x ´; **C**, 400 x ‘) the latter reveals superficial dilated capillaries with perivascular chronic inflammatory infiltrate of plasma cells (empty arrows) and mast cells (empty arrowhead) and deep fibroblastic proliferation. Fibroblasts (**D**, 400 x ‘) are randomly distributed between collagen fibers.

DISCUSSION

Since the first reported case of idiopathic intestinal dilation by Marshall et al (1955) (15), to this date it has been a challenge to accurately determine the pathophysiology of this disease, maintaining it as an exclusion diagnosis. It is most frequently identified in ileum or colon, but rarely in jejunum or duodenum. (16)
There is a lack in the literature concerning idiopathic intestinal dilation in adult patients. A systematic review by Rotigliano et al (2019) (14) found a total of 64 case reports in pediatric patients and 18 more cases in adults, of which 7 (39%) were women and 11 (61%) men, ranging from 25 to 72 years of age with a median of 48 years. This pathology is more frequently identified in pediatric population under 10 years of age, prompting a possible congenital origin, no signs of congenital disorders were found on our patient, showcasing correlation with congenital alterations of the midgut that could cause intrauterine obstruction such as omphalocele, malrotation or Meckel's diverticulum in 50% of cases (1). Another hypothesis suggests an idiopathic neuromuscular dysfunction of a localized loop responsible for the shortfall on peristaltic movements. (9) Approximately 80% of cases coexist with anemia secondary to gastrointestinal bleeding of variable magnitudes, (8) due to ulceration within the dilated segment. The latter is hypothesized to be the result of ectopic mucosa (which has been found in few cases) (4) or mechanical ischemia due to intermittent volvulus and intussusception (1). 50% of cases, patients may also present with abdominal pain related to intestinal obstruction (8). In 89% of reported cases the diagnose was made by intestinal

transit, 17% by CT, and in few by sonography (14). Even though evidence suggests this is a detectable pathology by imaging, pre-operative diagnosis is complicated due to unspecific signs, usually mimics tumors, intestinal diverticula, scleroderma, inflammatory bowel disease, infiltrative diseases, Cocoon syndrome or dilatation proximal to injury. Therefore, definite diagnosis depends on exclusion of the other conditions. The treatment of choice is segmental resection and transit restitution (9).

Evidence of IDLI at histopathological analysis describes a small intestine wall with diminished width in all its layers without alterations in its plexuses. (10) Nevertheless, during the surgical intervention of this case we observed a singular macroscopic anatomy with localized thickening of the visceral peritoneum without an apparent plane of dissection, as well as dilation of the distal ileum. Subsequent analysis described thickening of the subserosa with chronic inflammatory changes and ruled out infiltrative, tumorous, or autoimmune etiologies, as well as adherent and obstructive processes

CONCLUSION

Idiopathic localized dilation of the ileum (ILDI) is a rare pathology specially on adults, in children is often associated to other mid-gut congenital anomalies, gastrointestinal blood loss, with or without chronic anemia, and small-bowel obstruction, ILDI should be considered as a exclusion differential diagnose of unrelated intestinal obstruction. The recommended treatment of ILDI in symptomatic patients is surgical resection of the dilated segment. Nerve plexuses and ganglion cells remain unaltered in the pathological analysis of the resected tissue to meet the criteria for ILDI.

REFERENCES

[1] Ratcliffe J, TaitJ, Lisle D, Leditschko JF, Bell J. Segmental dilatation of the small bowel: report of three cases and literature review. Radiology 1989; 171:827-830

[2] Bell MJ, Ternberg JL, Bower RJ. Ileal dysgenesis in infants and children. J Pediatr Surg. 1982 Aug;17(4):395-9. doi: 10.1016/s0022-3468(82)80497-1

[3] Sawada S, Izumi S. A case report of mega-ileum. Getkahokan (Arch Japan Chirg) 1962; 31:792

[4] Leinster SJ, Hughes LE. Segmental mega-iloum presenting as anaemia. BrJ Surg 1981 68:41 7-419

[5] Lee J, Lee S, Hwang J. Segmental dilatation of the ileum: a report of two cases. Acta Radiologica. 2006;47(5):525-528.

[6] Brown A, Carty H. Segmental dilatation of ileum. Br J Radiol 1984; 57:371–4.

[7] Irving IM, Lister J. Segmental dilatation of the ileum. J Pediatr Surg 1977; 12:103–12.

[8] Javors BR, Gold RP, Ghahremani GG, et al. Idiopathic localized dilatation of the ileum in adults: findings on barium studies. AJR Am J Roentgenol 1995; 164:87–90.

[9] Usselman JA, Ghahremani GG, Bordin GM, et al. Idiopathic localized dilatation of the ileum in adults. Gastrointest Radiol 1981; 6:313–7.

[10] Simpson S, Hollinshead J, Katelaris PH. Idiopathic localized dilatation of the ileum. A rare cause of gastrointestinal haemorrhage in an adult. J Gastroenterol Hepatol 1998; 13:1234–6.

[11] Kim YS, Kim Y, Park DW, et al. Idiopathic localized dilatation of the ileum: CT findings. Abdom Imaging 2002; 27:15–17.

[12] Yang DM, Kim HC, Jin W, et al. Imaging findings of idiopathic localized dilatation of the ileum. Br J Radiol 2008;81: e48–9.

[13] Kim HS, Kim YW. Idiopathic localized dilatation of the ileum. Intern Med 2012; 51:2981–2.

[14] Rotigliano, N., Sauter, M., von Flüe, M., & Steinemann, D. C. (2019). Idiopathic localised dilatation of the intestine in adults. BMJ Case Reports 2019, 12(8), e230443. doi:10.1136/bcr-2019-230443.

[15] Michel ML, Field RJ, Ogden WW. Meckel's diverticulum; an analysis of one hundred cases and the report of a giant diverticulum and of four cases occurring within the same immediate family. Ann Surg 1955; 141:819–29.

[16] Sakaguchi T, Hamada Y, Masumoto K, et al. Segmental dilatation of the intestine: results of a nationwide survey in Japan. Pediatr Surg Int 2015; 31:1073–6