Revista Española de Enfermedades Digestivas
The Spanish Journal of Gastroenterology

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DOI: 10.17235/reed.2023.9630/2023
Link: PubMed (Epub ahead of print)

Please cite this article as:
Borrego Rivas Sandra, Domínguez Carbajo Ana Belén, Martín Izquierdo Alia, Díez Ruiz Sandra, Latras Cortés Irene, Jorquera Plaza Francisco. When intestinal obstruction is not a surgical condition. Rev Esp Enferm Dig 2023. doi: 10.17235/reed.2023.9630/2023.

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CC 9630 inglés

When intestinal obstruction is not a surgical condition

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Conflict of interest: the authors declare no conflict of interest.

Keywords: Chronic. Pseudo-obstruction.

Dear Editor,

Chronic intestinal pseudo-obstruction is a syndrome characterized by episodes of intense, cramping pain, abdominal distention and vomiting, with radiological images simulating an obstruction, but with no underlying mechanical etiology. These symptoms may present acutely, simulating a real obstruction, or chronically. Its etiology is an abnormal intestinal motility, due to muscular or neurological disorders, or both.

As proposed in another letter to the editor published in this journal, the diagnostic algorithm for chronic pseudo-obstruction must include the possibility of a paraneoplastic manifestation of an unidentified tumor. They are uncommon, with often unrecognized syndromes, high morbidity and disabling complications, which require a high level of suspicion for a proper diagnosis.

Case report

We report the case of a 43-year-old male with a history of Parkes-Weber syndrome and appendectomy. He was hospitalized for surgery three times. The first one at 36

years old, with a computed tomography (CT) scan that suggested obstruction of the distal ileum; a laparoscopic adhesiolysis was performed. On the second stay at the hospital, the CT scan reported an occlusion in the distal ileum, probably caused by an intraperitoneal adhesion. Another laparoscopy was performed, which revealed dilatation of the jejunoileal loops with no cause. After the third admission, the patient was referred to Hospital Vall d'Hebron, where gastrointestinal manometry was performed, establishing the diagnosis of chronic intestinal pseudo-obstruction. Finally, two new intestinal subocclusive episodes occurred, which were resolved with prokinetics and laxatives.

Afterwards, considering the possibility of a visceral myopathy as an etiology of the pseudo-obstruction, a genetic study was carried out, which confirmed a heterozygous mutation of the ACTG2 gene, compatible with megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) or Berdon's syndrome. It is characterized by smooth muscle dysfunction affecting the bladder and the gastrointestinal tract, with heterogeneous phenotypes. The most common urological symptoms range from megacystis to "prune belly" syndrome in the most extreme forms. In the gastrointestinal system, it usually debuts as malrotations or microcolon. Adult patients most commonly suffer from chronic intestinal pseudo-obstruction. Acute pseudo-obstructive episodes can be managed with prokinetics, always avoiding intestinal resections. Thus, we must ensure an adequate nutritional intake.

Our patient's progression was satisfactory and no further hospitalizations were required. The diagnosis was concluded with a urodynamic study which confirmed a megacystis bladder. The patient is currently on maintenance treatment based on osmotic laxatives, prokinetics and non-absorbable antibiotics cyclically, to prevent bacterial overgrowth. Furthermore, nocturnal octreotide is included as a stimulant of the migratory motor complex and nutritional supplements. Episodes of abdominal pain are resolved with pyridostigmine at home.

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de Enfermedades Digestivas
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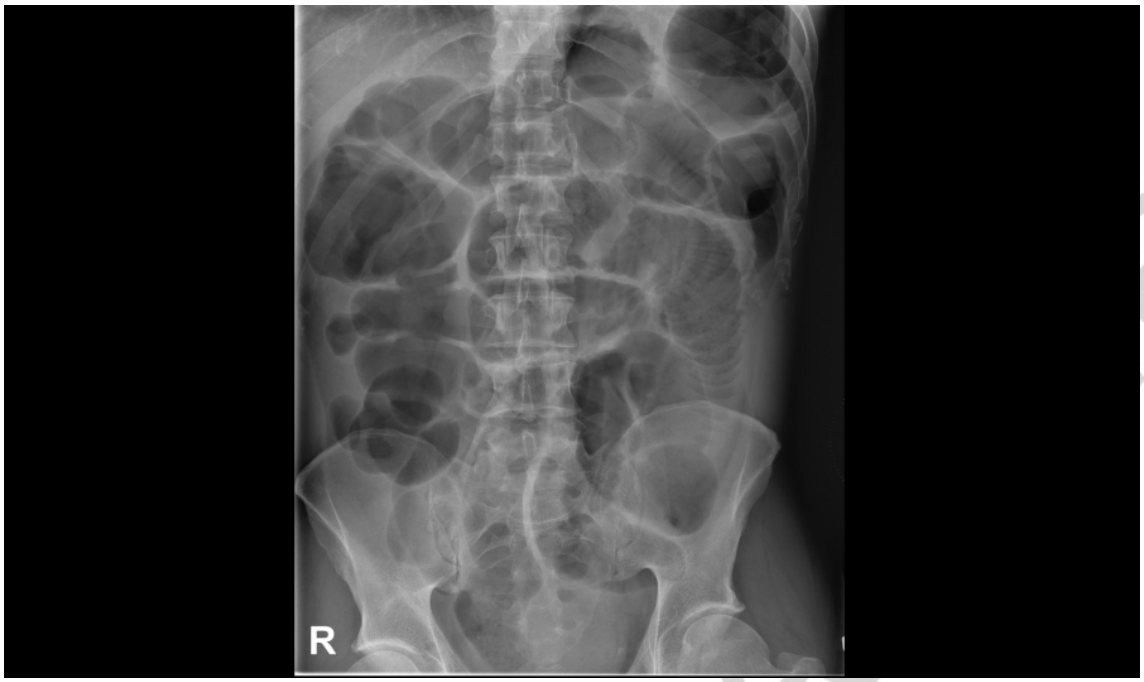

Fig. 1. Abdominal X-ray at the first hospital admission.