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Resolution of dysphagia due to secondary achalasia with neuropathic chronic intestinal pseudo-obstruction by peroral endoscopic myotomy

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Dear Editor,

Chronic intestinal pseudo-obstruction (CIPO) is a rare condition affecting the motility of the small intestine (SI), colon, or both, leading to symptoms and signs consistent with intestinal obstruction without a mechanical cause. Its etiology remains largely unknown, although nearly 50% of cases are associated with neurological, autoimmune, paraneoplastic, infectious, or metabolic disorders. Based on histological findings CIPO could be classified in enteric neuropathies, myopathies, mesenchymopathies, and in some cases it may coexist neuromyopathies. Achalasia is an uncommon esophageal

motility disorder characterized by the absence of lower esophageal sphincter (LES) relaxation and esophageal aperistalsis due to unknown functional loss of myenteric plexus ganglion cells.

Case report

We report the case of a 45-year-old woman who began evaluation in 2011 for chronic pain, abdominal distension, and weight loss, with no abnormalities detected on initial tests. She later developed a pseudo-obstructive syndrome, as demonstrated by abdominal computed tomography (CT), which revealed generalized dilation of SI loops (distal jejunum and proximal ileum) with air-fluid levels, free intra-abdominal fluid, and pneumoperitoneum. An exploratory laparotomy revealed an ileal perforation, managed with primary closure, with no evidence of mechanical obstruction. Due to poor progression and persistent pseudo-obstruction symptoms, she was referred to a specialized center for intestinal manometry (Fig.1A), leading to a diagnosis of neuropathic intestinal motility dysfunction. A transmural biopsy of the SI was recommended but declined by the patient. Since then, she has experienced several episodes of intestinal pseudo-obstruction.

In 2024, the patient was readmitted due to significant weight loss (15 kg) and vomiting, requiring parenteral nutrition (PN). Thoracoabdominal CT revealed SI loop dilation without evidence of mechanical obstruction, as well as marked pan-esophageal dilation. SPECT-CT demonstrated tracer retention in the distal esophagus with no passage into the gastric cavity. A timed barium esophagogram revealed a column height of >5cm at 5 minutes and threadlike emptying. Given the suspicion of achalasia, high-resolution esophageal manometry with impedance (HRIM) was performed, according to the Chicago v4.0 protocol, revealing a pattern consistent with type II achalasia (Fig.1B). A peroral endoscopic myotomy (POEM) was subsequently performed (Fig.1C), resulting in the resolution of dysphagia and enabling the discontinuation of PN. Five months after the POEM a control HRIM was performed to assess the esophageal motility, showing improved relaxation of the LES and a partial motility recovery of a middle esophageal segment (Fig.2), in the pre-test interview the

patient denied dysphagia with a weight gain (2kg) and only reported occasional heartburn.

Discussion

The coexistence of achalasia and CIPO is exceptional, with limited case reports in the literature (1,2,3), in these cases, the digestive symptoms arising from achalasia and CIPO are similar to those observed in our case. However, the sequence of symptoms differs depending on the onset of each condition. In all instances, while esophageal pathology improved with treatment of the LES, intestinal symptoms recurred despite treatment. These conditions may represent manifestations of a common idiopathic disorder of the myenteric plexus affecting both SI, esophagus and esophagogastric junction. In histological samples of CIPO due to idiopathic enteric neuropathy it could be find ganglion cell loss, even aganglionosis, as well as myenteric infiltrate of lymphocytic and/or eosinophilic cells. On the other hand, Achalasia is also associated with lymphocytic infiltrate in the myenteric plexus which causes the loss of ganglion cells. However, in this case the patient rejected a full-thickness biopsy, therefore without histological study this hypothesis remains unconfirmed. Currently, no curative treatment exists for CIPO, with most cases managed conservatively, although some patients may benefit from using immunosuppressants (4). In complex patients with secondary achalasia or concurrent CIPO, POEM offers an effective solution, ensuring adequate oral intake (5).

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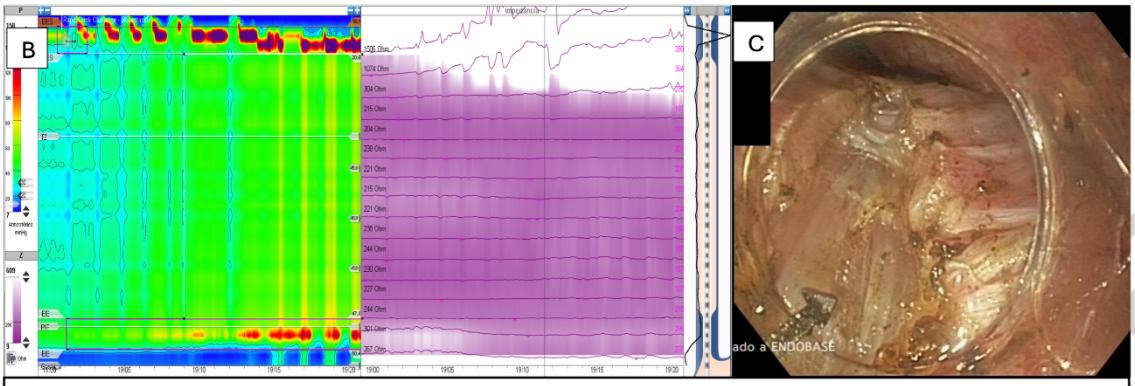

Figure 1A. Due to technical issues we were unable to obtain the images from the intestinal study, even though we have the medical report: during fasting, five phases of intense and regular activity (phase III) were observed, migrating normally through the intestine, with a burst of activity lasting 5 minutes. Postprandially, the recording continued during enteral nutrition administration into the duodenum, without conversion to the expected postprandial pattern. Instead, alternating bursts of activity and periods of absent contractility were noted, consistent with neuropathic type intestinal motility dysfunction.

Figure 1B. 36-channel solid-state high-resolution esophageal manometry with 16-channel of stationary impedanciometry (HRIM) (Unisensor®) (Rapid Drink Challenge): 100% failed peristalsis with incomplete transit is observed in both supine and sitting positions, swallows with panesophageal pressurization in most of them, with an integrated relaxation pressure (IRP) in supine position of 19 mmHg (at the high limit of normality) and in sitting position of 30.3 mmHg (well above the upper limit of normality). Findings consistent with achalasia type II (100% failed peristalsis with panesophageal pressurization in≥ 20% swallows).

Figure 1C. Gastroscopy: punctate esophagogastric junction was observed with significant resistance to endoscope passage, performing submucosal tunnel between 33 to 44cm from the dental arch (DA) and full-thickness myotomy from 35 to 44 cm from DA.

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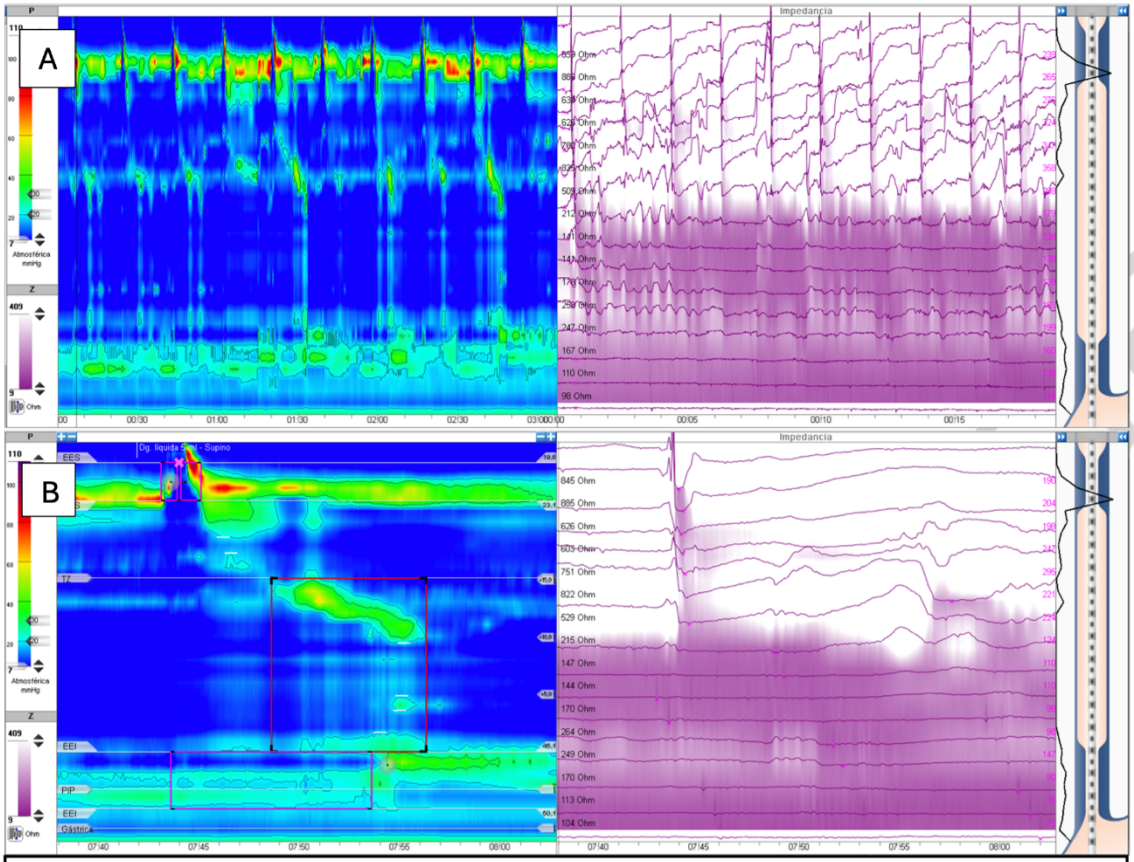

Figure 2. 36-channel solid-state high-resolution esophageal manometry with 16-channel of stationary impedanciometry (HRIM) (Unisensor®) (10 five ml wet swallows).

A. With 10 liquid swallows in the supine position, esophageal aperistalsis was observed in 100% of the swallows. Complete relaxations were present. The median integrated relaxation pressure (IRP4) was 9 mmHg (previously 19 mmHg) (normal reference value: less than 22 mmHg).

B. However, in a minority of swallows, there was a tendency to recover motility in a short segment of 3–5 cm in the mid-esophagus.