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Ball-Valve Syndrome, an Unusual Presentation of a GIST

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Keywords
Gastrointestinal stromal tumor · Gastric outlet obstruction · Ball-valve syndrome · Scarf-ring sign

Síndrome de *ball-valve*, uma apresentação incomum de GIST

Palavras-Chave
Tumor do estroma gastrointestinal · Obstrução ao esvaziamento gástrico · Síndrome de ball-valve · Sinal do cachecol

We present the case of an 87-year-old woman admitted to the emergency room due to persistent vomiting in the previous 4 days. She also complained of an intermittent abdominal pain and a weight loss of 10 kg with months of evolution. On physical examination, she had audible bowel sounds, painful palpation in the epigastrium without signs of peritoneal irritation. The laboratory panel showed only isolated neutrophilia, and the abdominal X-ray had no evidence of bowel distension.

The upper gastrointestinal endoscopy showed a distended gastric body and complete torsion of the gastric folds in the antropyloric region ("scarf-ring sign" shown in Fig. 1) causing obstruction to the passage of the endoscope.

After several attempts of aspiration, lavage and torque, it was possible to completely revert the torsion and reveal a bulky, semipediculated polypoid lesion with a large base of implantation on the anterior face of the distal body. Its surface was covered by congestive gastric mucosa with central erosion, suggestive of a submucosal lesion (Fig. 2, 3).

Following the endoscopy, the patient tolerated progressive diet with no recurrence of vomiting. She was discharged and referred to a surgical consultation. The patient later underwent a laparoscopic atypical gastrectomy. The pathological specimen was compatible with a 4 × 3.2 cm gastrointestinal stromal tumor (GIST), with a low mitotic index (1/50 HPF), suggestive of a low-risk tumor by the modified National Institute of Health classification [1] (Fig. 4).

"Ball-valve syndrome" is a condition caused by intermittent prolapse through the pylorus of a gastric lesion, first described in 1946 by Hobbs and Cohen [2]. The clin-

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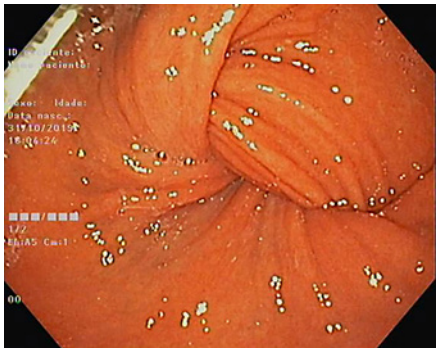

Fig. 1. Initial endoscopic image of complete torsion and invagination of gastric folds in the antropyloric region, compatible with the "scarf-ring sign."

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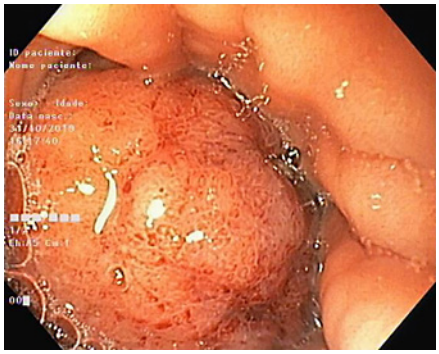

Fig. 2. After endoscopic resolution of gastric torsion, a bulky semipediculated polypoid lesion was observed.

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Fig. 3. Polypoid lesion with surface covered by congestive gastric mucosa with central erosion, suggestive of a submucosal lesion.

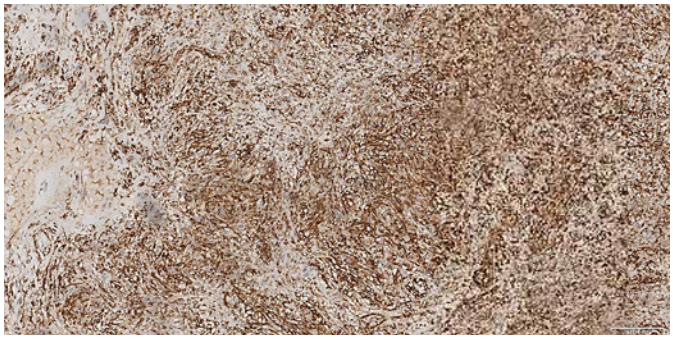

Fig. 4. Histology of the resected specimen with immunohistochemical study revealed CD117 positivity. Magnification ×20.

ical manifestations are variable according to the degree of obstruction, ranging from abdominal discomfort or intermittent abdominal pain, nausea, vomiting and loss of appetite. The diagnosis is supported by imaging and/or endoscopic findings. The observation of a "scarf-ring sign" on endoscopy is a valuable clue to the diagnosis [3]. This is a rare GIST presentation, with only 18 cases previously reported in the literature [4].

Statement of Ethics

Informed consent was obtained from the patient for the case publication.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Dr. Maria Margarida Carvalho wrote the paper. Dr. Carlos Quintana made and provided histological images. Dr. Ana Laranjo and Dr. Andreia Rei revised the paper. Dr. Lurdes Gonçalves and Dr. Isabel Medeiros revised the paper and approved the final version.

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