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Letter to Editor

Video technical notes for approaching a unique case of Juvenile Polyposis with massive gastric ingrowth

To the editor,

Hamartomatous polyps usually represent solitary findings in the gastrointestinal (GI) tract, yet they may occur as part of hamartomatous polyposis syndromes.[1,2] Among these, Juvenile Polyposis (JP) is an autosomal-dominant syndrome characterized by multiple hamartomatous polyps presenting with bleeding, anemia, and obstructive symptoms. Tailored endoscopic surveillance is mandatory since JP has an increased lifetime risk of developing GI cancer.[3,4] Furthermore, symptomatic cases require surgical treatment which is challenging due to the high variability of polyps' location, disease entity and clinical symptoms.

In 2020, a 50-years-old lady was referred to our center for recurrent vomiting, anemia, protein-losing gastropathy, severe weight loss, and an endoscopic diagnosis of hamartomatous polyposis. At first the patient was treated conservatively with laparoscopic intra-gastric removal of a large gastric polyp and contemporary resection of the first jejunal loop. JP was diagnosed based on SMAD4 mutations (c.1139+3A>G) which represented the first description of this pathogenic variant.[5] After nine months of

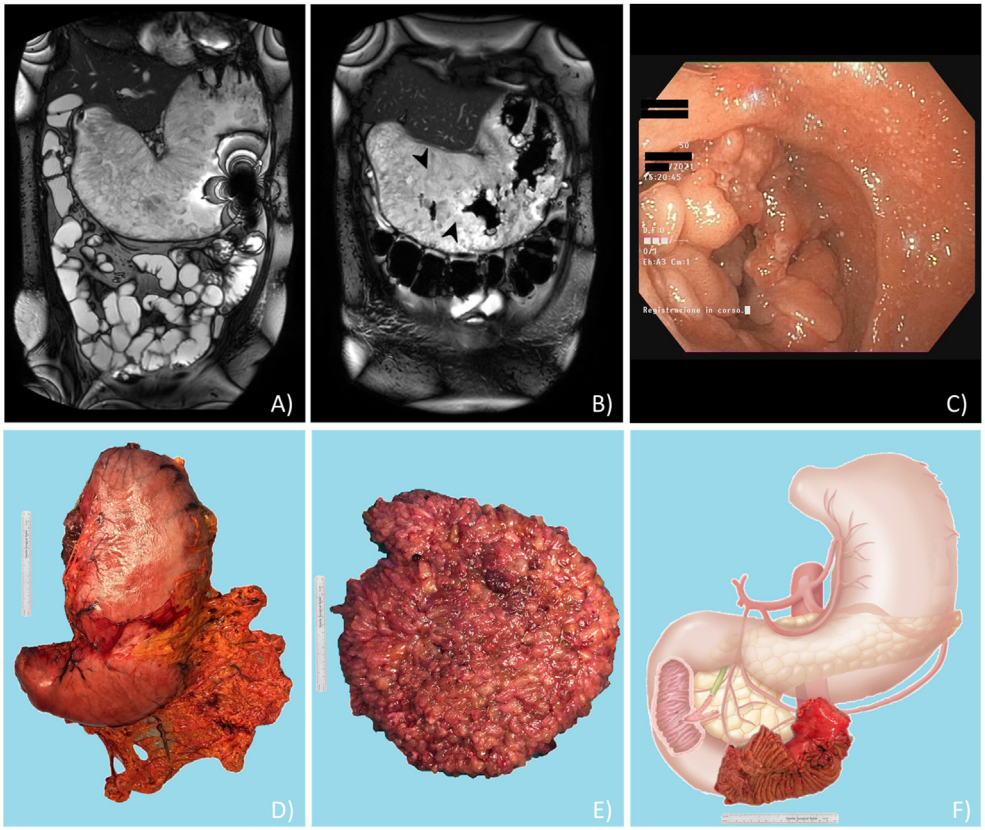

Fig. 1. preoperative MR enterography showing severe gastric distension with restriction of the gastric lumen (A) and thickened walls (B) and. A larger polyp located in the third duodenal portion was not amenable of endoscopic removal and required surgical resection (C). Gross photo of fresh total gastrectomy specimen showing the enlarged stomach measuring 39 cm in length (D). After opening the specimen, the whole gastric surface appeared subverted by a continuous layer of hamartomatous polyps (E). Gross photo of the opened specimen composed by the third duodenal portion and the previous duodeno-jejunal anastomosis (F).

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well-being, the patient reported progressive recurrence of dyspepsia, dysphagia, and food intake reduction. Hospital admission was required for hematemesis and anemia. Magnetic resonance enterography showed important gastric distension (Fig. 1A-B) since the number of polyps had rapidly increased explaining the recurrence of symptoms. Esophagogastroduodenoscopy highlighted replenishment of the stomach, few small polyps in the first and second duodenal portions, as well a voluminous hamartomatous polyp in the third duodenal portion (Fig. 1C). Thoracoabdominal CT scan, colonoscopy and video-capsule endoscopy were negative.

Since SMAD4-associated cases usually feature gastric and intestinal polyps, a tailored surgical procedure is required to treat bleeding and obstructive symptoms, to reduce the risk of malignant degeneration, and allow future endoscopic follow-up. As shown in this Video, the patient underwent open total gastrectomy extended to duodenal bulb (Fig. 1D-E) and distal duodenectomy comprising the previous duodeno-jejunal anastomosis (Fig. 1F). Preservation of the second duodenal portion and cephalad-pancreatic region was intended to avoid high-risk biliary and pancreatic anastomoses. Preoperative endoscopic resection of the polyps located in the second duodenal portion allowed its preservation. The transverse colon with vascular supply from the middle colic pedicle was chosen for the restoration of the upper GI tract. Overall, four anastomoses were fashioned: end-to-side circular stapled esophagus-colonic, handsewn end-to-end colo-duodenal, side-to-side handsewn duodeno-jejunal, and end-to-end colo-colic anastomoses.

No surgery-related complications occurred. Upper GI contrast study demonstrated no leakages and regular bowel emptying so enteral nutritional support was tapered, and the patient managed to meet adequate nutritional intake by postoperative day (POD) 15. Hospital-acquired pneumonia and Candidemia required intravenous antibiotic therapy, so the patient was discharged on POD30. Three months after surgery, the lady fully restored a hypercaloric fractionate diet with increase in bodyweight. Endoscopic and thoracoabdominal CT scan performed at 6 months showed no recurrence.

In conclusion, JP is a rare condition and surgery should be reserved to patients with severe symptoms or suspect of malignancy. After complete endoscopic and radiologic workup, a tailored surgical procedure is highly recommended to relieve symptoms, reduce risk of malignant degeneration, and maintain the possibility of endoscopic follow-up.

Declaration of competing interest

None to declare.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.asjsur.2022.07.058.

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Corrado Pedrazzani*
Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Unit of General and Hepatobiliary Surgery, University of Verona, Verona, Italy

Cristian Conti
Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Unit of General and Hepatobiliary Surgery, University of Verona, Verona, Italy

Angelo Di Vittori
Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Unit of General and Hepatobiliary Surgery, University of Verona, Verona, Italy

Giulia Turri
Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Unit of General and Hepatobiliary Surgery, University of Verona, Verona, Italy

Laura Bernardoni
Digestive Endoscopy Unit, The Pancreas Institute, University and Hospital Trust of Verona, Verona, Italy

Andrea Mafficini
Department of Diagnostics and Public Health, Section of Pathology, University and Hospital Trust of Verona, 37134, Verona, Italy

Claudio Luchini
Department of Diagnostics and Public Health, Section of Pathology, University and Hospital Trust of Verona, 37134, Verona, Italy

Armando Gabbrielli
Digestive Endoscopy Unit, The Pancreas Institute, University and Hospital Trust of Verona, Verona, Italy

Aldo Scarpa
Department of Diagnostics and Public Health, Section of Pathology, University and Hospital Trust of Verona, 37134, Verona, Italy

Alfredo Guglielmi
Department of Surgical Sciences, Dentistry, Gynecology and Pediatrics, Unit of General and Hepatobiliary Surgery, University of Verona, Verona, Italy

* Corresponding author. Unit of General and Hepatobiliary Surgery, University Hospital “G.B. Rossi”, Piazzale “L. Scuro” 10, 37134, Verona, Italy.
E-mail address: corrado.pedrazzani@univr.it (C. Pedrazzani).

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