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Case Report

Sleeve Gastrectomy in a Patient with Obesity and Ehlers-Danlos Syndrome: A Case Report

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ABSTRACT

Ehlers-Danlos syndrome (EDS) is an inherited connective tissue disorder with a huge variety of signs and symptoms. Gastrointestinal manifestations may be present in up to 50% of patients. We here report bariatric surgery in a patient with EDS, focusing on management challenges, preoperative assessment and one-year outcome. A 56-year-old woman with hypermobility-type (HM) EDS, BMI 42,5 kg/m² and hypertension underwent laparoscopic sleeve gastrectomy (LSG). She was uneventfully discharged on POD3. One-year after the operation her BMI was 28,3 kg/m² and hypertension receded. Postoperative upper GI series (POD-60) did show neither reflux nor esophageal dysmotility. Bariatric surgery in patients with EDS can be challenging due to the potential risks of wound healing. Proper preoperative assessment and follow up should be strongly recommended.

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Background

Ehlers-Danlos syndrome (EDS) is described by the Ehlers-Danlos National Foundation as a “heterogeneous group of inheritable connective tissue disorders characterized by articular hypermobility, skin extensibility and tissue fragility [1]. The incidence of EDS is approximately 1 in 5000 births. Formerly divided EDS into 11 subgroups according to clinical phenotype, the latest classification, published in 1998, recognizes 6 EDS subtypes, based on clinical characteristics, such as joint laxity, vascular manifestations, kyphoscoliosis, arthrochalasia and dermatosparaxis, pattern of inheritance, molecular and biochemical findings [1]. The most common subtype is the hypermobility type (HM, formerly type III) which comprises 90% of all diagnosed EDS patients. The main manifestations of EDS include hyperextensible skin, atrophic scars, easy bruising, joint hypermobility and variable involvement of internal organs.

Gastrointestinal involvement is a well-known complication of EDS. Patients can show either organic issues such as hiatal hernia, visceroptosis, rectoceles and rectal prolapse or functional problems such as altered gut motility. The association between HM-EDS and GI symptoms was first described by Hakim and Grahaem [2]. They found

that HM-EDS patients had a significant increase in GI symptoms compared to age and sex-matched controls (37% vs. 11%). The most common GI symptoms were nausea, abdominal pain, constipation and diarrhea. Direct evidence of the association between functional GI disorders and HM-EDS was firstly reported by a group of gastroenterologists [3]. The main upper GI complications are megaesophagus, esophageal, gastric or small bowel diverticula, hiatal hernia, gastric bleeding and ulcers, perforation or hematoma of the GI tract spontaneously or after surgery [4].

Case Report

A 56-year-old woman with HM-EDS and BMI of 42,5 kg/m² was referred to our Bariatric Unit for morbid obesity. Her only comorbidity was hypertension treated with diuretics and Ca channel blockers. She complained mild epigastric pain with episodic heartburn, without dysphagia. Pre-operatively, she underwent upper GI endoscopy that showed a small sliding hiatal hernia, chronic atrophic antral gastropathy with the absence of *Helicobacter pylori*, while a barium swallow (Figure 1) did not confirm the presence of hiatal hernia. No esophageal dysmotility but a regular esophageal clearance was evident.

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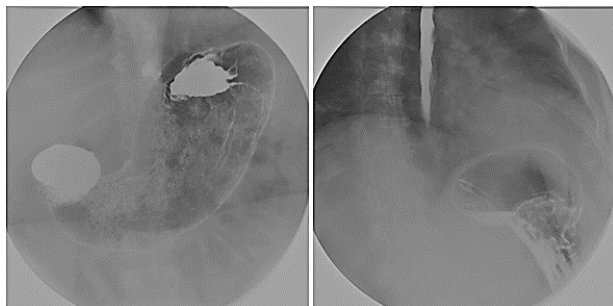


Figure 1: Preoperative upper GI series.

She underwent LSG. She was uneventfully discharged on POD3. Two months later she had a control barium swallow that did not reveal any reflux/dysmotility and normal gastric emptying (Figure 2). At 6-month follow-up she was asymptomatic for heartburn/reflux symptoms and quit antihypertensive therapy; her BMI was 31,6 kg/m² and EWL% 61,9%. At 1-year follow-up she was still off antihypertensive drugs and PPI, BMI, EWL% and TWL were 28.3 kg/m², 81% 34.5 kg, respectively.

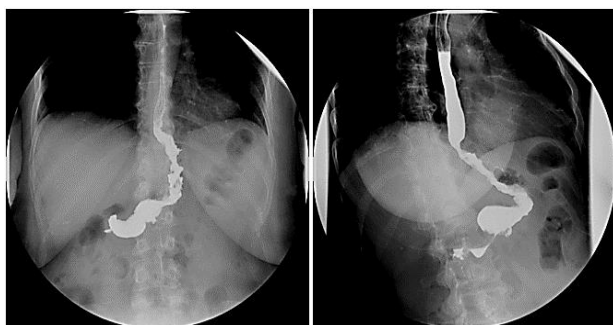


Figure 2: Postoperative upper GI series.

Discussion

EDS may be neglected, and past medical and family history are key issues in the assessment of a patient candidate for bariatric surgery. The GI symptoms vary with subtypes of EDS. Nelson *et al.* described nausea (44%), heartburn (38%), vomiting (24,7%), bloating (17%) and dysphagia as the most common upper GI symptoms. Constipation (42,4%), IBD-like symptoms (30,3%) and diarrhea (22,5%) the most common lower GI symptoms [5].

Surgical procedures represent a real challenge when dealing with EDS patients, given the natural intrinsic frailty of treated tissues and their impaired “healing capacity”. Moreover, several cases of troublesome intraoperative bleeding and massive amounts of intraperitoneal adhesions have been reported [6]. General cautions should be considered during surgery in order to minimize surgical dissection, retraction and suturing. In fact, one of the most frequent issues is wound rupture even several weeks after surgery. Moreover, patients should be properly positioned in order to avoid joint dislocation and cutaneous injuries and particular care should be adopted during induction and awakening anaesthesia phases [7].

The most frequent early postoperative complications are related to anastomotic dehiscence and bleeding [8]. Iatrogenic oesophageal perforation after endoscopic procedures is reported as more frequent in

EDS with predominant vascular type, less in the remaining subtypes [9]. In our case, no complication occurred. Considering late complications, spontaneous oesophageal perforation, oesophageal diverticula, megaesophagus, gastric atony, megaduodenum, small bowel dilation, megacolon and delayed gastric emptying are reported [1, 10, 11].

In our case we chose LSG taking into consideration the clinical conditions and past medical history of the patient. This operation is considered less invasive than Roux-en-Y gastric bypass, involving less tissue manipulation, no anastomosis but only a single long staple line. Moreover, the patient had no history of type 2 diabetes, and obstructive sleep apnoea syndrome and matched the selection criteria for bariatric surgery [12].

Despite the potential challenges posed by surgery on EDS patients, 1-year outcome was very satisfactory, as for comorbidity resolution (hypertension) and weight loss. Further research, longer follow-up periods and more cases are needed to confirm the safety and efficacy of LSG on EDS patients with obesity.

Conclusion

Bariatric surgery on patients with EDS can be challenging and stimulating at the same time. A patient with EDS and morbid obesity may increase the engagement and commitment of the multidisciplinary team, well aware of the potential surgical risks but less aware of the management of EDS-related surgical complications. Accordingly, a strict follow-up policy is advisable in order to assess any eventual postoperative issues.

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