The BIG Stretch: A Report of Three Cases of Gastroparesis as an Uncommon Complication of Hypermobile Ehlers-Danlos Syndrome

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Introduction

Ehlers-Danlos syndromes (EDS) are a group of inherited connective tissue disorders that affects the gastrointestinal system, skin, joints, and vasculature. Hypermobile EDS (hEDS) or Type III is a subset of EDS with a prevalence of 1 in 15,000 people[1]. Major GI features of hEDS include abdominal pain, nausea, constipation, heartburn, and irritable bowel syndrome-like symptoms.

Gastroparesis is a debilitating disorder defined as delayed gastric emptying in the absence of a mechanical obstruction[2]. However, hEDS as a cause of gastroparesis is not well established in the literature[3]. Here we report three cases of gastroparesis in patients with hEDS to demonstrate its uncommon and varying presentations.

Case 1

A 20-year-old woman diagnosed with hEDS at a young age and then developed severe epigastric postprandial abdominal pain with bloating, chronic nausea, early satiety, and unintentional weight loss. She had multiple hospital admissions over the past few years for similar symptoms. Physical examination revealed a young female in no distress with hyperflexible joints, normal bowel sounds, with soft and nontender abdomen. Lab values were within normal parameters. CT abdomen/pelvis showed no evidence of obstruction. EGD and gastric emptying studies both confirmed Grade III gastroparesis. Patient was initially started on metoclopramide and ondansetron without improvement of her symptoms. However, she could not achieve adequate symptom control with pharmacotherapy so a nasogastric tube was eventually placed for enteral feeding and she is currently being evaluated for a gastric stimulator.

Case 2

A 24-year-old man diagnosed with hEDS at an early age and initially presented with complaints of recurrent postprandial abdominal pain, bloating and heartburn, but no weight loss. CT abdomen/pelvis was negative for mechanical obstruction. Patient underwent EGD and was found to have gastritis, hiatal hernia and erosive esophagitis (Figure 2). He subsequently completed a gastric emptying scintigraphy which later confirmed Grade II gastroparesis (Figure 3). His symptoms improved with dietary modifications as well as metoclopramide.

Case 3

A 26-year-old woman with known history of hEDS who initially presented with mild intermittent postprandial abdominal pain and bloating. Prior work-up with EGD and gastric emptying scans confirmed diagnosis of Grade I gastroparesis. Fortunately, her symptoms clinically resolved with both inhaled and oral formulations of cannabis.

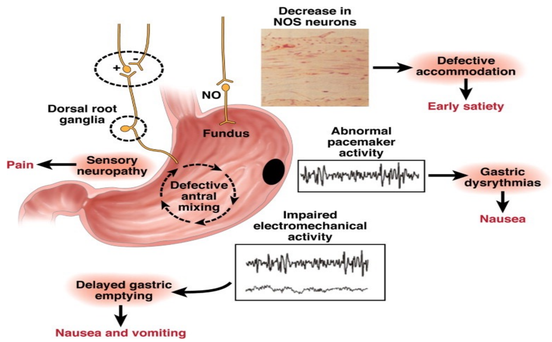

Figure 1. Pathogenesis of symptoms in gastroparesis[9].

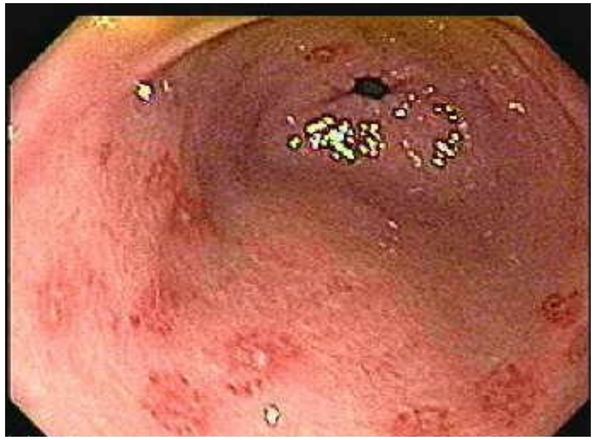

Figure 2. Esophagogastroduodenoscopy (EGD) of the stomach revealing erosive gastritis.

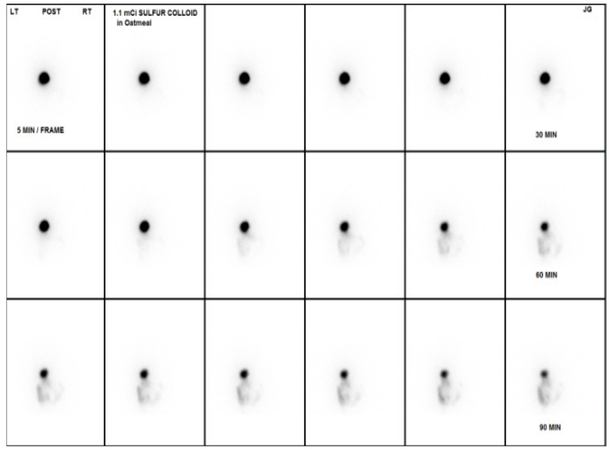

Figure 3. Gastric Emptying Scintigraphy demonstrating gastroparesis with a 25% gastric retention.

Discussion

- hEDS is a multisystem connective tissue disorder that can involve the entire GI tract in terms of structure and function[4]. Possible mechanisms behind the intestinal dysmotility seen in hEDS include autonomic neuropathy, mixed enteric neuro-myopathy, and alterations in the composition of the extracellular matrix in which the other components of the gut wall are embedded[5]. Hence, it can potentially affect any part of the GI tract leading to the variety of GI symptoms as seen amongst our patients.
- Gastroparesis was a common finding in our patients with hEDS who had different symptomatology that can be explained by the complex pathogenesis seen in Figure 1.
- The grades of gastroparesis are mild (Grade I), compensated (Grade II), and gastric failure (Grade III) which are based on symptom control, ability to maintain weight/nutrition, and also frequent hospitalizations[6].
- Initial treatment consists of dietary modifications, hydration, prokinetics, and antiemetics such as metoclopramide [7]. Refractory symptoms are treated with venting gastrostomy tube or decompression. Lastly, surgical options include gastric peroral endoscopic myotomy (G-POEM) or gastric electrical stimulation[8].

Conclusions

These cases serve to highlight the many unique clinical presentations and grades of gastroparesis in patients with a rare variant of EDS. We aim to educate more clinicians about hEDS and its strong association with functional bowel disease.

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