CASE REPORT

Jackhammer oesophagus in a case of linitis plastica

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SUMMARY

A 52-year-old Caucasian woman presented with progressive nausea and vomiting, weight loss, and burning epigastric and chest pain. Initial oesophagogastroduodenoscopy (OGD) with biopsies demonstrated gastritis in absence of *Helicobacter pylori*. A gastric emptying study, CT scan with pancreatic protocol and a colonoscopy revealed no concerning findings. Calcium channel blockers and proton pump inhibitors offered little improvement. A high-resolution oesophageal impedance manometry was performed, which was significant for jackhammer oesophagus with maximum distal contractile index 11 052 mm Hg-s-cm. Another OGD was carried out for Botox injection to oesophagogastric junction. Repeat gastric biopsies reported gastric adenocarcinoma, further diagnosed as stage 4 linitis plastica with metastatic peritoneal carcinomatosis. To the best of our knowledge, this is the only case of linitis plastica associated with jackhammer oesophagus.

BACKGROUND

A hypercontractile oesophagus or jackhammer oesophagus is identified on high-resolution oesophageal impedance manometry (HROIM) where distal contractile index (DCI) value is measured >8000 mm Hg-s-cm (single or multi-peaked contractions) in any single or multiple swallow.¹ It is not seen in the normal population and is sometimes associated with oesophagogastric outflow obstruction.² A few animal model studies have demonstrated that oesophageal peristalsis can be modulated by bolus volume and outflow obstruction.³ Gyawali and Kushnir,⁴ in their high-resolution oesophageal manometry studies on distal obstruction of the oesophagus due to mechanical versus functional causes, noted that the functionally obstructed oesophagus (manifesting as incomplete lower oesophageal sphincter relaxation) is associated with a shift in the force of contraction to the third segment of the oesophagus. A case report has described jackhammer oesophagus noted in a post-lung transplant patient, which later resolved.⁵ Similarly, Tolone *et al*⁶ reported jackhammer oesophagus triggered by radiofrequency catheter ablation in a patient with atrial fibrillation. Eosinophilic oesophagitis with muscularis propria infiltration was recently reported in association of hypercontractile oesophagus in Japan,⁷ and was successfully treated with a peroral endoscopic myotomy (POEM) procedure. This evidence suggests that a hypercontractile oesophagus may sometimes be a secondary manifestation of another disease process, as demonstrated in the following case report.

CASE PRESENTATION

A 52-year-old Caucasian woman with a medical history of hypertension, degenerative joint disease, fibromyalgia and depression, presented to her family physician's office, with symptoms of severe nausea and epigastric pain of recent onset. She described 'pressure in the chest' after meals, nocturnal burping, reflux of clear fluid and intermittent dysphagia. There was history of occasional non-steroidal anti-inflammatory drug use for fibromyalgia and arthritis, and moderate alcohol consumption, however, there was no history of smoking. Family history was negative for any known gastrointestinal (GI) malignancy. The patient was started on proton pump inhibitors (PPI) with no significant relief, and was hence referred for a gastroenterology evaluation.

An outpatient OGD demonstrated diffuse moderately erythematous and congested mucosa in the gastric fundus. Histopathology revealed oxyntic mucosa with chronic gastritis and negative *Helicobacter pylori*.

The patient was continued on PPI daily with nocturnal H2 blocker, however, her symptoms progressively worsened. More worrisome was her unintentional weight loss of more than 13 pounds over 5 months. A gastric emptying study showed normal emptying time. Given her symptoms of epigastric pain and weight loss, a CT scan with pancreatic protocol and colonoscopy was also performed, without concerning findings. A trial of diltiazem 60 mg daily for suspicion of diffuse oesophageal spasms did not improve her symptoms. In addition, she developed early satiety. Subsequently, HROIM, in October 2013, with liquid swallows was consistent with jackhammer oesophagus based on the Chicago Classification, with two swallows recording a DCI value >8000 mm Hg-s-cm. The maximal DCI value was 11 052 mm Hg-s-cm (figure 1), mean DCI was higher than normal at 5562 (normal 500–4300) with 50% of the swallows recorded as hypertensive peristalsis (DCI >5000 mm Hg-s-cm). The mean lower oesophageal sphincter relaxation pressure was normal at 5.6 mm Hg with no oesophagogastric junction obstruction in supine posture.

Three months after her prior abdominal imaging, the patient was admitted to a teaching hospital, with the symptom of haematuria. By this time, her GI symptoms had limited her dietary intake to only popsicles or occasional protein shakes. Abdominal examination elicited diffuse tenderness. Repeat OGD displayed a hypertonic lower oesophageal sphincter (figure 2), which was treated with 100 units of botulinum toxin in the same procedure. Diffuse severely erythematous and granular

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Figure 1 High-resolution oesophageal manometry (HROM) image demonstrated a jackhammer oesophagus pattern with DCI >8000 mm Hg-s-cm. DCI, distal contractile index.

appearing mucosa with bleeding was found in the entire stomach, with normal duodenum (figure 3); biopsies were taken. Pathology was reported as poorly differentiated gastric adenocarcinoma with signet ring features, confirmed by immunohistochemical staining with cytokeratin AE1/3, positive E-cadherin (figures 4 and 5). *H. pylori* remained negative. A subsequent staging CT scan of the abdomen/pelvis was significant for diffuse thickening of the stomach in a linitis plastica-like fashion (with diffuse infiltration) (figure 6). A soft tissue mass in the left adnexal region was concerning for a metastatic implant and likely was the cause of left-sided hydronephrosis with haematuria with which the patient had presented to the hospital.

Pathology of peritoneal biopsies was also reported as metastatic poorly differentiated adenocarcinoma with signet ring cell features compatible with gastric origin confirming the diagnosis of metastatic peritoneal carcinomatosis. A chemotherapy regimen with docetaxel, cisplatin and 5-FU for stage IV gastric adenocarcinoma was initiated by oncology. Nutritional support was provided by placement of a jejunal feeding tube.

OUTCOME AND FOLLOW-UP

The patient initially did well with nutritional supplementation and after the first few cycles of chemotherapy. Three-month

Figure 2 Retroflexed view in OGD showing erythematous and granular mucosa and tight gastro-oesophageal junction. OGD, oesophagogastroduodenoscopy.

Figure 3 Repeat OGD showing erythematous, granular, friable and haemorrhagic mucosa in gastric body in a patient with linitis plastica. OGD, oesophagogastroduodenoscopy.

follow-up CT scans did not reveal further metastatic disease and demonstrated decrease in the carcinomatosis-associated intraperitoneal field. However, 6 months after initial diagnosis, the patient had a sudden decline in status. She was transferred to hospice care and expired shortly afterwards.

DISCUSSION

The field of high-resolution manometry has allowed us a unique perspective of oesophageal functional disorders. The true significance and management of several patterns we recognise, including jackhammer (hypercontractile) oesophagus, remain in evolution. True oesophageal spasmodic disorders are uncommon diagnoses, and sometimes clinically indistinguishable from other conditions such as gastro-oesophageal reflux disease.⁸

Further research is needed to establish if there is a need to understand oesophageal hypercontractility as 'primary' (due to increased cholinergic drive) versus 'secondary' (co-existing conditions causing hypercontractility); or perhaps coexisting. Most importantly, we need to keep the patient's clinical improvement as a benchmark to assess if further investigations should be pursued. Our patient suffered from progressive symptoms affecting her quality of life even though a clear diagnosis remained elusive. The jackhammer oesophagus found on HROM was the initial culprit, however, the patient did not

Figure 4 Abdominal CT scan showing diffusely thickened appearance of the stomach in a linitis plastica-like fashion.

Figure 5 Pathology image from gastric biopsies (low power, 40×). The image showing gastric mucosa with normal overlying foveolar epithelium (red arrow) with diffuse infiltration of the lamina propria by diffuse signet ring cell of gastric carcinoma (asterisks).

respond to a trial of calcium channel blockers. The classic Trojan horse analogy in clinical practice is noted when our focus on treating a symptom makes us oblivious of its potential as a harbinger of a more sinister diagnosis. This case is a

Figure 6 Pathology image from gastric biopsy (high power, 400×). The image showing gastric mucosa with normal overlying foveolar epithelium (red arrow) with diffuse infiltration of the lamina propria by diffuse signet ring cell of gastric carcinoma (black arrow).

learning point for us. The first OGD and biopsy of our patient were negative for malignancy and therein lies her misfortune. The diagnostic accuracy of endoscopic biopsy is poor in patients with linitis plastica, since these tumours infiltrate the submucosa and muscularis propria rather than the superficial mucosa. A combination of bite and strip biopsies demonstrates better yield for cases of linitis plastica.⁹ Endoscopic ultrasound (EUS) can also be a useful diagnostic modality in such cases if it shows the clue of an abnormally thickened submucosal layer.

Several treatment modalities have been employed for oesophageal hypercontractility disorders, including calcium channel blockers, nitrates, myotomy and botox injection, with varying success.^{10 11} The POEM procedure was recently reported as successful treatment in a case of hypercontractility caused by eosinophilic oesophagitis.⁷ The primary pathophysiology is believed to be muscular asynchrony caused by increased cholinergic drive.² Reports in the literature provide evidence of symptom resolution both with treatment of hypercontractility by itself,¹² or of the condition that may have 'induced' it.^{5 11}

Gastric cancer has been attributed as one of the causes of pseudoachalasia,¹³ and hence may be responsible for other motility disorders, too. The most interesting comparison came to our attention with an old case report of gastric carcinoma and oesophageal hypercontractility in which there was autopsy evidence of invasion of the myenteric plexus by the malignancy, which likely led to hypercontractility.¹⁴ Even the most astute of physicians among us can overlook a rare diagnosis on the initial presentation. However, if the clinical improvement of a patient remains our compass, then it can often point our diagnostic 'detective' senses in the right direction.

Learning points

- ▶ Jackhammer or hypercontractile oesophagus can be either primary or induced. Further investigations of a secondary cause of jackhammer oesophagus should be considered if a patient does not demonstrate clinical improvement with therapy.
- ▶ Failure of improvement in a patient's symptomatology remains a vital guiding principle in clinical care, and should prompt the physician to search further for contributing causes.
- ▶ Gastric cancer is associated with pseudoachalasia, however, it can cause other oesophageal motility disorders as well.
- ▶ Linitis plastica is difficult to diagnose with conventional oesophagogastroduodenoscopy and biopsy, and hence is often picked up at an advanced stage of the disease.

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