It’s not just a heartburn and reflux disease: a case report of distal oesophageal spasm and review of literature

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CASE REPORT

An 87-year-old man was initially presented to a different hospital with chronic heartburn (>12 months) despite being treated with high dose PPI, antacids and prokinetic agent. He described a constant burning sensation at the epigastric region which radiated to the retrosternal area, associated with water brash in the mouth and occasional dysphagia. The symptoms occurred during the day and can significantly disturb his quality of sleep at night. He had no other alarming symptoms and no personal/family history of gastrointestinal malignancy or connective tissue disease. He had been extensively investigated by cardiologist and that included a normal finding of coronary angiogram. In addition, he underwent multiple esophagogastroduodenoscopy (OGD) examinations which all reported as normal. At our centre, we performed a 24-hour pH monitoring and high-resolution oesophageal manometry (HRM) examinations. A 24-hour pH monitoring showed no evidence of acid reflux disease (normal DeMeester score - see Figure 1A) and HRM’s result was suggestive of DES with an integrated relaxation pressure (IRP) of 12.3 sec and delayed latency (DL) at 0.7 sec, albeit lack of evidence of simultaneous contraction on the manometry (Figure 1B).

Subsequently, barium swallow examination revealed uncoordinated peristalsis with simultaneous tertiary contraction at multiple points at the distal half of thoracic oesophagus that supported the diagnosis of DES. OGD was performed which showed a typical appearance of corkscrew oesophagus (Figure 2B). The patient was then counselled regarding the diagnosis and offered various treatment options which include oral pharmacological agents, botulinum toxin A (Botox) injection and surgery. Due to the age of the patient and preference, diltiazem 30mg twice a day was commenced. At two-month follow-up, he reported improvement of his symptoms. Over the next 10 months, his symptom was under controlled, with just occasional ‘flare’ of heartburn and dysphagia.

However, one year later, the patient started to complain of worsening of his heartburn along with retrosternal chest pain which did not respond to the addition of PPI and antacid. Subsequently, an OGD was performed together with the injection of Botox (25 units per quadrant) to the four quadrants proximal to the lower oesophageal sphincter. At the routine follow-up three months later, he reported a complete resolution of the symptoms (heartburn and dysphagia) and fully satisfied with the treatment outcome. A followed-up HRM revealed an improvement of manometry parameters (IRP 4.9 sec, DL 10.6 sec - see Figure 1C) as compared from the previous values.

DISCUSSION

DES is a primary oesophageal motility disorder. It is a rare condition, with an estimated prevalence of 3-5%. Historically, the term ‘diffuse oesophageal spasm’ has been used, but it was proposed to be changed to ‘distal oesophageal spasm’ instead. Sperandio et al., found that simultaneous contractile activity in patients with diffuse oesophageal spasm was restricted almost exclusively to the distal esophagus. The pathophysiology of DES is thought due to impairment of the esophageal inhibitory pathway, which leads to premature contractions in distal muscularis propria.

Clinically, patients usually present with dysphagia, regurgitation, heartburn and/or chest pain. DES can be diagnosed by HRM, which shows ≥20% of simultaneous contraction with DL <4.5 seconds and mean IRP of <13mmHg. Other modalities such as barium swallow and
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Fig. 1: A: Acid reflux composite score analysis from the 24-hour pH monitoring test, B: High-resolution manometry of pre and C: Post-Botox injection for DES.

Fig. 2: A: Normal gastro-oesophageal junction, B: Appearance of Corkscrew oesophagus.
OGD are often helpful in aiding the diagnosis as well as rule out other differential diagnoses. Barium swallow findings for DES include nonspecific tertiary contraction and corkscrew or rosary-bead appearance of esophagus. OGD finding is however less specific (might show corkscrew appearance of esophagus); but its use is pertinent to rule out malignancy, esophagitis and eosinophilic esophagitis. Ambulatory pH monitoring is another important test to do since dysmotility on manometry can sometimes be due to pathological reflux. In addition, various degree of GERD might co-exist with DES, therefore, need to detect and treat at the same time.

Treatment of DES is mainly directed towards symptomatic relief. It can be divided into medical, endoscopic and surgical treatments. Nitrates, calcium channel blockers and low-dose tricyclic agents have been used, but with limited efficacy; and therefore, usually used in mildly symptomatic patients. Botox injection acts by blocking the release of acetylcholine at the neuromuscular junction, causing relaxation of the lower esophageal sphincter. A randomised, placebo-controlled, crossover trial involving 22 patients with DES or nutcracker esophagus, revealed 50% of patients were responders at 1 month after Botox injection, compared to 10% after saline injection. In a retrospective study involving Botox injection, four out of six DES patients had sustained response of more than six months. Another endoscopic treatment is esophageal dilatation with either bougie dilators or through-the-endoscope balloons. No rigorous study regarding this method on DES patients has been done, and therefore not widely practiced. Surgical treatment for DES includes Heller myotomy (which was not suitable for our case in view of age). It is typically used to treat achalasia but has also been used in treating DES. Peroral endoscopic myotomy is relatively a new technique in treating achalasia. However, now its indications have expanded to non-achalasia motility disorders that include DES. From multiple case series, it appears promising; but will need further studies before a strong recommendation can be given.

Our case highlights the importance of considering esophageal motility disorders in patients complaining of chronic and refractory heartburn, especially with the presence of dysphagia/chest pain. Since HRM is not easily available to many clinicians, a timely referral to gastroenterologists (with available resources) is paramount. Combination of diagnostic modalities is essential since manometry alone might be misleading. Oral pharmacological agents can be used as a first-line therapy, however, most of the patients will need more advanced endoscopic or surgical treatments. Our patient is currently doing well after his first Botox injection. Despite this, he is likely going to ‘relapse’ in the future – in which repeat Botox injection still can be offered.

REFERENCES