

## Case Report

# Dysphagia and oesophageal dilation in giant hiatus hernia: angulation of the gastro-oesophageal junction or achalasia?

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**Received:** 20 August 2023

**Revised:** 14 September 2023

**Accepted:** 15 September 2023

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## ABSTRACT

The co-existence of achalasia and large para oesophageal hernia (PEH) is uncommon. The anatomical disruption of the GOJ mechanism by hiatal herniation may result in chronic angulation and secondary partial oesophageal obstruction. A unique case is described where the diagnostic uncertainty of coexistent achalasia and PEH with angulation causing chronic oesophageal obstruction existed. This case highlights the importance of multimodality workup of oesophageal disorders utilising endoscopy, fluoroscopy and manometry. Accurate identification of physiological abnormalities allowed appropriate design of operative intervention.

**Keywords:** Hiatus hernia, Dysphagia, Dysmotility disorders

## INTRODUCTION

The co-existence of achalasia and large para oesophageal hernia (PEH) is uncommon.<sup>1,2</sup> Achalasia is an oesophageal motility disorder characterized by abnormal sphincter relaxation and loss of oesophageal peristalsis.<sup>3</sup> The anatomical disruption of the GOJ mechanism by PEH may result in chronic angulation, secondary partial obstruction of the oesophagus and subsequent dilatation with apparent loss of peristalsis.<sup>4</sup> A diagnostic dilemma existed as to the cause of dilatation of the oesophagus was consequence to chronic obstruction or a primary motility disorder.

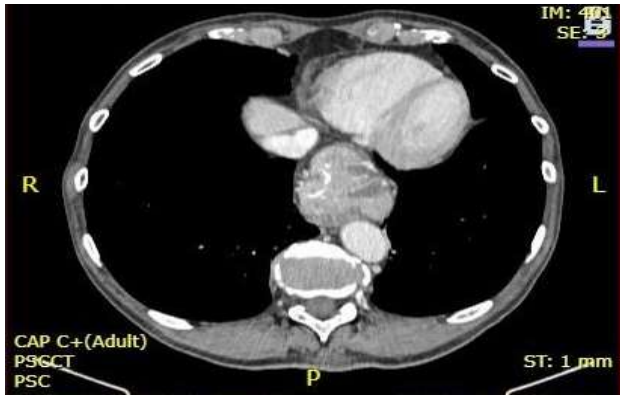
A unique case is described of apparent oesophageal obstruction causing progressive dysphagia with acute angulation of the GOJ in the presence of PEH oesophageal dilatation.

## CASE REPORT

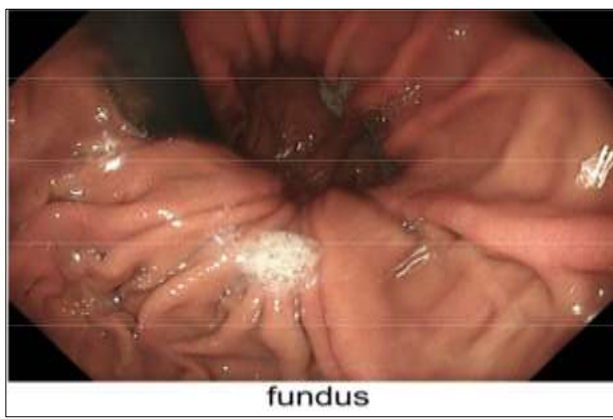
An 88-year old male presented with progressive dysphagia with substantial weight loss of 15 kg and early satiety.

There was regurgitation and retrosternal heartburn without respiratory symptoms. Past medical history included atrial fibrillation, hypertension, hyperlipidaemia and a longstanding pancreatic ganglioneuroma under radiological surveillance. Biochemistry demonstrated normal white cell count of  $11.1 \times 10^9/l$ , haemoglobin 153 g/l, eGFR of 68 ml/min/1.73 m<sup>2</sup> and no electrolyte abnormalities. The baseline chest X-ray findings showed a normal cardiomeastinal contour and clear pleural spaces. A background of intermittent dysphagia symptoms had been previously investigated with a computed tomography (CT) scan of the abdomen and pelvis, which demonstrated a moderate hiatus hernia associated with a thick walled, distended oesophagus reportedly suggestive of progressive reflux oesophagitis (Figure 1).

Gastroscopy demonstrated a tortuous oesophagus, a large hiatus hernia and a short area of narrowing at the level of the GOJ (Figure 2). Cardio-oesophageal dilatation up to 14 mm had not been symptomatically beneficial. There was no endoscopic evidence of oesophagitis.



**Figure 1: CT abdomen and pelvis: moderate hiatus hernia.**

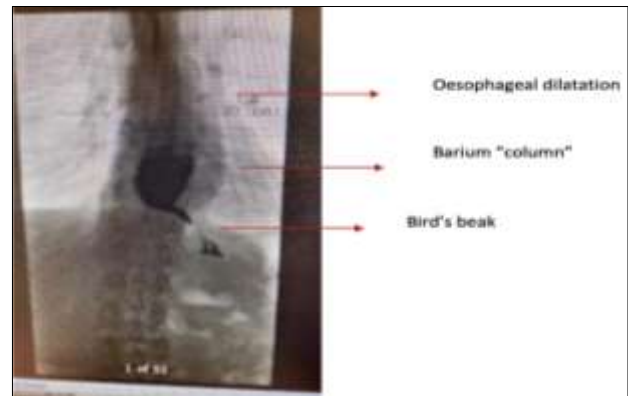


**Figure 2: Endoscopy: retroflexed view of large hiatus hernia.**

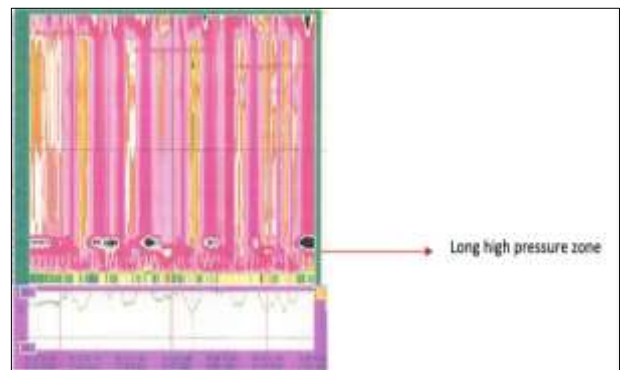
The patient underwent a barium study, which demonstrated gross oesophageal dilatation with a typical bird's beak appearance at the lower oesophageal sphincter (LOS) raising the prospect of there being coexistent disease processes (Figure 3). Passage of contrast was so slow a gastric study could not be performed to outline the hiatus hernia. Oesophageal manometry was then performed which showed no peristalsis and an inadequately relaxing LOS with no evidence of hiatal herniation. The thoracic oesophageal background pressure was raised above intra-abdominal pressure and there were episodes of pan pressurization (Figure 4). The findings were predictive of achalasia type II. The provisional diagnosis was synchronous achalasia type II and large hiatus hernia.

Surgery was undertaken to repair the hiatus hernia and treat the achalasia. At laparoscopy, the hiatal opening was large and a large paraoesophageal hernia was found which contained 40% of the stomach with cardia-oesophageal angulation. After dissection of the hernia sac and reduction, the hiatus was repaired anteriorly and posteriorly with 0-ethibond. A Heller's myotomy and a 270-degree anterior composite fundoplication was performed with posterior fixation of the COJ and lesser

curve of the stomach to the median arcuate ligament and right crural pillar.



**Figure 3: Barium swallow demonstrating bird's beak appearance of achalasia.**



**Figure 4: High resolution oesophageal manometry.**

IRP: 60.8 (n<15 mmHg), pan-pressurisation and basal end expiratory pressure: 65.1 (n=10-30 mmHg).

Post-operative recovery was uneventful and at follow up at nine months, there was an easily opening LOS at 40cm, intact fundoplication and no oesophagitis. LOS criteria were IRP 5.7 (decreased from 60) and basal EEP 10.2 (decreased from 65). There was no heartburn, regurgitation or dysphagia.

## DISCUSSION

The occurrence of achalasia and hiatus hernia simultaneously has a reported incidence of 1.2-20.8% of cases.<sup>1,5-7</sup> Studies do not differentiate between paraoesophageal or sliding hiatal hernias (SHH). PEH, unlike SHH is less frequent and more likely to cause an angulation of the cardia resulting in oesophageal obstruction, dysphagia and abnormal peristalsis.<sup>4,8</sup> The pathophysiology of chronic obstruction leading to oesophageal dilatation and associated appearance of pseudoachalasia has also been demonstrated in gastric banding; another situation where chronic oesophageal obstruction may mimic oesophageal dysmotility.<sup>9</sup> The use of barium meal in this case was helpful in that it was suspicious and was followed up by high resolution manometry.

Guidelines for the investigation of dysphagia recommend three tests - endoscopy, barium swallow and manometry.<sup>11,12</sup> In the diagnosis of achalasia, high resolution manometry is the current gold standard test.<sup>12</sup> In co-existing achalasia and large hiatus hernias (>5 cm), Roman et al showed that hiatal hernia resulted in lower mean GOJ pressures and a slower contractile front velocity which is possibly a reflection of the anatomical changes, but the distribution of motility disorders was shown to be unaffected.<sup>4</sup> Manometry may falsely under report peristalsis in oesophageal dilatation by lack of probe contact in the dilated lumen leading to inappropriate treatment. Barium study was therefore complimentary.

Incomplete LOS relaxation similarly may be confounded by GOJ angulation in HH causing lateral increased pressure on manometry sensors falsely showing elevated basal sphincter pressures and abnormal relaxation (integrated relaxation pressure=IRP). Such abnormalities are frequently seen during investigation of PEH. The presumed diagnosis of a peptic stricture or gross angulation appeared radiologically different from the tapered “bird’s beak” of achalasia. Demonstration of such findings assisted the probability of diagnosis and determination of an appropriate surgical strategy.

The management options include medical, endoscopic and surgical approaches. The surgical standard of care for achalasia in appropriately selected patients is a laparoscopic heller myotomy (LHM) with fundoplication.<sup>13</sup> In 2010, the perioral endoscopic myotomy (POEM) was introduced and had comparable short-term dysphagia outcomes to LHM. Although POEM offers good control of dysphagia symptoms, there is a high rate of post-POEM reflux, which has been well-established by meta-analysis.<sup>14</sup> In the presence of PEH, destruction of the LOS by myotomy or balloon without hernia repair was thought likely to expose the patient to severe reflux. In the setting of co-existing hiatus hernia, it has been recommended patients undergo laparoscopic Heller myotomy with a partial fundoplication.<sup>15</sup>

## CONCLUSION

This case highlights the value of thorough workup of oesophageal disorders with endoscopy, fluoroscopy and manometry. Differentiation of oesophageal dilatation and abnormal peristalsis in the setting of chronic obstruction or co-existing achalasia alters operative management for best outcome and avoidance of multiple procedures. In the context of achalasia and co-existing PEH, a laparoscopic hernia repair and Heller’s myotomy may be considered as the treatment of choice.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

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**Cite this article as:** Wong P, Seyfi D, Herba K, Falk G. Dysphagia and oesophageal dilation in giant hiatus hernia: angulation of the gastro-oesophageal junction or achalasia? *Int Surg J* 2023;10:1684-6.