Case report

Coexisting achalasia and paraoesophageal hiatal hernia

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Abstract

Disorders of the oesophagus present a diagnostic and therapeutic challenge. The presenting symptoms of dysphagia, reflux, pain and vomiting are almost universal, irrespective of the underlying pathology. A combination of endoscopy, barium studies, pH studies and manometry are often required to determine the exact diagnosis and to plan the most effective treatment. Paraoesophageal hiatal hernia is an uncommon condition, present in 14% of all hiatal hernias, which requires urgent correction to prevent life-threatening complications. It is unusual for other oesophageal disorders to coexist. We present a case where achalasia and a paraoesophageal hiatal hernia probably coexisted.

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1. Case report

A 67-year-old lady presented with a 6 month history of heartburn, vomiting, regurgitation and malaise. Her past medical history included hypertension and parathyroidectomy for hypercalcaemia and previous abdominal surgery for both renal and gynaecological surgery. Chest X-ray showed the presence of a large paraoesophageal hernia lying behind the heart shadow (Fig. 1). This was confirmed on barium swallow. Oesophageal manometry (Fig. 2) was performed. This showed a very short (1 cm) lower oesophageal sphincter with a mean tone of 32 mmHg. Although relaxation of the sphincter was felt to be co-ordinate, it was noted to be incomplete on some swallows. This was interpreted as representing pressure from the large para-oesophageal hiatus hernia that was definitely present. Oesophagoscopy was performed and showed early oesophagitis.

The patient was judged fit for surgery and we decided to proceed via a left thoracotomy in accordance with our usual practice. We were reluctant to consider the abdominal approach in view of the previous abdominal surgery. The large para-oesophageal hiatus hernia was reduced and, in view of the history of heartburn and regurgitation and the endoscopic findings of oesophagitis, an anchored Nissen’s total fundoplication was performed. A loose 1 cm fundoplication was performed a 50 Malloney Dilator being present in the oesophagus at the time, to ensure that the wrap was not tight. Whilst the oesophageal wall was noted to be slightly thickened at time of operation, no suspicions were raised by this observation. Post-operative recovery was uncomplicated and the patient was discharged on the 7th post-operative day on a ‘soft’ diet.

The patient attended the Out-patient clinic for review, unfortunately complaining of persisting dysphagia for some foods, such as bread bolus or seafood, although most swallowing was satisfactory. Although this was felt to be related to her recent Nissen’s fundoplication, her symptoms did not settle as expected, and she underwent repeat endoscopy in more than one occasion, which failed to reveal any anatomical cause for her symptoms. However her dysphagia worsened, and the decision to repeat the oesophageal studies was made.

An initial barium swallow study suggested that the fundoplication performed had slipped into a thoracic position, causing her symptoms of dysphagia due to technical failure of the anti-reflux procedure. Oesophageal manometry was attempted but, when some difficulties were encountered in passing the manometry catheter, the barium swallow was reviewed and the procedure abandoned as the diagnosis appeared certain. She was offered correctional surgery.

At re-operation, the approach to the hiatus was through the previous thoracotomy incision scar. It was evident that the repair had not failed, but the body of the oesophagus was now grossly thickened, suggesting a distal obstruction. The diaphragm was opened away from the hiatus and the
presence of an intact anti-reflux wrap confirmed. Attempts to take down the wrap resulted in perforation of a narrowed and thin-walled section of distal oesophagus and eventually a distal oesophagectomy was performed, the stomach being used as a replacement organ. Post-operative recovery was uneventful and a contrast swallow performed on the 7th post-operative day showed no leak and no obstruction to the flow of contrast.

Subsequent histological examination of the resected specimen revealed the typical appearances of oesophageal achalasia, with absence of ganglion cells above the gastro-oesophageal junction.

The patient unfortunately continued to have problems with persisting dysphagia and vomiting and was found at endoscopy to have disordered gastric motility with apparent reverse peristalsis. She was treated with NJ feeds and later with a feeding jejunostomy. She was allowed free oral intake and had no dysphagia or regurgitation whilst the jejunostomy was in situ. After 4 years, although she is able to take food and fluids orally without vomiting, she prefers to rely on her jejunostomy for nutrition.

2. Discussion

Paraoesophageal hiatal hernia is an uncommon condition, present in 14% of all hiatal hernias, which requires urgent correction to prevent life-threatening complications [1]. Sliding hiatal hernias coexist in approximately 10% of patients with achalasia [2]. The presence of oesophageal diverticula with achalasia has also been described [3]. Carcinomas are known to occur in both achalasia and paraoesophageal hiatal hernia. However, we believe that this is the first description of achalasia coexisting with a paraoesophageal hiatal hernia.

Before elective repair of a paraoesophageal hiatal hernia is undertaken, oesophageal manometry and pH studies are recommended to determine whether or not an additional anti-reflux procedure should be performed [4]. Manometry was performed in this case, and, although the diagnosis of coexisting achalasia was considered, the abnormal manometry findings were thought to be due to the presence of the large paraoesophageal hiatus hernia. There were felt to be insufficient incomplete relaxations of the LOS to immediately suspect early achalasia, an opinion later found to be incorrect.

If a fundoplication is performed with the wrap too tight, the motility of the body of the oesophagus will deteriorate with time. In this case, however, initial swallowing was as expected following after a complete fundoplication and repeat endoscopies did not raise the possibility of a tight wrap, nor did dilatation improve the situation. Subsequent progress of the patient has suggested a more widespread upper gastrointestinal motility disorder.

Re-operation was performed as a result of a barium study that confidently stated that the fundoplication had slipped into the chest, something found to be incorrect at surgery. If this examination had not been misleading, pneumatic bag dilatation would have been attempted, but with concern following the previous fundoplication and posterior crural repair. By the time the true situation was revealed at operation, resection was considered the safest surgical option.

Although coexisting achalasia and paraoesophageal hiatal hernia is extremely rare, clinicians should be aware that oesophageal disorders can coexist. The oesophagus should be fully investigated whenever possible. When unex-
pected or unusual findings occur, or the investigations do not correlate, then the presence of a coexisting disorder should always be considered. Experience from this case has resulted in reluctance to perform complete fundoplication following reduction of paraoesophageal hiatal hernia, being sensitive to criticism that the wrap could have been too tight in this case. In addition, where dysphagia is encountered following hiatal hernia repair, the possible causes and solutions should be examined more closely and investigation should be more thorough.

References