Diagnostic dilemmas in dysphagia aortica


aDepartment of Cardiothoracic Surgery, Northern General Hospital, Herries Road, Sheffield S5 7AU, UK
bDepartment of Diagnostic Imaging, Northern General Hospital, Herries Road, Sheffield S5 7AU, UK

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Abstract

Objective: Dysphagia aortica describes swallowing difficulty caused by external compression from a tortuous or aneurysmal aorta. We present 5 cases in which dysphagia to solids accompanied a localised high pressure barrier (HPB) on manometry suggestive of dysphagia aortica, and explore other investigation modalities useful to confirm the diagnosis. Methods: Four females and 1 male with a median age of 56 years (range 47–58) were investigated. All underwent investigation with endoscopy, chest radiography, CT scanning, barium swallow, and video solid bolus swallow in addition to oesophageal manometry and 24 h ambulatory pH monitoring. Results: Median basal pressure rise at the mid oesophageal HPB was 45 mmHg (range 40–80). In addition to the HPB, 4 patients had manometric abnormalities of swallow activity and 2 patients had significant gastroesophageal reflux disease (GORD). Contrast enhanced computed tomography and barium swallow were normal in all cases. Video bolus swallow showed pronounced obstruction to transit at the aortic arch in 2 cases and excluded significant dysphagia aortica in 2 others. Conclusions: Dysphagia aortica commonly coexists with motility disorders and GORD. Video solid bolus swallow allowed us to determine the clinical significance of a manometric HPB in 4 out of 5 patients suspected of dysphagia aortica where standard evaluation would have failed. We recommend its use in those patients with a manometric HPB suggestive of dysphagia aortica in whom standard barium swallow is normal. © 1997 Elsevier Science B.V.

Keywords: Dysphagia aortica; Manometry; Imaging; Video bolus swallow

1. Introduction

The term dysphagia aortica was first used by Pape in 1932 to describe difficulty in swallowing caused by external compression from an ectatic, tortuous, or aneurysmal aorta as a result of age related degeneration [19]. Typical patients are elderly, hypertensive females, often of short stature with kyphosis [12]. Low sternal dysphagia is caused by the diseased aorta pushing the oesophagus anterolaterally against the crural slings of the diaphragm [16]. Compression at the arch of the aorta is less common [4].

On standard chest radiography the typical findings in dysphagia aortica are an unfolded aortic arch below which lies a tortuous dilated aorta, the course of which is variable [10,12]. Barium swallow may show one or more of the following features: partial oesophageal obstruction; a flattened contour of the left margin of the oesophagus; and pulsatile movement of the barium synchronous with aortic pulsation [3]. Oesophageal manometry reveals a localised high pressure barrier with superimposed oscillations synchronous with aortic pulsation typically seen in only one transducer [14,20]. The endoscopic findings may be stenosis, band like pulsatile extrinsic compression, or kinking of the oesophagus [10,12].

Unfortunately, these classical findings for dysphagia aortica are subjective and inconstant. In some cases the radiographic or endoscopic features are minimal or absent [1,10,13]. Whilst barium swallow may be the
standard diagnostic tool, the occurrence of false negative results when investigating dysphagia to solids using liquid contrast is well documented [2,14,21] but often overlooked. In addition, tortuosity of the thoracic aorta is a common radiological finding in the elderly population without dysphagia [12], vascular pulsation is often seen on endoscopic evaluation of the normal oesophagus, and the characteristic manometric tracing of dysphagia aortica is also described in normal patients [14], as well as those with other causes of extrinsic vascular compression [5,11,17].

We have investigated a number of patients in which dysphagia to solids was associated with a high pressure barrier on manometry suggestive of dysphagia aortica. All have undergone extensive radiological and endoscopic investigation and their relative value in reaching a definitive diagnosis is discussed.

1.1. Materials and methods

Patients were collected for the study from a cohort who had been investigated in our oesophageal laboratory for symptoms of dysphagia and were found to have a localised high pressure barrier (HPB) of the type described in dysphagia aortica on oesophageal manometry. In addition to details regarding their dysphagia, information on reflux symptoms was gathered using a modified DeMeester type questionnaire, and all patients were asked about their medical history and drug therapy.

Manometry was performed using triple intraluminal catheters and transducers with 120° angulation and 5 cm spacings between ports (Gaeltec, Dunvegan, Isle of Skye). Recordings were made on a Multitrace IV chart recorder using Lectromed recording equipment (Lectromed, Letchworth Garden City, Herts) All patients underwent a Multitrace IV chart recorder using Lectromed recording equipment (Lectromed, Letchworth Garden City, Herts) All patients underwent a 24 h ambulatory pH monitoring off all acid suppressing medication, and data was analysed using Gastrosoft Oesophagram software (Synectics, Enfield, Middlesex).

All patients underwent radiological evaluation with plain chest X-ray, barium swallow and video bolus swallow using both marshmallow and barium. Intravenous contrast enhanced Computerised Tomography of the thorax and flexible upper gastrointestinal endoscopic evaluation were also performed in all cases.

2. Results

Four females and 1 male were included in the study with a median age of 56 years (range 47–58). All complained of dysphagia to solids. In 3 patients this was mild, but in the remaining 2 patients it was troublesome and required significant dietary modification. All patients were of below average stature and 2 were below the third centile for height. Patient demographics, severity of dysphagia and other relevant medical conditions are given in Table 1.

On manometric assessment all patients had a localised HPB similar to that shown in Fig. 1. In 3 patients it was located around the level of the aortic arch (13–15 cm from the lower oesophageal sphincter), and in the remaining 2 it was located more distally in the oesophagus. The median basal pressure rise at the HPB was 45 mmHg (range 40–80 mmHg). The manometric findings are shown in Table 2. Plain chest radiography, contrast enhanced computed tomography, and barium swallow were normal in all cases, except for patient 3 in which an unfolded, tortuous aorta was demonstrated. The results of the imaging and endoscopic findings are summarised in Table 3.

In patients 1 and 2 video bolus swallow demonstrated clear hold up of marshmallow at the level of the aortic arch (Fig. 2). Both cases required several swallows of liquid barium before passage of the marshmallow occurred through the obstruction. The site of hold up of the bolus in these cases was consistent with the manometric level of the HPB, and reproduced the patients usual dysphagic discomfort. In addition patient 2 had a marked kyphosis and patient 1 had a visible pulsatile barrier on endoscopy. Collectively, these findings were felt to be diagnostic of dysphagia aortica. Both patients had abnormal swallow activity in the distal oesophagus on manometry and patient 2 had documented reflux on pHmetry, however video swallow...
failed to demonstrate any hold up nor were there any symptoms as the bolus passed these levels, therefore a significant primary motility disorder or gastroesophageal reflux disease (GORD) as the cause of dysphagia was felt to be excluded. Patient 1 underwent oesophageal dilatation with Mallony bougies to size 50 Fr with significant relief of her symptoms. Patient 2 had mild symptoms which were well controlled following dietary advice only.

In patient 3 the cause of dysphagia could not be clearly determined. She had a marked thoracic kyphosis and an unfolded aorta on chest radiography with a typical HPB and normal motility on manometry. Video bolus swallow was normal, but unfortunately was performed following endoscopy and therapeutic dilatation of a benign mid oesophageal stricture. As there was no evidence of reflux at endoscopy or 24 h pHmetry, the stricture may have been due to stasis oesophagitis, but with no video evidence of hold up or symptoms at the level of the manometric HPB we were unable to confirm dysphagia aortica. Following dilatation she remains asymptomatic.

Cases 4 and 5 were considered not to be dysphagia aortica. In patient 4 there was no demonstrable hold up of bolus at the level of the manometric HPB, no transmission of vascular pulsation, and no compliant of dysphagia. It’s presence was therefore felt to be not clinically significant. Grade 2 distal oesophagitis was found at endoscopy, and disordered lower oesophageal sphincter relaxation was noted on manometry. This was clearly confirmed on video swallow by symptomatic delay of the bolus at the sphincter before passage into the stomach. The cause of the dysphagia was therefore felt to be a motor disorder secondary to oesophagitis. She was commenced on ranitidine with some relief of symptoms. In case 5 there was no focal hold up of bolus at the HPB, but passage was sluggish throughout the oesophagus. This patient had previously undergone cardiac transplantation and had significant GORD on pHmetry and his dysphagia was therefore felt to be due to reflux and secondary impaired motility. His symptoms remain mild and intermittent on maintenance omeprazole.

3. Discussion

The condition of dysphagia aortica is reminiscent of the Churchillian paraphrase ‘A riddle wrapped in a mystery inside an enigma’ and it’s differential diagnosis includes numerous common structural and neuromuscular abnormalities. Many authors have documented a
Table 2
Manometric findings and results of ambulatory pHmetry

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basal pressure rise at level of H.P.B. (mmHg)</td>
<td>50</td>
<td>40</td>
<td>80</td>
<td>45</td>
<td>40</td>
</tr>
<tr>
<td>Distance from H.P.B. to L.O.S. (cm)</td>
<td>14</td>
<td>15</td>
<td>10</td>
<td>13</td>
<td>11</td>
</tr>
<tr>
<td>Distance from teeth to L.O.S. (cm)</td>
<td>38</td>
<td>40</td>
<td>40</td>
<td>40</td>
<td>42</td>
</tr>
<tr>
<td>U.O.S. tone (mmHg)</td>
<td>200</td>
<td>Normal</td>
<td>20</td>
<td>Normal</td>
<td>150</td>
</tr>
<tr>
<td>L.O.S. tone (mmHg)</td>
<td>20</td>
<td>14</td>
<td>10</td>
<td>18</td>
<td>15</td>
</tr>
<tr>
<td>U.O.S. relaxation</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>L.O.S. relaxation</td>
<td>Normal</td>
<td>Normal</td>
<td>Peristaltic, slightly reduced</td>
<td>Peristaltic, non-motensive</td>
<td>Normal</td>
</tr>
<tr>
<td>Swallow activity</td>
<td>Low amplitude, asyn-tactic distal</td>
<td>Normal</td>
<td>Peristaltic, slightly reduced</td>
<td>Peristaltic, non-motensive</td>
<td>Normal</td>
</tr>
<tr>
<td>Ambulatory pHmetry %</td>
<td>Total 9.9 (upright 12.5)</td>
<td>Total 1.5</td>
<td>Total 2.3</td>
<td>Total 13.8 (up- right 21.4)</td>
<td></td>
</tr>
</tbody>
</table>

U.O.S., upper oesophageal sphincter; L.O.S., lower oesophageal sphincter; H.P.B., high pressure barrier. Ambulatory pHmetry % indicates percentage of time that probe placed 5 cm above gastro-oesophageal junction records pH < 4 over a 24 h period.

The coexistence between symptomatic external vascular compression of the oesophagus, manometric abnormalities of swallow activity [3,8,13,15,17], and gastroesophageal reflux disease (up to 50% in some reports). Some have suggested that external compression of the oesophagus may give rise to the disordered motility by nerve compression [8,15]. We consider that there may be an additive relationship between these conditions in that secondary disordered motility may be a possible final pathway causing previously subclinical oesophageal compression to become symptomatic. This may account for their frequent coexistence, the finding of vascular high pressure barriers in apparently normal subjects [13], and help explain the finding that most patients with dysphagia aortica have mild, intermittent symptoms.

Reflux oesophagitis and motility disorders are common causes of mild dysphagia [18]. The coexistence of such associated pathologies and the lack of sensitivity and specificity of standard investigations make the diagnosis of dysphagia aortica difficult to establish with certainty, thus it is often one of exclusion [1,3]. Mittal et al., [14] have shown that the classical dysphagia aortica manometric HPB also occurs in normal subjects, hence this modality alone is insufficient to confirm the diagnosis. We therefore, believe that it is necessary to demonstrate a structural obstruction at the same level as the HPB before the diagnosis can be made with any confidence. Only then can other pathologies such as a motility disorder be excluded as the primary cause of the dysphagia.

In 4 of our 5 cases, the use of video solid swallow was of great help in establishing such a diagnosis when standard barium swallow was normal. Video evaluation with solid and liquid contrast is a high yield procedure for both structural and functional disorders [6,7]. Momentary hold up at sites of anatomical narrowing is considered normal when using solid contrast, but prolonged hold up requiring many liquid swallows to clear is deemed to be pathological [9]. In 2 patients this technique allowed us to establish a diagnosis of dysphagia aortica by demonstrating significant hold up of

Fig. 2. Composite photograph of liquid and solid swallows in patient 2. The standard barium swallow (left) shows indentation of the oesophagus at the level of the aortic knuckle (within normal limits), but free flow of barium past this area. The solid bolus swallow (right) demonstrates clear hold up of the contrast at this level.
### Table 3
Results of imaging investigations, endoscopy, and clinical diagnosis given to each patient

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest radiograph</td>
<td>Normal</td>
<td>Normal</td>
<td>Unfolded, tortuous aorta</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Computerised tomography</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Barium swallow</td>
<td>Normal</td>
<td>Hold up at aortic arch</td>
<td>Normal</td>
<td>Hold up at aortic arch</td>
<td>Hold up at aortic arch</td>
</tr>
<tr>
<td>Video solid bolus swallow</td>
<td>Normal</td>
<td>Hold up at aortic arch</td>
<td>Normal</td>
<td>Normal</td>
<td>Hold up at L.O.S. only</td>
</tr>
<tr>
<td>Endoscopy</td>
<td>Prominent pulsation left lateral wall of oesophagus</td>
<td>Normal</td>
<td>Benign stricture at 25 cm from teeth</td>
<td>Areas of confluent, non circumferential, distal oesophagitis (Grade 2)</td>
<td>Gastritis</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>Dysphagia aortica</td>
<td>Dysphagia aortica</td>
<td>Oesophageal stricture, uncertain aetiology</td>
<td>GORD/disorder of LOS function</td>
<td>GORD</td>
</tr>
</tbody>
</table>
solid contrast at the level of the aortic arch, confirming the manometric and subjective level of obstruction. In 2 further patients we were able to confidently exclude dysphagia aortica as prolonged hold up could not be demonstrated at the level of the manometric HPB.

The presence of an obstructing aortic vascular barrier at endoscopy may also be considered diagnostic, and allows therapeutic dilatation to be performed. It is suggested that dilatation gives relief in such cases by stretching scar tissue induced by stasis oesophagitis above and at the level of the aortic barrier [13]. If extrinsic vascular compression is demonstrated at endoscopy, contrast enhanced computed tomography or arch aortography should be considered to exclude the presence of abnormal vascular rings [22], or dysphagia lusoria (due to aberrant origin of the right subclavian artery) [17] which give rise to similar manometric and endoscopic features as dysphagia aortica. We found computed tomography to be of no value in assessing compression of the oesophagus by the aorta.

The treatment of dysphagia aortica depends on the severity of symptoms. Most cases are mild in nature and can be treated by avoiding foodstuffs likely to stick in the oesophagus [4,10,16], the treatment of any associated cardiac failure or hypertension may also significantly reduce symptoms [4,12–14]. More troublesome symptoms may respond to oesophageal dilatation[13]. Many surgical procedures have been described for the management of resistant cases. Anterolateral transposition of the distal oesophagus with posterior crural repair [13], mobilisation of the distal oesophagus from the aorta above the obstruction and creation of a posterior pleural sling [14], Hellen’s oesophagomyotomy [13,16], division of the right crus of the diaphragm to free the oesophagus combined with Nissen fundoplication [1], and aortic resection in the presence of aneurysmal dilatation [10] have all been used with some success. In patients unfit for a major procedure feeding gastrostomy has also been advocated [3].

When presented with a dysphagic patient in whom dysphagia aortica is considered, we suggest that manometry should be performed to confirm a localised, non circumferential, pulsatile, HPB and it’s level noted. Contrast swallow should then be performed to establish that symptomatic obstruction occurs at this same level and is related to the aorta. If barium swallow is negative, particularly in dysphagia to solids, we believe it is essential to proceed to video solid bolus swallow if the diagnosis is not to be missed. If manometry shows the typical HPB but the obstruction is not related to the aorta at contrast swallow, vascular imaging techniques should be used to exclude other causes of extrinsic vascular compression.

Neoplasia, reflux, and motility disorders remain the most common causes of dysphagia rather than vascular compression. Endoscopy must always be performed where barium swallow suggests a mucosal lesion if malignancy is not to be missed.

References