Congenital tracheo-oesophageal fistula in the adult

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ABSTRACT A case of congenital tracheo-oesophageal fistula without oesophageal atresia in an adult is presented. Surgical repair via a right thoracotomy was successfully undertaken. The 13 previously reported cases in English publications are reviewed.

Two subsequent bronchoscopies have been performed for recurrence of cough. On each occasion, part of a silk stitch was found at the repair site and removed. There has been no recurrence of the fistula and the patient is well. She eats a normal diet without difficulty.

Discussion

There are five main categories of congenital tracheo-oesophageal fistula and oesophageal atresia (table 1). The incidence of fistula without atresia varies and has been reported as ranging from 1.8% to 8.3%. This defect is a rare anomaly in infants and its persistence into adult life is indeed uncommon. The patient often presents with chronic lung disease of unknown aetiology.

There are 13 previously reported cases in English publications. The first was in 1929 and was an incidental postmortem finding. The second case reported underwent no treatment, and two later cases died from aspiration pneumonia. Subsequent cases have been successfully treated and are reported in table 2.

The criteria for diagnosing the defect as congenital in origin are based on: (1) respiratory symptoms over many years; (2) histological evidence of epithelial lining of and some muscularis mucosae in the fistulous tract; (3) the absence of an aetiological factor—for example, inflammation, neoplasm, Table 1 Classification by anatomical characteristics

<table>
<thead>
<tr>
<th>Classification</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atresia without fistula</td>
<td>9</td>
</tr>
<tr>
<td>Atresia with proximal fistula</td>
<td>1</td>
</tr>
<tr>
<td>Atresia with distal fistula</td>
<td>87</td>
</tr>
<tr>
<td>Atresia with proximal and distal fistula</td>
<td>1.2</td>
</tr>
<tr>
<td>Fistula without atresia</td>
<td>1.8</td>
</tr>
</tbody>
</table>
foreign body.

The patients' ages at the time of diagnosis have ranged from 15-63 years and there were equal numbers of males and females. Treatment has usually been closure via a thoracotomy.

A chronic cough and recurrent respiratory infections are almost always present and characteristically solid food is better tolerated than liquids. Increasing obliquity of the course of the fistula with age (because of differential growth rates of the oesophagus and trachea) explains the trivial symptoms in early life and consequent progression of the undiagnosed defect into adulthood.

Diagnostic confirmation of a fistula may be difficult. The surgeon must be aware of the possibility and contrast studies should demonstrate the fistula and not just reveal the presence of barium in the bronchial tree. With the rapid pharyngeal phase and upper oesophageal phase of swallowing, demonstration of a fistula may be difficult with conventional barium swallow examination. Cineradiography in the prone position with oblique views may be rewarding. Endoscopy is essential to exclude other pathology. It may not be possible to identify the oesophageal opening (as in this case) but contrast material or a catheter passed from the tracheal orifice will assist. If fibreoptic broncho-

Table 2  Summary of reported cases of congenital tracheo-oesophageal fistula in adults

<table>
<thead>
<tr>
<th>Author</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Negus (1929)4</td>
<td>45</td>
<td>M</td>
<td>Died—carcinoma lung</td>
</tr>
<tr>
<td>McKinney (1933)</td>
<td>58</td>
<td>F</td>
<td>Nil</td>
</tr>
<tr>
<td>Krausey (1958)</td>
<td>38</td>
<td>F</td>
<td>Died—pneumonia</td>
</tr>
<tr>
<td>Caldwell (1954)</td>
<td>54</td>
<td>M</td>
<td>Died—pneumonia</td>
</tr>
<tr>
<td>Mathey (1954)6</td>
<td>16</td>
<td>F</td>
<td>Thoracotomy repair</td>
</tr>
<tr>
<td>Demong (1959)9</td>
<td>15</td>
<td>F</td>
<td>Pneumonectomy repair</td>
</tr>
<tr>
<td>Ferguson (1959)10</td>
<td>23</td>
<td>F</td>
<td>Thoracotomy repair</td>
</tr>
<tr>
<td>Landsen (1960)11</td>
<td>63</td>
<td>F</td>
<td>Cervical repair</td>
</tr>
<tr>
<td>Zack (1967)12</td>
<td>32</td>
<td>M</td>
<td>Thoracotomy repair</td>
</tr>
<tr>
<td>Tenta (1967)13</td>
<td>16</td>
<td>M</td>
<td>Thoracotomy repair</td>
</tr>
<tr>
<td>Bertelsen (1970)14</td>
<td>48</td>
<td>M</td>
<td>Thoracotomy repair</td>
</tr>
<tr>
<td>Acosta (1974)15</td>
<td>48</td>
<td>M</td>
<td>Cervical repair</td>
</tr>
<tr>
<td>Stephens (1976)16</td>
<td>26</td>
<td>M</td>
<td>Thoracotomy repair</td>
</tr>
<tr>
<td>Present case</td>
<td>50</td>
<td>F</td>
<td>Thoracotomy repair</td>
</tr>
</tbody>
</table>

Figure  Barium swallow demonstrating the tracheo-oesophageal fistula outlining the right main bronchus (a) and anterior segment of the right upper lobe (b).
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Scopy is performed under general anaesthesia with an endotracheal tube the tracheal opening may be missed, so thorough examination of the trachea with a rigid bronchoscope is advised.

Congenital tracheo-oesophageal fistula without atresia is commonly known as the “H” type defect, indicating direct communication. As stated earlier, the opening in the trachea is usually situated more cephalad than the opening into the oesophagus. In some cases, a mucosal tag at the oesophageal stoma acts as a valve. Both these anatomical features minimise the inhalation of secretions and explain the low morbidity in these patients for long periods of time. It is this variant of the “H” type fistula one would expect to see in the adult group as it is compatible with prolonged survival. A useful terminology in the adult is “N” type fistula. This depicts more accurately the anatomical features and has some prognostic significance for the behaviour of the fistula.

I would like to thank Dr C Lomas for permission to report this case and for his support and the Department of Radiology for their diagnostic assistance.

References

4 Negus VE. Esophagus from a middle aged man, showing a congenital opening into the trachea. J Laryngol Otol 1929;44:184.
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