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Case Report

Congenital Tracheoesophageal Fistula Presenting in an Asymptomatic Adult- A Case Report

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Abstract:

Introduction: Congenital tracheoseophageal fistula (TEF) involve an abnormal pathological connection between the oesophagus and trachea. Congenital TEF presenting in adults is extremely rare and literature is limited to case series of significantly symptomatic patients.

Case presentation: An asymptomatic 41 year-old gentleman presented to clinic, having been diagnosed with a congenital tracheoesophageal fistula 10 years ago on a routine oesophagogastroduodenoscopy (OGD) in Brazil. He underwent a barium contrast swallow revealing contrast in the left main bronchi and significant pooling in the upper trachea. A computerised tomography (CT) scan of the neck and thorax with contrast revealed a small fistula measuring 6mm in diameter between the trachea and the oesophagus at the level of the second thoracic vertebral body. A microlaryngobronchoscopy and examination under anaesthesia visualised a fistula measuring 1.8cm to 2cm superior to inferiorly and similar in transverse dimension, 6-7cm below the level of the glottis.

Management and outcome: He opted to have surgical treatment to prevent symptoms developing in the future. A trans-cervical approach was performed, and the oesophageal defect and tracheal defect underwent primary closure with sutures. The oesophageal defect repair was oversewn with the sternohyoid. He recovered well post-operatively but due to the considerable rotation and retraction of the trachea, he suffered a vocal cord palsy. This is improving at the latest follow-up.

Discussion: Otolaryngologist (ENT) surgeons need to be aware of asymptomatic adult patients presenting with congenital tracheoesophageal fistulas and the investigation and management options available.

Keywords: tracheoesophageal fistula; gastro-oesophageal reflux

1. Introduction:

Tracheoesophageal fistulae (TEF) involve an abnormal communication between the oesophagus and trachea. These are usually diagnosed in childhood amongst other congenital abnormalities with a common incidence rate of 1 in 3500 births [1], or can be acquired in adults due to iatrogenic causes or malignancy. They can cause significant morbidity and mortality related to aspiration of saliva and food through the fistula. Congenital TEF presenting in adults is extremely rare and the literature is limited to case series and reports of significantly symptomatic patients [2].

Here, we present a rare circumstance of an asymptomatic gentleman diagnosed with a congenital tracheoesophageal fistula as an adult.

2. Case Presentation:

An asymptomatic 41-year-old Brazilian male, previously fit and well, presented to clinic with a congenital tracheoesophageal fistula, diagnosed incidentally 10 years ago.

As a child, he suffered from recurrent chest infections and symptoms consistent with aspiration, but this was never investigated further. He remained asymptomatic until 10 years ago where in Brazil a routine oesophagogastroduodenoscopy (OGD) for gastro-oesophageal reflux symptoms revealed a tracheoesophageal fistula, thought to be congenital. At the time, he opted for conservative management over surgical repair as his reflux symptoms settled.

He presented to our clinic in the UK with quiescent symptoms for many years, keen to finally have the TEF repaired. He otherwise had a fairly unremarkable medical history and his only surgical history of note was a laparoscopic inguinal hernia repair.

A barium contrast swallow revealed contrast in the left main bronchi after the second swallow (Figure 1). It also revealed significant pooling of contrast in the upper trachea when the patient was in the prone position (demonstrated by the arrow). As part of his work-up, an OGD was done which was surprisingly unremarkable, with it being difficult to visually demonstrate a tracheoesophageal fistula.

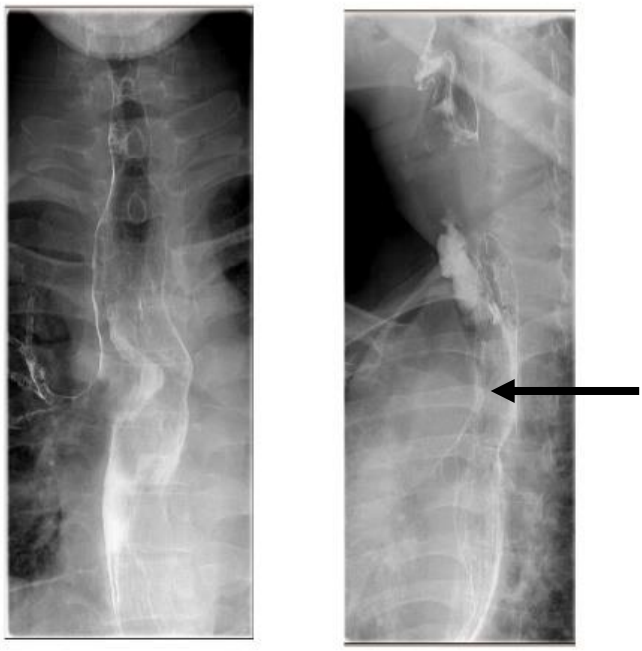

Figure 1: Barium contrast swallow images. Of note, there is evidence of contrast going into the left main bronchus and significant pooling in the trachea.

A computerised tomography (CT) scan of the neck and thorax with contrast revealed a small fistula measuring 6mm in diameter between the trachea and the oesophagus at the level of the second thoracic vertebral body (demonstrated by the arrow in Figure 2). Contrast was shown within the trachea and bronchi and an area of penetration into the posterior segment of the right upper lobe. Interestingly, mild bronchiectasis of the right middle lobe and a congenital butterfly vertebral anomaly of the 11th thoracic vertebrae were noted.

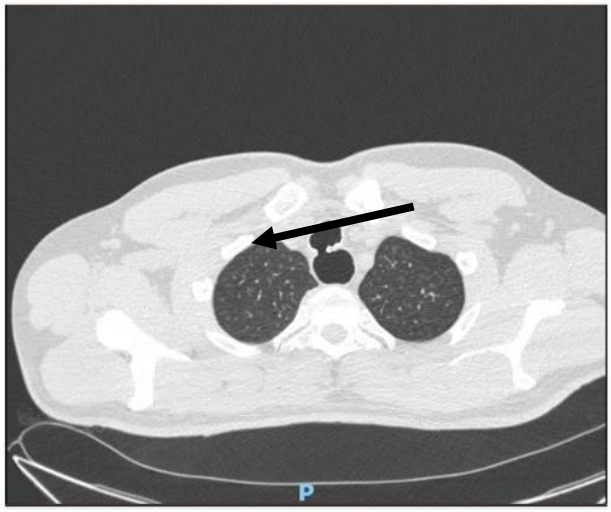

Figure 2: Computerised tomography (CT) scan of the neck and thorax with contrast. Of note there is a 6mm fistula between the trachea and oesophagus at the level of the second thoracic vertebral body (T2).

After discussion in the Upper Gastrointestinal (UGI) MDT, he was referred to the cardiothoracic and the ENT teams for consideration of tracheoesophageal fistula closure to avoid future aspiration pneumonia. Prior to the operation, he underwent a direct laryngobronchoscopy and examination under anaesthesia. The fistula was visualised measuring 1.8cm to 2cm in cranio-caudal dimension, 6.5cm distal to the glottis and 4cm proximal to the carina.

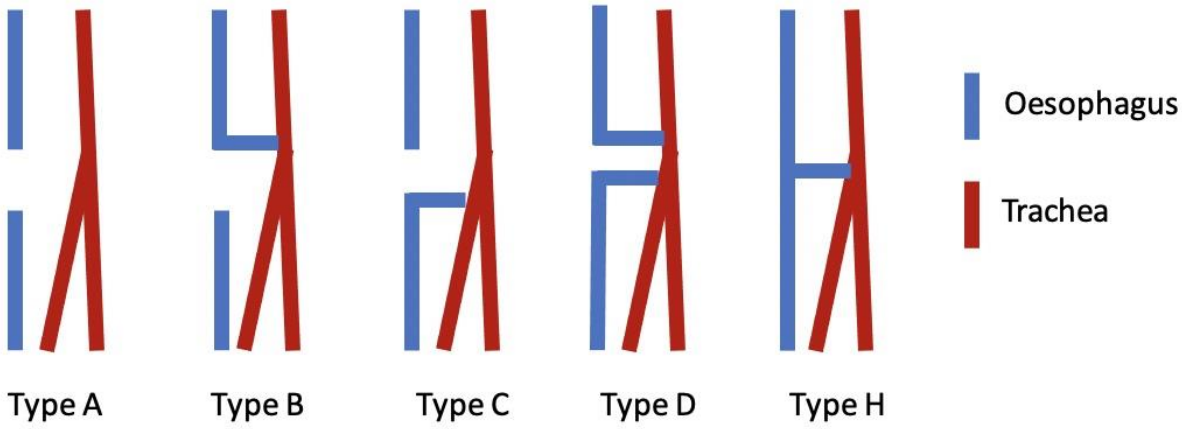

Figure 3: The five main types of congenital tracheaoesophageal fistulae. Type H fistulae are described as seen above.

Surgical repair was undertaken using a transcervical approach. Subplatysmal flaps were raised and dissection performed between sternocleidomastoid and strap muscles of the neck. The trachea was followed distally in the neck and the fistula delineated. The oesophageal defect and tracheal defects were both oversewn. The sternohyoid was dissected out, delivered into the mediastinum and then sewn over the oesophageal repair. He recovered well post-operatively, suffered no immediate post-operative complications, and was discharged home. Unfortunately, he suffered a vocal cord palsy due to the considerable rotation and retraction of the trachea involved. At follow-up one month post-operatively, there remained a flicker of movement of the left arytenoid. After undergoing speech therapy, his voice has markedly improved as of his most recent follow-up at 4 months post-operatively. There have been no swallow sequelae.

3. Discussion:

This is an interesting case of a congenital tracheoesophageal fistula diagnosed and treated in adulthood.

Congenital TEFs are commonly associated with oesophageal atresia, and as such present with severe morbidity and mortality in the neonatal population [4]. The prevalence of the condition is 1 in 3,500 births [1], usually as part of wider congenital syndromes e.g. VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) [4]. There are five types of congenital TEF (seen in figure 3), all of which are associated with oesophageal atresia apart from the H-type constituting 4% of all congenital TEFs [5]. This type of abnormality commonly escapes childhood diagnosis and presents in adults. There tends to be a history of recurrent aspiration pneumonia or coughing symptoms and if severe, result in chronic lung conditions like bronchiectasis [6]. Thus far in the literature there have been no identified asymptomatic adult patients presenting with a congenital TEF [7].

Due to the relatively low incidence rates of this condition in adults, there is no gold standard for diagnosis. A combination of clinical, radiological and endoscopic techniques are used. A literature review of patients aged 5 and over revealed the most sensitive tests to be CT chest with contrast and bronchoscopy, demonstrating over 80% chance of finding the defect on the first attempt [7]. Certainly, for our patient, both of these investigations were successful in identifying of the fistula. The diagnostic laryngobronchoscopy revealed accurate dimensions of the defect when compared with CT. Interestingly, in the same review, endoscopy was only successful in delineating the fistula in 50% of cases [7]. Certainly in our patient although the fistula was initially diagnosed on an OGD 10 years previously, his most recent OGD was unable to visualise it. Similarly, the barium contrast swallow strongly suggested a fistula with pooling of the contrast in the left main bronchi and upper main trachea, however the fistula itself was not identified. Multi-centred studies have suggested a combination of fluoroscopy and imaging increases diagnostic potential and accuracy [5,8].

Bronchiectasis is one of the most common presenting diagnoses of adults with congenital tracheoesophageal fistulae, at over 50% quoted in the literature [9]. Despite our patient denying any symptoms, his CT did reveal mild bronchiectasis of the right middle lobe. This factored into his decision to opt for surgical treatment as it would have been likely to progress.

Surgical management for fistulae around the T2-4 level such as this generally tends to favour a transcervical approach [10, 11], over transthoracic, with demonstrably good operative outcomes. The main advantages stem from a reduction in anaesthetic time and decreased post-operative respiratory dysfunction in respiratory-compromised patients compared to other surgical approaches. The oesophagus lacks serosa, therefore repair of the defect requires a local muscle flap [12] ranging from sternohyoid [13] to intercostal and pectoralis major flaps, as well as pleural, azygous vein and pericardial flaps [14]. Newer therapies include endoscopic over the scope clipping [15], local injection occlusive therapies [16] and laser, with argon coagulation used to promote re-epithelialisation [17]. Wider use of these newer therapies in adults have yet to demonstrate reproducible and clinically beneficial results.

Conclusion

The identification of congenital TEF in adults is possible but only if it is considered a likely diagnosis and appropriate investigations are performed. Management using the above technique can be successful however availability of thoracic surgeons intraoperatively is important, particularly for low fistulae. Often, a multi-faceted method of diagnosis and management is required and congenital TEF in adults should be a consideration in the differential diagnosis of bronchiectasis.

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