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Case Report

Triple Lumen Esophagus; A Rare Congenital Anomaly Presenting with Dysphagia

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ABSTRACT

Alimentary tract duplications are rare congenital malformations that can occur at any level of the gastrointestinal (GI) tract from mouth to anus. Triple lumen esophagus is even rarer occurrence and has not been reported previously according to our literature search. The diagnosis is often confused with esophageal tract duplication secondary to perforation or surgeries leading to Gastroesophageal fistula formation. A middle-aged female presented to us with an intermittent history of dysphagia and gastroesophageal reflux disease (GERD) and was diagnosed with a triple lumen esophagus radiographically, endoscopically which was confirmed and managed surgically.

Keywords: Triple lumen esophagus, alimentary tract duplication, esophagus, Dysphagia.

Introduction

The esophagus is normally a single lumen tube running between the pharynx and the stomach. As suggested by Gross [1], current nomenclature relies on the anatomic location of the duplication in relation to the normal GI tract and does not rely on the histologic features of the mucosal lining. Duplication may be defined as the presence of a secondary or tertiary tract located in or adjacent to the wall of the GI tract, possessing smooth muscle in the wall, and lined by alimentary tract mucosa. The lining mucosa may or may not be like that of the adjacent segment of the GI tract. Approximately two-thirds of all duplications are discovered within the first two years of life

and the ileum is the most common site of duplication. The esophagus is the second most common location. Although the exact incidence is unknown, in 1961 Potter suggests two reported cases in more than 9000 fetal and neonatal autopsies [2]. About 80% of duplications are found in the abdomen and 20% in the thoracic cavity. According to our literature review, there have been no previous reports on triple lumen esophagus and to this date, approximately eleven cases of the double lumen esophagus have been recorded [3]. We are reporting the endoscopic, radiological, and operative findings of a patient with triple lumen esophagus.

Case Report

A 45-year-old female, referred by a gastroenterologist, presented to the department of Thoracic surgery at Ojha Institute of Chest Diseases, Karachi, complaining of on and off retrosternal burning for 15-20 years, relapsing periods of dysphagia with odynophagia for six years and weight loss for four years. She came to our facility from a remote area and was inititally diagnosed with dyspepsia. Her vitals were within normal limits and physical exam was unremarkable. Her baseline investigations, as well as her Chest X-ray, were normal. Her medications included occasional anti-acids and some herbal remedies. A barium swallow was planned by our multi-disciplinary team, but since the patient was unable to tolerate oral intake due to severe odynophagia and dysphagia, we proceeded with endoscopy. Endoscopy revealed friable and severely ulcerated and abnormal-looking mucosa in the proximal part of the esophagus.

The middle and distal parts of the esophagus could not be visualized because the scope could not be negotiated further. Multiple biopsies were taken, and histopathology showed slough without any mucosal tissue. As the patient was unable to tolerate oral feeding and Percutaneous Endoscopic Gastrostomy (PEG) was not possible, a feeding jejunostomy was made. There was an improvement in her dysphagia after two weeks later. A barium swallow was performed which revealed double-lumen of the esophagus in the upper part with contrast in an irregular, ulcerated elongated blind-ending false tract lying posterolaterally, compressing the main lumen (Figure 1a). Chest CT was performed which revealed a single esophageal tube with two lumens opacified with oral contrast in the neck and chest just below the hypopharynx (Figure 1b).

The smaller tract on the left side extended between C7-T5 and measured 8.9cm in length. The second tract on the right side was 23cm in length and extended from C7-T12. Both tracts appeared to be intercommunicating at the level of T2 and T3 vertebrae. The true esophagus was compressed between the trachea and the smaller false tract. No extravasation of contrast was noted. The case was discussed at the multidisciplinary team meeting and a repeat endoscopy was advised. Endoscopy showed a single esophageal tube with two lumens, one large and one small starting at 17 cm from incisors.

The mucosa of the large lumen was normal, opened at the gastroesophageal (GE) junction while the smaller lumen had a tight ostium. An ultra slim scope was negotiated over a savory wire into the smaller lumen. As the scope was continued further into the smaller lumen, a third lumen was noted at 28 cm from incisors and the scope could not be negotiated into it. Here, a diagnosis of a triple lumen esophagus was made. The patient underwent McKeown esophagectomy (three-stage esophagectomy; laparotomy, thoracotomy, and neck exploration). Per operatively, a single firm esophageal tube was seen which was densely adherent to the prevertebral fascia. This tube was dissected in the chest and neck. At 17cm from incisors, normal esophagus with a single tube was found and mobilized. The stomach was anastomosed with the esophagus in the neck. After resection, radiological and endoscopic findings were confirmed by dissecting the resected esophagus (Figure 1c, d). The patient was allowed orally on the seventh postoperative day after excluding anastomotic leaks by contrast study. She has remained on regular follow-ups since discharge and is doing well at three months postoperatively.

A variety of congenital and acquired anomalies can affect the alimentary tract. One such entity is "alimentary tract duplication", first described in 1941 by Ladd, who comprehensively compiled many descriptive terms such as enteral cysts, ileal duplications, and other giant diverticula [2]. The exact etiology remains elusive, though there are several different theories about the origin of alimentary tract duplications. The aberrant luminal recanalization theory is the most widely accepted and explains that during embryologic development, the esophagus is partially solid. Its final lumen is established by the formation of vacuoles that coalesce. Sometimes vacuoles may fail to become connected with the esophageal lumen and result in duplication. Esophageal duplications account for the second most common duplications [4].

Fleming et al reported that esophageal duplication may be a post-endoscopy complication, although it is extremely rare. His team reported their endoscopic experience of 24 years and 37,808 endoscopies and only one patient had this complication [5].

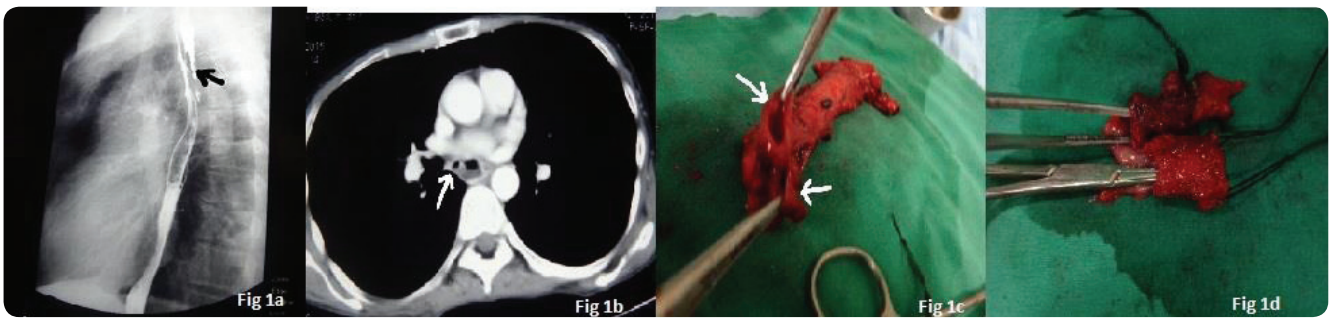

Figure 1: Contrast enhanced CT scan of the abdomen showing a large soft tissue mass (arrow) within the anterior abdominal wall in the epigastric region.

Congenital esophageal tract duplication can be differentiated from double esophageal lumen secondary to a fistula or perforation on the basis of location with the pathologic double-lumen occurring in the lower third of the esophagus often close to the GE junction whereas a congenitally duplicated tract is found in the proximal esophagus, usually along the entire length of the normal esophagus. The duplication may be extremely short or may include a large amount of the length of the normal GI tract. It may thus be described as either cystic or tubular in nature, with the tubular type being much rarer than the cystic type. In contrast to the cystic variety, the tubular variety may communicate with the normal mucosa at one or several points along the common wall [6]. In our patient, a single esophageal tube had 3 different lumens and they all shared the same mucosal wall.

Our patient was a middle-aged female. There have been other case reports about esophageal duplications in middle-aged patients [2]. Common symptoms include retro-sternal pain odynophagia and dysphagia. Likewise, our patient complained of retrosternal burning for about two decades before she presented to us. There was a complete symptom resolution post-operatively and she remains well at 3 months follow up. Diagnosis of such an entity is incidental and may be found during radiological investigations like barium studies or chest CT, which may show a double/triple-barreled esophagus with a longitudinal septum separating the different lumens [6]. Radiologically, these findings may be confused with alimentary tract diverticula or fistula formation. We did not find much literature on this topic due to the rarity of this pathology. We found reports of patients presenting with a double-lumen esophagus in the lower third secondary to untreated GERD or secondary to Nissen fundoplication procedure complicated by gastroesophageal fistula formation. Hussain et al [7] treated their patient's symptoms by surgical reconstruction of the distal esophagus using a novel stapling technique through a gastrostomy. In our patient, almost the entire esophagus was found to be abnormal except for the proximal 2cm (15cm from incisors till 17cm) and her disease could not be treated with medications or endoscopic procedures hence we had to resect and reconstruct proximal 2cm of the normal esophagus with stomach tube in the neck.

Conclusion

Triple lumen esophagus is rare and has not been frequently reported in the literature. Our aim was to highlight the importance of anatomical variations in people. Anomalies of the upper GI tract should always be kept in mind in patients who present with intermittent or recurrent dysphagia.

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