Communicating tubular esophageal duplication combined with broncho-esophageal fistula

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Abstract

Esophageal duplication (ED) is rarely diagnosed in adults and usually asymptomatic. Especially, ED which is connected to the esophagus through a tubular communication and combined with broncho-esophageal fistula (BEF), is extremely rare and has never been reported in English literature. This condition is very difficult to diagnose as well. Although some combinations of several modalities, such as upper gastrointestinal endoscopy, esophagography, CT, MRI and EUS can be used for diagnosis, they might be inconclusive. Here, we report a patient with communicating tubular ED that was incidentally diagnosed on the basis of the endoscopy and esophagography during post-operative evaluation of BEF.

Key words: Esophageal duplication; Broncho-esophageal fistula; Bronchial fistula; Esophageal fistula
INTRODUCTION

Gastrointestinal tract duplications are uncommon congenital anomalies and found anywhere along the alimentary tract from the mouth to the anus. Especially, esophageal duplication (ED) is rarely diagnosed in adults. Estimated incidence of congenital ED is 1:8200, with male sex predominance.\textsuperscript{1, 2} Almost all these cases were detected with respiratory distress manifestation in early days of life. Most EDs are found in the distal third of the esophagus and are frequently incidental findings at routine chest radiography. ED is divided into three types: cystic (most common type), tubular and diverticular forms.\textsuperscript{3} ED can be associated with other congenital anomalies, such as small intestine duplications, esophageal atresia, broncho-esophageal fistulas (BEF), and spinal abnormalities.\textsuperscript{4} Less than 20\% of the EDs can communicate with the main lumen by a tubular tract.\textsuperscript{5} Also, ED could communicate with the tracheo-bronchial tree and make fistulae.

Communicating tubular ED is extremely rare and only one case report of communicating tubular ED combined with BEF in an adult was published in 1994 in Japanese literature.\textsuperscript{6} There was no report of such case in English literature.

Here, we report a case of communicating tubular ED combined with BEF diagnosed on the basis of the endoscopy and esophagography during post-operational evaluation of BEF.

CASE REPORT

A 49-year-old male was admitted to our hospital's emergency center due to herbicide poisoning. After drinking several glasses of alcohol and herbicide, he presented persistent nausea and vomiting. He had been drinking about 54 grams of alcohol daily and had 50-pack-year history of cigarette smoking. He had a history of pulmonary tuberculosis and received anti-tuberculosis medication 8 years ago. At that time, he was also diagnosed with BEF and alcoholic liver...
disease. Intermittent and persistent cough had developed for a while. At admission, initial physical examination revealed no specific finding except an elevated respiratory rate of 28/min. The results of laboratory tests were as follows; white blood cell count 15,660/mm$^3$, hemoglobin 12.9 g/dL, hematocrit 38.1%, platelet count 155,000/mm$^3$, total protein 7.9 g/dL, albumin 3.7 g/dL, aspartate aminotransferase (AST) 80 U/L, alanine aminotransferase (ALT) 37 U/L, prothrombin time (INR) 1.03 and C-reactive protein (CRP) 3.48 mg/dL. The results of blood gas were pH 7.513, pCO$_2$ 35.2 mmHg, pO$_2$ 80.5 mmHg and HCO$_3$ 27.7 mmol/L. Chest X-ray demonstrated a consolidation in the lower left zone of the lungs. After being diagnosed with aspiration pneumonia, the patient was treated with conservative management. On the second hospital day, he developed severe persistent cough. On endoscopic and bronchoscopic examinations, a fistula opening was found at the mid-esophagus and the opening had whitish surface with slightly screwed pattern without inflammatory sign or discharge (Fig. 1A). We couldn't localize the other opening or a fistula tract. Previous esophagography (Fig. 1B) taken at a local hospital revealed a communicating fistula tract between the bronchus and the lower esophagus (BEF), but chest computed tomography (CT) also taken at the local hospital didn't show discrete BEF. On the 6th hospital day, his pneumonic condition was well improved and he could ambulate. On the 10th hospital day, he took a surgical operation for the repair of the BEF. During the operation, a 2cm long fistula tract between the lower esophagus and the medial basal segment of the left lung was noted, which was successfully removed and repaired. His condition was stable and had no complication. On the 3rd post-operational day, a follow-up esophagography for the post-operational evaluation revealed a contrast leakage at the left side of the mid-esophagus and drainage to the distal esophagus (Fig. 2). We thought that the leakage resulted from a missing condition, such as ED with esophago-esophageal fistula (EEF) (Fig.2). On a follow-up endoscopic examination, the proximal fistula opening that was different from previously noted BEF opening was found at the mid-esophagus and the distal opening was found on the cardia of the stomach, located in the hiatal hernia (Fig.3). On chest CT, about 7mm
sized air-filled tract was noted, one end of which was connected with the distal esophagus and the other end was connected with the cardia of the stomach (Fig. 4). The final diagnosis of the patient was communicating tubular ED with BEF (Fig. 5). Initially he was operated for the symptomatic BEF but a communicating ED was detected incidentally later during a post-operational follow-up. Symptoms of BEF were improved without any unusual complication, and he wanted to be discharged without any further treatment or surgery. For 2 years of follow-up, he has been doing well without any complication of ED with BEF.

DISCUSSION

The only case of communicating tubular ED with BEF was published in Japanese literature. The patient was 51-year-old woman who had chronic cough when drinking water. Esophagoscopy revealed tubular ED with BEF and endoscopy showed one small opening of ED. She was operated to remove the BEF with ED.

Most EDs are found in the distal third of the esophagus and are frequently asymptomatic, although dysphagia, respiratory distress, recurrent pneumonia, vomiting, failure to thrive, gastro-esophageal reflux, melena and anemia may be accompanied in rare cases.\(^1\,\,^7\)

At the time of admission, our patient had had chronic intermittent cough and knew about his BEF. Pneumonia might be considered as a complication of drug intoxication or BEF. On admission, persistent cough developed abruptly and the patient wanted evaluation and treatment of his disease. Chest CT and endoscopic exam didn't reveal communicating ED at that time, although post-operational, follow-up esophagography for confirmation of fistula closure revealed another remaining fistula tract. This fistula tract communicated not with the bronchus but with the distal esophagus as a tubular structure, which is an evidence of the possibility of ED. Therefore, endoscopic re-examination and follow-up chest CT were proceeded and we finally confirmed ED.
Later we incidentally diagnosed that the patient had a communicating tubular ED with BEF, which we couldn't find on the initial evaluation. The ED was initially not detected because we thought the cough symptom was caused by the BEF that was detected on esophagography at previous hospital. Another possible reason is that the ED was not obvious on the esophagography. The ED was congenitally accompanied by the BEF and they shared the same opening, which might have brought relatively less influx of the contrast media into the ED than into the BEF, making the ED undetectable on the esophagography. According to the assumption from interdisciplinary discussion, it is highly likely that the removal of the BEF might have made the ED easily accessible by the contrast media and thus more detectable on esophagography. This is also supported by the fact that ED was suspected upon the review of the previous CT images (images not presented here).

One major limitation of this report is that the possibility of an esophago-gastric fistula, not an ED, could not be confirmed by histology. Another case report of an ED similar to our case has verified ED by histology. However, as the endoscopic findings suggest (Fig. 3B), the orifice inside the hiatal hernia appears to be covered by the epithelium of the esophagus, suggesting the possibility of an orifice from the esophageal duplication, rather than an esophagogastric fistula. A histological examination would be helpful for differential diagnosis in patients showing similar findings in the future.

Identifying an ED is very difficult. Upper GI endoscopy can make a diagnosis with direct vision but this is not possible to determine whether it is a communicating one or a non-communicating one. In our case, initial endoscopic exam for confirmation of BEF couldn't find communicating tubular ED. But the result of the follow-up esophagography revealed the possibility of communicating tubular ED and we conducted repeat endoscopy carefully. Follow-up endoscopy found two openings of ED. As can be seen in Figure 3, the distal orifice of the ED was at the sliding hiatal hernia sac and it was very hard to find. The opening could be seen along
the whole esophagus as a whitish or pinkish hole not discriminated from the surrounding opening tissue.

Several imaging studies for diagnostic work-up were also needed. Radiological methods can be helpful in the diagnosis and localization of the disease. Esophagography may show displacement of the esophagus by a para-esophageal mass but tubular ED may not influence the esophagus.⁹ Contrast esophagographic study could be useful preoperatively to diagnose a communicating ED and during the postoperative follow-up period to confirm that a leak is not present. CT could demonstrate the exact anatomic position of the fistular tract and influence the decision making about resectability. MR imaging, endoscopic ultrasonography (EUS) and Tc-99m pertechnetate scintigraphy could be helpful for the diagnosis of this disease.⁸ However, these studies are mainly inconclusive.¹⁰,¹¹

Management of ED is dependent on the type and size of ED and severity of symptoms. Complete resection is a well-documented treatment for duplication of the esophagus but conservative management can be an option for non-symptomatic patients. Symptomatic ED has mainly been managed with extensive surgery via a thoracotomy.¹¹ With recent advances in minimally invasive surgery, surgical treatment has gained in efficacy and offers advantages to patients. Clear exposure and identification of the duplication are important for successful operation. After surgical excision, the prognosis is encouraging. As our patient received primary repair of BEF without knowledge of ED and his symptoms disappeared, we didn't conduct additional surgery for ED. After surgery, he has been doing well during the follow-up period.

In conclusion, identifying a communicating ED with BEF is difficult. On the examination for diagnosis of BEF, the possibility of ED should be considered and careful endoscopic examination is also important for detection of fistula orifices.

Conflicts of interest
All authors declare no conflict of interests for this article.

References


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duplication cyst (with video). Gastrointest endosc 2006;64:288-289.

Figure legends

Figure 1. A. Initial upper GI endoscopy reveals an opening (arrow) in the mid-esophagus. B. Esophagography taken at another hospital shows a broncho-esophageal fistula (arrow) between the mid-esophagus (arrow head) and the lower left bronchus.

Figure 2. Follow-up esophagography. - Suspicious contrast leakage at the left side of the mid-esophagus and drainage to the distal esophagus were noted.

Figure 3. Follow-up upper GI endoscopy. A. At mid-esophagus, proximal opening of ED was seen (arrow). The location of the proximal opening was different from the previous examined area. B. Distal opening of the ED was found at the cardia in hiatal hernia sac (arrow).

Figure 4. Chest CT found about 7mm sized air-filled tract (arrow). A. Axial section view. B. Coronal section view.

Figure 5. A schematic diagram of patient's pathological anatomy.