Incomplete Duplication of the Esophagus: A Case Report

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A case of incomplete duplication of the esophagus diagnosed in a 20-month-old girl with an esophageal perforation is presented. X-ray films of the chest showed pneumomediastinum and subcutaneous emphysema. A water soluble contrast esophagogram revealed a contrast leakage from the cervical esophagus to the thoracic inlet. The endoscopic findings are described. Complete resection of the esophageal duplication was effective.

Keywords: *Esophagus, Incomplete esophageal duplication, Mediastinitis, Pneumomediastinum*

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Duplications in the alimentary tract are unusual congenital anomalies. They may be found anywhere between the mouth and anus. The ileum is the most commonly affected site, followed by the thoracic esophagus, usually in the lower third (60%). A cervical esophageal duplication is extremely rare[1]. The symptoms are obstruction, pain, local hemorrhage, respiratory distress or perforation. The duplications are commonly classified into two types, tubular and cystic.

Case Report

A 20-month-old girl presented to her local hospital with a 4-day history of vomiting and dysphagia which had developed into a swelling in the left side of her neck and chest with dyspnea and fever. The physical examination found a BP of 100/70 mmHg, PR 180/min, RR 36/min, and BT 39 C. There was subcutaneous emphysema in her neck and chest. X-ray films of the chest showed pneumomediastinum and subcutaneous emphysema (Fig. 1). A water soluble contrast esophagogram revealed a contrast leakage from the cervical esophagus to the thoracic inlet (Fig. 2). (At birth the infant had been asymptomatic)

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Following transfer to the authors' institution, a computed tomographic (CT) examination of her neck and chest demonstrated a fluid-filled tubular structure alongside the esophagus beginning in the left posterior hypopharynx, extending into the left posterior mediastinum, distal margin immediately adjacent to the native esophagus at the carina level, and pneumomediastinum (Fig. 3). Endoscopy revealed, in the upper third of the esophagus, thin septa, arising in opposite

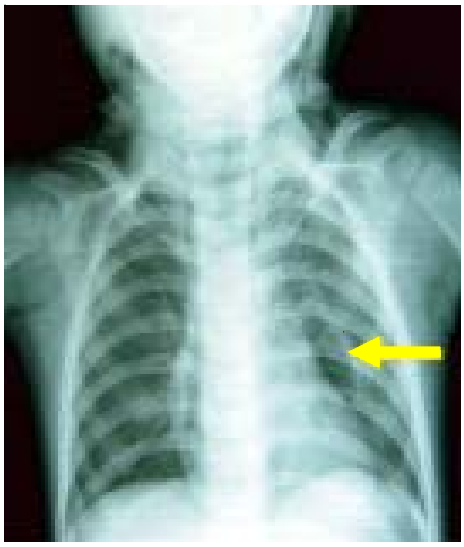


Fig. 1 Chest X-ray shows the pneumomediastinum and subcutaneous emphysema

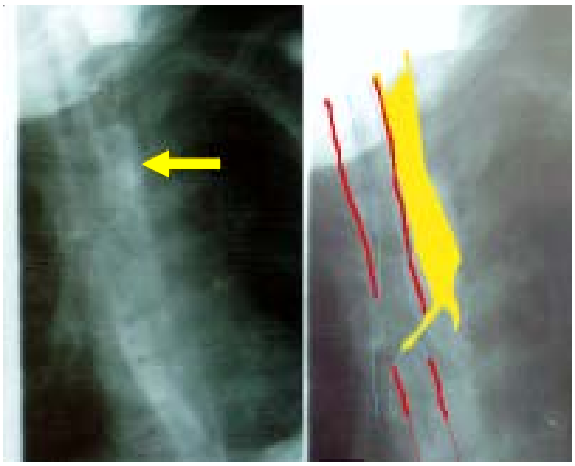
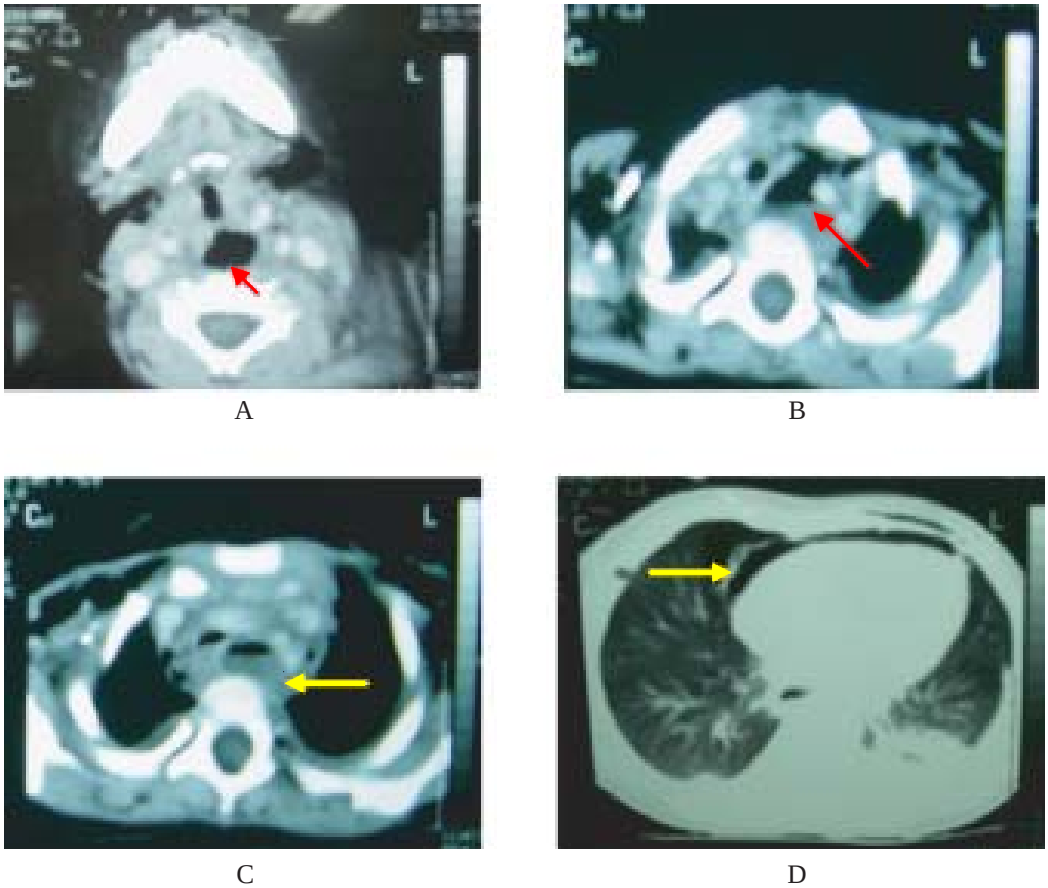


Fig. 2 The esophagogram shows the left tubular esophagus

walls, which were occasionally confluent, giving rise to a double lumen of the esophagus (Fig. 4). A gastroscope was easily introduced into both esophageal lumen. The mucosal lining of the septum was macroscopically no different from the normal lining of the esophageal wall, but the end pouch was inflamed.

After resuscitation and pre-operative management, the lesion was approached via a left transverse cervical incision, retracting the sternomastoid muscle and vessels laterally (Fig. 5). The duplication was resected and the esophagus was repaired with interrupted absorbable sutures. A right thoracotomy was opened for mediastinal drainage and distal esophageal duplication evaluation. A Stamm's gastrostomy was performed to allow the patient to eat. The excised esophageal duplication consisted of a squamous cell epithelium and inflammatory cells infiltrated into the mucous membrane (Fig. 6). Postoperatively, 3 months



A B C D

Fig. 3 Computed tomographic images outlining the duplication in the neck and the chest. In the upper neck (A), the beginning esophageal duplication arises to the left of the esophagus. In the thoracic inlet (B), the duplication is larger(2cm), contains an air fluid level and free air. In the upper thorax (C), the duplication contains an air fluid level. In the lower thorax (D), is pneumopericardium

Fig. 4 Esophagoscopy shows the double esophageal lumen

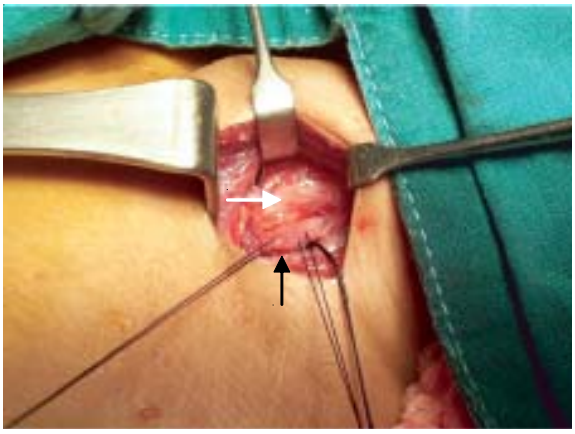


Fig. 5 The left cervical exploration, the duplication (black arrow) arises to the left of the esophagus (white arrow)

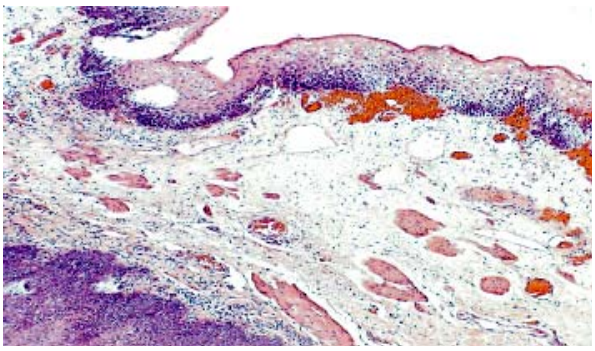


Fig. 6 Histopathological finding of the tubular duplication showing stratified squamous epithelium with submucosa and muscular layer. H&E, x 100

later the patient was doing well and able to eat a regular diet.

Discussion

Alimentary tract duplications are unusual events in the pediatric population. Only 10-15% of alimentary tract duplications involve the esophagus, with approximately 60% of these being in the distal portion. Embryologically, duplications result from a defect in the tubulation (vacuolization) of the esophagus, normally occurring in the sixth gestational week. As the foregut epithelium develops, it elongates, develops a lumen, and undergoes dextro-rotations. Thus, the majority of esophageal duplications occur distally and on the right. In the present case the esophageal duplication was proximal to the cervical part and on the left.

The duplication of the esophagus may be of three forms: 1) a cystic form[2-13] that may or may not communicate with the esophageal lumen; 2) a tubular form[14,15]; or 3) a diverticular form[16,17]. In one study[18], only six tubular duplications of 44 esophageal duplications were identified. In another review of alimentary tract duplications[3], 22% were intrathoracic, and all were cystic rather than tubular. The present report reveals a case of an incomplete tubular duplication of the esophagus.

The duplication of the hollow structures of the digestive tract manifest three essential points: 1) they are in intimate contact with the alimentary tract; 2) they are lined by a mucous membrane similar to other such membranes at the same level of the digestive tract; and 3) they are provided with a smooth muscle coat, though an esophageal duplication without muscle fibers has been reported[19]. In the present case the wall dividing the esophagus lumen into two was simply a mucous membrane.

An esophageal duplication can be an isolated anomaly or can be seen with other malformations of the intestinal system such as esophageal atresias[9-13], fistulae, bronchopulmonary foregut malformations[4], duplication of the bowel or stomach, diaphragm, spinal abnormalities, including scoliosis, hemivertebrae, and fusion. They are usually associated with vertebral anomalies due to a similar embryologic development, but rarely other duplications of the digestive tract.

Esophageal duplications are found incidentally in asymptomatic patients; however they become symptomatic when complications such as hemorrhage, rupture, pain, vomiting, displacement of adjacent organs, obstruction, respiratory distress, infection or perforation occur. In the present patient, the symptoms, including vomiting, dysphagia, fever, pneumomediatinum and mediastinitis, may have been caused by infection of the esophageal duplication and inflammation inducing the perforation of the esophagus.

The diagnosis of esophageal duplication is complicated by their rare occurrence, and by the

variety of locations they can exhibit within the mediastinum. CT and MRI have been used to demonstrate esophageal duplication and vertebral anomalies. A barium swallow is effective for the detection of a tubular duplication. Endoscopy may reveal the opening in tubular duplications, while cystic duplication may show only esophageal obstruction, due to extraluminal compression. Esophageal US may demonstrate a lesion, or a technetium pertechnetate scan can demonstrate gastric mucosa in the duplication.

The definitive diagnosis of esophageal duplication requires pathological evaluation. The lesion should meet the pathological criteria of a connection with the esophageal wall, two layers of muscularis propria and squamous epithelium.

To summarize, the presented patient had an incomplete tubular esophageal duplication at the left cervical level with perforation and mediastinitis which was revealed after an infection. Complete resection of the esophageal duplication was effective.

References

1. Wrenn EL, Hollabaugh RS. Alimentary tract duplications. In: Ashcraft KW, ed. Pediatric surgery. Philadelphia: Saunders, 2000: 527-39.
2. Nakahara K, Fujii Y, Miyoshi S, Yoneda A, Miyata M, Kawashima Y. Acute symptoms due to a huge duplication cyst ruptured into the esophagus. Ann Thorac Surg 1990; 50: 309-11.
3. Bajpai M, Mathur M. Duplications of the alimentary tract: clues to the missing links. J Pediatr Surg 1994; 29: 1361-5.
4. Kitano Y, Iwanaka T, Tsuchida Y, Oka T. Esophageal duplication cyst associated with pulmonary cystic malformations. J Pediatr Surg 1995; 30: 1724-7.
5. Karahasanoglu T, Ozbal A, Alcicek S, Goksel S, Altun M. Giant intra-abdominal esophageal duplication cyst. Endoscopy 1997; 29: S54-5.
6. Bhutani MS, Hoffman BJ, Reed C. Endosonographic diagnosis of an esophageal duplication cyst. Endoscopy 1996; 28: 396-7.
7. Snyder ME, Luck SR, Hernandez R, Sherman JO, Raffensperger JG. Diagnostic dilemmas of mediastinal cysts. J Pediatr Surg 1985; 20: 810-5.
8. Michel JL, Revillon Y, Montupet P, Sauvat F, Sarnacki S, Sayegh N, et al. Thoracoscopic treatment of mediastinal cyst in children. J Pediatr Surg 1998; 33: 1745- 8.
9. Fernandes ET, Hollabaugh RS, Boulden T. Mediastinal mass and radiolucent esophageal foreign body. J Pediatr Surg 1989; 24: 1135-6.
10. Yamagiwa I, Obata K, Ouchi T, Sotoda Y, Shimazaki Y. Heterotopic pancreas of the esophagus associated with a rare type of esophageal atresia. Ann Thorac Surg 1998; 65: 1143-4.
11. Hemalatha V, Batcup G, Brereton RJ, Spitz L. Intrathoracic foregut cyst (Foregut duplication) associated with esophageal atresia. J Pediatr Surg 1980; 15: 178-80.
12. Narasimharao KL, Mitra SK. Esophageal atresia associated with esophageal duplication cyst. J Pediatr Surg 1987; 22: 984-5.
13. Snyder CL, Bickler SW, Gittes GK, Ramachandran V, Aschcraft KW. Esophageal duplication cyst with esophageal web and tracheoesophageal fistula. J Pediatr Surg 1996; 31: 968-9.
14. Cantallops JG, Adrover AO, Fernandez JM, Fernandez AB. Incomplete duplication of the esophagus: one case. Endoscopy 1981; 13: 46-8.
15. Dresler CM, Patterson GA, Taylor BR, Moote DJ. Complete foregut duplication. Ann Thorac Surg 1990; 50: 306-8.
16. Ohbatake M, Muraji T, Yamazato M, Higashimoto Y, Nishijima E, Tsugawa C. Congenital true diverticula of the esophagus: a case report. J Pediatr Surg 1997; 32: 1592-4.
17. Gorenstein A, Serour F, Bujanover Y. Unusual presentation of esophageal communicating duplication in a child. J Pediatr Surg 1999; 34: 1430-1.
18. Peiper M, Lambrecht W, Kluth D, Huneke B. Bleeding esophageal duplication detected in utero. Ann Thorac Surg 1995; 60: 1790-1.
19. Knight J, Garvin PJ, Lewis E Jr. Gastric duplication presenting as a double esophagus. J Pediatr Surg 1983; 18: 300-1.

รายผู้ป่วยมีหลอดอาหารซ้ำซ้อนแบบ Incomplete duplication

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รายงานกรณีศึกษาเด็กผู้หญิงอายุ 20 เดือน มาด้วยอาการอาเจียน และ mediastinitis ผลจากการรักษาพบการอักเสบและทะลุของปลาย incomplete tubular esophageal duplication ซึ่งมีความยาวจาก cervical esophagus ถึง thoracic inlet ได้รับการผ่าตัดออกทั้งหมด ผู้ป่วยหายดี กลืนอาหารปกติ