CASE REPORT

doi: 10.5455/medarh.2015.69.425-427

Med Arh. 2015 Dec; 69(6): 425-427

Received: September 25th 2015 | Accepted: November 05th 2015

© 2015 Tamer Sekmenli, Ilhan Ciftci, Mesut Sivri, Mustafa Koplay

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Short-gap Isolated Esophageal Atresia Causing Stridor Due to Compression of the Trachea

Tamer Sekmenli, Ilhan Ciftci, Mesut Sivri, Mustafa Koplay

Department of Pediatric Surgery, Medical Faculty, Selcuk University, Konya, Turkey
Radiology department, Medical Faculty, Selcuk University, Konya, Turkey

Corresponding author: Tamer Sekmenli. E-mail: dr_sekmenli@hotmail.com

ABSTRACT

Isolated esophageal atresias are reported always to be associated with long gap in the literature. In this manuscript, we aimed to discuss the imaging and surgical treatment methods of an isolated esophageal atresia case with 'short gap' who had stridor due to compression of the trachea by dilated upper esophageal pouch and had not identified previously in the literature.

Key words: stridor, compression of trachea, short-gap isolated esophageal atresia.

1. INTRODUCTION

Esophageal atresia (EA) is a congenital anomaly characterized by the lack of continuity within the esophageal lumen due to a disruption in the embryological development of esophagus. Anatomically, it is divided into five categories. The most common type is proximal atresia with tracheal fistula accompanying distally (1). Clinical symptoms such as regurgitation, inability to feed and foamy salivation from mouth and nasal cavity draw attention at first feeding. Cough and respiratory distress may be present due to aspiration of the saliva collected in the proximal blind pouch. Another rare cause of respiratory distress might be the compression of trachea by dilated upper pouch with saliva. Clinical findings, prenatal ultrasonography (USG), radiography, fluoroscopy and cross-sectional imaging methods are used in diagnosis and classification (2, 3). It is important to know the gap length between the proximal and distal atresic segments, in terms of, choosing the appropriate surgical procedure.

In this manuscript, we aimed to discuss the imaging and surgical treatment methods of an isolated esophageal atresia case with 'short gap' who had stridor due to compression of the trachea by dilated

upper esophageal pouch and had not identified previously in the literature.

2. CASE REPORT

A female newborn whom fetal ultrasound at 7th month showed polyhydramnios with no visible fluid in the stomach was delivered with a birth weight of 1660g, and admitted with the prediagnosis of esophageal atresia. On physical examination, saliva were spreading from her mouth and nose, in addition to evident stridor breath sounds were decreased, and the abdomen was noticed to be scaphoid. A nasogastric tube could not be advanced to stomach. Straight X-Ray demonstrated a curled tube within the air filled proximal esophageal pouch, and absence of gas in the abdomen (Figure 1). In order to visualize atresic pouch for a definitive diagnosis, a contrast study by injecting 2 mL of contrast medium through the nasogastric tube was performed. A water-soluble contrast agent (Ultravist) was used, since there was a risk of aspiration. Tracheobronchial tree was visible due to aspiration of contrast material, which was overflowed from the upper pouch. Proximal esophageal pouch filled with contrast medium was significantly compressing the trachea (Figure 2). These findings

Figure 1. Isolated esophageal atresia absence of gas in the abdomen

Figure 2. Proximal esophageal pouch filled with contrast medium was significantly compressing the trachea

promoted the diagnosis of isolated esophageal atresia. Other examinations showed no additional anomaly. Initially we justified performing a feeding gastrostomy and surgical exploration demonstrated a microgastri. We performed a stamm type gastrostomy, and the patient was started on enteral feeding. Around two months of age, contrast studies in Trendelenburg's position were performed to show the length of the distal esophagus. However, distal esophagus and the gap could not be

viewed in this way. Since long gap is seen usually in isolated cases, patient was presumed to have long-gap atresia and additional investigation was not done. When the patient was 3 months of age, a right thoracotomy through 5th intercostal space was performed. Surgical intervention demonstrated that distal esophagus and trachea were attached with a fibrous band without any passage (Figure 3 and 4). Following division of distal esophagus from trachea, in contrast to our expectation, there was no significant gap between the proximal pouch and distal esophagus, which enabled a tension-free end-to-end anastomosis. Enteral feeding was started via gastrostomy on postoperative day 2, contrast study on postoperative day 8 demonstrated no leakage from anastomosis. Patient was discharged on postoperative day 12.

3. DISCUSSION

Etiology of esophageal atresia is still poorly understood. There are additional anomalies in 30-70% of cases, and cardiovascular anomalies (VSD, ASD, PDA, tetralogy of fallot, coarctation of the aorta) accompany esophageal atresia most commonly. Of the cases, 25% have gastrointestinal (anorectal malformation, rotation anomalies, annular pancreas, duodenal-jejunoileal atresia, pyloric stenosis), 20% have genitourinary (renal dys-

Figure 3. Distal esophagus and trachea were attached with a fibrous band

Figure 4. Proximal to distal esophageal blind pouch end

plasia, renal agenesis), 13% have skeletal and 3-5% have nervous system anomalies (5).

Prenatal ultrasonography may detect 20-40% of patients during antenatal period (2). Esophageal atresia should be kept in mind when fetal USG shows polyhydramnios, lack or absence of fluid in stomach (microgastria), anechoic structure in the middle part of fetal neck, enlarged esophageal pouch. The earliest finding in the neonatal period is excessive secretions from mouth due to the accumulation of secretions in the posterior pharynx. Typically, first feeding leads to cough, regurgitation and respiratory distress. The main cause of respiratory distress is aspiration of saliva accumulated in the upper pouch into the trachea. Although very rare, another reason for the respiratory distress is tracheal compression by dilated upper pouch as represented in this case. When the literature was reviewed, stridor caused by tracheal compression due to dilated upper pouch does not involve in typically defined in clinical symptoms of patients with esophageal atresia. Stridor was the dominant respiratory problem in this case. Abdomen is distended in cases of distal TEF, whereas infants who have isolated esophageal atresia and proximal fistula have scaphoid abdomen. Absence of gas in abdominal X-ray is a typical sign for isolated esophageal atresia (6). Inability of to pass nasogastric tube to the stomach provides diagnostic support, but pharyngeal pseudo-diverticula and laryngo-tracheo-esophageal clefts can also block the tube going further down, so they should be considered in the diagnosis (4). Radiological display of the upper pouch filled with contrast material was given through catheter confirms the diagnosis (2). This procedure also provides information about the level of the proximal esophagus.

Preoperative preparation is very important on survival after diagnosis was confirmed. Infants should be kept in semi-fowler position and proximal pouch should be aspirated continuously with Replogle catheter which has double-lumen to reduce the risk of aspiration and pneumonia. Concomitant abnormalities should be investigated. Preoperative start of prophylactic broad-spectrum antibiotics and fluid support are important.

Initial surgical treatment often includes feeding gastrostomy, because of long gap. Level of bottom pouch and gap length are tried to be determined by giving retrograde contrast material through gastrostomy during operation. Although not common but since extension techniques won't be need in patients with a short gap distance as in our case, one should not forget to determine the gap length first. In this regard multi-slice imaging modalities such as computed tomography and magnetic resonance imaging may be quite useful.

Tension-free anastomosis is often not possible in classical treatment because of the longer distance between the upper and lower esophageal segments. For this reason, the majority of pediatric surgeons perform gastrostomy tube before the latter primary repair. Early enteral feeding from gastrostomy supports the growth of stomach. Growth of the stomach allows the use of it for reverse gastric tube or gastric transposition purposes in the next stage. We did not use cross-sectional imag-

ing methods because of the high possibility of long gap in isolated atresia in this case. Contrast imaging in the Trendelenburg's position did not provide any clue about distal esophagus when our patient was 2 months old. Stomach and distal esophagus were expected to grow into some degree after opening gastrostomy with classical approach and feeding for 12 weeks through gastrostomy tube in our case. Then the second operation was planned.

One of the methods used in isolated esophageal atresia is upper pouch buginage defined by Howard and Myers. Primary anastomosis is performed after 6-12 weeks of advancing a weighted dilator orally up to upper pouch once or twice a day (7). In Hendren and Hale's approach, bullet shaped electromagnetic metals are placed both in the upper pouch and in the esophagus through the gastrostomy then fistulization is waited by advancement of these metals to each other (8). Method defined by Foker suggests making a thoracotomy first and place traction sutures on upper pouch and the lower esophageal pouch and apply traction in opposite directions to elongate the esophagus and perform primary anastomosis after 10-14 days when appropriate length is established (9).

4. CONCLUSION

In conclusion, one should kept in mind that respiratory distress and stridor may develop from the pressure of upper pouch on trachea in patients with isolated atresia. In addition, preoperative determination of the gap length with the help of cross-sectional imaging methods would be very useful in determining the approaches for the surgical treatment.

CONFLICT OF INTEREST: NONE DECLARED.

REFERENCES

1. Harmon CM, Coran AG. Congenital anomalies of the esophagus. In: O'Neill JA, Rowe MI, Grosfeld JL, et al (eds): *Pediatric Surgery* Vol 1 (ed 5.). St Louis, MO, Mosby-Year Book. 1998: 941-967.
2. Harmon CM, Coran AG. Congenital Anomalies of the Esophagus. In: JL. Grosfeld, EW. Fonkalsurd and AG. Coran (eds), *Pediatric Surgery* (Vol. 1). Philadelphia: Mosby Elsevier, 2006; 1051-1081.
3. Fitoz S, Atasoy C, Yagmurlu A. et al. Three-dimensional CT of congenital esophageal atresia and distal tracheoesophageal fistula in neonates: preliminary results. *Am J Roentgenol.* 2000; 175 (5): 1403-1407.
4. C B: Konjenital Özofagus Atrezisi ve Trakeo-özogageal Fistül In B. C (ed), *Bebek ve Çocukların Cerrahi ve Ürolojik Hastalıkları* (Vol. 1). Ankara: Palme Yayıncılık 2006: 311-352.
5. Solomon BD, Pineda-Alvarez DE, Raam MS, et al. Analysis of component findings in 79 patients diagnosed with VACTERL association. *Am J Med Genet A.* 2010; 152: 2236-2244.
6. Pandit SK, Rattan KN, Budhiraja S. Esophageal atresia with blocked distal tracheo-esophageal fistula. *Indian J Pediatr.* 1998; 65: 763-765.
7. Mahour GH, Woolley MM, Gwinn JL. Elongation of the upper pouch and delayed anatomic reconstruction in esophageal atresia. *J Pediatr Surg.* 1974; 9: 373-383.
8. Hendren WH, Hale JR. Esophageal atresia treated by electromagnetic bougienage and subsequent repair. *J Pediatr Surg.* 1976; 11: 713-722.
9. Foker JE, Linden BC, Boyle EM Jr, Marquardt C. Development of a true primary repair for the full spectrum of esophageal atresia. *Ann Surg.* 1997; 226: 533-541.