Persistent esophagotrachea: description of a case

M. ROGGINI, I. CARBONE*, S. BOSCO**, D.A. COZZI***, G. ANDREOLI*,
P. CAPOCACCIA

Radiology Department of Pediatric Clinic, *Department of Radiology, **Department of Experimental Medicine and Pathology, and ***Pediatric Surgery Unit
"La Sapienza" University - Rome (Italy)

Abstract. – Esophagotrachea is the most severe form of laryngo-tracheo-esophageal cleft. This congenital anomaly is due to the anomalous differentiation of the primitive cephalic gut into trachea and esophagus. We present a case of a new born female with a common tracheoesophageal canal up to the carina. Atresia ani, a vulvo-vestibular fistula, sacral ipoplasia and others associated anomalies were also present. The baby underwent surgery after a laringo-tracheoscopy and a barium study of the esophagus. The prognosis of this extremely rare malformation is generally poor and the baby died on the fifth day after surgery for a serious ipertensive pneumothorax.

Key Words: 
Esophagotrachea, Laringo-trachea-esophageal cleft, Skeletal malformation.

Introduction

Esophagotrachea is a rare and severe malformation of the gastrointestinal tract due to the anomalous differentiation of the primitive cephalic gut into the trachea and the esophagus.

This malformation occurs between the 21st and the 27th day of the gestational period, caused by the unsuccessful development of the tracheoesophageal septum. Its clinical presentation depends on the anatomical severity of the malformation and the possible associated anomalies.

Symptomatology includes: enhancing dyspnea, caugh and cyanosis episodes. During oral feedings symptoms worsen with a serious aspiration associated with prolonged crises of apnea.

This congenital anomaly, described in the following case report, is the most severe form of laryngotracheoesophageal cleft.

Case Report

We present the case of a 35-week premature female newborn weighting 1960 g. She arrived at our Department 3 hours after birth for respiratory distress and anal atresia with a vulvo-vestibular fistula.

The Roentgenographic studies of the chest and the abdomen showed scattered bilateral shadows due to aspiration, vascular markings, and an enlargement of the cardiac silhouette. Sacral hypoplasia and numerous butterfly vertebras were present. There was an increased intestinal meteorism with no air in the rectum.

The anal examination showed atresia ani with a vulvo-vestibular fistula.

The intubated baby’s breathing parameters were decreasing and failed to reach high ventilation values. Therefore a laryngo-tracheoscopy was carried out showing the presence of a common esophagotracheal canal due to the fusion of the anterior laryngo-tracheal wall with the posterior esophageal wall. A radiological barium study was performed under general anesthesia. The study showed a common tracheoesophageal canal up to the carina with the distal esophageal lumen originating under the carina (Figure 1). For these reasons
the baby underwent surgery. A fundoplicatio
by Nissen, a gastrostomy, and multiple polypsplenectomies were performed. On the 5th
day after surgery the baby died for serious hy-
pertensive pneumothorax. The autopsy con-
firmed the presence of a common esophago-
tracheal canal up to the carina (Figure 2).

Discussion

Persistent esophagotrachea has a simpto-
matology similar to that of esophageal atresia.
There is an increased salivation with, some-
times, accumulation of mucus in the hy-
popharynx and the mouth. Because of the ab-
sence of the laryngeal posterior wall, aphon ia
is also often present. A spiration syndromes
arise early and are more severe than those oc-
curring in patients with a tracheoesophageal
fistula. The chest X ray shows a diffuse air
trapping, vascular markings, and a few opaci-
ties within the lungs due to aspiration pneu-
monia. The intestinal meteorism is usually in-
creased, with a uniform dilation of intestinal
loops. The stomach, as in the tracheoes-
ophageal fistula, is dilated and the position-
ing of a naso-gastric tube is usually very easy.

When the suspicion of severe laryngotra-
cheoesophageal communications is raised,
the radiographic study should be done under
fluoroscopy and with a small quantity of bar-
um introduced by a hypopharyngeal tube.
The study will generally show a common
canal up to the carina, made by the anterior
laryngotracheal walls and by the posterior
wall of the esophagus. Generally, there are
no fistulas and it is not possible to separate
the shadow of the proximal esophagus from

---

Figure 1. Bronchogram and esophageal barium study. There are a few butterfly vertebrae in the dorsal spine tract.
The sacrum is dismorphic. The contrast medium opacifies a common esophagotrachea; the bronchial tree and the pe-
ripheral alveologram is visualized. The distal esophagus originates below the carina, a gastric balloon is seen in the
fundus.
the trachea filled with contrast. The bronchial tree opacity as well as the visualization of the esophagus all the way down to the stomach will appear. An endoscopic exam will easily show the laryngeal malformation and the entity of the possible associated tracheomalacia.

Esophagotrachea is a very uncommon disease and there is a few number cases described in the literature until now. The interest of this case is due to the absence almost complete of the tracheoesophageal septum.

The prognosis is generally poor and death will occur in the early days of life. Surgery would be successful when the tracheoesophageal communication is limited and when there are no other severe malformations.

References

1) Griscom NT. Persistent esophagotrachea; the most severe degree of laryngotracheo-oesophageal cleft. AJR 1966; 97: 211-215.


Acknowledgements

The authors thank Egildo Santarelli, pediatric X-ray technologist, for his help in performing the Radiological exam of this case.