Foregut Duplication Cyst Associated with Esophageal Atresia—One-Stage Neonatal Surgical Repair

R. I. Spataru · M. C. Popoiu · M. Ivanov

Received: 24 March 2014 / Accepted: 9 June 2014 / Published online: 18 June 2014
© Association of Surgeons of India 2014

Abstract Foregut duplication cysts can occur anywhere along the foregut-derived structures. Its association with esophageal atresia (EA) and tracheoesophageal fistula (TEF) is exceptionally rare, the majority of cases being diagnosed at an older age due to symptoms or complications. We present the third reported case in the literature, when diagnosis and treatment were done at the time of repair for EA-TEF.

Keywords Foregut duplication cyst · Esophageal atresia · Tracheoesophageal fistula

Introduction

Foregut duplications can occur anywhere along the embryological foregut-derived structures. Most lesions are cystic and found in the thorax or intraabdominal. Their association with esophageal atresia (EA) is extremely rare with only 16 cases reported so far [1–6]. In only two patients, the associated foregut cyst was removed at the time of repair of EA and tracheoesophageal fistula (TEF) [2, 6].

R. I. Spataru (✉) · M. Ivanov
Department of Pediatric Surgery, “Maria Skłodowska Curie”
Children’s Hospital, University of Medicine and Pharmacy “Carol
Davila”, Bucharest, Romania
e-mail: radu_spataru@yahoo.com

M. Ivanov
e-mail: mqmivanov@yahoo.com

M. C. Popoiu
Department of Pediatric Surgery, “Louis Turcanu” Children’s
Hospital, University of Medicine and Pharmacy “Victor Babes”,
Timisoara, Romania
e-mail: mcpopoiu@yahoo.com

Case Report

A 2,820 g female newborn delivered at 40 weeks of gestation was transferred to our hospital 4 h after birth, with suspected EA because of a failure to pass a nasogastric tube. The esophagography performed at admission established the diagnosis of EA, the upper esophageal pouch ending at the level of third thoracic vertebra. The presence of gas into the abdomen suggested the presence of a distal TEF. Preoperative additional workup for associated anomalies (including cardiac, abdominal, and transfontanelar ultrasonography) was negative.

A right extrapleural thoracotomy was performed in the same day. Near the TEF, the lower esophagus caliber was apparently greater than the upper pouch, with an asymmetrical positioning of the distal fistula. The gap between the two separated ends of the esophagus was approximately 2 cm. A well-defined 2 cm cyst-like structure with a clear fluid was identified close to the fistulous segment, in the muscular wall of the distal esophagus (Fig. 1). The cyst, well delineated and having no communication with the esophageal lumen, was removed. The mucosa and most of the muscular wall of the adjacent esophagus was preserved. An end-to-end single layer anastomosis between the esophageal segments was performed.

The histopathological examination demonstrated a foregut duplication cyst with a three-layer structure: a well-developed muscle wall with two layers of smooth muscle (circular and longitudinal) covered by a mucosa with respiratory epithelium.

The postoperative evolution was uneventful. The control esophagography at 6 months of age showed no stenosis at the anastomosis level.

Fig. 1 Line diagram with intraoperative findings. *PE* proximal esophagus; *DE* distal esophagus; *T* trachea; *TEF* tracheo-esophageal fistula; *ED* esophageal duplication

Discussion

Foregut duplication cysts are the most common mediastinal cysts, accounting for about 20 % of mediastinal masses. They are probably a result of aberrant development of the primitive foregut. They are epithelial-lined structures, defined by histologic features rather than by location. Foregut cysts may be bronchogenic, enterogenous (which include esophageal duplication), neurenteric, or mixed [7]. The treatment of choice of the foregut duplication is total resection, due to the risk of compression if the cyst is only aspirated, hemorrhage and anemia if gastric mucosa is left in place, recurrent pancreatitis, rupture or malignant degeneration [3, 6].

The association between congenital foregut duplications and EA is extremely rare, only a few cases being reported so far (Table 1). There are experimental studies suggest that there is a common pathogenesis for these two entities, related to the failure of the foregut to detach from the notochord at the normal time [8].

Reviewing the literature from 1956 to present, we found 17 cases (including ours) of EA associated with foregut duplication (Table 1). Kluth (1976) presented four cases of EA associated with esophageal duplication [1]. Since then, 12 more cases were reported [2–6]. Eight patients (47 %) had EA with distal TEF, five patients (29.5 %) had EA type V

Table 1 Reported cases of esophageal atresia associated with foregut duplications [1–6]

Authors and year	Type of EA Associated anomalies	Localization of the duplication	Pathological findings
1. Present case, 2014	AE, distal TEF	Intramural cyst on lower esophagus, close to TEF	Respiratory epithelium, two layers of smooth muscle
2. Knod et al. 2013	AE distal TEF Trisomy 21	Intramural foregut cyst on lower esophagus, close to TEF	Ciliated pseudostratified epithelium with portions of columnar and mucin producing epithelium, double layer of smooth muscle
3. Escobar Jr et al. 2012	EA distal TEF Vascular ring	Large congenital diverticulum of the distal esophagus	Complex bronchopulmonary foregut malformation (so-called “esophageal lung”) with focal ectopic pancreatic tissue
4. Inan et al. 2007	EA distal TEF	Esophageal duplication located on distal segment of the esophagus, communicating with the lumen of esophagus	Squamous epithelium with bundles of the smooth muscle in the wall
5. McNally et al. 2001	AE distal TEF	Bilocular abdomino-mediastinal	Mixed components of bronchogenic, esophageal duplication and pancreatic enterogenous cyst
6. Yamagiwa et al. 1998	Type V, according to Kluth	The blind ending of the proximal esophageal segment reached 1.5 cm below the tracheal carina. The fistula arising from the distal segment ends high on the trachea	A small mass at the distal end of the proximal esophagus, representing heterotopic pancreatic tissue
7. Snyder et al. 1996	TEF and esophageal web Tracheomalacia	Y-shaped duplication of the esophagus in the upper thorax and lower cervical region	Cystic duplication lined by squamous epithelium
8. Janik et al. 1988	EA and proximal TEF Cleft lip and palate	Noncommunicating esophageal duplication in the right posterior mediastinum	Double muscle layer, myenteric plexus, respiratory and squamous epithelium surrounded by mucous glands

Table 1 (continued)

Authors and year	Type of EA Associated anomalies	Localization of the duplication	Pathological findings
9. Santamaria et al. 1988	AE, distal TEF	Cystic duplication on distal esophagus	Heterotopic pseudostratified columnar epithelium
10. Narasimharao and Mitra 1987	AE, distal TEF	Cystic duplication on distal esophagus, 2 cm from the fistulous opening	Ciliated pseudostratified columnar epithelium with bundles of smooth muscles in the wall
11. Kirks and Filston 1981	AE proximal TEF	The cyst (4 cm) was attached by a fibrous stalk to the membranous portion of the trachea, just above the carina	Stratified columnar epithelium and smooth muscles tissue
12. Hemalatha et al. 1980	EA distal TEF	1. 10×4 cm cyst in lower posterior mediastinum and intimately involving the basal segments of the lower lobe of the right lung 2. tubular duplication along the entire greater curvature of the stomach	1. unavailable 2. multiple duplications containing pyloric-type gastric mucosa with aberrant pancreatic tissue, smooth muscle tissue
13. Hemalatha et al. 1980	Isolated EA	Large cyst in the upper part of the posterior mediastinum, without communication with the esophagus	Mixture of squamous and transitional ciliated epithelium, double layer of smooth muscle
14. Bienayme 1967	Type V, according with Kluth	Segmental thoracic duplication. The fistula arising from the distal segment ends high in the trachea	unavailable
15. Minnis et al. 1962	Type V, according with Kluth	“here is a muscular septum between the proximal and the distal segments of the esophagus. The fistula arising from the distal segment ends high on the trachea”	unavailable
16. Yahr et al. 1962	Type V, according with Kluth	“here is a fairly long overlapping of the proximal blind pouch and the distal esophagus. Along practically the whole segment of duplication, both segments share a common muscular wall”	unavailable
17. Wiersema 1956	Type V, according with Kluth	Same with Minnis	unavailable

according to Kluth classification [1, 3], two cases (11.7 %) had EA with proximal TEF [3, 4], one case (5.9 %) isolated EA [3], and one case (5.9 %) had esophageal web and distal TEF [3]. The foregut duplication was intrathoracic, excepting three cases: bilocular thoraco-abdominal cyst; along the greater gastric curvature; on abdominal esophagus.

In ten cases, the duplication was cystic. In three patients, there was a communication between the esophagus and the duplication. In five patients, the duplication contained pancreatic tissue. A 23.5 % of the cases presented different associated anomalies (trisomy 21, vascular ring, tracheomalacia, cleft lip and palate).

The diagnosis of foregut duplication is often delayed in cases associated with EA [6], because of its asymptomatic presentation, small-size, or its position out of the surgical exploration field.

We identified only two cases of EA with distal TEF, where the foregut duplication cyst, situated on the distal esophagus, was an intraoperative finding and resected in one stage together with EA-TEF repair (Narasimharao et al. [2], Knod et al. [6]). Our case is the third reported. Local anatomy allowed us to preserve the adjacent esophageal structure. Knod et al. [13]

had a similar therapeutic approach, but a short segmental resection was required secondary to ischemia of the adjacent esophageal wall, while Narasimharao resected the esophageal segment containing the cyst [2].

Conclusion

When dealing with this extremely rare association between foregut duplication cysts and EA-TEF, it is preferable to treat both anomalies in a one stage neonatal surgery. Whenever possible, preservation of the adjacent esophageal segment will improve the success rate of the primary anastomosis and lower the possibility of short- or long-term complications.

References

1. Kluth D (1976) Atlas of esophageal atresia. *J Pediatr Surg* 11:901–919. doi:10.1016/S0022-3468(76)80066-8

2. Narasimharao KL, Mitra SK (1987) Esophageal atresia associated with esophageal duplication cyst. *J Pediatr Surg* 22:984–985. doi:[10.1016/S0022-3468\(87\)80487-6](https://doi.org/10.1016/S0022-3468(87)80487-6)
3. Inan M, Basaran UN, Aksu B, Dereli M, Dortdogan Z (2007) Esophageal duplication associated with esophageal atresia and tracheoesophageal fistula in a child. *Int J Pediatr Otorhinolaryngol Extra* 2:202–205. doi:[10.1016/j.pedex.2007.05.010](https://doi.org/10.1016/j.pedex.2007.05.010)
4. Janik JS, Hernandez AM, Lilly JR (1988) Esophageal atresia with proximal pouch tracheoesophageal fistula and noncommunicating esophageal duplication. *Pediatr Surg Int* 3:70–73. doi:[10.1007/BF00177086](https://doi.org/10.1007/BF00177086)
5. Escobar MA Jr, Welke KF, Holland RM, Caty MG (2012) Esophageal atresia associated with a rare vascular ring and esophageal duplication diverticulum: a case report and review of the literature. *J Pediatr Surg* 47:1926–1929. doi:[10.1016/j.jpedsurg.2012.07.051](https://doi.org/10.1016/j.jpedsurg.2012.07.051)
6. Knod JL, Garrison AP, Frischer JS, Dickie B (2013) Foregut duplication cyst associated with esophageal atresia and tracheoesophageal fistula: a case report and literature review. *J Pediatr Surg* 48(5):E5–E7. doi:[10.1016/j.jpedsurg.2013.02.071](https://doi.org/10.1016/j.jpedsurg.2013.02.071)
7. Strollo DC, Rosado-de-Christenson ML, Jett JR (1997) Primary mediastinal tumors. Part II. Tumors of the middle and posterior mediastinum. *Chest* 112(5):1344–1357
8. Qi BQ, Beasley SW, Williams AK (2001) Evidence of a common pathogenesis for foregut duplications and esophageal atresia with tracheo-esophageal fistula. *Anat Rec* 264(1):93–100. doi:[10.1002/ar.1125](https://doi.org/10.1002/ar.1125)