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A rare variant case of pure esophageal atresia with an atretic segment

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ABSTRACT

Pure esophageal atresia is typically characterized by a long gap between the upper and lower pouches, with a gasless abdomen and no fistula. The association of pure esophageal atresia with an atretic segment is extremely rare. We report a rare variant case of pure esophageal atresia in which the two blind esophageal pouches were joined by an atretic segment. Excision of the atretic segment and primary anastomosis were performed successfully.

Pure esophageal atresia (PEA) is typically characterized by a long gap between the upper and lower pouches, with a gasless abdomen and no fistula [1]. PEA is found in about 5–7% of all cases of esophageal atresia (EA) [2]. A variety of anatomical subtypes have been reported in PEA [3]. The association of PEA with an atretic segment is extremely rare, and only a few cases have been reported [2,4,5]. We report a rare case of PEA corresponding to subtype II_3 EA in Kluth's atlas [3], with two blind esophageal pouches connected by an atretic segment.

1. Case report

A 2.8-kg male baby was born at 36 weeks of gestation by Cesarean section. Prenatal ultrasonography showed polyhydramnios and absence of a fetal gastric fluid bubble, suggesting EA of Gross type A. Apgar score was 8 at both 1 and 5 min. He presented with drooling of saliva and a flat abdomen, but no associated anomalies. In the neonatal intensive care unit, a nasogastric tube was not able to be passed into the stomach, and radiological examination showed a gasless abdomen. On the basis of these findings, EA of Gross type A was diagnosed. Temporary gastrostomy was performed and bronchoscopy revealed no evidence of tracheoesophageal fistula (TEF) on the first day of life. Contrast study via the gastrostomy suggested that the distance between the esophageal segments was at least two vertebral bodies (Fig. 1). The decision was made at the time to delay attempted primary anastomosis until after elongation of the upper pouch by manual bougienage. By 3 months old, both ends of the esophageal segments overlapped on radiography (Fig. 2). Mediastinal exploration was performed through a right extrapleural thoracotomy. The upper esophageal pouch appeared markedly enlarged. The blind ends of the esophagus were confirmed to be connected by an atretic segment about 20 mm long and 2 mm thick (Fig. 3). A feeding tube inserted by the anesthetist was not able to be passed through the beginning of this segment. The atretic segment was excised, the blind ends opened and primary end-to-end esophageal anastomosis was performed. On histological examination, the atretic segment

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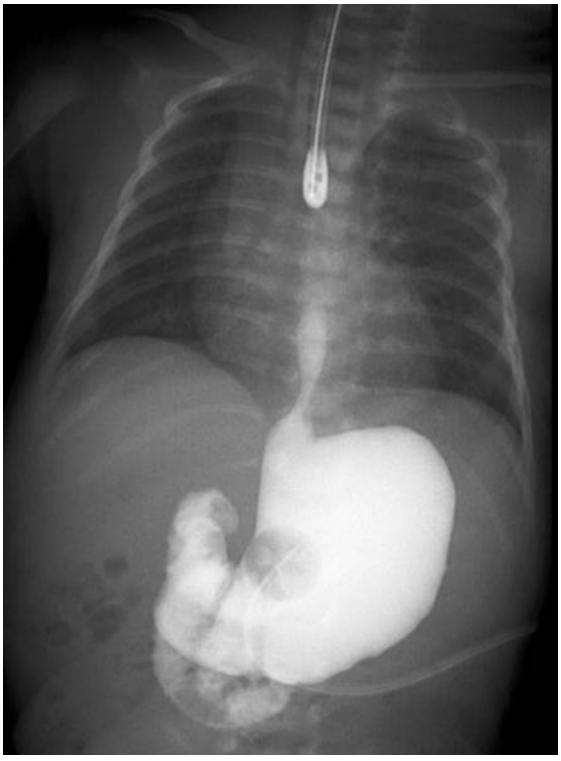

Fig. 1. Preoperative contrast study via gastrostomy demonstrates a gap of at least two vertebral bodies between the upper and lower esophageal pouches.

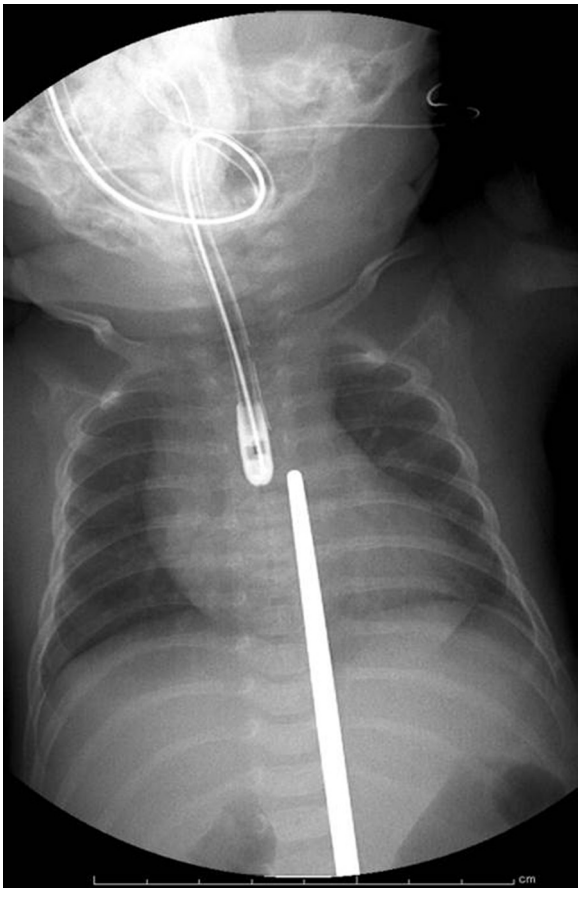

Fig. 2. Chest radiography demonstrates both ends of the esophagus overlapping at 3 months old.

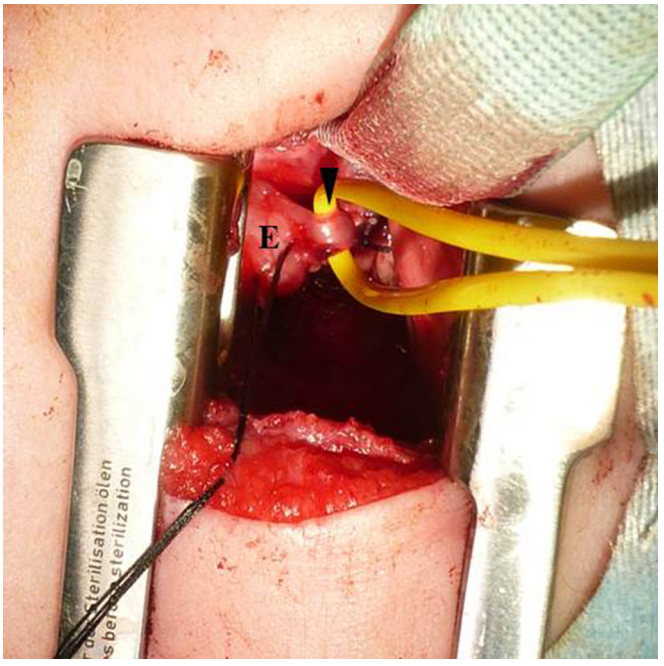

Fig. 3. Intraoperative photograph shows the upper esophageal pouch (E) and an atretic segment (arrow) that connects both esophageal pouches.

consisted of smooth muscle layers with a fibrous core and no lumen. These findings were consistent with subtype II_3 in Kluth's atlas of EA (Fig. 4) [3]. Although the postoperative course was uneventful, a contrast study demonstrated mild anastomotic stenosis and gastroesophageal reflux on postoperative day 10. The patient was maintained on antireflux medication after the surgery to decrease the risk of gastroesophageal reflux disease. Oral feeding progressed gradually from postoperative day 14. The patient was discharged on postoperative day 40, and as of 5 months old appears to be developing well.

2. Discussion

EA is characterized by interruption to the continuity of the esophagus with or without fistula to the trachea. The incidence of EA is generally 1 in approximately 2500–5000 live births [6]. The commonly used classifications are described by Vogt [7] and Gross [8]. A variety of anatomical subtypes of EA not covered by these

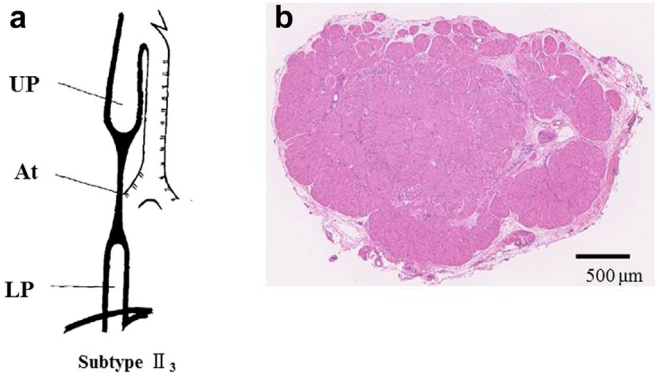

Fig. 4. Diagrammatic representation of the variant anatomy of type II_3 EA according to the Kluth atlas (a) and histologic examination of an atretic segment (b) showing smooth muscle layers with a fibrous core without any lumen. Our case was classified as this type of PEA. Hematoxylin and eosin staining. UP, upper esophageal pouch; At, atretic segment; LP, lower esophageal pouch.

Table 1
Summary of patients with variant subtype II_3 EA.

Case	Reported year	Author	Sex	Age at definitive surgery	Preoperative elongation	Treatment	Length of atretic band	Histological findings of atretic band	Postoperative course
1	1984	Wada et al.	F	5 mo	Mechanical elongation	Delayed primary anastomosis	NA	Smooth muscles without a lumen	NA
2	1999	Katsuno et al.	F	5 mo	NA	Delayed primary anastomosis	3.7 cm	Smooth muscles without a lumen	NA
3	1999	Katsuno et al.	M	3 mo	NA	Delayed primary anastomosis	2.5 cm	Smooth muscles without a lumen	NA
4	2007	Sanal et al. [2]	M	0 d	No treatment	Early primary anastomosis	1 cm	Smooth muscles, nonfunctional lumina and blood vessels	Good
5	2009	Dutta et al. [4]	M	2 d	No treatment	Early primary anastomosis	2.4 cm	Striated muscles groups without a lumen	Good
6	2010	Torikai et al.	NA	5 mo	Mechanical elongation	Delayed primary anastomosis	NA	NA	Good
7	2011	Yokoi et al.	NA	2 mo	No treatment	Delayed primary anastomosis	NA	NA	NA
8	2012	Nishi et al. [5]	M	3 mo	No treatment	Delayed primary anastomosis	2 cm	Smooth muscles and tracheal cartilage without a lumen	Good
9	2015	Present report	M	3 mo	Mechanical elongation	Delayed primary anastomosis	2 cm	Smooth muscles and nerve fibers without a lumen	Good

M, male; F, female; mo, months; d, days; NA, not available.

classifications have been reported. Kluth published a complete listing of all described variations of EA in 1976, including 10 separate classes and additional subclasses [3]. He placed the forms of PEA as type II. In this group of esophageal anomalies, he classified 5 subtypes. In our case, the upper and lower esophageal pouches were connected by an atretic segment without a lumen (Fig. 3). According to Kluth's classification, this type of PEA would be classified as subtype II_3, in which the midportion of the esophagus is atretic without a TEF. This type of PEA is extremely rare. We identified 8 other cases of Kluth type II_3 described in the literature, with the findings summarized in Table 1 [2,4,5]. Ages at definitive surgery ranged from 0 days to 5 months (mean, 2.8 months) and the length of the atretic segment ranged from 1.0 cm to 3.7 cm (mean, 2.2 cm). Six cases underwent delayed primary repair following initial gastrostomy, whereas two cases underwent curative surgery without gastrostomy shortly after birth. The most accepted treatment strategy for PEA is delayed primary repair following initial gastrostomy for feeding purposes, because of the long gap between pouches. In three cases, the upper esophageal pouch was able to be sufficiently elongated by manual bougienage preoperatively, so primary esophageal anastomosis was performed after excising the atretic segment. In all cases, esophageal anastomosis was successfully achieved, and the postoperative course was uneventful.

Various theories have been put forward, but the pathogenesis of PEA with an atretic segment are unclear. Mechanical hypotheses involving factors such as intraembryonic pressure, vascular occlusion, failure of recanalizaion, and disproportionate growth of the lateral folds do not appear sufficient to explain the whole spectrum of this anomaly [9]. Kluth and Habenicht [10] suggested that the pathogeneses of PEA and EA with TEF differ from those of the combined form of EA with fistula between the distal pouch and trachea. They demonstrated that PEA is probably due to partial necrosis of the esophagus caused by local microcirculatory disorders. According to the classification of intestinal atresia, this type of PEA with an atretic segment is similar to type II atresia of the small intestine, which involves two atretic blind ends connected by a band of fibrous cord [4,5]. The pathogenesis of this variant could be explained on the basis of the prenatal vascular accident theory proposed by Louw and Barnard for intestinal atresia [11]. In 1955, they demonstrated in experiments that interruption of blood flow caused intestinal atresia to various degrees in dog fetuses. Tsujimoto et al. [12] verified in an experiment that ischemic intestine causing a vascular obstruction became necrotic and was resorbed, with the wall comprising fibrous tissues and smooth muscle. As a result, we think that such a lesion becomes an atretic segment with the viable ends connected by a fibrous cord. However, only one case with tracheal cartilage in the atretic segment occurring in association with PEA has been described [5]. Such findings suggest that the present type of PEA may be caused by disturbance of the blood supply at an early stage in embryonic development and foregut malformation.

In conclusion, we have presented a rare variant of PEA with an atretic segment, corresponding to subtype II_3 EA in Kluth's atlas. In such cases, esophageal anastomosis may be performed successfully and lead to good prognosis.

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None.

Conflict of interest

The authors declare that they have no conflicts of interest.

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