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Case Report

Rare presentation of esophageal atresia with supra-esophageal atresia: a case report and management challenges

Wiwin Suhandri*, Muhammad Hericos

Department of Obstetrics and Gynecology, Faculty of Medicine, University of Riau/Arifin Achmad General Hospital, Pekanbaru, Indonesia

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***Correspondence:**
Wiwin Suhandri,
E-mail: wiwinsuhandri@lecturer.unri.ac.id

ABSTRACT

Esophageal atresia (EA), often regarded as 'the epitome of modern surgery', typically presents with tracheoesophageal fistula (TEF) and occurs in about 1 in 2400 to 1 in 3000 newborns, frequently accompanied by other congenital malformations. Rarely, EA is associated with congenital esophageal stenosis (CES), a condition first identified by Spitz. EA and TEF result from the incomplete development of the esophagus and trachea, with polyhydramnios in the third trimester being a potential indicator. This case study described a 43-year-old multiparous woman who, during an antenatal care examination, was found to have a fetus with abnormalities and excessive amniotic fluid, necessitating two amnioreduction procedures. Despite the interventions, the fetus exhibited signs of supra-esophageal atresia, leading to an elective cesarean section. The newborn, a girl with a birth weight of 3200 grams and an Apgar score of 5/6, could not be fitted with an orogastric tube, although karyotyping showed no chromosomal abnormalities. The discussion highlights the association of EA with various tracheobronchial anomalies and emphasizes the need for preserving the native esophagus in cases of CES. Gastroesophageal reflux disease (GERD) is common in EA patients, often requiring long-term follow-up and possible antireflux surgery. The case underscores the importance of diligent monitoring and intervention in high-risk pregnancies, with regular ANC examinations and accurate diagnoses being crucial for effective management.

Keywords: Esophageal atresia, Tracheoesophageal fistula, Congenital esophageal stenosis, Gastroesophageal reflux disease

INTRODUCTION

Esophageal atresia (EA) is often regarded as the epitome of modern surgery. In 1936, Thomas Lanman conducted the first primary extrapleural anastomosis for this condition, although the child survived only for 3 hours.[1,2] The first staged survivors were reported by Leven and Ladd in 1939, and Cameron Haight achieved the first successful primary anastomosis in 1941. EA, often accompanied by various types of tracheoesophageal fistula (TEF), occurs in approximately 1 in 2400 to 1 in 3000 newborns. Most infants with EA have additional congenital malformations, and it is rare for EA to be associated with another esophageal anomaly such as congenital esophageal stenosis (CES).[3] Spitz was the first to identify a congenital basis for distal esophageal stenosis associated with EA, showing tracheobronchial remnants in an esophagectomy specimen. EA and TEF arise from the incomplete separation or development of the esophagus and trachea. Polyhydramnios in the third trimester of pregnancy can be a sign of EA.[4]

CASE REPORT

A 43-year-old multiparous woman came to the Arifin Achmad Hospital for an ANC examination, then an

ultrasound examination was carried out, and abnormalities in the foetus were found, accompanied by a large amount of amniotic fluid coming out. Based on these findings, the patient underwent two amnioreduction procedures. The first amnioreduction procedure was carried out on 5 September 2023 by removing 1000 cc of amniotic fluid. The second amniotic reduction was carried out on 19 September 2023, by removing 2000 cc of amniotic fluid. The patient was then sent home with recommendations for routine examinations at the polyclinic until the gestational age reached term. On ultrasound examination, foetal intra-abdominal ascites were visible; the size of the unfilled stomach was 1.11 cm; the amnhiotic fluid measure MVP was 16.43 cm; and there was an impression of supra-esophageal atresia. It was decided to undergo an elective caesarean section, and a baby girl was born with a birth weight of 3200 grams and an Apgar score of 5/6. A carcaryotyping examination was carried out to detect abnormalities, but the results did not reveal any abnormalities in the chromosomes.

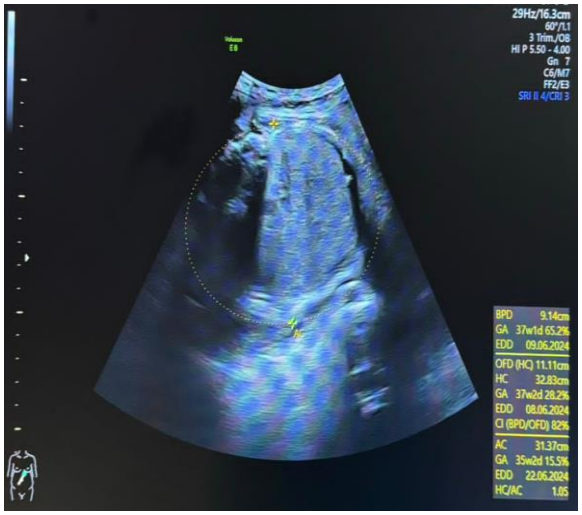

Figure 1: Ultrasound the foetus stomach appears empty.

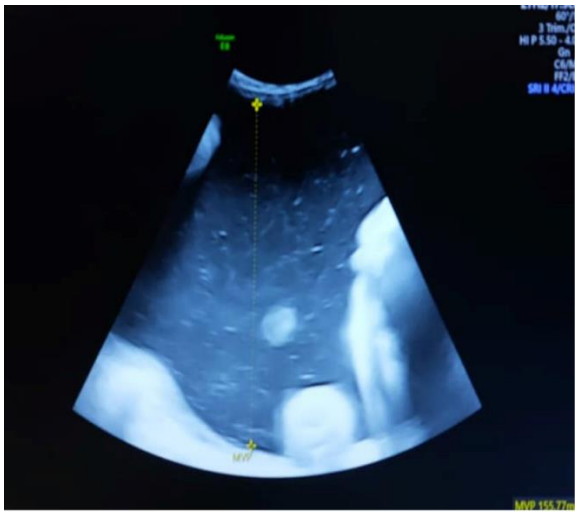

Figure 2: Ultrasound polyhydramnios found.

Figure 3: The baby after birth cannot be fitted with orogastric tube.

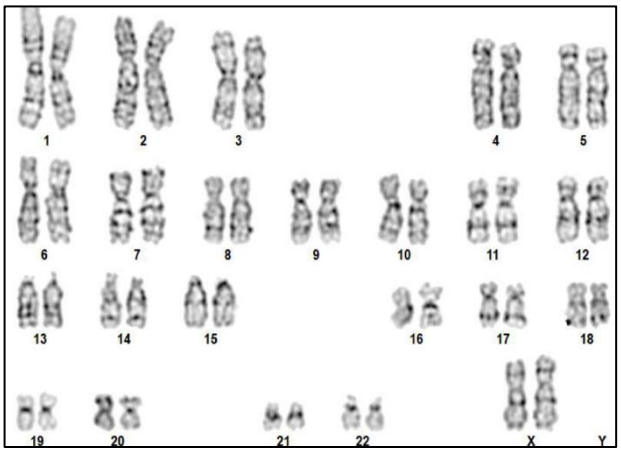

Figure 4: Caryotyping result.

DISCUSSION

EA is often accompanied by various tracheobronchial abnormalities, including tracheal agenesis, laryngeal atresia, congenital tracheal stenosis, and pulmonary agenesis. Other anomalies, such as ectopic bronchi, trifurcated trachea, and the absence of the right upper lobe bronchus, can also occur. The presence of a trifurcated trachea in EA patients supports current embryological theories.[5] The pathogenesis of EA stems from abnormal embryogenesis affecting the laryngo-tracheoesophageal groove and septum formation, resulting in a persistent connection between the trachea and esophagus. In cases where esophageal stenosis is also present, preserving the native oesophagus is advised, involving stenting, evaluation, and potential intervention as needed.[6,7] GERD is common in EA patients, often requiring long-term follow-up due to potential complications. Anti-reflux surgery, such as Nissen fundoplication, may be necessary, with some surgeons preferring partial wraps or modifications. This case emphasizes the rare association of EA-TEF with congenital esophageal stenosis and highlights the importance of managing GERD in EA patients.[8] Gastroesophageal reflux occurs in 30-82% of

patients after EA and TEF repair, with 30% of these patients needing an anti-reflux operation. Additionally, factors like maternal parity, age, and ethnicity can influence the risk of esophageal atresia in infants.[9,10]

CONCLUSION

This case underscores the importance of medical intervention and careful monitoring during high-risk pregnancies to ensure a successful outcome. Routine ANC examinations and a correct diagnosis can determine what actions to take next.

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