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CASE SERIES

Crossover Upper Pouch in Type C Esophageal Atresia: An Uncommon Variant Causing Diagnostic Dilemma

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ABSTRACT

Esophageal atresia with tracheoesophageal fistula (EA-TEF) is a well-known congenital anomaly and Type C variety of gross classification is the most common. Even for Type C variety, anatomy of upper pouch and lower pouch is not always the same. We are presenting three cases of Type C EA-TEF with unusual anatomy. In this type, upper pouch crosses over the lower pouch for a significant length. The cases are described here to highlight this variant of Type C EA-TEF which produces diagnostic dilemma. An early diagnosis of this variant, prevents morbidity and mortality.

Key words: Esophagus; Neonate; Pneumonia; Tracheoesophageal fistula

INTRODUCTION

Esophageal atresia with tracheoesophageal fistula (EA-TEF) is a common congenital anomaly with well-established investigation protocols. Gross classification is commonly used to classify EA-TEF and divide it into five types. The Type C variety (EA with distal TEF) is most common and accounts for approximately 86% of cases [1]. The diagnosis in these cases is straightforward with classical test to diagnose EA-TEF in a newborn being inability to pass orogastric tube with resistance at 10–12 cm [2]. Once the diagnosis is confirmed, surgical treatment of EA-TEF is considered as urgent [3]. If the diagnosis is delayed, many infants that present with EA-TEF exhibit pneumonitis due to late referral and it affects survival [4,5].

There are few scenarios where the passage of nasogastric tube is fallacious and gives a false impression of intact esophagus. We are presenting a rare variant of Type C, in which orogastric tube goes well beyond 18–20 cm and gives a false impression that the tube is in stomach and creates a diagnostic dilemma in emergency room. We present three such cases with the aim to enlighten the reader about such possibility so that timely diagnosis and management leads to a better outcome.

CASE SERIES

Three cases of newborn EA-TEF presented at day 2, 5, and 6 of life with excessive salivation and respiratory distress. The second and third babies were under treatment for pneumonia in another hospital and were being investigated. In all three children, the orogastric tube was negotiated well beyond the 12 cm mark and was seen near the diaphragm (Figure 1). These findings had given a false impression of intact esophagus and child continued to receive treatment for pneumonia. It was only after a contrast study, which indicated anatomical abnormality and then child was referred to pediatric surgeon. On admission with the surgical team, the patients underwent bronchoscopy and esophagoscopy. The bronchoscopy confirmed the presence of fistula and esophagoscopy showed a blind-ending pouch. All three patients underwent thoracotomy. Intraoperatively, there was a well-developed upper esophageal pouch which was reaching up to diaphragm (Figure 1). The lower pouch fistula was seen just above carina in all three patients. An end-to-end esophageal anastomosis was done after fistula ligation. There was significant discrepancy in the luminal diameter which was dealt with spatulation of lower pouch. Remaining management was as per our ICU protocol for EA-TEF. Out of three children, only one child (2 days old) survived and another two children died of septicemia.

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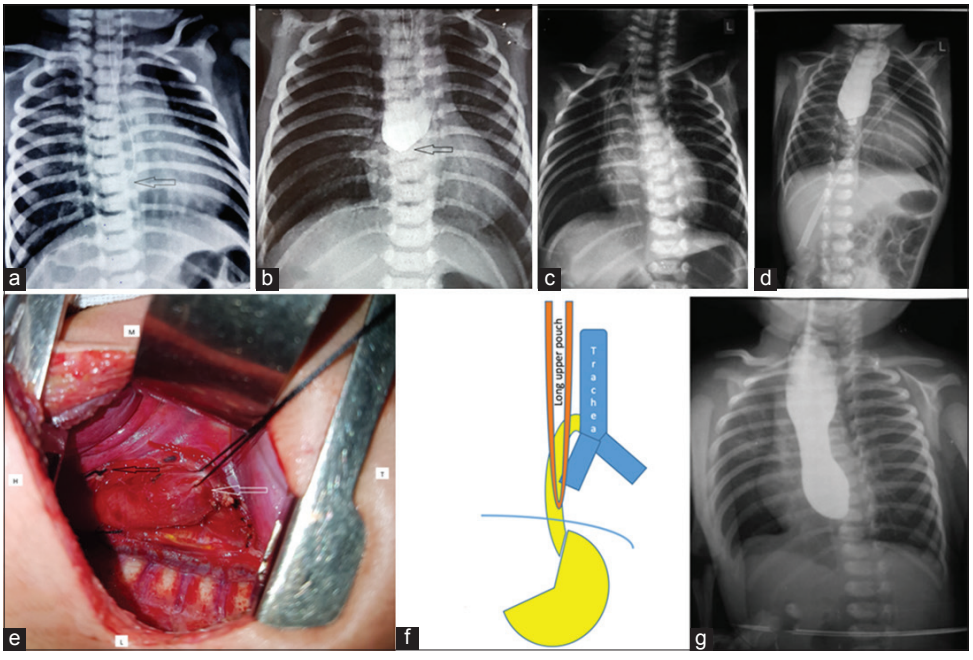

Figure 1: Composite figure of the three cases (a and c) showing the orogastric tube reaching well below T4 level. (b, d, g) Showing the upper pouch on a dye study. Note the pouch is wide and well developed, reaching up to diaphragm. (e) Upper pouch as seen intraoperatively. (f) Schematic diagram showing the anatomical arrangement

DISCUSSION

Diagnosis of EA-TEF is straight forward with failure to pass an orogastric tube, which usually gets stuck at 10–12 cm from the mouth and is usually visible at T3-T4 level on a plain X-ray [1]. There are some fallacies of this test, which one has to keep in mind. These are very extraordinary and rare situations. The tube can be accidently inserted in trachea and it can then travel to stomach through fistula giving a false impression of intact esophagus [6]. The second scenario is that of an upper esophageal pouch perforation where the tube exits through the perforation site and goes into mediastinum giving again a false impression of intact esophagus. The third scenario is of congenital esophageal stenosis, where orogastric tube can go beyond 12 cm. The former two situations will lead to delay in diagnosis. With these misleading findings, caregivers can allow feeding and worsen the condition.

The diagnostic confusion due to this type of situation often prompts the treating physician to do contrast esophagogram, as happened in our two cases. Contrast study shows the blind-ending pouch well below the T4 level and extends up to the level of diaphragm. Although this can clear the diagnosis, still, we do not recommend contrast study for the fear of aspiration/pneumonitis.

Whenever there is diagnostic dilemma even after orogastric tube test, we recheck it with red rubber catheter (stiff and wide) and still not clear then we proceed with esophagobronchoscopy. The esophagobronchoscopy will show the fistula and the blind-ending esophageal pouch and establish the diagnosis. This was followed in all three cases before they underwent thoracotomy.

Intraoperatively, problem in this type of crossed over esophageal pouch is that they have a very thin lower esophagus and handling the luminal discrepancy is tricky. This can be handled by lower esophagus spatulation.

In 1976, Dietrich Kluth described 95 types of EA and 20 subtypes in EA-TEF [7]. The variant which described here is the Type IIIb4 of Kluth's classification. Since Type C is the more common type in the gross classification, there are likely chances that one may come across this variant of Type C more frequently, especially in high volume centers.

To conclude, the most common variety of EA-TEF can present with unusual anatomical relationship which leads to diagnostic dilemma.

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