Case Report

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Combined oesophageal atresia with upper pouch fistula and meconium peritonitis

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ABSTRACT

Upper pouch tracheoesophageal fistula occurs is less than 1% of all oesophageal atresia variants. Meconium peritonitis is a rare neonatal condition with an incidence of 1:30 000 live births. In this case report, we describe the presentation, clinical findings and management of a patient diagnosed with an oesophageal atresia with upper pouch fistula as well as meconium peritonitis. To the best of our knowledge, this is the first case such as this described in published literature.

Key words: Oesophageal atresia, meconium peritonitis, tracheoesophageal fistula

INTRODUCTION

Combined oesophageal atresia with upper pouch fistula and meconium peritonitis: A case report discusses the presentation, clinical findings and management of a neonatal case which had a combination of an oesophageal atresia with upper pouch fistula and meconium peritonitis. A brief discussion on oesophageal atresia with upper pouch fistula and meconium peritonitis follows the case study. As far as we are aware, there have been no cases previously published with this combination of rare conditions.

CASE REPORT

Baby BA was born at 35 weeks of gestation via an uncomplicated elective caesarean section. An antenatal ultrasonographic diagnosis of oesophageal atresia had already been established. Good APGAR scores of nine and ten out of ten were recorded for the female infant at one and five minutes, respectively. Clinically, she was maintaining her airway, breathing spontaneously and did not require ventilation. On palpation, her abdomen was soft, not distended or peritonitic. The neurological, cardiovascular (including echocardiography) and genitourinary systems appeared within normal limits, and no other congenital malformations were noted.

After insertion of a replogle tube, an anteroposterior chest radiograph demonstrated the tip to be at the level of the second thoracic vertebrae [Figure 1] and an abdominal view revealed a gasless abdomen with multiple diffuse calcifications [Figure 2]. The former finding was diagnostic of either a pure oesophageal atresia or of an oesophageal atresia with an upper pouch fistula, whilst the latter confirmed meconium peritonitis secondary to an antenatal perforation.

The neonate was scheduled for an open insertion of a gastrostomy tube and at laparotomy, extensive adhesions with multiple areas of calcification were found in the peritoneal cavity, with no evidence of perforation or small bowel atresia. An extensive adhesiolysis was

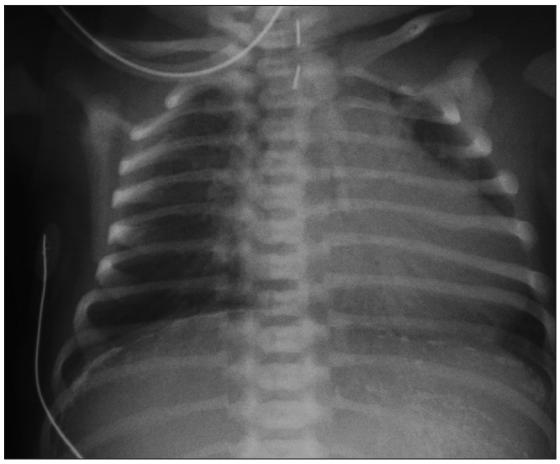


Figure 1: Supine chest radiograph on the first day of life shows tip of replogle tube in the upper oesophageal pouch and no distal bowel gas consistent with oesophageal atresia and no distal TOF. Curvilinear peritoneal calcifications are noted below the diaphragm

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necessary to adequately access the stomach.

Endoscopy of both the trachea and oesophagus at the time was suspicious for an upper pouch fistula on the posterior surface of the trachea; however, this was not visible from the oesophageal lumen and we were unable to cannulate the fistula at the time. A subsequent contrast study of the upper pouch also failed to demonstrate the fistula.

A gap-o-gram at ten days of age showed the upper and lower pouches of the oesophagus to be just over one vertebral body apart [Figure 3], and we thought that exploration with a primary anastomosis would be feasible.

Despite extensive mobilisation of the two oesophageal pouches, and not visualising an upper pouch fistula, the two ends could not be brought together for a primary anastomosis. It was decided to place Foker's sutures in an attempt to stimulate growth and allow a delayed primary repair.[1]

A chest X-ray after seven days of traction demonstrated a gap of less than 1 cm; however, exploration revealed that unfortunately, despite excellent growth of the lower pouch, the upper pouch had remained tethered high in the mediastinum, precluding any chance of anastomosis. At this point, we elected to follow a more traditional approach and fashioned a proximal oesophagostomy during which a cervical tracheoesophageal fistula (TOF) was identified. The fistula was divided and oversewn. The child is currently thriving and awaiting oesophageal replacement.

DISCUSSION

Oesophageal atresia is characterised by the absence of normal patency and continuity of the oesophagus. It is a common congenital abnormality with an incidence of 1 : 2 500 to 1 : 3 500 live births.[2,3] There are five described subtypes dependent on the presence of atresia and the presence and location of a fistula between the trachea and oesophagus. The incidence of associated anomalies is approximately 48%.[4]

The variety described, atresia with an upper pouch fistula is much less common, occurring in less than 1% of the total incidence of oesophageal atresias.[5] Unusually, the fistula in this case was in the proximal pouch, high in the neck, whilst its classical position is described as being near the distal end of the pouch.[6]

The aetiology of oesophageal atresia is poorly understood. The trachea, oesophagus and lung are foregut derivatives, and during the fourth week of embryonic life, the foregut divides into ventral respiratory and dorsal oesophageal components. Although the precise mechanism of separation is not clear, current consensus attributes the spectrum of anomalies of oesophageal atresia to have a multifactorial origin, including the involvement of genetic and environmental factors.[7] The majority of cases appear to be sporadic and non-syndromic.[3]

The management of oesophageal atresia where primary repair and anastomosis is not feasible remains problematic, with numerous alternatives described. The 'Foker technique'[1] provides an excellent alternative to waiting for differential growth, circular myotomy and flap procedures, or oesophageal replacement,



Figure 2: Supine abdominal radiograph shows a gasless abdomen with diffuse punctate and curvilinear calcifications along the peritoneal reflections of the abdomen consistent with meconium peritonitis

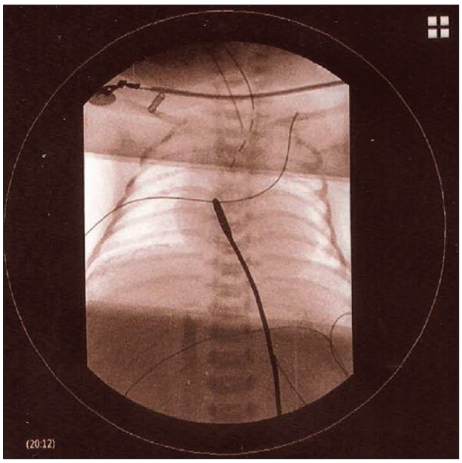


Figure 3: Gap-o-gram: Demonstrating the gap between the upper and lower pouch by using a replogle tube for the upper pouch and a Blake's dilator for the lower pouch

presumably on the basis that lengthening of the oesophageal segments is achieved by true growth and not just as a result of stretching of the respective pouches.[8] This hypothesis is yet to be proven.

In our case, the presence of the missed upper pouch fistula resulted in the tethering of the upper pouch, preventing the growth required to achieve a delayed primary anastomosis. This emphasises the importance of diagnosing and repairing the fistula prior to placing traction sutures.

Meconium peritonitis is defined as a non-bacterial, chemical peritonitis occurring during the intrauterine or early neonatal life secondary to an abnormal communication between the bowel lumen and peritoneal cavity.[9] It is a rare neonatal condition occurring in 1 : 30 000 live births.

There are four recognised clinical subtypes, including adhesive meconium peritonitis, giant cystic meconium peritonitis, meconium ascites and infected meconium peritonitis.[10] Each having different clinical and operative findings, these subtypes are dependent on the time of the antenatal perforation and whether the perforation seals or not.[11]

Decreased blood flow from any number of possible causes with its consequent intestinal ischaemia is thought to be the main cause for perforation.[12] Intestinal obstruction was associated with 53% of cases with meconium peritonitis, while 66% of those cases were due to intestinal atresia.[13] In keeping with other previously published reports, the cause or the site of the perforation causing the meconium peritonitis could not be determined in this case.[12,14,15] Once the meconium comes into contact with the peritoneum, it induces an intense inflammatory reaction and numerous intra-abdominal adhesions form as a consequence.[16] This reaction can be localised or involve the entire peritoneal cavity. The resultant abdominal calcifications are common radiographic and intraoperative findings.[17]

To summarise, oesophageal atresia with proximal pouch TOF and meconium peritonitis have separate aetiologies, none of which are completely understood. Due to the extremely low incidence of both of these conditions, it is most unusual to find them in combination. As far as we are aware, this is the first case describing this combination of rare conditions.

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