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Case Report
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Unusual Presentation Of Tracheoesophageal Fistula With Meconium Aspiration Syndrome In A Preterm Infant

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Abstract

Congenital malformations usually occur during organogenesis and result in complete or partial absence of an anatomical part or alteration in its normal configuration. Major structural anomalies occur in 2-3% of live births. The reported global incidence of tracheoesophageal fistula is roughly 1 in 2,500 live births and in Pakistan, incidence is only reported by those tertiary care centers which have paediatric surgery facilities available. We report a case of esophageal atresia (OA) with tracheoesophageal fistula (TEF) associated with meconium aspiration syndrome (MAS) in an infant. Reporting this anomaly highlights the importance of early diagnosis and thorough clinical examination of a newborn, signifying that a meticulous prenatal workup should be conducted. TEF/OA should be suspected in any newborn presenting with respiratory distress, especially cough, emesis and/or cyanosis during feeds, history of polyhydrominos and inability to pass nasogastric tube. The parents should also be counseled regarding future pregnancies as it carries a 1% risk of recurrence.

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INTRODUCTION

Thomas Gibson, is credited for describing the first case of oesophageal atresia (OA) and tracheoesophageal fistula (TEF) and Thomas Hill, noted the second case of OA with an associated anomaly of an imperforate anus. While some newborns may have TEF, about 4% of newborns with TEF do not have OA.1 the incidence of OA is 1 in 2500 live births and 86% of cases are associated with TEF.2 There is 0.5 to 2% risk of recurrence among siblings of affected child.3 However, in Pakistan, only tertiary care centers which have paediatric surgery facility report anomaly cases4.

Here we present a case TEF with OA, associated with meconium aspiration syndrome (MAS) in an infant. We aim to highlight the importance of conducting a comprehensive antenatal and prenatal workup in newborns to avoid management difficulties after birth and prevent an avoidable fatality. Also, to highlight an early diagnosis if a baby has respiratory distress and ther is failure to pass nasogastric tube and an X-ray which indicates a coiled tube in esophagus and intestinal air suggesting TEF.

CASE REPORT

A 2 day old female, weighing 2.3 kg, born at 34 weeks, presented in the emergency department with complaints of fever, drooling with breathing and feeding difficulties. She had been delivered in a private clinic.

Her mother was a primigravida with no significant prenatal history except hypertension in second trimester with use of antihypertensive drugs. There was no use of contraceptives pills or exposure to estrogen and progesterone during pregnancy. She had been vaccinated with tetanus toxoid. Mother made a total of three antenatal visits, with an ultrasound performed in last trimester showing that the baby had intra uterine growth retardation. She gave history of premature rupture of membranes 12 hours prior labor with Grade I meconium-stained liquor, leading to emergency C-section due to fetal distress. Baby had a weak cry at the time of birth and developed severe respiratory distress with peripheral and central cyanosis, with an Apgar score of 3 and 4 at 1 and 5 minutes respectively, hence she was admitted in the Neonatal Intensive Care Unit of a private hospital. The infant's condition did not improve after admission and she was referred to our center for ventilator support.

On examination, the baby was sick looking, lethargic, with poor reflexes. She had a respiratory rate of 82/min, heart rate of 180/min, a temperature of 100o C, was cyanosed with yellowish staining of nails, but no anemia or jaundice. No signs of dehydration were present and her lymph nodes were not palpable.. Chest examination revealed subcostal and intercostal recessions with bilateral crepitation. First and second heart sounds were audible with no murmur. Eye examination was normal, no chonal atresia, gross genital or ear anomalies; hence checked CHARGE syndrome (Coloboma, Heart anamoly, choanal Atresia,Retardation, Genital and Ear anomaly). On inspection, her abdomen was normal and on auscultation gut sounds were audible, but there was no anal opening. Samples were drawn for complete blood picture, which showed leukocytosis and arterial blood gases signifying metabolic acidosis.

After suctioning of oropharynx was performed we passed a nasogastric tube into the baby gently but beyond a certain length the tube couldn't pass. At the same time chest X-ray was done which showed hyperinflation of the lungs with patchy infiltrates in lower lung fields and coiling of nasogastric tube in the oropharynx. It was also seen while trying to pass urinary catheter that the child passed stool through vagina, hence a working diagnosis of tracheoesophageal fistula with imperforate anus with rectovaginal fistula and sepsis was made.

The child was placed on oxygen, given IV hydration and antibiotic therapy. As her condition improved, a barium esophagogram with contrast was performed to confirm the diagnosis which showed: distended stomach and proximal small bowel loops; nasogastric tube curling over itself at D4 level; and evidence of a blind ending proximal esophageal pouch that dilated with contrast medium. No evidence of air or leak was seen in the rectum. Upper oesophageal atresia associated with tracheoesophageal fistula between the trachea and lower oesophageal segment was hence considered in this patient.

Suddenly, during the procedure the baby collapsed; She was cyanosed, gasping for breath and was placed on the ventilator immediately. She required frequent suctioning onwards due to excessive secretions and a call was sent to another tertiary care center with paediatric surgery department but due to unavailability of ventilator support there; baby could not be shifted.

On the 10th day of life, the baby's condition deteriorated further despite all efforts. Unfortunately, she stopped breathing, her pupils became fixed and dilated and all efforts to resuscitate her failed.

This sad news was conveyed to the parents and they were counseled for future pregnancies regarding the risk of recurrence.

DISCUSSION

Congenital malformations usually occur during organogenesis resulting in complete or partial absence of an anatomical part or alteration of its normal configuration. Major structural anomalies occur in 2-3% of live births and an additional 2-3% are recognized by the age of 5.4

This case report presented has shown TEF with blind ending esophageal pouch with a fistula from trachea o distal esophagus.The pathogenesis is poorly understood; the trachea, oesophagus and lungs are foregut derivatives that divide into ventral respiratory and dorsal oesophageal components. Sometimes the oesophagus fails to separate from trachea leading to TEF.

Mastroiacovo et al5 found that out of 92 malformations studied, 39 malformations including TEF/ OA were more common in twins than in singletons and in males. Chromosomal anomalies like trisomy 18 and 21 also predispose to this condition. Even more recently, three genes associated with TEF/OA in humans have been identified.1 In our case, the child was a singleton, female and no other obvious dysmorphic features were seen. CHARGE and VACTERL were excluded. Karyotyping and any other chromosomal analysis couldn't be done due to lack of funding.

According to Waterston classification our case was type B and according Spitz classification she was falling into category II.6

The incidence of other anomalies associated with TEF/OA is reported to be 30-60%.7 Children with TEF have a higher incidence of pyloric stenosis 3..The most common anomaly is that of cardiovascular(11-49%) followed by genitourinary(24%), gastrointestinal (24%), musculoskeletal(13%)6. In our case, associated malformation was that of gastrointestinal.

Sparey et al8 reported that prenatal detection of TEF/OA was possible in 10% of the cases, which is very low compared to other congenital anomalies. The positive predictive value of both polyhydrominos and a small or absent stomach bubble has been reported to be only 56%, making prenatal diagnosis even more difficult,9 same was the scenario in our case: the mother had normal ultrasound scans. This call for more aggressive attempts to conduct prenatal exams and close follow ups of such a pregnancy with delivery in a specialist center.

In the postnatal period TEF/OA should be considered if newborn has pooling of saliva, feeding and breathing difficulties, and passes nasogastric tube with a pain; however chest x-ray to demonstrate the coiled tube in the esophagus is more confirmatory as was the case in our patient except that she also had Grade 1 MAS. Additional diagnostic modalities include upper pouch esophagogram (UPEG), tracheobronchoscopy and contrast esophagogram with fluoroscopic control. Even endoscopic procedures like bronchoscopy and oesophagoscopy can be used. However, esophagogram with contrast and barium studies offer the best visualization of TEF/OA.10

Surgical management includes Neonatal Intensive Care Unit and appropriate anesthesia, which is available in developed countries and has an almost 100% survival rate as compared to low-income settings that exist in countries like ours, where all discipline facilities including prompt surgical intervention, ventilator support and prevention of septicemia are lacking, resulting in a high morbidity and mortality ratio.4,11

The outcome is generally better for term babies 12 than preterm. The death in our case reported was most probably due to low birth weight, prematurity, septicemia and inadequate management modalities.

CONCLUSION

This case highlights the significance of conducting a comprehensive clinical examination in newborns; meticulous prenatal workup is also necessary to reduce fatalities. TEF/OA should be suspected in any newborn presenting with respiratory distress, persistent drooling, a history of polyhydrominos and inability to pass nasogastric tube. Ventilator support and other management modalities should be mandatory in a tertiary care setup. The parents should also be counseled about future pregnancy as it carries a 1% risk of recurrence.

Consent: Written informed consent was obtained from the parents of the baby for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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