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Research Paper

Congenital Esophageal Atresia with Tracheo Esophageal Fistula and its related review of literature.

Medical Science

KEYWORDS : Esophageal atresia, Tracheoesophageal fistula, Orogastric tube, Thoracotomy

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ABSTRACT

We report here, an interesting and rare case of a female neonate born spontaneously at 38 weeks of gestation, with Type H congenital esophageal atresia with trachea-esophageal fistula. Atresia is a condition in which a body orifice or passage in the body is abnormally closed or absent.In Esophageal Atresia the proximal and distal portions of the esophagus do not communicate.Tracheoesophageal fistula is an abnormal communication between the trachea and esophagus. Blockade of the esophagus occurs with an incidence of 1 in 3500 to 1 in 4000 live births.(C Shaw-Smith, 2006,J Med Genet43:545–554;Ashwin Ashok Jaiswal et al,doi:10.1016/j.ejenta.2013.12.007) Review of literature showed similar cases in only 4% patients. (C Shaw-Smith, 2006,J Med Genet43:545–554; Paulo Fernando Martins Pinheiro et al,2012,World J Gastroenterol 18(28): 3662–3672.)

INTRODUCTION:

Esophageal atresia and tracheo-oesophageal fistula (OA/TOF) are common life-threatening malformations having an incidence of approximately 1 in 3500 to 1 in 4000 births.(C Shaw-Smith,2006,;Ashwin Ashok Jaiswal et al, doi:10.1016/j.ejenta.2013.12.007).Approximately 92% of patients with OA have a tracheo-oesophageal fistula (TOF), and about 4% of patients with TOF do not have OA.

In oesophageal atresia there is congenital complete interruption of the esophageal lumen, and tracheoesophageal fistula is a congenital fistulous connection between esophagus and the trachea or main bronchus.

The early history of esophageal atresia was documented by Ashcraft and Holder in 1969 and extensively reviewed by Myers in 1986. The first recorded case was in 1670 by Durston. However,Thomas Gibson in 1697 , documented the first classical description of an esophageal atresia with a distal fistula.(Lewis Spitz,1996; Paulo Fernando Martins Pinheiro, 2012) The next case reported by Thomas Hill, came about 150 years later.(Lewis Spitz,1996)

More than 50% of newborns presenting with OA/TOF have other associated anomalies, many of them leading to considerable morbidity and mortality. A variety of congenital anomalies have been described in association with OA/TOF: a well-known acronym is the VACTERL association (vertebral, anorectal, cardiac, tracheo-esophageal,renal, limb abnormalities). Rarely, OA/TOF may be associated with the Holt–Oram syndrome, the DiGeorge syndrome, polysplenia, and the Pierre–Robin syndrome. (C Shaw-Smith, 2006; Paulo Fernando Martins Pinheiro et al,2012)

Early diagnosis, improved anesthesia and surgical technique, good ventilatory support, advanced intensive care management, early treatment of associated anomalies, responsiveness of post operative complications have influenced survival positively. With few exceptions, most infants with esophageal atresia and/or tracheoesophageal fistula survive in the current era. (Engum SA et al ,1995)

DEVELOPMENT:

Though in the recent times clinical and surgical management has shown significant improvement, still the knowledge of development of these anomalies is not clear.

The esophagus and trachea are derivatives of the primitive foregut.Around the 4th -5th week of intrauterine life the esophagus develops immediately caudal to the pharynx and the trachea develops ventral to it, from that part of respiratory diverticulum , that lies between the point of its bifurcation and the larynx. The partitioning of the trachea from the esophagus is by the tracheoesophageal septum. Esophageal Atresia results from deviation of the tracheoesophageal septum posteriorly; as a result , there is incomplete separation of the esophagus from the laryngotracheal tube and results in concurrent tracheo esophageal fistula.

CASE REPORT:

A female neonate having a birth weight of 2.3kgs was born to 36 year old $G_3P_2L_1$ women and was asymptomatic at the time of birth. The patient cried normally and had a APGAR score of 9 at one minute. After the first feed she vomited,coughed, became cyanosed and hypothermic .Detailed history taken from the mother revealed that mother was on antibiotics for upper respiratory tract infection in the second trimester of pregnancy. Amniotic fluid index was 16cms at 2/3rd gestational scan. There was a family history of consanguineous marriage, but the other siblings of the patient had no similar anomalies.

Chest radiogram taken in antero-posterior and lateral views showed air in the upper part of the esophagus and trachea was shifted anteriorly.(Fig 1) In tube esophagogram, the contrast was seen in the esophagus and the trachea with a fistula in between.In the contrast chest radiogram the dye was visible in the esophagus and in the trachea suggesting Type H Esophageal Atresia with tracheoesophageal fistula.(Fig 2) No other associated congenital anomaly was found.Oral feeds were stopped and right sided thoracotomy was done on 3rd day of birth and patient was under antibiotic coverage in the post operative period. Nasogasrtic feeding was started from the 2nd post operative day .Breast feed was started from 7th post op day.Post operative period was uneventful and contrast radiogram showed that the dye passed through the esophagus into the stomach and the fistula was closed.(Fig 3) Review check up after 4 weeks showed weight gain of 500 grams and no complaints of fever, respiratory problem or regurgitations.

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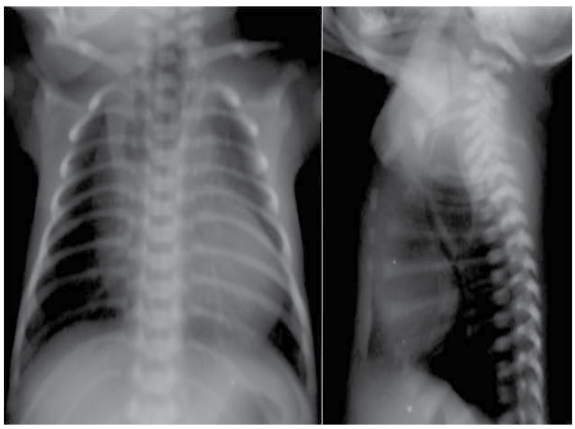

Fig 1: Plain Xray showing air in esophagus and trachea shifted anteriorly

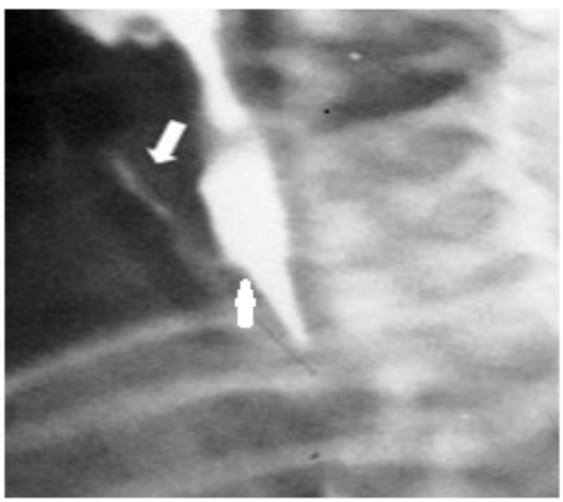

Fig 2: contrast xray showing dye in esophagus and trachea

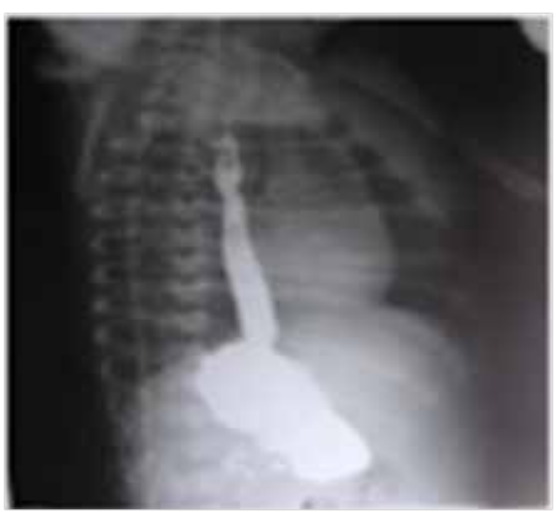

Fig 3: post op contrast xray showing dye passing into stomach uninterrupted.

DICUSSION:

Esophageal atresia with trachea esophageal fistula is a serious life threatening challenge to the pediatric surgeons. It has been anatomically classified into type A(7.8%) –eosphageal atresia without trachea esophageal fistula, type B(0.8-3%)- esophageal atresia with proximal trachea esophageal fistula, type C(85.8%)-esophageal atresia with distal fistula, type D(1-1.4%)- esopahageal atresia with proximal and distal fistula, type E/H(4.2%)-trachea esophageal fistula without esophageal atresia.(C Shaw-Smith,2006; Gross RE. Surgery of infancy and childhood. Philadelphia, PA: WB Saunders, 1953 ;Paulo Fernando Martins Pinheiro,2012)

Type H esophageal atresia with trachea esophageal fistula can miss an early diagnosis because the esophagus is patent, but atresias with a proximal fistula are diagnosed immediately after birth beacuase of typical clinical presentation. (Muhammad Riazulhaq and Elbagir Elhassan, 2012) The above patient was diagnosed after she showed symptoms after her first feed and was investigated promptly.

H-type osophageal atresia with trachea esophageal fistula is associated with other congenital anomalies in about 30% of cases, including VACTERL/VATER, CHARGE syndrome, Goldenhar's syndrome, esophageal stenosis, and syndactyly . Our case has none of these associations.(Muhammad Riazulhaq and Elbagir Elhassan,2012;Genty E et al, 1999; Paulo Fernando Martins Pinheiro et al,2012; Spitz L,2006)

Warren et al,1979, were the first to estimate offspring risk in patients with OA/TOF. They studied 79 patients who had undergone operations for the condition between 1947 and 1959. Fifteen of these patients had produced a total of 28 children. One of these children was affected. The sibling recurrence risk was similarly low, with one instance of recurrence in the 130 siblings of the 79 study patients.In the present case also the siblings of the patient were unaffected and were healthy.(Warren J,1979; C Shaw-Smith 2006)

The birth of a child with OA/TOF into a family without a previous history of the condition is associated with a low recurrence risk, of the order of 1%. The twin concordance rate for OA/ TOF is likewise low, at around 2.5%. These data do not indicate a major role for genetic factors in the pathogenesis of OA/TOF.(C shaw-Smith,2006;Harper PS,2004;Robert E et al,1993)However few studies indicate increased risk in twins.(Paulo Fernando Martins Pinheiro et al,2012) In the case reported here also there is no history of twining and no significant family history is seen, though the parents had consanguinous marriage.

There can be post operative complications like recurrence of fistula,stenosis and leakage of anastomosis, respiratory problems, thoracic wall deformities like scoliosis(L Agarwal et al,1999; Kovesi T and Rubin S. 2004; Spitz L,2006;Mortell AE and Azizkhan RG, 2009; Paulo Fernando Martins Pinheiro et al,2012)In our case the post operative period did not show any complications and the patient showed progressive improvement

In the case reported by Dwayne C. Clark et al , 1999,it is seen that there was a history intake of antibiotics(azithromycin) by mother during pregnancy.In the index case also the mother was on antibiotics during second trimester for treatment of upper respiratory tract infection. DWAYNE C. CLARK, LCDR, MC, USNR, Cherry Point Naval Hospital, Cherry Point, North Carolina *Am Fam Physician.* 1999 Feb 15;59(4):910-916. Esophageal Atresia and Tracheoesophageal Fistula

CONCLUSION:

Antenatal diagnosis of esophageal atresia with trachea esophageal fistula, by ultrasound examination is possible. It should be suspected if a newborn has difficulty in clearing saliva, repeated episodes of coughing and choking, or transient cyanosis shortly after birth.Infants may present with a sudden onset of respiratory distress when given an oral feed. Nasogastric tube cannot pass through easily because of distal resistance at the blind end

of the upper oesophageal pouch.Early diagnosis, effective clinical management and referral to tertiary health care centres in the absence of other congenital malformations can lead to good prognosis and survival rates.

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