ejpmr, 2022,9(8), 185-189

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Review Article

ISSN 2394-3211

EJPMR

A CASE OF ANAL ATRESIA, ESOPHAGEAL ATRESIA WITH TRACHEOESOPHAGEAL FISTULA (TOF) AND BICORNUATE UTERUS IN A NEONATE

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Article Received on 14/07/2022 **Article Revised on 04/08/2022** **Article Accepted on 24/08/2022**

ABSTRACT

Anorectal malformations(ARM) and Esophageal atresia with Tracheoesophageal fistula (TOF) are usually found congenital anomalies of VACTERL association that are seen in pediatric surgery, occurring in 1:2,000–1:5,000 and 1:3,000–1:5,000 live births, respectively. Bicornuate uterus, a Müllerian duct abnormality has a prevalence of 0.2-0.6 %. Association between VACTERL syndrome and genitourinary anomalies are frequent. We present a case of anal atresia, TOF and Bicornuate uterus. Patient had only 2 of the component from the VACTERL association failing to diagnose under VACTERL association. But still patient had two significant anomalies along with Bicornuate uterus. Surgical management was done for the TOF and anal atresia.

KEYWORDS: Anal atresia, Anorectal malformation, Bicornuate uterus, Esophageal atresia, Tracheoesophageal fistula, VACTERL.

INTRODUCTION

Anorectal malformations(ARM) and Esophageal atresia with Tracheoesophageal fistula (TOF) are usually found congenital anomalies of VACTERL association that are seen in pediatric surgery, occurring in 1:2,000–1:5,000 and 1:3,000–1:5,000 live births, respectively.[1,2] ARM with associated anomalies are found in 50 % of the patients.[3] Most frequent associated anomalies are[4]

1) Genitourinary anomalies (40-50%)
2) Cardiovascular (30-35%)
3) Spinal cord tethering (25-30%)
4) Gastrointestinal anomalies (5-10%)
5) VACTERL anomalies (4-9%)

Incomplete fusion of Müllerian ducts leads to formation of Bicornuate uterus. Uterus with two cavities, each containing uterine lining is formed when partial fusion of Müllerian ducts occur at proximal portion.[5]

We present a rare case of anal atresia and Esophageal atresia with Tracheoesophageal fistula (TOF) with Bicornuate uterus.

Case description

A 3 day old female neonate was referred to our hospital in view of difficulty in feeding and non-passage of stool after birth. Baby was delivered through vaginal delivery. Birth weight was 2609 gm. After the birth neonate had feeding difficulties and also child did not pass first stool. Suspicion of TE fistula and anal atresia were considered and patient was referred to our tertiary care center for further management. On arrival radiological investigations were done followed by immediate surgical management.

Imaging findings

On radiograph, Coiling of the nasogastric tube was noted at T2-T3 thoracic vertebrae level. Gas was noted in the stomach and abdomen. (Fig 1)

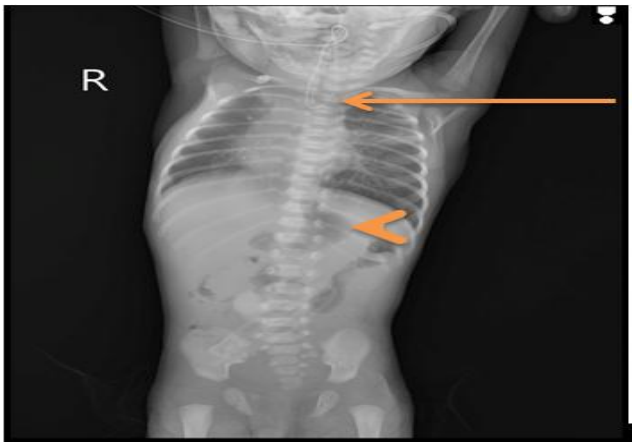

Fig. 1: Frontal supine x ray of baby, coiling of the nasogastric tube noted. (Arrow).Gas bubble noted in stomach.(arrow head).

On Invertogram, the gas shadow was noted 3.6 cm from the marker s/o high ARM. (Fig 2)

Fig. 2: Image 1 (Invertogram)-Showing distal air bubble(arrow) 3.85 cm from metallic marker(arrow head) placed at external anal opening indentation. Image 2-Showing external anal indentation but no opening.(arrow)

On abdominal Ultrasonography, biliary channel dilatation was noted in liver along with some echogenic material within. Gall bladder was over distended. (Fig 3)

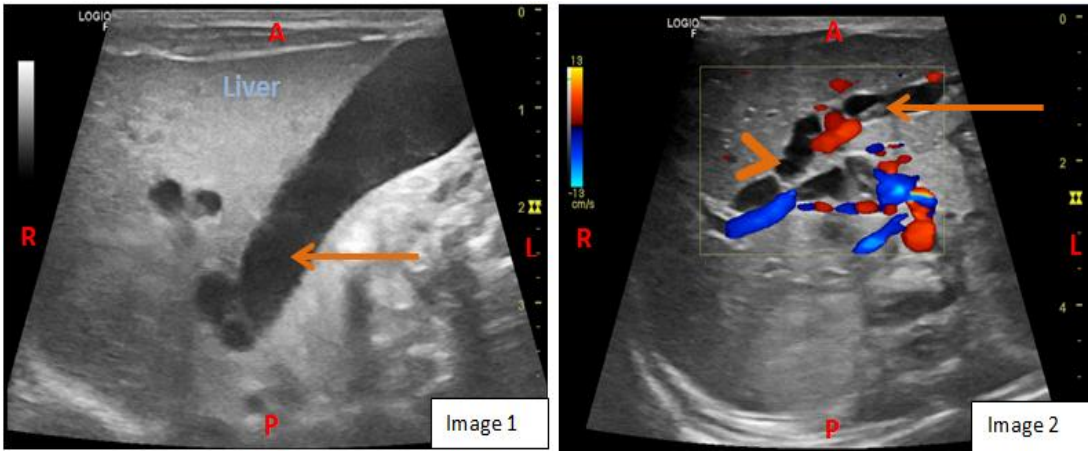

Fig. 3: [USG images on linear probe (8 khz)]:Image 1(Axial image of Liver and Gall bladder)-Overdistended Gall bladder(arrow).Image 2(Axial Colour doppler image of Liver) –Hepatic biliary radicles dilatation(arrowhead) with echogenic material within(arrow).{A-Anterior, L-Left ,P-Posteriro,R-Right}

CBD measured 2.8 mm in diameter. Bowel loops were filled with the content with average diameter of small bowel loop was 1 to 2.8 cm. On examination of pelvis two separate cornu of uterus were noted s/o Bicornuate uterus. (Fig 4) Colon was seen posterior to the uterus which showed distal tapering. (Fig 5)

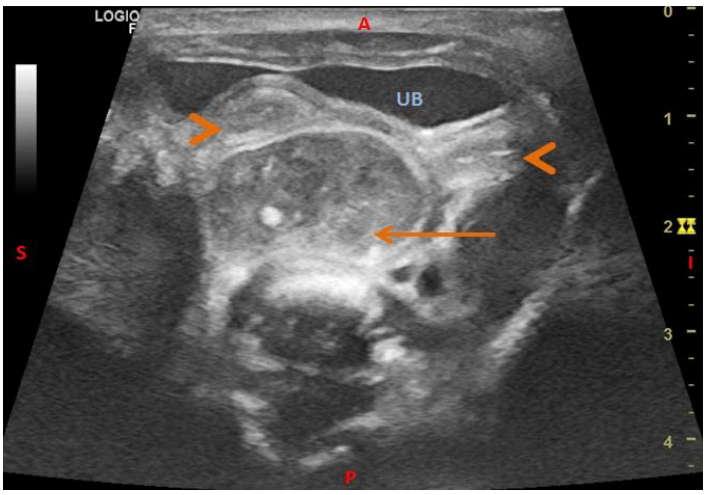

Fig. 4: [USG image on linear probe (8 kHz)]: Axial image at the level of pelvis showing, Two cornu of uterus (Arrowheads) and distended rectum(arrow),UB-Urinary bladder. {A-Anterior, I-Inferior,P-Posteriro,S-Superior}

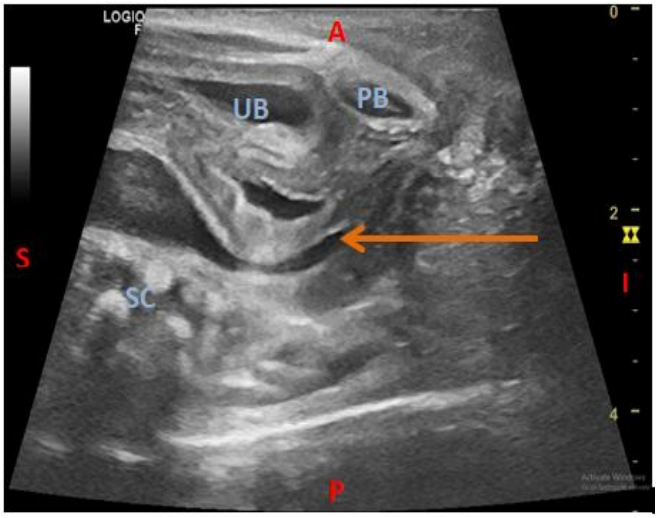

Fig. 5: [USG image on linear probe (8 kHz)]:Sagittal USG image at the pelvis showing distal end of the bowel and atretric segment further(Arrow).PB-Pubic bone,UB-Urinary bladder,SC-Sacrum{A-Anterior, I-Inferior,P-Posteriro,S-Superior}

Management

Patient was operated for Tracheoesophageal fistula and anal atresia. Type C tracheoesophageal fistula was noted on surgery. (Fig 6) Surgeon tried to reconnect the ends of the esophagus after resection of fistula, but was unsuccessful due to large gap between the two end. So oesophagostomy was done at the left cervical region for the proximal end of the esophagus. Anal atresia (approximately for 3-4 cm) was confirmed intraoperative. For Anal atresia, first stage operation was done. Colostomy was done in left iliac fossa.

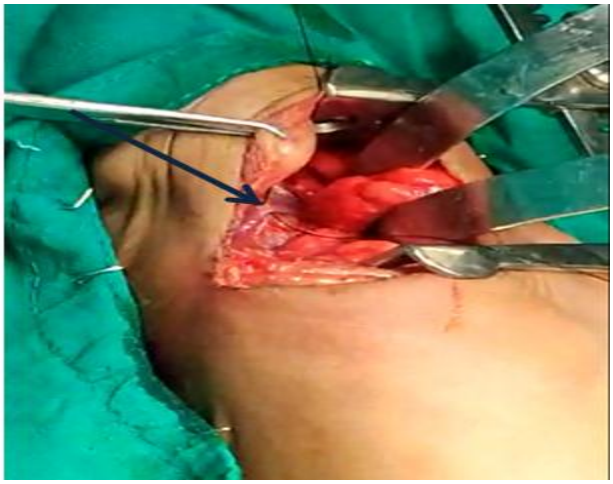

Fig. 6: Intraoperative image showing fistulous connection between Trachea and Oesophagus. (Arrow)

DISCUSSION

We presented a case of anal atresia and esophageal atresia with tracheoesophageal fistula (TOF type C) with bicornuate uterus.

ARM with associated anomalies are found in 50 % of the patients.[3] Most frequent associated anomalies are[4]

1) Genitourinary anomalies (40-50%)
2) Cardiovascular (30-35%)
3) Spinal cord tethering (25-30%)
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5) VACTERL anomalies (4-9%)

Congenital uterine anomalies are usually incidentally discovered on imaging. Prevelence of 5.5% was found for the congenital uterine anomalies in general population, out of which Bicornuate uterus accounted for 0.2 -0.6 % in a recent systematic review. Bicornuate uterus results because of the partial lateral fusion of the Müllerian duct at around 10th week of intrauterine development. Cause of the incomplete fusion is not known.[6]

Previously one case of bicornuate uterus with VACTERL association has been published in which patient had Congenital anal atresia with recto vestibular fistula, scoliosis, unilateral renal agenesis, and finger defect (VACTERL association) in a patient with partial bicornuate uterus and distal vaginal atresia.[7]

Also 2 cases of VACTERL association with Mayer-Rokitansky-Kuster-Hauser syndrome has been published by Mayer et al in which both patient had vertebral defect, anal atresia, cardiac defect, tracheoesophageal fistula/esophageal atresia, renal defect, and limb defect association.[8]

Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) syndrome (OMIM 277000) which is a congenital disorder of the Müllerian ducts,is characterized by agenesis or aplasia of the uterus and upper two-thirds of the vagina in females with normal karyotype.(46,XX). Females with MRKH have normal thelarche and pubarche (secondary sex characteristics). [9] It is seen in 1 in 5000 live female births.[10,11]

Our case was unique in which patient had Anal atresia and Esophageal atresia with Tracheoesophageal fistula (TOF) with Bicornuate uterus. Patient had only 2 components from the VACTERL association, failing to diagnose under the VACTERL association. But still patient had two significant anomalies along with Bicornuate uterus.

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