Content available at: https://www.ipinnovative.com/open-access-journals

IP Journal of Surgery and Allied Sciences

Journal homepage: https://www.jsas.co.in/

Case Report

A rare variant of tracheoesophageal fistula type -3 with opening at left bronchus

Aditya Prasad Padhy[1], Abdul Rehman Siddiqi[1], Swetamayee Dash[1,*]

[1] *Dept. of General Surgery, Kalinga Institute of Medical Sciences, Bhubaneshwar, Odisha, India*

ARTICLE INFO

Article history:
Received 06-12-2021
Accepted 18-12-2021
Available online 16-03-2022

Keywords:
Tracheo esophageal fistula
Nasogastric tube
Ppulmonary Infiltrates
Aspiration
Atresia

ABSTRACT

Esophageal atresia is an anomaly which is usually congenital but can be acquired in which there is disruption in the continuity of the oesophageal lumen resulting in an upper & lower segment. We present the case of a rare sub-type of Oesophageal atresia Type 3 with distal opening at left bronchus. This is the first case reported in literature in the Kalinga hospital Of Medical Sciences, Bhubaneswar. A 3 day old term male baby who presented as an emergency with difficulty breathing, fever and refusal to feed. Initially managed as early onset neonatal sepsis with aspiration pneumonia in which a diagnosis of oesophageal atresia was finally made.A high index of suspicion for Oesophageal atresia/trachea-oesophageal fistula should prevail when faced with a neonate with the triad: respiratory distress during feeds, regurgitation and persistent frothy salivation. The case discusses about a rare variant of Type 3 oesophageal fistula where the opening is present at the left bronchus.

This is an Open Access (OA) journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprint@ipinnovative.com

1. Introduction

Oesophageal atresia (OA) is a congenital malformation in which the oesophageal lumen is found to be interrupted, resulting in an upper and lower segment. While a vast majority of patients (92%) usually have a trachea-oesophageal fistula (TOF), about 4% of patients with TOF do not have OA.[1] This occurs in 1/2500 – 4500 live births.[1–3] The aetiology of oesophageal atresia is poorly understood.

2. Case Report

A 3 day term male baby was admitted in paediatric surgery with difficulty in breathing, fever and refusal to feed following which Nasogastric tube was inserted which coiled returning towards oral cavity and a repeat Chest and Abdominal X-ray with tube in place showed mild pulmonary infiltrates with no air in the stomach and duodenum respectively which was very suggestive of tracheo esophageal fistula. Later in order to confirm a CECT was done which revealed tracheo esophageal fistula H variant with narrow communication between distal esophagus with left mainstem bronchus at D5 level just beyond carina.

After analysing the findings the child underwent surgery. Post surgery child was transferred to NICU where he was kept for 5 days and extubated on POD 5. The baby was kept nil per oral for inital 6 days and was on iv fluids and antibiotics and on day 7 was shifted to oral feeds. Later a dye study was done which showed no leakage as well. The duration of stay in the hospital being 1 month.

The child was followed up and at the age of 3 years visited OPD with chief complaints of vomiting 12 episodes. Child was admitted for furthur work up and was advised for barium swallow which was suggestive oesophageal stricture. Advice from gastroentrology side was taken and an upper GI endoscopy was done which revealed foreign body obstruction which was eventually removed and was

* *Corresponding author*.
E-mail address: docadityapadhy@gmail.com (S. Dash).

https://doi.org/10.18231/j.jsas.2022.005
2582-6387/© 2022 Innovative Publication, All rights reserved.

advised for regular dialtation of the narrowing.

The child visited the OPD again at the age of 4 years with chief complaint of foreign body sensation following which I\UPPER GI ENDOSCOPY was done showing mild mid esophageal narrowing with no stricture. Child was shifted to liquid diet the next day and later onto solid diet.

3. Discussion

Oesophageal atresia (OA) is a congenital malformation in which the oesophageal lumen is found to be interrupted, resulting in an upper and lower segment. While a vast majority of patients (92%) usually have a trachea-oesophageal fistula (TOF), about 4% of patients with TOF do not have OA.[1] Incidence is 1/2500 – 4500 live births.[1–3] The aetiology of oesophageal atresia is poorly understood. It is suggested that there is an alteration in the migration of the lateral folds or growth arrest at the time of evagination. TOF mainly occurs between the third and sixth week of gestation.[4] Twin concordance rate for OA/TOF is about 2.5%.[1] The above information suggests that genetic factors play a minor role in the pathogenesis of OA/TOF, though chromosomal anomalies like trisomy 18 and 21 predispose to this condition.

Anatomically, there are five subtypes (Gross & Vogt classification) of Oesophageal atresia and this is based on their relative frequencies; Type A: OA with distal TOF (86%), Type B: Isolated OA (8%), Type C: Isolated TOF (4%), Type D: OA with proximal TOF (1%), Type E: OA with double TOF (1%). Our case constitutes a rare subtype whose relative frequency is ~1%.[1,2] Our diagnosis was however suggestive, due to the suctioning of maternal milk from the trachea and lung infiltrates found on chest x-ray. Several phenotypic variants have been identified; the VACTERL association (vertebral, anorectal, cardiac, trachea, esophageal, renal and limb abnormalities) has been described. Though rare, a number of syndromes have also been associated like Holt-Oram syndrome, the DiGeorge syndrome, polysplenia and the Pierre-Robin syndrome.[1]

The diagnosis of OA/TOF can be made in the antenatal period with the help of an Ultrasound scan which will show polyhydramnios and the proximal dilated blind ending oesophageal pouch.[5,6] In our case, this was missed as the mother had no ultrasound scan prenatally due to financial constraints. In the post natal period, OA and TOF should be suspected if a newborn is noted to have difficulty in clearing saliva, repeated episodes of coughing and choking (especially following feeds, as was the case in our patient), or transient cyanosis shortly after birth. Inability to pass a rigid nasogastric tube down the oesophagus can confirm the suspicion with a plain abdominal X-ray showing the chest to demonstrate the coiled tube in the oesophagus being more confirmatory. Additionally, the presence of air in the stomach and intestine is suggestive of the presence of TOF and its absence rules it out (isolated OA) and a dilated upper pouch is more suggestive of TOF. Upper pouch TOF occurs in less than 1% of cases and could easily be missed immediately after birth. In the presence of upper pouch esophagogram (UPEG) and tracheobronchoscopy, the diagnosis could be made. Thus recently, contrast Oesophagogram with fluoroscopic control and even endoscopic procedures like bronchoscopy and oesophagoscopy are being used, though the former must be done by an experienced radiologist and in a setting with adequate emergency neonatal resuscitation facilities due to the risk of aspiration pneumonia & lung injury from the contrast.[7] Barium offers best visualization as contrast but extraluminal barium can cause fibrous and granulomatous reactions leading to fibrous mediastinitis. Hyperosmolar agents are usually contraindicated as they could cause irritation and pulmonary oedema if aspirated. In diagnosis of the rare proximal fistuala, barium swallow may fail to demonstrate this anomaly but videofluoroscopic studies during cautious filling of the proximal pouch would visualize it.[8] Magnetic Resonance Imaging has very little role in diagnosis of EA and TOF but 3D CT scan has 100% sensitivity and specificity for oesophageal atresia and hence most reliable.[9] The diagnosis of this condition in developed world rarely exceeds 20 hours[10] as opposed to 4.4 days in our low-income setting.[11]

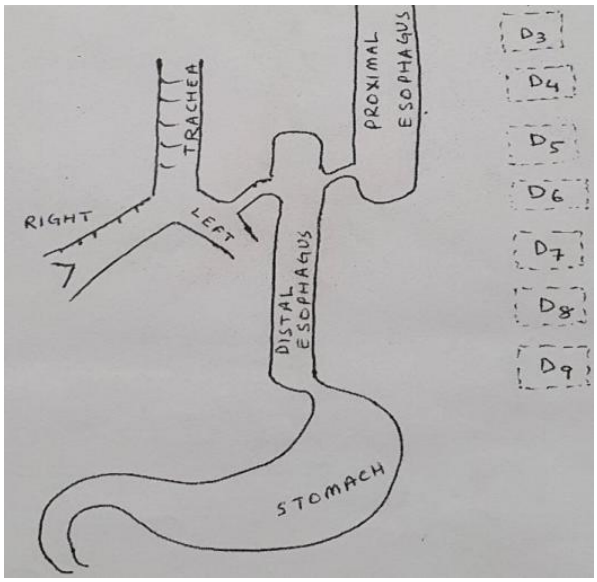

Fig. 1:

Surgical correction is adviced to be urgent as delay increases the risk of aspiration of saliva from the upper pouch or reflux of gastric acid through the lower pouch and a TOF causing pneumonitis. Cardiac ultrasonography is of prime importance to demonstrate presence of cardiovascular anomaly that could affect anaesthetic management or surgical approach as thoracotomy is usually performed opposite to the side of the aortic arch.[12] Surgical

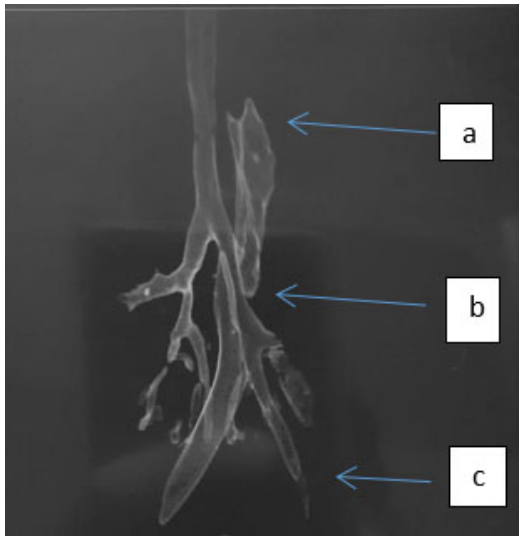

Fig. 2: a) Proximal esophagus eeeesophagus **b)** Left bronchus **c)** Distal esophagus

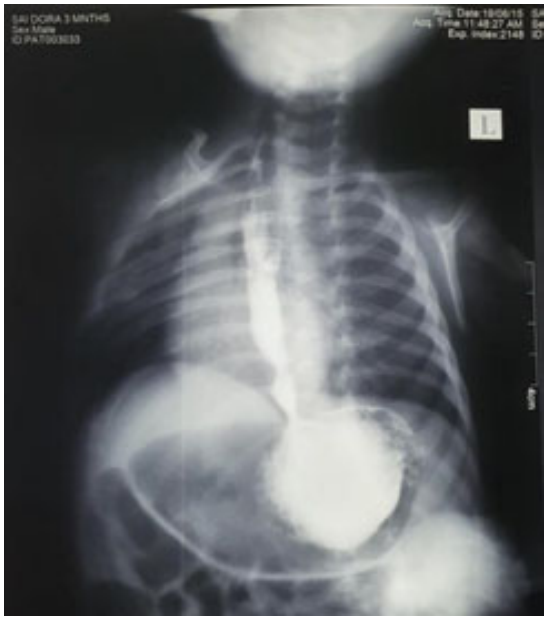

Fig. 3: Site of repair showingno leakage from the site of anastomosis following dye study

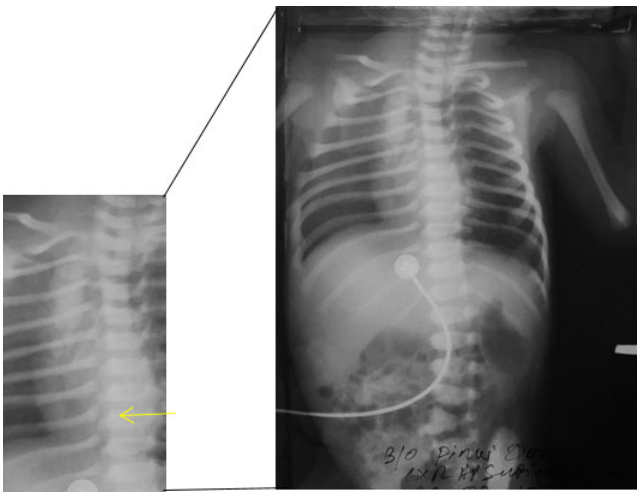

Fig. 4: Showing coiling of tube

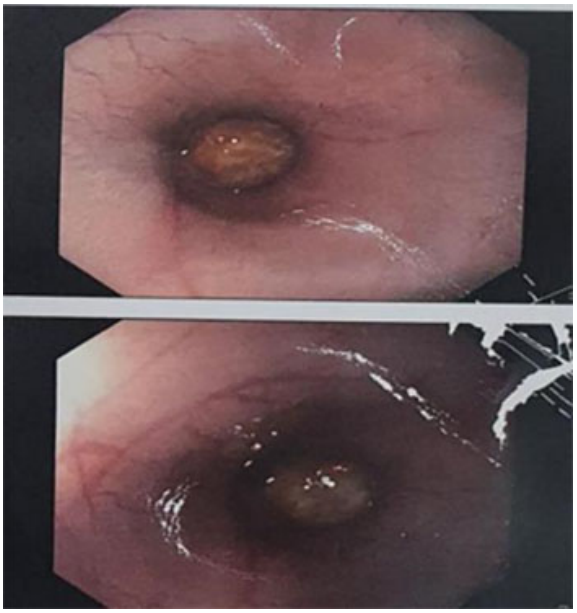

Fig. 5: Foreign body obstruction

management would involve a setting with an upto date Neonatal ICU with appropriate anaesthesia. These are readily available in the developed world giving current survival rates of almost 100%. This is not the case in low-income settings like ours, without all the above facilities, as morbidity and mortality rates are still high with some patients even dying before surgery.[11]

In the early post-op period, they range from tracheomalacia, repeated chest infections, anastomotic leak (11-21% of patients) and upto 50% developing oesophageal stricture, and pneumothorax from disruption. Gastro-oesophageal reflux (GOR) occurs in 35-58% of patients.[13] Late complications range from respiratory (46%) with 19% being recurrent pneumonia and 23% having repeated episodes of aspiration. It should be noted that these respiratory complications are secondary to GOR (74%), tracheomalacia (13%), recurrent TOF (13%), or oesophageal stricture (10%). Generally, recurrence of TOF occurs in about 9% of cases, typically 2-12 months after surgery and is more likely if there was excessive mobilization of the oesophagus during surgery, anastomotic leak and oesophageal stenosis.[14] Generally, the mortality rate for OA/TOF remains on the decline in the developed world currently <1.5% for patients without major cardiac

anomalies and with birth weight of >1500 g. The outcome is generally better for term babies than preterm. The fatality in the case reported was most probably due to delay in diagnosis, prematurity and inadequate management modalities.

4. Conclusion

This illustrates the importance of detailed clinical examination in newborns. Thorough prenatal work-up is also important as the Oesophageal atresia may have been diagnosed earlier by ultrasound scan. Oesophageal atresia with proximal TOF is rare, and its rarer association with MAS together with absence of up to date diagnostic and management facilities, most likely led to delay in diagnosis in our case. A high index of suspicion for TOF should exist when faced with a newborn with the triad: respiratory distress especially during feeds, regurgitation and persistent frothy salivation.

5. Source of Funding

None.

6. Conflict of Interest

The author declares that there is no conflict of interest.

References

1. Shaw-Smith C. Oesophageal atresia, trachea-oesophageal fistula, and the VACTERL association: review of genetics and epidemiology. *J Med Genet*. 2006;43(7):545–54. doi:10.1136/jmg.2005.038158.
2. Garcia H, Gutierrez MF. Multidiscplinary management of patients with Esophageal atresia. *Bol Med Hosp Infant Mex*. 2011;68(6):432–9.
3. Depaepe A, Dolk H, Lechat MF. The epidemiology of tracheooesophageal fistula and Oesophageal atresia in Europe: EUROCAT working group. *Arch Dis Child*. 1993;68(6):743–8. doi:10.1136/adc.68.6.743.
4. Moore KL, Persaud T. TVN: Embriologia Clinica. McGraw-Hill Interamericana; 2001. p. 288–9.
5. Vijayaraghavan BS. Antenatal diagnosis of esophageal atresia with tracheoesophageal fistula. *J Ultrasound Med*. 1996;15(5):417–9. doi:10.7863/jum.1996.15.5.417.
6. Pretorius DH, Drose JA, Dennis MA, Manchester DK, Manco JM. Tracheoesophageal fistula in utero. Twenty-two cases. *J Ultrasound Med*. 1987;6(9):509–13. doi:10.7863/jum.1987.6.9.509.
7. Riazulhaq M, Elhassan E. Early recognition of H-type tracheoesophageal fistula. *APSP J Case Rep*. 2012;3(1):4.
8. Houben CH, Curry JI. Current status of prenatal diagnosis, operative management and outcome of esophageal atresia/tracheo-esophageal fistula. *Prenat Diagn*. 2008;28(7):667–75. doi:10.1002/pd.1938.
9. Fitoz S, Atasoy C, Yagmurlu A. Three-dimensional CT of congenital esophageal atresia and distal tracheoesophageal fistula in neonates: preliminary results. *AJR Am J Roentgenol*. 2000;175(5):1403–7. doi:10.2214/ajr.175.5.1751403.
10. Tambo MF, Nonga BN, Andze OG, Chiabi A, Minkande JZ, Ngowe MN, et al. Problematique de la prise en charge de l'atresie de l'oesophge en pays sous medicalises. *Mali Med*. 2010;4:36–8.
11. Al-Rawi O, Booker PD. Oesophageal atresia and trachea-oesophageal fistula. *Contin Educ Anaesth Crit Care Pain*. 2007;7(1):15–9. doi:10.1093/bjaceaccp/mkl062.
12. Tsai JY, Berkery L, Wessen DE, Redo SF, Spigland NA. Esophageal atresia and tracheoesophageal fistula: surgical experience over two decades. *Ann Thorac Surg*. 1997;64:778–83. doi:10.1016/S0003-4975(97)00752-2.
13. Kovesi T, Rubin S. Longterm complications of congenital esophageal atresia and/or tracheoesophageal fistula. *Chest*. 2014;126(3):915–25. doi:10.1378/chest.126.3.915.
14. Lopez PJ, Keys C, Pierro A, Drake DP, Kiely EM, Curry JI, et al. Oesophageal atresia: improved outcome in high-risk groups. *J Pediatr Surg*. 2006;41:331–4. doi:10.1016/j.jpedsurg.2005.11.009.

Author biography

Aditya Prasad Padhy, Senior Resident

Abdul Rehman Siddiqi, Junior Resident

Swetamayee Dash, Junior Resident

Cite this article: Padhy AP, Siddiqi AR, Dash S. A rare variant of tracheoesophageal fistula type -3 with opening at left bronchus. *IP J Surg Allied Sci* 2022;4(1):25-28.