

Etiological Factors and Patterns of Clinical Presentation of Esophageal Fistulas

Devalkumar Manharbhai Patel¹, Gaurav Gattani², Jigneshkumar D. Damor³, Prakash Kumar Mansangbhai Chaudhary⁴

¹Assistant Professor, Department of General Surgery, Geetanjali Medical College, Udaipur, Rajasthan

²Assistant Professor, Department of General Surgery, Geetanjali Medical College, Udaipur, Rajasthan

³Assistant Professor, Department of General Surgery, Geetanjali Medical College, Udaipur, Rajasthan

⁴Senior Resident, Department of General Surgery, Geetanjali Medical College, Udaipur, Rajasthan

Received: 29-03-2023 / Revised: 28-04-2023 / Accepted: 30-05-2023

Corresponding author: Dr PrakashKumar Mansangbhai Chaudhary

Conflict of interest: Nil

Abstract:

Background and Aim: Esophageal Atresia (EA) with or without associated tracheo-esophageal fistula (TEF) is a common congenital anomaly occurring in 1/2500 to 1/4000 live births. This is life threatening if not managed early as it can lead to significant morbidity and mortality. Aim of the present study was to analyze the various etiological factors and patterns of clinical presentation of esophageal fistulas.

Material and Methods: The present study was done in the department of the surgery, medical college & associated hospital. All the patients who were diagnosed with esophageal fistula during the study period were included in the study. Each patient was closely monitored and detail clinical course was recorded. The etiology of the fistula, the investigations done and undergone by the patients and the treatment done for each of the patients were recorded. Each patient's natural course in the hospital was observed and analyzed.

Results: A total of 92 patients with the esophageal fistula were included in the study. The total included patients were divided into two groups: one with malignant etiologies and other one with nonmalignant etiologies. The carcinoma esophagus was the cause of malignant tracheoesophageal fistula in all the 62 cases. The ECOG score was used for the assessment of performance status of patients. The score was found to be good (1-2) in 24 patients. The poor performance was found in significant number of 38 patients. In the present study, there were 30 patients with non-malignant fistulas. The most common etiological factor was found to be foreign body ingestion. A majority of patients had the typical swallow-induced cough the so-called Ono's sign. Sixteen patients had complained of coughing of ingested food.

Conclusion: Esophageal fistulas are complex and heterogeneous disorders. The mode of clinical presentation varies depending on the etiology. With respect to malignant tracheoesophageal fistulas, stenting of the esophagus and/or the airway is the current standard of care.

Keywords: Carcinoma Esophagus, Esophageal Atresia, Tracheo-Esophageal Fistula, Foreign Body Ingestion.

This is an Open Access article that uses a funding model which does not charge readers or their institutions for access and distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>) and the Budapest Open Access Initiative (<http://www.budapestopenaccessinitiative.org/read>), which permit unrestricted use, distribution, and reproduction in any medium, provided original work is properly credited.

Introduction

Esophageal atresia with tracheo-esophageal fistula (EA with TEF) is a well-known congenital anomaly with an incidence of 1 in 2,400–4,500 live births. The abnormality was uniformly fatal throughout the world in first half of the twentieth century. Ever since Cameron Haight's first report of successful surgical correction in 1941, the survival of neonates with EA with TEF has dramatically improved. Now, the anomaly is regarded as an eminently correctable congenital lesion with survival rates more than 90%. [1-3]

Esophageal fistulas are a major cause of morbidity and mortality necessitating complex clinical evaluation and decision making for optimal management. It is best treated in a specialty tertiary care setting by a multidisciplinary team approach. Etiology of esophageal fistulas is multifactorial and the presentation can vary from simple external cervical esophageal fistulas to complex tracheoesophageal fistulas and esophago-pleurocutaneous fistulas.[4,5]

Esophageal Atresia (EA) with or without associated tracheo-esophageal fistula (TEF) is a common congenital anomaly occurring in 1/2500 to 1/4000 live births. This is life threatening if not managed early as it can lead to significant morbidity and mortality. Early diagnosis and referral to pediatric surgeon for surgical intervention is important in reducing morbidity and mortality and plays vital role in short term as well as long term prognosis.[6,7]

Outcome depends upon time of diagnosis, birth weight, respiratory complications, chromosomal abnormality, sepsis, socioeconomic status, prematurity, associated congenital anomalies and anastomotic leak.[6,8] Hence the aim of the present study was to analyze the

various etiological factors and patterns of clinical presentation of esophageal fistulas.

Materials and Methods

The present study was done in the department of the surgery, medical college & associated hospital. The ethical committee was informed about the study and the ethical clearance certificate was obtained prior to the start of the study. All the patients who were diagnosed with esophageal fistula during the study period were included in the study. The included patients were informed about the study and written informed consent was obtained prior to the inclusion in the study.

The detail history of the included patients was recorded during the study period. The demographic data of the included patients was recorded and documented. The proper recorded history was verified by the expert and recorded systemically in the tabulated form.

Each patient was closely monitored and detail clinical course was recorded. The etiology of the fistula, the investigations done and undergone by the patients and the treatment done for each of the patients were recorded. Each patient's natural course in the hospital was observed and analyzed.

In general, patients with malignant tracheoesophageal fistula were taken up for endoscopic esophageal stent placement. Before stent placement, the site of the tracheoesophageal fistula (TEF) was assessed by upper gastrointestinal endoscopy which is performed with topical lidocaine spray. A guide wire was inserted through the endoscope and under fluoroscopic guidance, it is passed distal to the tumor and the site of the fistula and a covered metallic stent was placed. In the event of unavailability of stents, they were offered other treatment. For patients with acquired nonmalignant fistulas, each patient was individually assessed and

treatment decisions taken by multidisciplinary team.

Results

Owing to the aim and objective of the research analysis; a total of 92 patients with the esophageal fistula were included in the study. The total included patients were divided into two groups: one with malignant etiologies and other one with non-malignant etiologies. Of the included total 92 patients; 62 patients had tracheoesophageal fistula due to malignancy and the other 30 patients were due to non-malignant etiologies. (Table 1) The carcinoma esophagus was the cause of malignant tracheoesophageal fistula in all the 62 cases.

The histopathological diagnosis of the entire included malignant fistula was of squamous cell carcinoma. None of the cases included in the study were diagnosed with bronchogenic carcinoma and adenocarcinoma. The ECOG score was used for the assessment of performance status of patients. The score was found to be good (1-2) in 24 patients. The poor performance was found in significant number of 38 patients. (Table 2)

All the patients were subjected to upper gastrointestinal endoscopy. The location of fistula was in the middle third of esophagus in almost all patients (44/62 patients). In the remaining 18 cases, the exact location of fistula was not made out as the growth was occupying the entire lumen not admitting the passage of endoscope. In patients in whom the fistula site was visualized, it was less than 1 cm in 24 patients and large (> 1cm) in 6

patients. The esophageal stenting with the success rate of 86% was seen in the patients. The dysphagia score improved from 3.4 to 1.8 after the stenting procedure. Majority of the patients had clinical relief of dysphagia. In the present study, there were 30 patients with non-malignant fistulas.

The most common etiological factor was found to be foreign body ingestion. A majority of patients had the typical swallow-induced cough the so-called Ono's sign. Sixteen patients had complained of coughing of ingested food. Eight patients had chest pain. Four patients had dyspnea and hemoptysis. Eight patients had reported history of foreign body ingestion and four patients had suicidal ingestion of corrosive liquid. There were three deaths among the non-malignant fistula group. One patient had tracheal dehiscence with respiratory failure. One patient with corrosive induced tracheoesophageal fistula had recurrent lower respiratory infection and succumbed to the same. Another patient died of mediastinal sepsis. The specific surgical procedure carried out was also a recorded. The tract was excised in three patients and sent for histopathology. The esophageal defect was closed in single or two layers using 3-0 or 4-0 polyglactin 910 (vicryl).

There were three deaths among the non-malignant fistula group. One patient had tracheal dehiscence with respiratory failure. One patient with corrosive induced tracheoesophageal fistula had recurrent lower respiratory infection and succumbed to the same. Another patient died of mediastinal sepsis.

Table 1: Etiology of Esophageal Fistula among study participants

Etiology of esophageal fistula	Number	Percentage (%)
Malignancy	62	67.39
Nonmalignant reason	30	32.60
Total	92	100

Table 2: Distribution according to ECOG Score

ECOG Score	Number	Percentage (%)
Good	24	38.70
Poor	38	61.29
Total	62	100

Discussion

Esophageal fistulas are complex clinical problems encountered predominately in tertiary care centers.[9] In our hospital, we encountered a total of 92 cases with airway-esophageal fistulas. Among them two-thirds were due to malignancy and one-third are acquired non-malignant causes. This is true even in Western literature as we can observe a majority of cases are due to malignancy. However, acquired causes are important treatable but clinically challenging situations requiring multidisciplinary team management.[10] Among the patients with malignant TEF, 59% presented with fistula at the time of primary diagnosis. However, 49% patients had fistulisation following radiotherapy or chemotherapy or both. It is established in the literature that chemo radiotherapy for cancer esophagus can result in TEF, especially in locally advanced cases with tracheobronchial tree invasion.[11]

The in-hospital mortality rate was 19% among malignant TEF patients with majority of death occurring due to pulmonary sepsis. All patients were followed up for a mean duration of 8 months, and among those patients who underwent stenting, sustained improvement in dysphagia and respiratory infection was observed.[12] In our series, 39% of cases were due to acquired non-malignant causes. The most common cause is foreign body ingestion (31%) closely followed by Boerhaave syndrome (20%). Other causes include postintubation TEF, corrosive e ingestion and post-surgical (16% each). Rarely did we encounter a patient with tuberculous TEF and another patient had a spontaneous TEF without any obvious etiology.

Surgery was done with adherence to all the principles of surgery of TEF as outlined by Grillo et al. In patients with postintubation TEF, the tract is dissected; disconnected and esophageal closure was done in two layers. As one of the patients had coexisting subglottic stenosis, resection of tracheal segment consisting two rings was done and end-to-end anastomosis was done with 3-0 vicryl in interrupted fashion.[13] The operative results in this group were satisfactory with all patients operated for Boerhaave's syndrome doing very well at the end of 1 year follow-up.[14,15] The four patients operated for postintubation TEF had good recovery and feeding orally, but one patient is still on tracheostomy. Two patient with retained foreign body- related bronchoesophageal fistula, after removal of the denture, esophageal and bronchial end was closed. The patient recovered and was discharged.

Conclusion

Esophageal fistulas are complex and heterogeneous disorders. The mode of clinical presentation varies depending on the etiology. With respect to malignant tracheoesophageal fistulas, stenting of the esophagus and/or the airway is the current standard of care.

References

1. Nagdeve, N.; Sukhdeve, M.; Thakre, T.; Morey, S. J. J. o. N. S. Esophageal atresia with tracheo-esophageal fistula presenting beyond 7 days. 2017; 6.
2. Nakayama, D. K. J. J. o. P. S. Vignettes from the history of pediatric surgery. 2020; 55: 1-37.
3. Nakayama, D. K. J. J. o. P. S. The history of surgery for esophageal atresia. 2020; 55: 1414-1419.

4. Thakkar, H.; Hewitt, R.; Cross, K.; Hannon, E.; De Bie, F.; Blackburn, S.; Eaton, S.; McLaren, C.; Roebuck, D.; Elliott, M. J. P. S. I. The multi-disciplinary management of complex congenital and acquired tracheo-oesophageal fistulae. 2019; 35: 97-105.
5. Salik, I.; Paul, M.: Tracheoesophageal Fistula. In StatPearls [Internet]; StatPearls Publishing, 2021.
6. Nisar, M. U.; Khan, N. A.; Javed, N.; Sikander, S.; Chaudhry, M. A.; Chaudhry, A. R. J. P. J. o. M. R. Esophageal Atresia: Management and Outcome in Resource Limited Settings. 2019; 58: 159-164.
7. Cross, K.; Smith, J.; Walker, I. A. J. N. a. Thoracoabdominal and general surgery. 2015; 225-269.
8. Gupta, D. K.; Sharma, S. In Tilte2008; Elsevier.
9. Madhusudhanan, J. Study of Presentation, Management and Outcome of Esophageal Fistulas. Madras Medical College, Chennai, 2013.
10. Galle, P. R.; Forner, A.; Llovet, J. M.; Mazzaferro, V.; Piscaglia, F.; Raoul, J.-L.; Schirmacher, P.; Vilgrain, V. J. J. o. h. EASL clinical practice guidelines: management of hepatocellular carcinoma. 2018; 69: 182-236.
11. Wang, H.; Zhang, N.; Li, D.; Zou, H.; Zhang, J.; Luo, L.; Ma, H.; Tao, M.; Zhou, Y. J. T. A. J. o. t. M. S. Airway covered metallic stent based on different fistula location and size in malignant tracheoesophageal fistula. 2015; 350: 364-368.
12. Davari, H. A.; Hosseinpour, M.; Nasiri, G. M.; Kiani, G. J. J. o. r. i. m. s. t. o. j. o. I. U. o. M. S. Mortality in esophageal atresia: assessment of probable risk factors (10 years' experience). 2012; 17: 540.
13. Mathisen, D. J. J. C. p. i. s. Surgery of the trachea. 1998; 35: 453-542.
14. Shen, G.; Chai, Y.; Zhang, G.-F. J. W. J. o. G. W. Successful surgical strategy in a late case of Boerhaave's syndrome. 2014; 20: 12696.
15. Han, D.; Huang, Z.; Xiang, J.; Li, H.; Hang, J. J. B. r. i. The role of operation in the treatment of Boerhaave's syndrome, 2018.