Type D Tracheoesophageal Fistula : Double the Trouble

Zulhisyam Z [1], Zubaidi SA[2], Tuan Mas Ain[1], , Faizatul AD[1] , Roshafiza JH[1], M.N. Tarmizi[3] , Zakaria Zahari[4] , Z. Habibullah[5] , Nik Khairani Nik Mohd[6]

[1] Pediatric Surgery Unit, Department of Surgery, Hospital Sultanah Nur Zahirah, Terengganu, Malaysia

[2] Paediatric Surgery Unit, Department of Surgery, University of Malaya Medical Centre (UMMC), Malaysia

[3] Department of Paediatric Surgery, Hospital Raja Perempuan Zainab II, Kelantan, Malaysia

[4] Department of Paediatric Surgery, Hospital Tunku Azizah, Kuala Lumpur, Malaysia

[5] Department of Anaesthesiology, Hospital Sultanah Nur Zahirah, Terengganu , Malaysia

[6] Department of Otorhinolaryngology, Hospital Sultanah Nur Zahirah, Terengganu, Malaysia

Introduction

Oesophageal atresia with tracheoesophageal fistula (EA/TOF) is a rare disease that occurs in 1/3,000 live births. Various types of EA/TOF and the incidence of each of them have been described in details by Filston et al. Amongst them, type D is one of the least common with incidence less than 1% [1]. However, this incidence is underestimated [2]. The actual incidence of upper pouch fistulas is rather higher than previously reported, in view of some of them are missed upper pouch fistula which mistakenly diagnosed with recurrent TOF [3]. It is very important to diagnose the upper pouch fistulas as early as possible to prevent morbidities and even mortalities to the patient as the consequences of persistent TOF mainly due to recurrent aspiration pneumonia and respiratory failure. We report a case of missed type D TOF at initial presentation and the detection of upper fistula was detected post operatively by bronchoscopy and upper contrast study.

Case Summary

A term newborn female, weight 2.29kg diagnosed with type C TOF. She presented with copious amount of secretion and coiling of ryles tube in the upper oesophageal pouch. The usual right thoracotomy, fistula ligation, and primary anastomosis performed. Postoperatively, she had persistent left lung collapse which requires high setting ventilation. Thus decided for bronchoscopy which revealed proximal fistula 2cm below the vocal cord. Gastrostomy creation was done in the same setting. Findings further supported with upper gastrointestinal contrast study. Patient then underwent delayed proximal oesophageal fistula ligation via cervical approach. She had an uneventful recovery after the surgery.

Discussion

EA represents a spectrum of anomalies with a fistula to the respiratory system present in the vast majority of cases. Numerous variants of EA/TOF have been thoroughly described [4], wherein the vast majority of cases are of type C. Fistulas between the upper oesophageal segment and the trachea have been infrequently recognized and missed.

There are believe that contrast studies and bronchoscopy unable to identify the fistulas however will be found during dissection of the proximal oesophageal pouch anyhow [4]. Nevertheless, there are reports showed misdiagnosis of the disease even during the operative procedure as reported in this case and was suspected recurrent post operatively instead of missed congenital upper pouch fistula [3].

The importance of preoperative bronchoscopy had been reported as it can confirm the diagnosis, delineate the exact site of the fistula and also report associated upper airway anatomical abnormalities [5,6]. It can be done at the same setting with the thoracotomy as preoperative assessment in guiding the surgeon regarding the anatomy and site of fistula.

In our local setting, bronchoscopy was reserved for pure OA (type A). In order to prevent missed fistula, routine preoperative bronchoscopy should be performed regardless the type of TOF.

Conclusion

Type D TOF was commonly missed and under reported. Therefore we propose routine preoperative bronchoscopy in all TOF patients to prevent catastrophic complications.

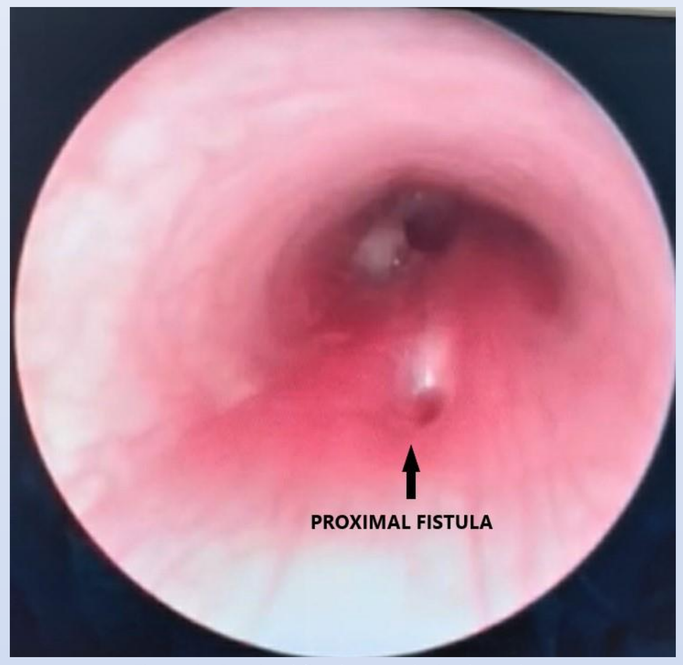

Figure 1 : Proximal fistula seen during bronchoscopy

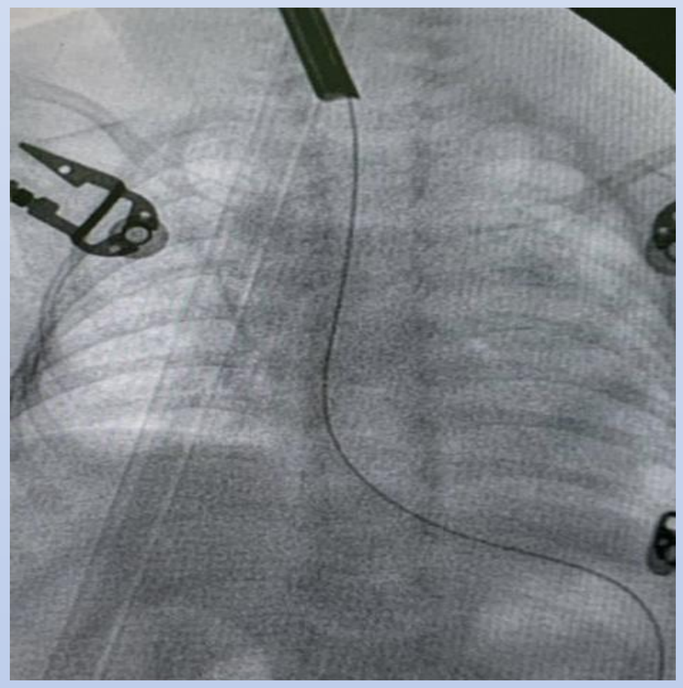

Figure 2: Fogarty catheter size 2fr was introduce during bronchoscopy via proximal fistula into oesophagus

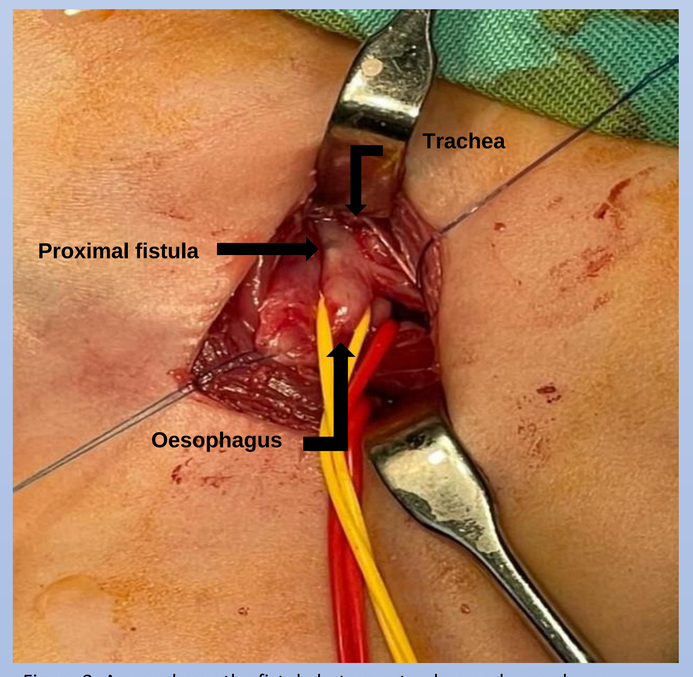

Figure 3: Arrow shows the fistula between trachea and oesophagus

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