Case Report/*Olgu Sunumu*

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A Laryngeal Web with Tracheo-Esophageal Fistula: Anaesthesiologist Skating on a Thin Ice

Trakeo-Özofageal Fistüllü Bir Laringeal Web: İnce Bir Buz Üzerinde Paten Yapan Anestezist

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ABSTRACT

Tracheo-esophageal fistula (TEF) in association with subglottic laryngeal web is a rare entity. Often the diagnosis of laryngeal web in the presence of TEF is missed and it is identified during intubation attempts. The triad of neonatal age, TEF and laryngeal web can even cause the most experienced anaesthetists concern about the mortality and morbidity that may develop due to the inability to secure the airway. In this case report, we aimed to share our experience on the airway management of a newborn with laryngeal web.

Keywords: Congenital anomaly, laryngeal web, neonate, tracheo-esophageal fistula

ÖZ

Subglottik laringeal web ile birlikte trakeoözofageal fistül (TÖF) nadir görülen bir durumdur. Çoğu zaman TÖF varlığında laringeal web tanısı atlanır ve entübasyon girişimleri sırasında saptanır. Yenidoğan yaşı, TÖF ve laringeal web üçlüsü, en deneyimli anestezistlerde bile hava yolunun güvenliğinin sağlanamamasına bağlı olarak gelişebilecek mortalite ve morbidite nedeniyle endişe yaratabilir. Bu olgu sunumunda, laringeal webi olan bir yenidoğanda hava yolu yönetimi deneyimimizi paylaştık.

Anahtar sözcükler: Konjenital anomali, laringeal web, yenidoğan, trakeo-özofageal fistül

INTRODUCTION

Congenital airway lesions involving larynx pose significant anaesthetic challenges in the form of difficult and emergent airway management. Congenital laryngeal webs form only 5% of the congenital laryngeal anomalies; the incidence has been estimated to be 1:10000 births (1). Of these 75% are glottic webs while sub-glottic webs constitute only 1.5% (2). The concomitant presence of trachea-esophageal fistula (TEF) with laryngeal web is even rarer to encounter in clinical practice. Only few case reports have been published till date (3). We are describing the difficult airway management due to presence of an un-expected laryngeal web in a neonate posted for TEF repair. Written informed consent has been obtained from the parents of the baby and all efforts have been made to hide the identity of the patient.

CASE REPORT

We present a case of a 1-day old male baby weighing 2.3 kg who was born in our institute following normal vaginal delivery. There was a weak cry at birth and the nasogastric tube could not be passed beyond 10 cm and its coiling was seen in the upper oesophageal pouch on subsequent antero-posterior view of chest radiography. The baby had a heart rate of around 150-160 beats min^{-1} and a saturation of 88% on room air. The baby was kept on oxygen using nasal canula at 5 L min^{-1} with a target saturation of 94% and was titrated accordingly. There were no added sounds in lung field or audible murmur on auscultation. On 2-D echocardiography, dextroversion was found with no major vessel anomaly. There was no other associated visible congenital abnormality. After obtaining written informed consent from the parents, the baby was shifted to paediatric surgery operation theatre on incubator and oxygen for ligation of TEF and its repair. Standard

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monitors like electrocardiogram, non-invasive blood pressure and pulse oximeter were attached, and drugs were prepared as per the age and weight of the baby and senior paediatric anesthetist was present for securing the airway. Baby was given iv fentanyl 2 µg kg^{-1} and induced with iv propofol 2.5 mg kg^{-1} and iv cisatracurium 0.15 mg kg^{-1}. A continuous nasal oxygen was kept attached and a laryngoscopy attempt was made. The vocal cords were visible but the black marking on the size 3.0 uncuffed endotracheal tube (ETT) was not going beyond the vocal cord and a sensation of mechanical obstruction was felt. On reexamining by laryngoscopy, a white glistening membrane type of structure could be visualised beyond the vocal cord with limited visibility. A neonatal bougie was then passed under direct laryngoscopy but the 2.0 mm size uncuffed ETT couldn't be rolled over it any further as it was getting kinked. The baby started desaturating and bag mask ventilation with minimal tidal volume and high rate was given to avoid gastric distension. It was decided to perform a diagnostic rigid bronchoscopy of size 3.5 mm (10339 DD DOESEL-HUZLY, KARL-STORZ SE & Co. KG, Germany) to find the reason behind mechanical obstruction. On bronchoscopy, a laryngeal web was found with a small hole at its center (Figure 1), through which the bougie was able to pass. Just as a part of trial method and to avoid tracheostomy, we used serial malleable dilators of increasing sizes (10, 12, 14 and 16 Fr) to enlarge the central hole (Figure 2). Following which, the 3.0 mm uncuffed tube was able to pass with a stylet within. Chest was auscultated for the correct placement of the tube and was then ventilated with air oxygen mixture. After stabilizing the baby and fixing the ETT, the diagnostic sheath of cystoscope no. 8 (KARL-STORZ SE & Co. KG, Germany) was passed through the ETT (due to non-availability of such small size bronchoscope to get negotiated with 3 mm size ETT) and the correct placement of tube was confirmed. Further the fistula was located on right main bronchus. The surgery was done and the baby was shifted post-operatively to neonatal intensive care unit for elective mechanical ventilation. Baby was extubated after 2 days upon meeting the desired requirements and discharged on 5th post-operative day.

DISCUSSION

Neonates are no doubt the "difficult patients" for anaesthesiologists due to their unique anatomical and physiological properties. It is even more challenging to manage a neonate with TEF due to their typical anatomical variation of the airway and associated complex congenital anomalies like vertebral defects, anal atresia, cardiac defects, TEF, renal anomalies, and limb abnormalities (VACTERL syndrome) (4). They are prone for desaturation and bradycardia both during intubation and during surgery. Only few authors have described the presence of laryngeal web along with TEF (3). Tracheo-esophageal fistula patients usually present with respiratory distress and weak cry. Therefore, laryngeal web can be easily missed in the preoperative period as its presentation mimics that of a TEF and its diagnosis is usually made by radiological imaging like MRI which is not routinely done as a part of pre-operative investigations for TEF.

Tracheo-esophageal fistula in association with laryngeal web is a rare but interesting entity from the perspective of embryogenesis and due to posing difficulty in airway

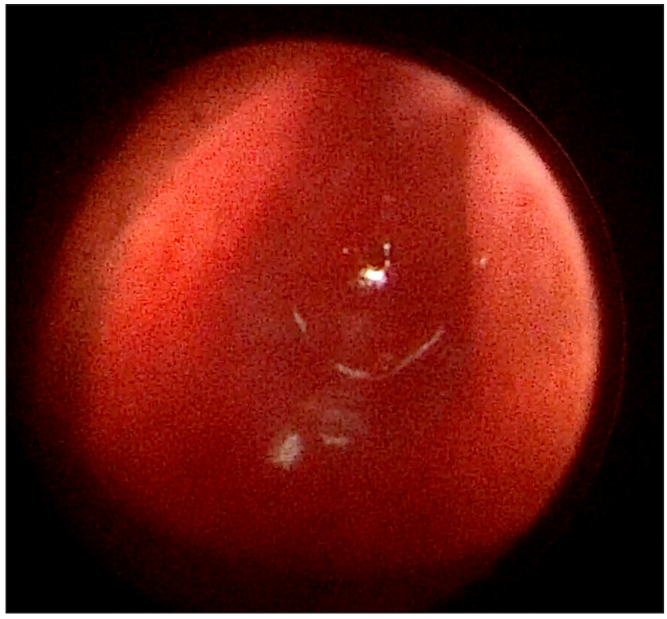

Figure 1. Bronchoscopic view of subglottic web.

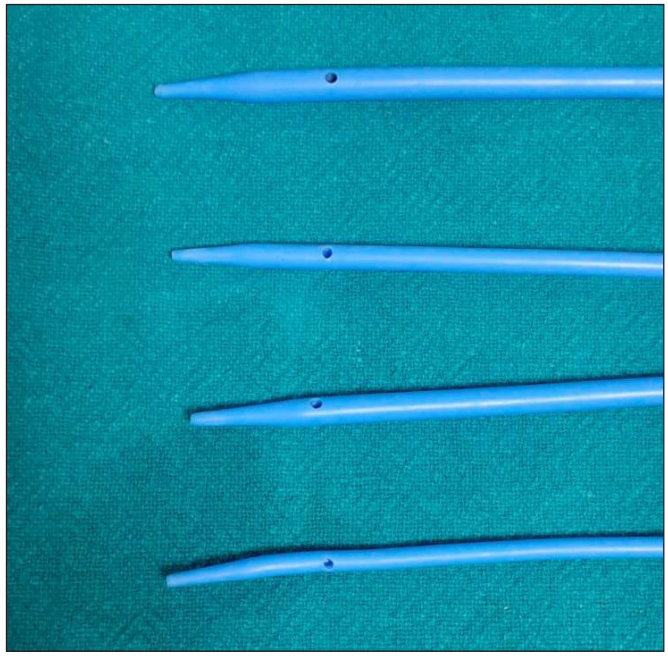

Figure 2. Malleable dilators of increasing sizes.

management. Classically, the esophagus and trachea develop from the primitive digestive tube (PDT), which first develops from the primitive endoderm. From the distal to the proximal ends of PDT, trachea and esophagus are separated by a mesenchymal septum, which is created in the coronal plane of the PDT. If this process is unsuccessful, a trachea-esophageal deformity, such as an esophageal atresia, tracheal atresia, or TEF develops (5,6). The laryngeal lumen gets temporarily blocked by an epithelial lamina during development, which dissolves at 10th week of gestation. Laryngeal web is formed due to incomplete recanalization of this primitive laryngeal airway (7).

Sub-glottis is the narrowest part of the neonatal airway. Any condition like presence of a web will further decrease its lumen thereby increasing the airflow resistance in the exponent of four. In previously reported cases, airway was secured with neonatal tracheostomy (3,8). Since tracheostomy in a neonate has a potential risk of subsequent complications, we attempted dilatation first. The malleable dilator which we used had a smooth surface with an opening at the tip. First, we used the smallest available dilator, which could be passed through the central hole of the web. Following this, we used serial dilators of increasing sizes, to create enough space for an uncuffed tube of size 3 mm to pass through it. In our case, the laryngeal web was like a thin membrane, so we were able to dilate it using serial dilators. However, thicker and more extensive webs often have subglottic cartilaginous involvement which cannot be dilated using these dilators and require emergency tracheostomy to secure the airway (9).

CONCLUSION

Tracheo-esophageal fistula with a laryngeal web is a rare but extremely difficult airway situation. A close collaborative effort between anaesthesiologist, pediatric surgeon and neonatologist is the pre-requisite for a better patient outcome.

AUTHOR CONTRIBUTIONS

Conception or design of the work: AR, R

Data collection: AS, PS, KD

Data analysis and interpretation: R, AR

Drafting the article: AR, R

Critical revision of the article: SC, SPS

All authors (AR, AS, R, PS, SC, SPS, KD) reviewed the results and approved the final version of the manuscript.

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