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Blue Baby and Association of Tracheal Anomalies- A Rare Entity

Dr Pragya Gupta, Dr Arin Choudhury, Dr Virendar Kumar

Abstract: ***Tetrology of Fallot is a congenital heart disease.It was first reported in 1673 by Nils Stenson, a Danish anatomist. Later on, associated symptoms such as tracheal anomalies came into existence and we called them Blue Baby with difficult airway. We present a rare case of a 12-year old boy with Tetrology of Fallot, posted for intracardiac repair with incidental finding ofa supraglottic glottic web at level of vocal cord that posed an anesthetic challenge for intubation.***

Keywords: Tetrology of Fallot, Tracheal Anomalies, Blue Baby, Difficult airway, Supraglottic web

1. Introduction

Tetrology of fallot (TOF) is one of the most common congenital heart disorders often associated with congenital tracheal anomalies. In literature, syndromes have been described of heart defects associated with deletion of chromosome 22q11 with tracheal anomalies such as DiGeorge syndrome, velocardiofacial (Shprintzen) syndrome and CATCH22 (Cardiac defect, abnormal facies, thymus hypoplasia, cleft palate, hypocalcemia, chromosome 22 deletion).[1] Among the congenital anomalies of the larynx, supralglottic webs are a rarity with an incidence of 1 in 10,000.[2] We describe here a case of difficult intubation in a 12year old boy after induction of anesthesia caused by an unexpected supraglottic web.

2. Case Report

A 12year old boy presented to us with cyanosis, dyspnea since birth.He had been experiencing increasing exercise intolerance, frequent episodes of cyanosis and substantial shortness of breath upon exertion with no history of chest pain or palpitations. His past history was otherwise notable only for left temporoparietal brain abscess and cyanotic spells for which he was on conservative management. No other congenital abnormalities were detected from history. He had not undergone any surgical treatment for his cardiac condition before because of limited access to medical care.

General physical examination revealed weight and height to be normal according to age.Central cyanosis of tongue,lips and clubbing Grade III present. Cardiac auscultation revealed a systolic ejection murmur best heard at left upper sternal border and radiating to axilla and back. His respiratory, abdominal and neurologic examinations were normal.

Airway assessment was done and it was found to be an easy airway. All routine investigations within normal limit.

Chest x-ray revealed boot shaped heart. Echocardiography and Computed Tomography scan finding confirmed the diagnosis of TOF and Major Aortopulmonary Collateral Arteries (MAPCAs) were coiled.Post coiling saturation was 82% on room air.

Patient was posted for elective intracardiac repair after taking high risk consent from parents.All American Society of Anesthesiologists standard monitoring attached. Intravenous access secured.

Preoperatively patient wasconscious oriented, Pulse-78/min, Blood Pressure-100/62mmHg, Saturation- 82% on room air.Preoxygenation was done with 100% oxygen with adequate sized closed fitting mask for three minutes. Premedication was done with fentanyl. General anesthesia was induced with ketamine. After induction of anesthesia, no difficulty was encountered in bag mask ventilation so muscle relaxant, vecuronium was given. Easy mask ventilation was performed for three minutes.

On Direct Laryngoscopy with a Macintosh blade size 3, only posterior margin of vocal cords was seen clearly, a 7.0-mm internal diameter cuffed tracheal tube could not be advanced below the level of vocal cords because of resistance. Intubation was re-attempted after oxygenation by mask with trial of smaller tubes. Finally, a 5.0-mmid uncuffed tube was passed successfully through the vocal cords and secured in place in the third attempt. Because of the unexpected difficulties during intubation, surgery was deferred and an otolaryngologist was consulted to examine the larynx with flexible fibreoptic bronchoscope. Anterior glottic web at the level ofvocal cordsextending antero-posteriorly from the anterior commissure to the junction of the anterior 2/3rd and posterior 1/3rd of the vocal cords with minimal posterior opening, extending into sub-glottis and restricting airway by 35-50% seen. It was thick, white in color and had a smooth surface (Figure1 & 2). A diagnosis of congenital supraglottic web was made. The consultant advised no treatment because the web caused no apparent symptoms.

Surgery was resumed as scheduled. Patient shifted to intensive care unit for elective post-operative ventilation and monitoring and was extubated over airway exchange catheter the next day. Tracheostomy was kept as back up if prolonged intubation was expected. There was no respiratory problem noted during hospitalization. Patient was discharged from hospital one week later.

3. Discussion

Unanticipated laryngeal webs creating difficult intubating conditions in the operating room have been characterized as rare. It has been reported that 75% of laryngeal webs occur at the level of the vocal cords, and the remainder are in the subglottic or supraglottic location.[3]Supraglottic webs are

diaphragmatic growths of differing thickness that partially occlude supraglottic lumen, resulting from incomplete recanalization of larynx during embryogenesis. More than 90% of laryngeal webs are located anteriorly and extend towards the arytenoids.[4] A third of children with laryngeal webs have anomalies of the respiratory tract, most commonly subglottic stenosis. The webs vary in thickness from a thin structure to one that is thicker and more difficult to eradicate. Diagnosis may be made via flexible or rigid laryngoscopy, or radiographic films if subglottic or cricoid pathology is present.They are generally congenital defects but may be acquired secondary to a surgical procedure, intubation or infection. Congenital supraglottic webs are uncommon, constituting five percent of all congenital laryngeal lesions. Many anterior webs are associated with deletions of chromosome 22q11.[1] Clinical presentations of laryngeal webs vary from case to case. Patients may be asymptomatic and may present as unanticipated cases of difficult intubation. Most congenital webs present at birth or in the first few months of life with symptoms. Symptoms can vary from hoarseness of voice to respiratory distress, stridor, croup and dysphagia.[3]

Cohen categorized laryngeal webs (glottic webs) into four types based on their appearance and an estimation of the degree of airway obstruction. Type ! glottic webs have uniform thickness with no subglottic extension, compromises < 35% of the airway and usually no airway obstruction. Type 2 glottic webs are slightly thicker, with a significantly thicker anterior component. Subglottic involvement is minimal. The web restricts the airway by 35 to 50% and usually causes little airway distress, unless the patient has an acute infection or is traumatized during intubation. Type 3 glottic web is thick; the anterior portion of the web is solid, extends into the sub glottis and the true vocal cords are not well delineated. The web restricts the airway by 50 to 75%, and obstruction is moderately severe. Type 4 glottic web is uniformly thick extending into the subglottic area with resulting subglottic stenosis, occluding airway by75 to 90%. Respiratory obstruction is severe, and the patient is almost always aphonic.[5]

Treatment of laryngeal webs depends on the web thickness. Asymptomatic patients do not require treatment. Approximately 60% of patients require surgical intervention, depending upon the type of lesion. Treatment depends on the severity of airway obstruction, and may be single or multi staged. If a patient presents with difficulty breathing, the airway must be secured first. This can be done through endotracheal intubation, which can be converted into a tracheostomy if prolonged intubation is expected. Surgical division can be done using laryngeal knives, micro-scissors, galvanocautery or radiofrequency. Web excision results in two opposing surfaces with denuded epithelium which tends to heal together and form a web again. A keel (Silastic sheet) prevents this sequlae. Topical corticosteroid injections and local application of mitomycin C also prevents scarring.[6]

Learning Points

This case stresses the importance of evaluationof other congenital anomalies beforehand.Planning for awake intubation.Difficult airway cart including tracheal tubes of various sizes, video laryngoscope and fibreoptic should be kept ready. Tracheal tube should not be advanced when resistance is encountered in order to avoid trauma. The airway can be managed in patients with an unsuspected tracheal web by using different sizes of endotracheal tubes, using laryngeal mask airway, tracheostomy, or waking up the patient and postponing the surgery until further evaluation.

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Nil

5. Conflicts of interest

There are no conflicts of interest

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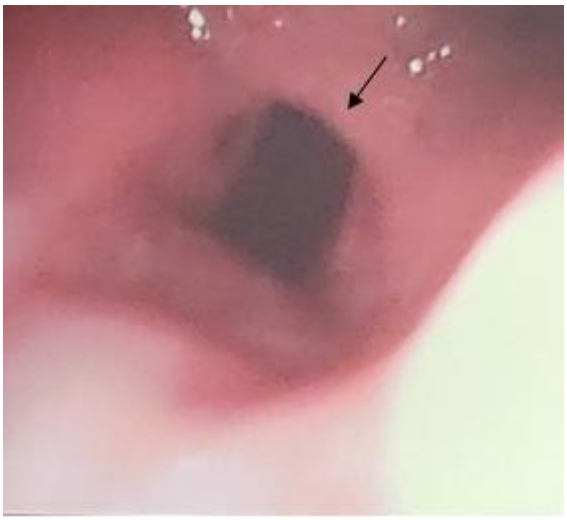

Figure 1

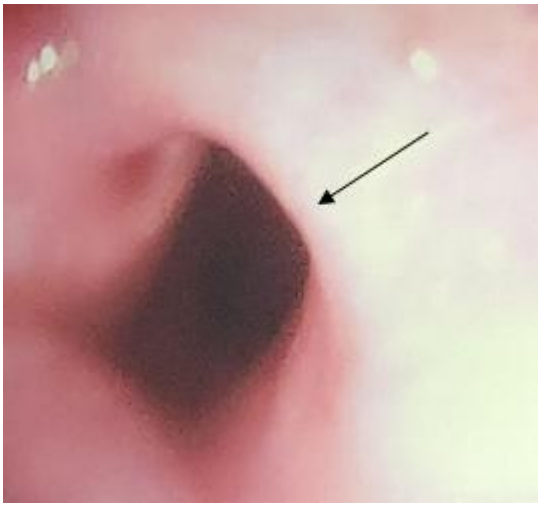

Figure 2

Figure1 & 2- Views of the larynx on flexible fibreoptic bronchoscopy showing supraglottic web