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Case Report

Anesthetic management of difficult paediatric airway in a case of large facial Neurofibromatosis – A case report

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ABSTRACT

Neurofibromatosis is tumor involving different parts of the body. Neurofibromatosis may potentially lead to difficult airway. These challenging cases need special considerations. Various anesthetic techniques and devices are described to secure the airway. In the present case we intubated a 11-year-old child with large left side facial tumor extending to the neck which had intraoral encroachment displacing the tongue and pharynx to the right. This is a case report illustrating intubation of difficult pediatrics airway using awake bronchoscopic intubation under conscious sedation.

1. Introduction

Neurofibromatosis is a group of an autosomal dominant disorders leading to the formation of neurofibromas or nerve sheath tumors all over the body. This disease occurs in 1:3000 births and it is the most common type of neurocutaneous syndrome.[1] Plexiform neurofibromas occur mostly in the head, neck, face and larynx, which makes bag mask ventilation and intubation difficult.[2] It can lead to the most dreaded complication of CVCI (Cannot ventilate cannot intubate). This compromises the safety of the patient and hence special precautions and safe exit strategies need to be followed while handling a large facial tumor specially in pediatric patients. We report a case which demonstrates the use of awake bronchoscopic intubation as a safe and valuable tool in managing a unique difficult pediatric airway case with negligible risk.

2. Case Report

A 11-year-old patient presented with a huge lobulated swelling in the left side of face and neck since last 10 years and it gradually progressed extending into the oral cavity displacing the tongue and pharynx to the right. The child suffered from difficulty in swallowing. There were no breathing complaints, stridor nor any change in voice. The child had good effort tolerance and had no associated comorbidities. He weighed 23kg, and his height was 137cm. The patient was scheduled for debulking of the mass which was diagnosed to be neurofibromatosis (Figure 1). Preoperative anesthesia evaluation revealed mouth opening 1.5 finger breadth, tongue and uvula deviated to the right. However, the neck extension and dentition were within acceptable limits (Figures 2 and 3). The parents were counselled for awake brochoscopic intubation with written informed consents explaining the alternative options and complications. As a part of the multidisciplinary approach the ENT team was informed for the potential need of emergency surgical tracheostomy.

In the preoperative room intravenous (IV) glycopyrrolate 0.1mg and IV midazolam 0.5mg were given for anti-

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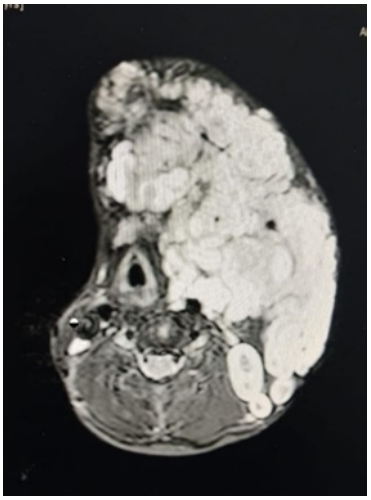

Figure 1: MRI of neck showing tumor encroaching the oropharynx and causing deviation of the glottis

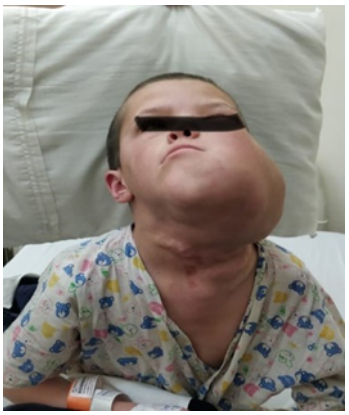

Figure 2: Extent of tumor on the face and neck

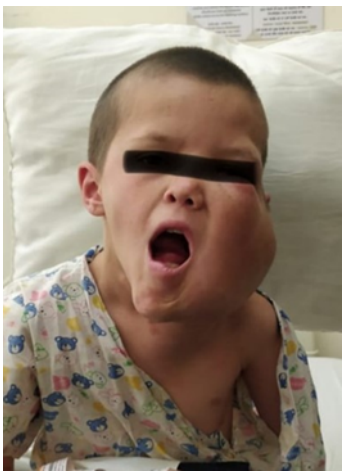

Figure 3: Airway examination

sialagogue and anti-anxiety action respectively. IV dexmedetomidine was started for conscious sedation in the dose 1mcg/kg over 10 min followed by infusion @ 0.5 mcg/kg/hr. Nebulization with Lignocaine 4% solution was done and the child was wheeled into the OR. Intraoperatively, ASA standard monitoring was used. Oxygen was administered with nasal prongs @ 4l/min. After administering IV Ketamine 10mg, a prelubricated Ambu aScope™ Broncho Slim (outer diameter 3.8mm) bronchoscope was introduced through the nares. Gentle bronchoscopy was performed using spray as you go technique using 2% lignocaine. After entering and identifying the trachea the railroaded ETT was introduced and advanced. The correct placement of ETT was ensured and Inj Atracurium 12.5mg administered. Anesthesia was maintained with controlled mechanical ventilation using sevoflurane, IV morphine 2.5 mg and intermittent IV atracurium. Radial artery was cannulated for invasive BP monitoring. The debulking surgery lasted for 8hrs and the patient was transferred to PICU and gradually weaned from the ventilator support. The patient was satisfactorily discharged on a later date after an uneventful management.

3. Discussion

Neurofibromatosis is an autosomal dominant disorder described as the congenital malformation of ectodermal tissue. It leads to abnormal deposition of excess neural tissue within the nervous system, viscera, endocrine system and skin. It is classified as Neurofibromatosis Type 1 (NF1) and Neurofibromatosis Type 2 (NF2). Incidence of NF1 is 1 in 3000 live births and NF2 affects 1 in 33000 – 40000 of the children. NF2 gene is present on chromosome 22 which form merlin or schwannomin, a cytoskeleton protein.[1] Screening MRI can rule out any disease of the thorax and abdomen which can lead to the compressive symptoms. As NF2 is relatively rare there are not enough cases reported about the difficulty faced during intubation of the pediatric patients specially involving the head and neck region. Neurofibromas involving the tongue, pharynx or larynx can lead to difficult intubation and requires specialized technique. Patients may show symptoms like dysphagia, dysarthria or stridor. Any change of voice should be considered as a warning sign which is suggestive of the difficult airway.[2]

Paediatric awake fibreoptic intubation is difficult because smaller airways limit the range of manipulation of tip of the bronchoscope. In addition to this, there is rapid desaturation due to increased oxygen consumption and shorter safely tolerated apnoea time. Awake fibreoptic intubation is recommended for intubation of patients with difficult airways.[3] Although this is feasible in adult patients, it is more difficult to perform on paediatric patients while awake because of lack of cooperation and risk of bronchospasm.[4]

Kholy JE et al performed successful awake fibreoptic intubation in a child with burn over face and neck region.[5] Sharma A et al reported the asleep fibreoptic intubation using sevoflurane in incremental dosage while maintaining the spontaneous breathing in a child with restricted mouth opening.[6] Asghar A et al used sevoflurane as an anaesthetic agent in case of awake fibreoptic in a child with bilateral temporomandibular joint ankylosis.[7] Mazlan et al. used dexmedetomidine in loading dose with midazolam and fentanyl in titrating dosages for smooth and successful intubation in a child with giant haemangioma of the tongue.[8] Nebulized lignocaine (4%) is used pre-operatively for smooth induction is particularly helpful.[9]

In our patient all efforts were aimed at maintaining spontaneous respiration throughout till ETT was secured. We used dexmedetomidine for conscious sedation. We used ketamine to prevent undue movements at the introduction of the bronchoscope. Nebulisation with inj. Lignocaine followed by spray as you go technique contributed to topical anaesthesia of airway mucosa and thus blunting the airway reflexes. This further aided smooth advancement of the bronchoscope and the ETT with adequate cooperation from the patient. Although the tumour was huge, we successfully prevented the airway trauma, bleeding and oedema and secured the airway in safest possible way. There are other methods of difficult airway management like video-laryngoscope assisted intubation, blind nasal intubation and tracheostomy described in the literature. The video-laryngoscope was not considered because of intraoral extension of tumour mass which deviated the tongue, uvula and pharynx to the opposite side. There was no room for supraglottic airway devices as a rescue tool. The Blind nasal intubation carries the risk of airway injury which could have been catastrophic. Tracheostomy was our last resort if the awake bronchoscopic intubation had failed because it is an invasive procedure with serious complications in paediatric age group.

4. Conclusion

We conclude that securing difficult pediatric airway using awake bronchoscopic intubation technique under conscious sedation in titrated doses yet maintaining spontaneous respiration after adequate airway preparation is a very safe technique. However, the steep learning curve for awake bronchoscopic intubation cannot be ignored. Well trained and equipped anesthesiologist with readily available assistance for emergent surgical airway access, if needed, is a pre requisite for conducting awake bronchoscopic intubation.

5. Source of Funding

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6. Conflict of Interest

None.

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