Case Report

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Anaesthesia Section

Navigating Airway Challenges in a Case of Neurofibromatosis: An Anaesthetic Perspective

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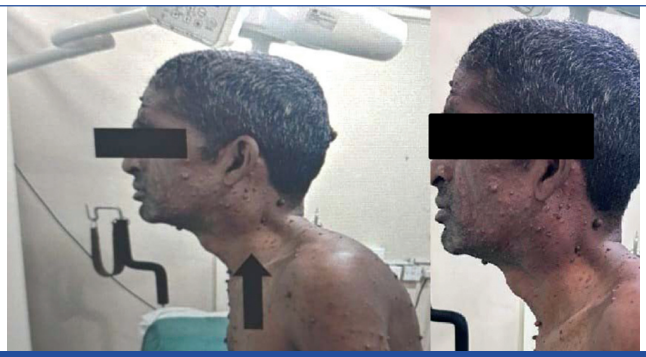
ABSTRACT

Neurofibromatosis type 1 (NF1) often involves multisystem manifestations, including pulmonary fibrosis, hypertension and neurofibromas in the larynx and oropharynx, leading to airway challenges. This case report highlights the anaesthesia and airway management of a 48-year-old male with NF1 and a large parapharyngeal mass causing hoarseness, dysphagia and airway compromise, displacing major neck structures; he was posted for excision of the parapharyngeal mass. Indirect videolaryngoscopy during preoperative evaluation showed a funnel-shaped epiglottis, significant oedema of the surrounding tissues and a fixed left vocal cord, with a Mallampati score of IV. Fibreoptic intubation was initially planned; however, elective fibreoptic intubation might fail even with prior recognition of intraoral pathology due to grossly distorted anatomy and bleeding from trauma. Utilising the airway preparation carried out for fibreoptic intubation, videolaryngoscopy using C-MAC (Karl Storz GmbH & Co. KG, Germany) was performed to assess the feasibility of intubation under sedation and spontaneous respiration, which yielded a Percentage of Glottic Opening (POGO) score of 50%, corresponding to a Modified Cormack and Lehane grade of 2b. The possibility of pressure effects was ruled out, as the tumour was located laterally. After assessing the possibility of intubation with C-MAC, nasal intubation was successfully achieved using a size 6.5 flexometallic tube under deep sedation, following the administration of Inj. Midazolam 1 mg, Inj. Fentanyl 80 µg, and Inj. Propofol 1 mg/kg. The tumour was excised without complications and the patient was extubated uneventfully. This report emphasises the importance of preoperative planning and a stepwise, adaptable approach to airway management in such complex scenarios.

Keywords: Awake intubation, Fiberoptic intubation, Multisystem involvement, Parapharyngeal mass

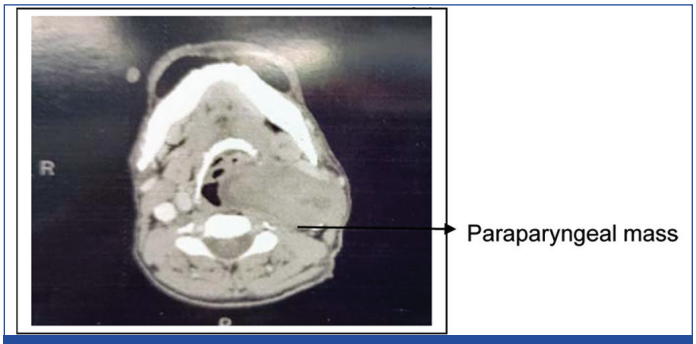
CASE REPORT

A 48-year-old male, weighing 56 kg and measuring 160 cm, who was scheduled for the excision of a parapharyngeal tumour. He reported swelling in the upper left neck for two years, along with hoarseness and dysphagia for one month. The patient was a known case of Neurofibromatosis (NF) with multiple neurofibromas since birth but had no other co-morbidities and was vitally stable [Table/Fig-1].

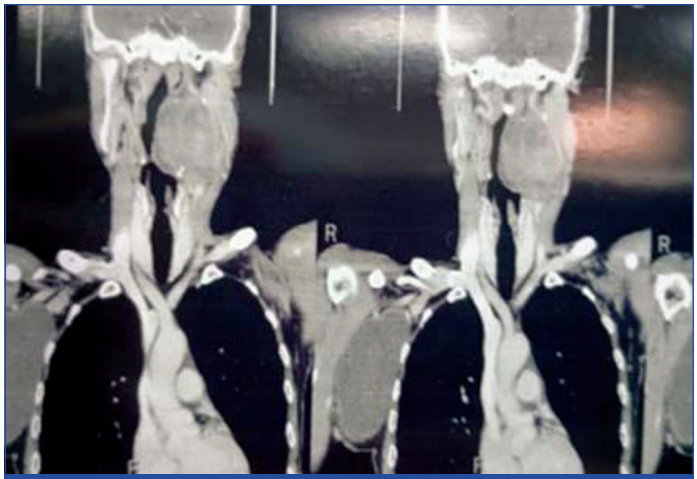


[Table/Fig-1]: Multiple Neurofibromas with left parapharyngeal mass swelling.

Airway assessment revealed a Mallampati score of IV. Magnetic Resonance Imaging (MRI) showed a left parapharyngeal mass measuring 75×69×62 mm, displacing the common carotid artery, as well as the internal and external carotid arteries and causing a mass effect on the oropharynx [Table/Fig-2,3]. Histopathological examination confirmed a schwannoma of nerve cells. Other investigations, including chest X-ray, ECG, 2D Echocardiogram (2D echo), and thyroid function tests, were normal. Videolaryngoscopy revealed a mass occluding the oropharynx, a displaced tonsil, a compressed, funnel-shaped epiglottis with significant surrounding oedema and a fixed left vocal cord [Table/Fig-4], indicating potential difficulties in ventilation and intubation. Fibreoptic intubation was initially planned.

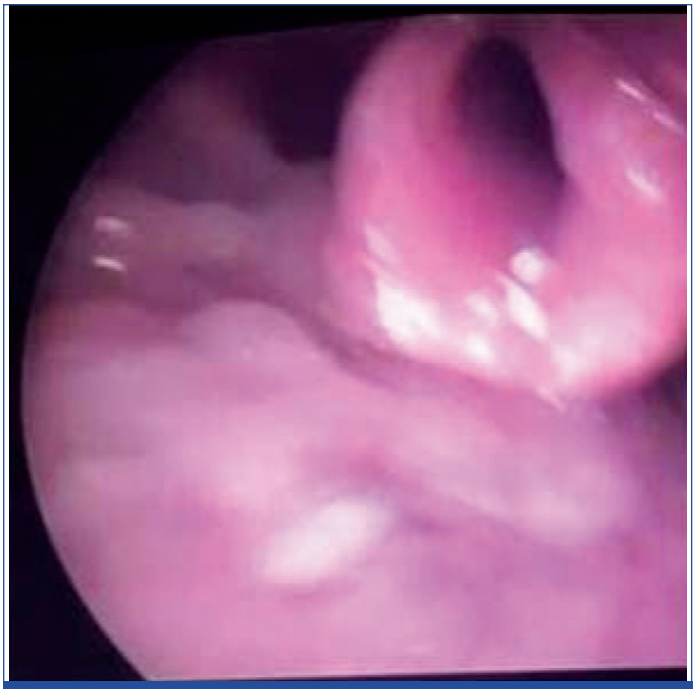


[Table/Fig-2]: Axial CT scan of neck (plain) showing parapharyngeal mass (arrows), possibly Schwannoma.



[Table/Fig-3]: Contrast enhanced coronal CT images of the neck and upper thorax showing well defines soft-tissue mass on left side of neck.

A difficult airway consent was obtained. Preoperative airway preparation included intravenous hydrocortisone 100 mg and ondansetron 4 mg. Glycopyrrolate 0.2 mg was administered

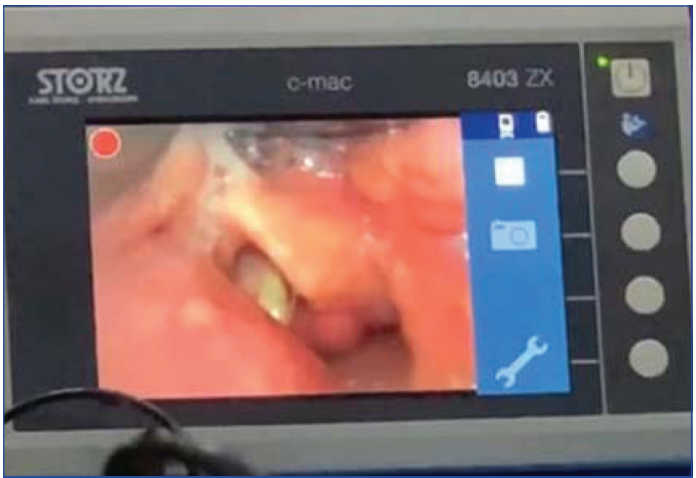


[Table/Fig-4]: Videolaryngoscopy view of epiglottis – With 70° endoscope during preanaesthesia check-up.

intramuscularly 30 minutes before induction. Analgesia included paracetamol 1 g and tramadol 50 mg. Nebulisation with dexmedetomidine 40 µg and 4% lignocaine (4 mL) was provided for 20 minutes. Nasal packing with 2% lignocaine-adrenaline solution was applied for 10 minutes.

In the operating theatre, American Society of Anaesthesiologists (ASA) standard monitors were attached. Xylometazoline nasal drops were instilled. Equipment for difficult intubation, including various endotracheal tubes, supraglottic airway devices, a C-MAC videolaryngoscope (Karl Storz GmbH & Co. KG, Germany), a cricothyroidotomy set, and a tracheostomy set was prepared. A transtracheal nerve block with 2% lignocaine was performed, and the posterior tongue and oropharynx were anaesthetised with a 10% lignocaine spray.

Elective fibreoptic intubation might fail even after recognition of intraoral pathology due to grossly distorted anatomy and bleeding from trauma. The airway preparation was utilised to perform awake videolaryngoscopy with the C-MAC blade to assess the possibility of intubation with spontaneous respiration. Awake videolaryngoscopy with C-MAC showed a POGO score of 50% (Modified Cormack-Lehane grade 2b) [Table/Fig-5]. The compression effect was ruled out, as the tumour was located laterally. Therefore, it was decided to proceed with nasal intubation under sedation, using C-MAC under spontaneous respiration.



[Table/Fig-5]: C-MAC videolaryngoscope view showing epiglottis and POGO score of 50%.

Preoxygenation was initiated with 100% oxygen. Sedation was achieved with midazolam 1 mg, fentanyl 80 µg, and propofol 1 mg/kg. A 6.5 mm flexometallic tube was used for nasal intubation. After confirming bilateral air entry, vecuronium 6 mg was administered. Mechanical ventilation was initiated in volume control mode and anaesthesia was maintained with sevoflurane, oxygen and nitrous oxide. The patient remained vitally stable throughout the intraoperative period, except for one episode of bradycardia (HR 45 bpm), which was managed with glycopyrrolate 0.2 mg, likely due to the proximity of the tumour to the carotid artery. The surgery was uneventful and extubation was performed successfully with 3.5 mg/kg of sugammadex.

DISCUSSION

NF is an autosomal dominant disorder, representing the most common neurocutaneous syndrome. It is categorised into NF1 and NF2, with NF1 presenting as cutaneous, nodular, or plexiform neurofibromas [1]. NF may be associated with multisystem involvement. Airway involvement, such as neurofibromas of the tongue, larynx, or pharynx, may cause difficult intubation. This can be anticipated based on symptoms like dysphagia, dysarthria, stridor, or voice changes. Respiratory system involvement may lead to pulmonary fibrosis or intrapulmonary neurofibromas, which may produce cough and dyspnoea and may be associated with right ventricular failure. Any associated kyphosis or scoliosis may compromise lung function. The presence of hypertension may be associated with phaeochromocytoma and renal artery stenosis in such cases. Signs of superior vena cava obstruction may indicate a mediastinal tumour. Involvement of the Central Nervous System (CNS) may cause epilepsy. Involvement of the musculoskeletal system, in the form of vertebral deformities or spinal cord tumours, may make neuraxial blocks difficult [1].

Other concerns include the possibility that these patients might have short stature, bone abnormalities and cardiovascular abnormalities such as congenital heart malformations and vasculopathy. A carcinoid tumour in the duodenum may result in jaundice and carcinoid syndrome. Cognitive impairments, as well as attention and hyperactivity problems, are two conditions that can co-exist [2]. Exceptionally, some cases have been described of altered sensitivity to neuromuscular blockers, giving rise to prolonged episodes of apnoea with an unexplained mechanism [1,3-6].

In index patient, a large parapharyngeal mass with associated glottic oedema, a fixed left vocal cord and anticipated airway distortion presented a major challenge during airway management. Although index patient had no systemic involvement, comprehensive preoperative evaluation and readiness for emergency front-of-neck access were ensured. Even after considering awake fibreoptic intubation initially and adequate airway preparation for the same, the risk of failure due to mucosal trauma and anatomical distortion prompted us to proceed with awake check videolaryngoscopy using the C-MAC. During airway assessment, the compressive effect of the mass due to gravity on the airway must be taken into consideration. Once it was confirmed that compression was not critical by videolaryngoscopy, and with a POGO score of 50%, which corresponded with a modified Cormack-Lehane grade 2b, nasal intubation was successfully carried out under sedation and spontaneous ventilation. Nagabuchi R et al., reported a similar case of a giant parapharyngeal mass with upper airway obstruction where awake intubation using a McGrath® MAC video laryngoscope was successfully employed to avoid invasive techniques and maintain spontaneous ventilation [7]. Watson WA and Cormack J, used a flexible-tipped bougie under videolaryngoscopic guidance to intubate a patient with a large parapharyngeal mass, demonstrating the benefit of alternative strategies when airway anatomy is compromised [8].

While awake fibreoptic intubation remains the gold standard in predicted difficult airways, it is not without limitations. A prospective cohort study in the UK showed a 1% failure rate and an 11% complication rate [9], and a US closed claims analysis highlighted risks associated with failed attempts [10]. Wulf H et al., reported a failed fibreoptic intubation despite vocal cord visualisation, which was attributed to glottic deviation and necessitated an urgent tracheostomy under local anaesthesia [11]. Vinayagam S et al., used a 70° rigid nasendoscope with a Frova introducer to railroad the endotracheal tube in two of their cases where fibreoptic attempts failed due to mass-induced dynamic airway obstruction or lateral glottic displacement [12]. These findings suggest that in settings of distorted airway anatomy or when fibreoptic devices are unavailable, awake videolaryngoscopy can serve as an effective and safer alternative.

In recent years, the widespread use of videolaryngoscopes, second-generation laryngeal masks, the possibility of reversing neuromuscular blockade with sugammadex and teaching surgical techniques for emergency front-of-neck access have changed the approach to the difficult airway [13]. Thus, in cases of NF where a tumour involves the oropharynx, tongue, or larynx, a stepwise approach is required to prevent complications.

CONCLUSION(S)

This case highlights the airway challenges in a patient with NF type 1 and a large parapharyngeal mass. Awake videolaryngoscopy with the C-MAC proved to be a safe and effective alternative to fibreoptic intubation, allowing for successful intubation under sedation with spontaneous respiration. It emphasises the importance of individualised airway planning, preintubation assessment and readiness with alternative strategies. Videolaryngoscopy can be a valuable option when fibreoptic techniques are limited by anatomical distortion or when they are unavailable.

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