Type-2 Neurofibromatosis Patient with Parasagittal Meningioma: A Challenging Airway

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Abstract

The anesthetic management of a type-2 neurofibromatosis patient with a difficult airway is quite challenging. Such a situation tests the limits of the anesthesiologist's preparedness in maintaining the balance of the cerebral protection strategies and securing the airway. The anticipated challenges include managing the airway, controlling the raised intracranial pressure, and maintaining stable hemodynamics in anticipation of the expected blood loss. This article focuses on the preoperative airway evaluation and intraoperative preparedness in case of a patient with type-2 neurofibromatosis scheduled for craniotomy and excision of intracranial meningioma.

Keywords

- type-2 neurofibromatosis
- difficult airway
- parasagittal meningioma
- craniotomy

Introduction

The neurofibromatosis (NF) is a group of disorders with autosomal dominant penetrance and presents with benign tumors of ectodermal and mesodermal origin. It is the most common cause of the various documented neurocutaneous syndromes.[1] Type-2 neurofibromatosis (NF-2) is usually familial and presents with vestibular schwannomas, meningiomas, glioma, juvenile posterior subcapsular cataract, and gradual progressive hearing loss. Type-2 neurofibromatosis involving the pharynx and larynx can complicate airway management; however, it is a rare occurrence. We present a successful management of a NF-2 patient scheduled for craniotomy and excision of parasagittal meningioma with upper airway involvement.

Case Report

A 23 year-old-male patient (weight: 54 kg, height: 168 cm) was diagnosed with parasagittal meningioma, scheduled for craniotomy and excision. The patient presented with dizziness, headache, and decreased vision, swelling in the left side of the neck, and hoarseness of voice for 1 year. Airway examination revealed a mouth opening of more than three fingers, Mallampati class-3, a left submandibular mass with restricted neck movements on the left side. Videostroboscopy showed left vocal cord palsy, tubular epiglottis, and no sulcus (►**Fig. 1A**). Magnetic resonance imaging (MRI) neck revealed a submandibular mass ($4 \times 3.2 \times 5.1$ cm), anterior and posterior cervical node enlargement with tonsillar hypertrophy, fatty infiltration of the left side of the tongue, and floor of the mouth, suggesting hypoglossal schwannoma (►**Fig. 1B**). MRI brain showed multiple schwannomas involving the lower cranial nerve, cervical spinal nerves, and parasagittal meningioma, $5 \times 5 \times 3$ cm in size (►**Fig. 1C**).

On the day of surgery, nil per oral status was confirmed. The patient was shifted to the operation room, and standard American Society of Anesthesiologists (ASA) monitoring was started. A difficult airway cart, including fiberoptic intubation (FOI), was prepared in view of the anticipated difficulty. The patient was induced with intravenous (IV) injection fentanyl 120 µg and propofol titrated to loss of verbal response (60 mg), followed by infusion at a rate of 150

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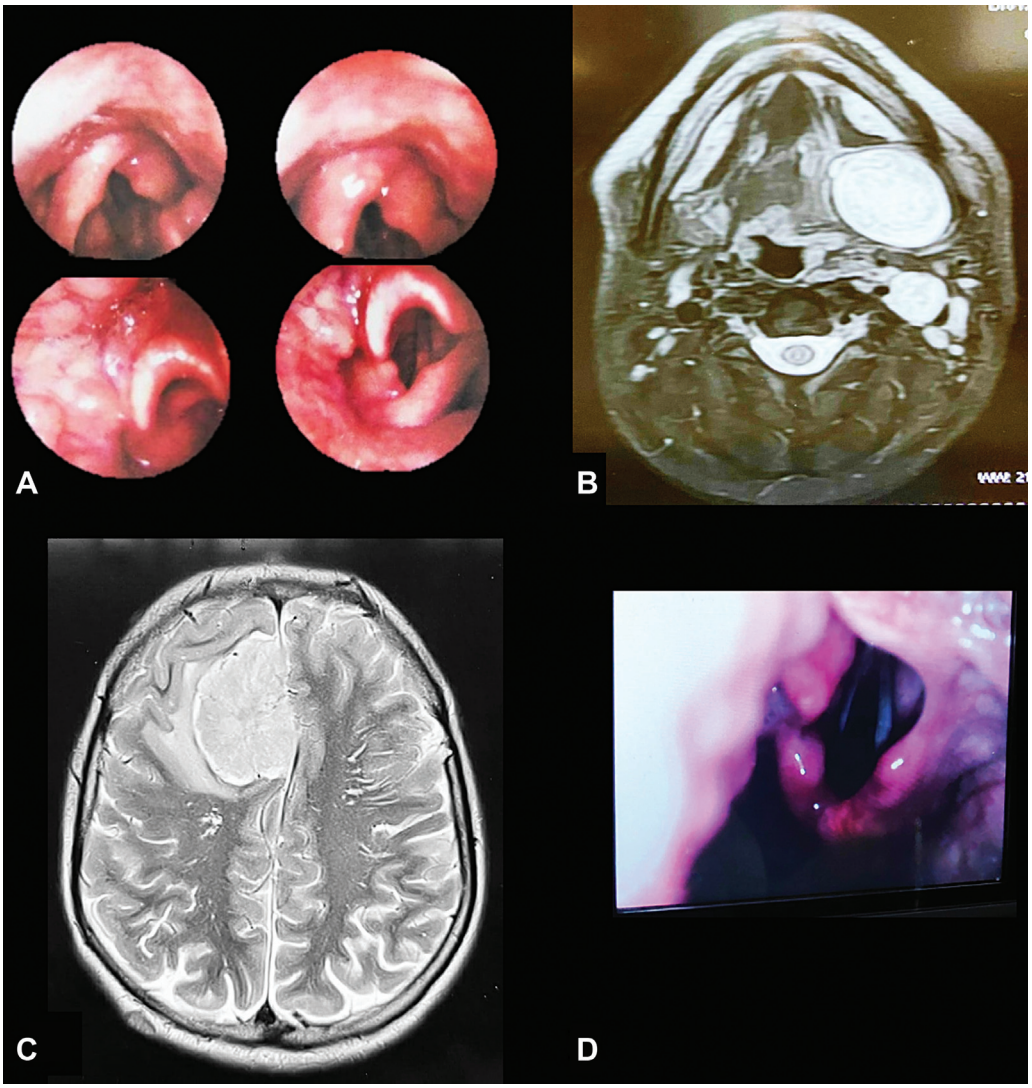

Fig. 1 (**A**) Videostroboscopy view showing left vocal cord palsy, tubular epiglottis, and no sulcus. (**B**) MRI neck with contrast showing hypoglossal schwannoma. (**C**) MRI brain showing parasagittal meningioma. (**D**) Videolaryngoscopy view showing neurofibroma involving corniculate and cuneiform cartilage.

µg/kg/min to achieve the desired depth of anesthesia (BIS value of 40–60). A check videolaryngoscopy was performed in anticipation of the neurofibromas in the pharynx and larynx. Neurofibromas involving corniculate and cuneiform cartilage were seen without any other abnormality (►**Fig. 1D**). Bag and mask ventilation was confirmed, and IV injection atracurium 30 mg was administered. The trachea was intubated with an 8.0 mm/cuffed/polyvinyl chloride endotracheal tube. The patient was ventilated using oxygen–air (1:1), intermittent IV injection vecuronium, fentanyl, and infusion propofol 150–200 µg/kg/min. The $EtCO_2$ was maintained between 30 and 35 mm Hg. The arterial line was secured in the right radial artery and a right subclavian was cannulated. Scalp block (20 ml of 0.5% plain bupivacaine (10 mL) and 2% lignocaine with adrenaline (10 mL) + 10 mL saline to make the desired volume of 30 mL) was given to blunt the pin response and postoperative analgesia. Intravenous injection of 20% mannitol (1 g/kg) was administered to achieve intraoperative brain relaxation. The arterial blood gas analysis showed pH-7.35, PO_2-120, PCO_2-30.6, and HCO_3-21.6. The intraoperative course was uneventful. Total blood loss was 1400 mL, which was replaced with one unit of packed red blood cells. After completion of the surgery, the patient was shifted to the neurosurgery ICU for elective ventilation. He was extubated on postoperative day one, and the course for 2 weeks was uneventful on follow-up.

Discussion

The incidence of meningioma in patients with NF-2 is ~50%, with a lifetime risk of ~75%.[2,3] In conjunction with intracranial involvement, patients can also present with multiple schwannomas at various sites of the body. Intraoral involvement in NF-1 has been previously reported to complicate airway management.[4,5] However, similar difficulties due to airway involvement in patients with NF-2 (acoustic neurofibromatosis) is rare. Neurofibromas involving the cervical region, oral cavity, tongue, pharynx, aryepiglottic folds, or arytenoids can pose a dreaded challenge to the anesthesiologist. NF-2 can cause painless cervical vertebrae dislocation,

requiring a preoperative radiographic examination. Thus, a meticulous examination avoids spinal cord damage during laryngoscopy and tracheal intubation. The possible airway management plan in neurofibromatosis patients includes but is not limited to direct laryngoscopy with a bougie, awake FOI, video laryngoscope including Airtraq, Rendell–Baker–Soucek mask with the left molar approach. Finally, if other conventional methods fail, transtracheal jet ventilation, cricothyroidotomy, and tracheostomy can be lifesaving.[6] Awake FOI is the preferred technique of airway management in an anticipated difficult airway, but it can result in a transient rise in intracranial pressure (ICP) in patients with supratentorial tumors and can also be associated with a failed trial in NF-2 patients. In our case, preoperative airway evaluation was performed with videostroboscopy.

A difficult airway cart, including a fiberoptic bronchoscope, was kept ready on the day of surgery. An intravenous agent (for induction and maintenance of desired depth of anesthesia) was preferred over inhalational agents to avoid any rise in ICP. The trachea was intubated uneventfully. Intracranial pressure controlling measures were implemented for better intraoperative brain relaxation. After surgery, the patient was shifted to ICU in anticipation of difficult extubation due to unilateral vocal cord palsy following recurrent nerve involvement. On postoperative day 1, the patient was planned to extubate, where the difficult airway cart was also kept ready in view of the challenging airway. The patient was assessed for adequate tidal volume (5 mL/kg) with regular respiration of at least 14 cycles/min. A tracheal cuff leak test was performed to exclude any vocal cord edema. The patient was extubated on a tube exchanger. After extubation, the patient was placed in a propped-up position with supplemental oxygen with a venturi mask (0.6 FiO_2@ 15 L/min) and gradually weaned off the oxygen.

In conclusion, although airway involvement is rare in NF-2, a detailed airway evaluation should be performed using videostroboscopy or indirect laryngoscopy. A further radiological evaluation may be sought if any lesion is found.

Conflict of Interest
None declared.

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