

Airway management of a neurofibromatosis type 2 with multicompartmental tumours: a case report

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Abstract

Neurofibromatosis type 2 (NF2) is a rare autosomal dominant disorder. We report a case of a 52-year-old woman with underlying NF2 who was scheduled for excision of cervical neurofibromas. The patient had four nerve sheath tumours affecting different parts of the airway, namely, two cervical neurofibromas with spinal cord compression, a large retrosternal goitre extending into the anterior mediastinal compartment causing central airway obstruction, and a large left thoracic paravertebral tumour in the posterior mediastinal compartment. After risk stratification and multidisciplinary discussion, awake nasal fiberoptic intubation with a contingency plan of rigid bronchoscopy and jet ventilation was decided. The patient was intubated successfully with target-controlled infusion remifentanyl as the sole sedative and airway topicalization with local anaesthetic. The patient was ventilated in a prone position intraoperatively with no desaturation. The tumours were successfully removed and the patient was discharged well. Awake nasal fiberoptic intubation is the choice of management in difficult airways affected by multicompartmental tumours in a centre that is devoid of extracorporeal membrane oxygenation service.

Keywords: airway management, central airway obstruction, mediastinal mass, neurofibromatosis type 2

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Introduction

Neurofibromatosis (NF) is an inherited autosomal dominant neurocutaneous disorder classified into three types: NF1, NF2, and schwannomatosis. NF1 comprises approximately 96% of all cases, while NF2 and schwannomatosis comprise 3% and < 1%, respectively.¹ NF1, also known as von Recklinghausen's disease, is associated with malignant tumours and vascular diseases that significantly contribute to the death of NF1 patients before the age of 40 years. Meanwhile, NF2 predisposes individuals to multiple nervous system tumours.¹ Central airway obstruction (CAO) is a pathological condition that leads to airflow limitation of the trachea, main stem bronchi, bronchus intermedius, or lobar bronchus which may result from intrinsic stenosis or extrinsic compression.² We describe a case of CAO in a NF2 patient who presented with complex multicompartamental tumours involving the airway. The patient required excision of the cervical neurofibromas in the prone position, which posed a considerable challenge in anaesthetic management.

Case presentation

A 52-year-old female was diagnosed with NF2 involving the cervical spine, thoracic spine, left elbow, and left lung for the past 13 years and was under neurosurgical team follow-up. The patient opted for conservative management as the tumours did not affect her daily activities. She had bilateral thyroid swelling but was clinically and biochemically euthyroid under conservative management. The patient's hypertension was well controlled with oral hydrochlorothiazide 25 mg once daily. Her body mass index was 25.8 kg/m² (weight 58 kg, height 150 cm). Due to progressive right upper limb weakness and bilateral lower limb numbness over the course of 1 month, she agreed to excision of the cervical neurofibromas.

Preoperatively, the patient was comfortable under room air with no obstructive symptoms. Her mouth opening was three fingers wide with a Mallampati score of class I. Range of motion of the neck and thyromental distance were normal. A diffuse neck swelling measuring 7.0 x 5.0 cm, larger on the right side with retrosternal extension and without bruit, was noted. There were no signs suggestive of superior vena cava compression. Indirect laryngoscopy performed by an otorhinolaryngologist in the clinic revealed normal vocal cords. Preoperative laboratory results including full blood count, renal and liver profile, thyroid function test, arterial blood gas, and electrocardiogram were normal. Chest radiography noted a large thyroid mass measuring 6.0 x 6.0 cm with tracheal deviation to the left (Fig. 1). Magnetic resonance imaging of the whole spine revealed four lesions: a lobulated right extradural lesion, extending from C3 to C5, measuring 2.7 x 1.8

x 3.6 cm with adjacent significant cord compression and narrowing of the spinal canal (Fig. 2a); a similar lobulated lesion in the left neural foramen extending into the central canal causing cord compression; a large heterogeneous lesion measuring 9.7 x 9.5 x 13.6 cm at the left thoracic paravertebral region (Fig. 2b, 2c); and a large right thyroid lesion with retrosternal extension measuring 4.2 x 6.6 x 6.8 cm causing CAO, shifting the trachea to the left with significant luminal narrowing of 4.0 mm at the level of C6 and compressing the left bronchus (Fig. 2d).

A comprehensive preoperative multidisciplinary team discussion was carried out among anaesthesiologists, otorhinolaryngologists, neurosurgeons, and radiologists. Awake nasal fibreoptic intubation was planned with an otorhinolaryngologist on standby in the operation theatre to initiate rigid bronchoscopy and jet ventilation if necessary. The airway management plan and risks of difficult airway were explained to the patient, followed by a written informed anaesthetic consent. The patient was fasted for 6 hours before being sent to the operation theatre. Her initial vital signs were stable, with a blood pressure of 115/70 mmHg, heart rate of 90 bpm, and oxygen saturation of 98% under room air. Intravenous (IV) glycopyrrolate 200 µg was given 30 minutes before induction as an anti-sialagogue. The right radial artery was cannulated under local anaesthesia and the patient was put on standard monitoring of blood pressure, electrocardiogram, and pulse oximeter.

The nasal cavity was packed with a 4% cocaine-soaked cotton applied to the left nostril for 10 minutes. It was followed by a combination of nebulised lignocaine 2% (5.0 ml) over 20 minutes and two puffs of lignocaine 10% sprayed onto the bilateral tonsillar pillars and uvula, respectively. The patient was subsequently preoxygenated with 100% oxygen for 5 minutes and oxygenation was continued with nasal prong 3 L/min. Target-controlled infusion (TCI) remifentanyl was initiated using the Minto model targeting the effect site at 0.5–1.5 ng/ml, followed by the spray-as-you-go technique during awake fibreoptic bronchoscopy. An aliquot of 2.0 ml lignocaine 2% was sprayed directly onto the epiglottis, arytenoid, and glottis inlet sequentially. A total of 4.8 mg/kg lignocaine was used. No cough was elicited during tracheal intubation with a 6.5-mm armoured endotracheal tube.

General anaesthesia was induced and maintained with TCI propofol using the Marsh model targeting the plasma site at 3.0–4.5 µg/mL and TCI remifentanyl 2.0–4.0 ng/mL, guided by Bispectral index monitoring at a value between 40 and 60. The right femoral vein was cannulated with a 7-French triple-lumen catheter. Blood pressure and heart rate were maintained within 15% from baseline after induction and positioning. Saturation was maintained at 100% throughout the surgery. No muscle relaxant was given throughout the operation. The cervical tumours were removed successfully with an estimated blood loss of 300 ml. The

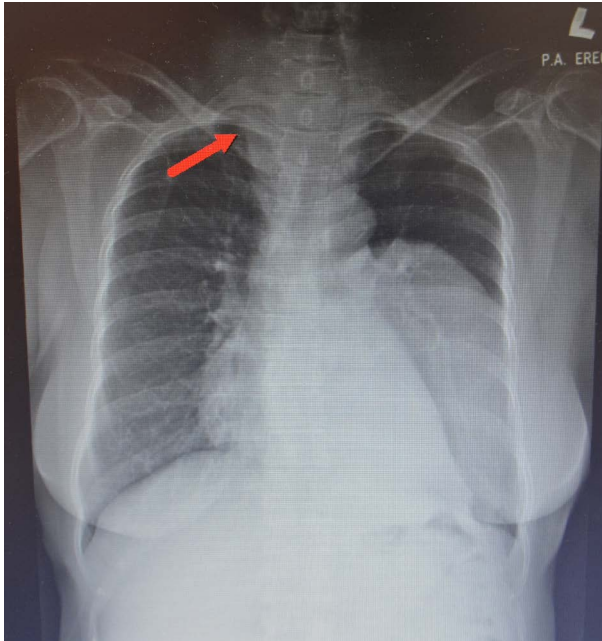


Fig. 1. Chest X-ray showing mediastinal widening with tracheal deviation to the right.



Fig. 2. Magnetic resonance imaging (MRI) of the spine. (a) Lobulated right extradural lesion at the right neural foramen with adjacent significant cord compression and narrowing of the spinal canal. (b) Anteroposterior and (c) lateral view of a large heterogenous lesion at the left thoracic paravertebral region. (d) Large right thyroid lesion with retrosternal extension and significant mass effect, shifting trachea to the left with significant luminal narrowing of 4 mm at level of C6 and compressing left bronchus

patient was mechanically ventilated overnight in the intensive care unit postoperatively. After a positive cuff leak test, the patient was extubated with no respiratory compromise. She was discharged well 3 days after the operation.

Discussion

The coexistence of cervical spine tumours with anterior and posterior mediastinal tumours is rare. This is the first case report involving these four tumours simultaneously to date. The first lesion involved two cervical neurofibromas with myelopathy that demanded minimal neck movement during airway management to prevent secondary spinal cord injury. The second tumour was a large, left, posterior mediastinal tumour that has been suggested to carry a low risk of anaesthetic complications. However, a case of large posterior mediastinal mass that required urgent transition to extracorporeal membrane oxygenation (ECMO) has been reported due to haemodynamic and respiratory decompensation upon induction of general anaesthesia.³ The last and most significant tumour was a large retrosternal goitre in the anterior mediastinum with airway compression that led to CAO and mandated spontaneous ventilation. This was to preserve the diaphragm in caudal position, thereby maintaining normal pleural pressure, keeping the airway dilated, and minimizing airway collapsibility due to airway compression by the mediastinal mass.⁴ In severe cases of CAO, ECMO has been used when there are doubts in maintaining oxygenation with either an endotracheal tube or a rigid bronchoscopy.⁵

Establishing an airway is challenging in patients with severe CAO as it may be distal and precludes the option of front of neck access.⁵ The decision for pre-emptive standby of ECMO as a salvage therapy can be difficult. Hence, it is useful to assign the patient a severity grade of safe, uncertain, or unsafe based on the classification of anaesthetic risks for mediastinal mass syndrome.⁶ Mediastinal mass syndrome denotes the clinical picture caused by a mediastinal mass in anaesthetized patients. It can occur at every stage of anaesthesia and spiral down into acute respiratory or hemodynamic compromise, or both.⁶ Management of adult patients is guided by the severity of symptoms and computerized tomography (CT) scan. Patients are deemed safe if they are asymptomatic and the CT scan shows tracheal/bronchial diameter > 50% of normal. Patients are deemed unsafe if severely symptomatic and the CT scan shows tracheal/bronchial diameter < 50% of normal. Patients are deemed uncertain if they are mildly or moderately symptomatic or asymptomatic, but the CT scan shows tracheal/bronchial diameter < 50% of normal. Grading of symptoms is further classified into mild, moderate, or severe. Patients graded as mild can lie supine with some cough/pressure sensation; those graded as moderate grade can lie supine for short periods but not indefinitely; and those graded as severe grade cannot tolerate a supine position.⁷

Our patient fell into the uncertain group as she was asymptomatic despite imaging showing compression of the tracheal diameter < 50% of normal by goitre. The recommended anaesthetic management for this risk group involves awake fibreoptic intubation, as it preserves spontaneous ventilation; determination of optimal positioning of patient during preinduction, as it will be the position that causes least compression; rigid bronchoscopy as rescue therapy; and monitoring for airway compromise postoperatively.⁷ Due to a lack of evidence from a similar case, the authors had planned the anaesthetic management based on this classification. Although the severity grading is intended to guide airway management for mediastinal mass, it has proven useful in managing multiple tumours affecting the airway, as happened in our case, which was treated in a centre devoid of ECMO.

In cases of critical airway obstruction, there is little evidence to suggest remifentanyl is superior to dexmedetomidine as the sole sedative agent for awake fibreoptic intubation.⁸ The option is based on individual experience and judgement on a case-by-case basis. Remifentanyl was chosen in our case as it has little effect on cognition, suppresses the airway reflex, and is easily titratable due to its unique pharmacokinetic characteristics.

Conclusion

Awake nasal fibreoptic intubation under monitored anaesthesia using a titrating dose of TCI remifentanyl 1–3 ng/mL and airway topicalization with local anaesthesia is the preferred mode of airway management for multicompartmental tumours. The classification of anaesthetic risks for mediastinal mass syndrome was useful in planning airway strategies for such a perplexing enigma.

Declarations

Informed consent for publication

The patient provided informed consent for the publication of the clinical data and images contained in this case report.

Competing interests

None to declare.

Funding

None to declare.

Acknowledgements

None to declare.

References

1. Tamura R. Current understanding of neurofibromatosis type 1, 2, and schwannomatosis. *Int J Mol Sci.* 2021;22(11):5850. <https://doi.org/10.3390/ijms22115850>
2. Guedes F, Branquinho MV, Sousa AC, Alvites RD, Bugalho A, Maurício AC. Central airway obstruction: is it time to move forward? *BMC Pulm Med.* 2022;22(1):68. <https://doi.org/10.1186/s12890-022-01862-x>
3. Anderson DM, Dimitrova GT, Awad H. Patient with posterior mediastinal mass requiring urgent cardiopulmonary bypass. *Anesthesiology.* 2011;114(6):1488-93. doi: <https://doi.org/10.1097/ALN.0b013e31821a8af1>
4. Datt V, Tempe DK. Airway management in patients with mediastinal masses. *Indian J Anaesth.* 2005;49(4):344-52.
5. Lin J, Frye L. The intersection of bronchoscopy and extracorporeal membrane oxygenation. *J Thorac Dis.* 2021;13(8):5176-82. <https://doi.org/10.21037/jtd-2019-ipicu-08>
6. Erdős G, Tzanova I. Perioperative anaesthetic management of mediastinal mass in adults. *Eur J Anaesthesiol.* 2009;26(8):627-32. <https://doi.org/10.1097/EJA.0b013e328324b7f8>
7. Hensley Jr, DE, Martin DE, Gravlee GP, editors. *A Practical Approach to Cardiac Anesthesia*, 5th edn, FA. Philadelphia, PA: Lippincott, Williams and Wilkins, 2013.
8. Johnston KD, Rai MR. Conscious sedation for awake fiberoptic intubation: a review of the literature. *Can J Anaesth.* 2013;60(6):584-99. <https://doi.org/10.1007/s12630-013-9915-9>