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Anaesthetic Management of a Patient with Neurofibromatosis and a Large Orbital Tumor: A Case Report

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Abstract

Neurofibromatosis is an autosomal dominant hereditary disease that is usually present for diverse surgical procedures, their management poses a challenge both to the surgeon and the anaesthetist because of its multisystemic affectation. Neurofibromatosis is associated with a variety of conditions often requiring anaesthesia for surgical treatment, including painful neurofibromas, severe kyphoscoliosis, pseudarthroses, hydrocephalus, intra-cranial tumors and other malignancies. The type and severity of systemic dysfunction must be considered while planning anaesthesia for a patient with neurofibromatosis.[1] This paper is a case report on the anaesthetic management of a patient with neurofibromatosis that had staged orbital tumor excision and parotidectomy.

Keywords: Neurofibromatosis, multisystemic, anaesthesia

INTRODUCTION

Neurofibromatoses are a group of hereditary diseases transmitted in an autosomal dominant fashion. They usually form tumors of ectoderm and mesoderm tissues, said to be the most common type of neurocutaneous syndrome.2 There are two main types based on clinical and genetic grounds, which include Type 1(NF1, Von Recklinghausen's, Peripheral Neurofibromatosis) and Type 2 (NF2, Bilateral Acoustic Neurofibromatosis, Central Neurofibromatosis). Neurofibromas are characteristic of the condition, found along all neurons paths, oropharynx, and larynx, which can cause difficulties with laryngoscopy. 3,4,5 Neurofibromatosis type 1 is caused by a mutation of the NF1 gene that results in a nonfunctional neurofibromin protein. Neurofibromatosis-1 affects most organ systems and present with complications of the central nervous, respiratory, cardiovascular, musculoskeletal, gastrointestinal and genitourinary systems. These systems all present with different considerations that pose challenges for anaesthesiologists.

CASE REPORT

A 36-year-old woman presented with increasing proptosis, craniofacial swelling, and difficulty breathing. Craniofacial swelling had been gradually increasing in size over 5 years, associated with an increase in the size of the eyes, more on the left than the right. She also had a recurrent history of blocked nostrils, heavy snoring, and difficulty in breathing when lying flat. She had no comorbidities, had a previous craniotomy 5years prior to presentation for brain tumor excision. There were no other significant findings in her history.

Examination revealed a young woman with facial asymmetry, bilateral proptosis which was greater on the left side. No neurological deficits, multiple nodules on the face, neck, upper half of the body, macroglossia with Mallampati of 3. The respiratory and cardiovascular systems were essentially normal. Pre- operative investigations were normal except for a low cortisol level, for which the patient was managed with prednisolone, and the cortisol level was repeated after a week, which was normal. High-risk anaesthesia consent was taken from the patient. Preparation for difficult intubation was made (MAC 5, Mc Coy and Videolaryngoscope were all available, bougie, and FONA

equipment). Venous access was with a 3-way femoral central line because of the multiple nodules in the upper part of the body.

Premedication given were Lidocaine(80mg), Dexamethasone(8mg), Tranexamic Acid(1g). Standard monitors were attached BP, Pulse oximeter, ECG, Temperature probe. She was pre-oxygenated for five minutes. Induced with Thiopentone (300mg), paralyzed with Suxamethonium (100mg), laryngoscopy was done using a MAC 5 aided with a bougie, a 7mm armoured ETT tube was railroaded into the trachea, and the placement was confirmed by auscultation and capnography.

Maintenance was TIVA using propofol (0.1 – 0.2 mg/kg/min) and oxygen. The analgesia used was Fentanyl and PCM. Paralysis was with Pancuronium (0.1mg/kg as an initial dose, then 1/3rd of the it for maintenance), 0.9% normal saline was used intra-operatively, and 2 pints of blood was given during surgery. There were two episodes of critical incidents intra-operatively: the first was bradycardia, the surgeons were notified, and this resolved spontaneously; the second was hypotension, which was managed with 10mg of Ephedrine. The surgery done was frontoorbitozygomatic craniotomy with tumor excision and maxillectomy.

The duration of surgery was 5hours 27minutes, while anaesthesia lasted 7hours 2minutes. She was extubated awake after surgery, transferred to HDU, moved to the open ward 2nd day post op, and was discharged home 6days after surgery. Patient re- presented 6weeks later for parotidectomy. There was no critical incident, and she was discharged 3 days after surgery.

DISCUSSION

Neurofibromatosis type 1 (NF-1) is a complex multisystemic genetic disorder associated with the mutation of a gene on chromosome 17. The changes in the neurofibromin protein produce multiple effects on ectodermal and mesodermal tissue, with the formation of tumors primarily in the nervous system and the skin.

Clinical manifestations are extremely variable, even among patients with the same genotype.3,4,5Therefore, a thorough evaluation of anticipated complications must be done when planning anaesthesia. The examination must include airway assessment, respiratory, cardiovascular, and neurological system examination and rule out vertebral anomalies. This case highlights the importance of careful assessment and pre-emptive management of neurofibromatosis, as we can encounter them at a point in time in our anaesthesia practice. In neurofibromatosis, macroglossia, abnormal formations in the tongue, presence of plexiform fibromas in the pharynx, larynx, and supraglottic region can prevent endotracheal intubation and cause upper airway obstruction during anesthetic induction. presence of macroglossia, macrocephaly, mandibular abnormalities and cervical spine involvement may contribute to difficulties in airway management.2 For these presentations, history of dysphagia, dysarthria, presence of stridor, and voice changes should be questioned in the patients prior to anaesthesia. 6,7 The patient had an history of stridor however, she had no history of dyspnea, macroglossia was also noted. A difficult airway was anticipated in this patient, therefore a MAC 5 laryngoscope was used, and the endotracheal tube railroaded with a bougie into the trachea. The majority of the patients with neurofibromatosis present with cranial or spinal tumors, this accounts for a major portion of the morbidity and mortality in them.1,2 Hypertension due to renal artery stenosis or occult phaeochromocytoma may be found in patients with neurofibromatosis, though this patient was not a known hypertensive. 4,7 During the anaesthesia rather than hypertension, we had hypotensive episodes due to the proximity of the tumor to the orbit which triggered occulocardiac reflex during the intra-cranial dissection by the surgeons. This was abated by immediately alerting the surgeons of the haemodynamic changes observed, which further emphasises the importance of vigilance in the monitoring of patients. The anaesthesia management of patients with neurofibromatosis is rare in our practice, however proper peri-operative evaluation, preparation, and vigilance to details will give the optimum outcome in these patients.

CONCLUSION

Anaesthesia in the presence of neurofibromatosis may be hazardous due to associated conditions and widespread organ involvement. Their diverse presentations present as a challenge for both anaesthetists and surgeons. The management of patients with neurofibromatosis in a resource-challenged environment like ours may pose difficulties for anaesthetists; however, adequate preoperative assessment, preparation, and vigilance during surgery will ensure a good perioperative outcome for the patient.

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