

Anaesthesia Implications in Neurofibromatosis Case for Emergency Cesarean Section

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Abstract: *Neurofibromatosis type 1 is a rare neurocutaneous condition that can have widespread and deleterious effects on various organ systems, including the spine with implications for the choice of anesthetic technique. Neurofibromas are the characteristic lesions of the condition and not only occur in the neuraxis but may also be found in the oropharynx and larynx; these may produce difficulties with laryngoscopy and tracheal intubation. We report the case of a parturient, who was received in the emergency room of the maternity ward of our hospital during the active phase of labor, without neuraxial imaging, in whom a cesarean section was indicated and performed under general anesthesia. Anesthesia for the parturient with neurofibromatosis is still not well documented, its management needs to be multidisciplinary, with a careful pre-conception planning.*

Keywords: Neurofibromatosis, Anaesthesia

1. Introduction

Neurofibromatosis is an autosomal dominant disease and is divided into two groups: neurofibromatosis type-1 (NF-1) and neurofibromatosis type-2 (NF-2). Type 1 neurofibromatosis, also known as von Recklinghausen disease, is the most common type and is characterized by café-au-lait spots and benign skin neurofibromas. Type 2 neurofibromatosis affects the central nervous system due to spinal cord tumors and bilateral vestibular schwannomas. In NF-1, neurofibromas in the tongue, pharynx, and larynx can prevent intubation by making the airway more constricted. For this reason, in pregnant women with NF1, difficult airway has been the main cause of anesthesia-related deaths. In these cases, it is important for anesthesiologists to perform airway examinations carefully. Increasing the risk of bleeding while applying regional anesthesia, there is an increased risk of hematoma and intracranial pressure. However, successful computer spinal anesthesia has been reported in patients by excluding the presence of spinal neurofibroma by brain computer tomography (CT) and magnetic resonance imaging (MRI). Neurofibromas are the characteristic lesions of the condition and not only occur in the neuraxis but may also be found in the oropharynx and larynx; these may produce difficulties with laryngoscopy and tracheal intubation. Pulmonary pathology includes pulmonary fibrosis and cystic lung disease. The cardiovascular manifestations of NF1 include hypertension, which may be associated with pheochromocytoma or renal artery stenosis. Neurofibromas may also affect the gastrointestinal tract and carcinoid tumours may be found in the duodenum. This review documents the aetiology and clinical manifestations of the neurofibromatoses and discusses their relevance to the anaesthetist.



Figure 1: NF lesion over spine

2. Case Study

36yr Female G4 P3 L1 IUFD2 35.3 wks by date, with breech presentation with uterine and umbilical artery Doppler changes with IUGR with neurofibromatosis proposed for emergency LSCS

K/c/o neurofibromatosis since age of 10yrs , aggravated since 2 yrs , not on any medication.

No h/o stridor/ dyspnea on supine position, no h/o wheeze / adventitious sounds, No h/o chest pain/palpitations , syncope

No h/o facial weakness, sensory / motor weakness in upper and lower limbs

No h/o constipation/diarrohoea, wt loss or wt gain

Recently diagnosed with hypothyroidism on tab thyronorm 25 mcg od since 15 days.

H/o haemorrhoids surgery ? SAB 8 yrs back

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General condition of patient conscious oriented afebrile, Pr 88bpm, Bp 120/80mmhg, Spo2 99% on room air

Spine – neurofibromas all over the spine and body

Ophthalmological evaluation suggestive of Lisch nodules present, fundus- WNL

Plan of anaesthesia -

Indirect laryngoscopy to rule out neurofibromas in supraglottic or glottic region,
Two wide bore IV cannulas

General anaesthesia as there were neurofibromas all over the spine and MRI of brain and spine wasn't available to know if there are neurofibromas in epidural or subarachnoid space

For intubation Conventional laryngoscope >VLScope with frova> tracheostomy standby >needle cricothyroidotomy

Nbm status was confirmed, Standard ASA monitors were attached, vitals checked and noted , PR 108 BPM , bp 130/80mmHg, in glycopyrrolate wasn't given as HR was above 108bpm, Anaesthesia induced with Propofol 2-3mg/kg iv slowly in graded dosage and injscoline 1.5mg /kg iv , induction dose of atracurium was given after intubation

Intubation by RSI technique

Attempt of VLS with FROVA failed >> Mac 3 blade was used, PCETT no. 6.5 inserted, tube fixed at mark 18 , AE checked , AEBE clear

Anaesthesia was maintained on O2 + air+ intermittent propofol on closed circuit with controlled mechanical ventilation

3. Conclusion

Anesthesia for the parturient with neurofibromatosis is not well documented. General anesthesia is currently the technique of choice if spinal status is unknown, but other alternatives are being evaluated. When deciding on the anesthesia method to be applied in a pregnant patient with neurofibromatosis, we should thoroughly evaluate the patient preoperatively, perform the physical examination completely, and use imaging methods if possible. Necessary precautions should be taken by taking into consideration the possibility of difficult airway, problems encountered in regional anesthesia, and multisystem complications that may develop.

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