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Anaesthetic Considerations in a Patient with Neurofibromatosis

Punam Raghove[1], Karampal Singh[1], Jatin Lal[1], Susheela Taxak[2], Geeta Ahlawat[1]
1.Assistant Professor, 2. Senior Professor
Dept. of Anaesthesiology, Pt.BDS PGIMS Rohtak
Corresponding Author: Karampal Singh (Karampal.d@rediffmail.com) Phone: 919050077120

About the Author: Dr Punam Raghove is working as assistant professor in Pt. BD Sharma PGIMS Rohtak. She did her post-graduation in Anaesthesiology & Critical care from Pt. BD Sharma PGIMS Rohtak and senior residency from JPN Apex Trauma Center, AIIMS, New Delhi.

Abstract: The neurofibromatosis is an autosomal dominant disease that has widespread effects on ectodermal and mesodermal tissue. The commonest member of the group is neurofibromatosis type 1 (NF1) which varies in severity but which can affect all physiological systems. Anaesthetic management of a forty five years old female patient with neurofibromatosis is discussed

Key Words: Neurofibromatosis, Von Recklinghausen disease, spinal, Perianal abscess

The neurofibromatosis is a group of hereditary diseases transmitted in an autosomal dominant fashion and are characterized by a tendency to formation of tumours of ectodermal and mesodermal tissues. They represent the most common example of the neurocutaneous syndromes. Two distinct forms have been recognized on clinical and genetic grounds. These are designated neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2). Neurofibromatosis 1 (NF1), also known as Von Recklinghausen disease, is characterized by multiple cafe-au-lait spots in the skin, multiple peripheral nerve tumours, and a variety of other dysplastic abnormalities of the skin, nervous system, bones, endocrine organs, and blood vessels.[1] The birth incidence of NF1 lies between 1 in 2500–3300 and its prevalence in the population is 1 in 5000[2].

Case Report: A 45 years old female came to emergency department with painful perianal abscess. She was scheduled for emergency incision drainage of abscess. On physical examination she was noted to have various epidemic, indolent neurofibromatosis nodules of different sizes over his entire body. Oral examination revealed lesions on her tongue. Suspecting deeper involvement of airways, we got indirect laryngoscopy examination of patient and nodular involvement of aryepiglottic fold was seen. Apart from neurofibromatic lesions over body rest of her physical examination was normal. She was 62 kg and 165 cm in height. Her blood pressure and pulse rate was WNL. She initially noted these neurofibromatic nodules over her body when she was fourteen years old and they were increasing in size and number since then. Her mother was also having similar lesion. She denied any other medical problem. Her lab investigations like Hb, BT, CT, blood Urea, blood Sugar,

and urine examination were within normal limits and in the electrocardiographic examination, an incomplete right branch block was found.

Considering the possibility of difficulty in airway management and patient wishes, it was decided to conduct the case under spinal anaesthesia. On arrival in the operating room intravenous line was secured and standard monitoring (ECG, SPO_2, automated blood pressure) was instituted. After administration of 500 mL lactated Ringer's solution; spinal anaesthesia was successfully performed in the left lateral position in L3-4 intervertebral spaces with a 26-gauge atraumatic spinal needle with 2 mL 0.5% hyperbaric bupivacaine. Position was changed to sitting once pain intensity decreased. After 5 min, level of anaesthesia was checked and surgery was started after 10 min.

The duration of the surgery was 30 minutes. All hemodynamic parameters were stable during surgery. After two hour observation in recovery room, patient was transferred to the surgical floor without any complications observed. No postoperative complications were observed and she was discharged from the hospital on the 3rd post-operative day.

Discussion: The neurofibromatosis is an autosomal dominant disease that has widespread effects on ectodermal and mesodermal tissue. The commonest member of the group is neurofibromatosis type 1 (NF1) which varies in severity but which can affect all physiological systems. 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. An estimated 5% of patients with NF1 have an intra-oral manifestation of the disease.[3] Discrete neurofibromas may involve the tongue[4] or the larynx.[5,6] When the latter is involved it is often the aryepiglottic fold or arytenoids that are affected, presumably reflecting those areas most rich in terminal nerve plexuses.[7] Airway obstruction after induction of anaesthesia has been reported in patients with a tongue neurofibroma[8] and a neurofibroma involving the laryngeal inlet.[9] Painless dislocation of cervical vertebrae also has been reported in a patient with multiple cervical neurofibromas and radiographic examination of the neck should be obtained before administering anaesthesia in these patients in order to avoid spinal cord damage during laryngoscopy and tracheal intubation.[10] Pulmonary pathology includes pulmonary fibrosis and cystic lung disease. The cardiovascular manifestations of NF1 include hypertension, which may be associated with phaeochromocytoma or renal artery stenosis.

Tumours of the central nervous system (CNS) account for the major portion of the morbidity and mortality of patients with neurofibromatosis. These patients have increased incidence of epilepsy, learning difficulties and possibility of undiagnosed CNS tumours. Involvement of brain stem structures by neurofibroma or glioma may result in central hypoventilation syndromes. Such patients may exhibit protracted weaning from mechanical ventilation postoperatively.[11]

The choice of anaesthetic technique in patients with neurofibromatosis deserves careful systemic evaluation. Factors influencing airway management, respiratory and cardiovascular problems, central nervous system involvement, and vertebral anomalies make the choice of either general or regional anaesthesia complex. For these patients who require surgery, general

anaesthesia has been the preferred method, as coexisting cranial or spinal neuromas might worsen the neurological status of even the asymptomatic patients during regional anaesthesia[12].

In our patient, we preferred spinal anaesthesia as neurofibromas were present on tongue and larynx and there was possibility of difficult airways because of this. We got an x-ray of lumbar spine to rule out any spinal lesion though CT scan or MRI are better investigation tools to rule out spinal involvement. Additionally a 26-gauge atraumatic spinal needle was used in order to prevent cerebrospinal fluid leakage that might possibly deteriorate neurological status. Spinal block may be extremely difficult in a patient with neurofibromatosis, as kyphoscoliosis or neurofibromas close to the needle puncture site may limit the safety of the procedure.

Conclusion: The neurofibromatosis is a group of conditions that vary in their severity but which have fundamental implications for the anaesthetist. NF1 is one of the most common genetically transmitted diseases and anaesthetists with a general practice are likely to encounter patients with the condition. Although the manifestations of NF1 are often mild, there may be associated pathology of direct relevance and importance to the anaesthetic management of patients with the disease. The anaesthetic management of patients with neurofibromatosis requires attention to all possible abnormalities and associated disturbances. Spinal anaesthesia may be a safe practice in thoroughly evaluated patients with neurofibromatosis. Special caution must be taken to avoid neurological squeal.

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