Anaesthetic Considerations in a Patient with Von Recklinghausen Neurofibromatosis

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Von Recklinghausen 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.[1] The type and severity of systemic dysfunction must be considered while planning anaesthesia for a patient with von Recklinghausen neurofibromatosis.

CASE REPORT

60 yrs old, 55 kg female with known case of von Recklinghausen neurofibromatosis (Fig 1) was scheduled for dynamic hip screw fixation of left femur and intramedullary interlocking nail for humerus. There was no history of previous surgeries and drug allergies. She was poorly built & systemic examination revealed a normal cardiovascular and respiratory systems. She had protruding upper incissors with adequate mouth opening, normal neck and temporo mandibular joint movements, Mallampati class-1 on oropharyngeal examination.

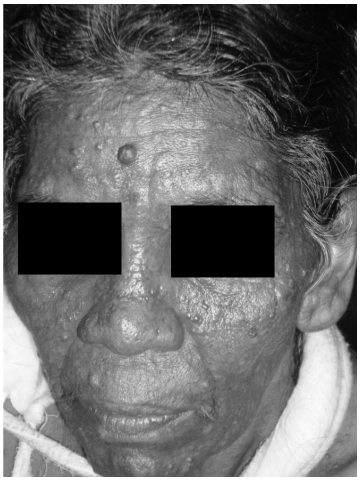
Pulmonary function tests revealed forced vital capacity (FVC of 1.5 lts/min), forced expiratory volume in first second (1.09lt/min) and FEV1/FVC of 91%. X-ray chest (PA), CT abdomen, CT scan of head & brain were normal and other blood investigations are within normal limits.

Regional anaesthesia was planned for both the surgical procedures. The anaesthetic procedures were explained to the patient and written informed consent obtained. She was advised oral Midazolam (10mg) at bed time on the night before and 5 mg on the morning of surgery following an overnight fast. In the operating room her heart rate was 88/min & blood pressure was 140/80 mmHg, oxygen saturation was 99%. Intravenous access with 18G canula was obtained. The patient was positioned for subarachnoid block in a sitting position. After preparation of back of the patient, appreciation of the tips of the spinous processes and interspinous spaces, sub-arachnoid space was located in L2-L3 space with midline approach. After free flow of cerebro spinal fluid, 3 ml of 0.5% Bupivacaine heavy was given into the sub-arachnoid space. The patient was put in supine position and oxygen supplementation was given with Hudsons mask at a flow rate 3 lts/min. After 5 min, level of anaesthesia was checked and patient is shifted on to the fracture table. The surgery lasted for 1½ hour during which 1500ml of crystalloids were given. The urine out put during the intra operative period was 900 ml. After the completion of hip surgery patient was shifted to another table.

The patient was explained about the procedure before giving brachial plexus block (25ml 1% lignocaine and 1 in 200000 Adrenaline) by supra clavicular route .After giving the block, the patient was monitored for 10 minutes and kept in supine position. The surgery lasted for 1 hr 20 min during which 500 ml of crystalloid & one unit of compatible blood was transfused. The urine output during the intra operative period was 600ml. Post operative period was uneventful and post-operative pain was managed with IV Tramadol hydrochloride (50 mg, 8 hourly)

DISCUSSION

Neurofibromatosis is autosomal dominant disease that have widespread effects on ectodermal & mesodermal tissue. The commonest member of the group is Neurofibromatosis type 1 (NF1) which varies in severity but affects 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



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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.[2]

Painless dislocation of cervical vertebrae has been reported in a patient with multiple cervical neurofibromas and it has been suggested that radiographic examination of the neck should be performed before administering anaesthesia in these patients in order to avoid spinal cord damage during laryngoscopy and tracheal intubation.[3]

Hypertension presenting in the young NF1 sufferer is usually because of renal artery stenosis, which may be bilateral.[4] The arterial lesions are of variable morphology with fusiform intimal narrowing or nodular or aneurysm formation.[5]

Tumors of the central nervous system (CNS) account for the major portion of the morbidity and mortality of patients with neurofibromatosis. Anaesthetic assessment of such patients should take into account the increased incidence of epilepsy, learning difficulties and possibility of undiagnosed CNS tumors.[2] Involvement of brain stem structures by neurofibroma or glioma may result in central hypoventilation syndromes. Such patients may exhibit protracted weaning from mechanical ventilation post-operatively.[6]

The genitourinary tract may be involved in NF1 and retroperitoneal neurofibromas may result in ureteric obstruction and hydronephrosis. Similarly out flow obstruction has been reported and bladder catheterization may be difficult.[7]

There have been many reports suggesting an increased sensitivity of patients with NF1 to non-depolarizing neuromuscular blocking drugs.[8,9]

This is especially pertinent in NF1 patients with renal impairment or those on concurrent medication (e.g. anticonvulsant drugs), which may interfere with the normal pharmacokinetics or pharmacodynamics of neuromuscular blocking drugs.[2]

In conclusion, the neurofibromatosis is a group of conditions that vary in their severity but which have fundamental implications for the anesthesiologists. Manifestations of neurofibromatosis are often mild; there may be associated pathology of direct relevance and importance to the anaesthetic management of patients with the disease. It is therefore important to have a working knowledge of the clinical manifestations of the disease, so that a systemic approach to the pre-operative assessment of these patients can result in rational perioperative management.

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