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CASE REPORT

UNEXPECTED FAILURE OF THE REGIONEL BLOCK IN NEUROFIBROMATOSIS: CASE REPORT

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ABSTRACT

Neurofibromatoses, mutation of the NF1 gene are autosomal disorders formed as neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and schwannomatosis. Neurofibromatosis type 1 is the most common of these disorders, seen 1 in 3500 individuals worldwide (1). Characteristic features of the disease; café au lait spots, neurofibromas, lisch nodules, macrocephaly, short stature, hypertelorism, thorax abnormalities, tumours of the central nervous system (optic and chiasm gliomas), orthopaedic abnormalities, endocrinological disorders, and malignancies (2). In this case report we describe the management of a patient with NF posted for urgent strongule herni with multiple neurofibromas common on his back and face causing difficulty both in regional anesthesia and general anaesthesia.

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INTRODUCTION

Neurofibramatozis type 1 was first described in 1882 by Frederich von Recklinghausen, is an autosomal disorder caused by mutation of the NF1 gene which results in a nonfunctional neurofibromin protein. Neurofibromatosis-1 exhibits complete penetrance, meaning 100% patients with the disorder have some phenotypic expression during their lifetimes. NF1 demonstrates variable expressivity, clinical features vary across individuals even with the same genotype. Preanesthetic evaluation of NF patients is of an extreme importance and anaesthesiologists must consider the multisystemic complications of this disorder at the stage the surgical approach to the patients planned. Anaesthesiologists must pay attention for the manifestations in the airways and neuroaxis to meet high performance degree (Charles *et al.,* 2012).

CASE REPORT

52 yrs old, 58 kg male patient was admitted to our emergency department with abdominal pain, vomiting and constipation that had started 72 h ago.

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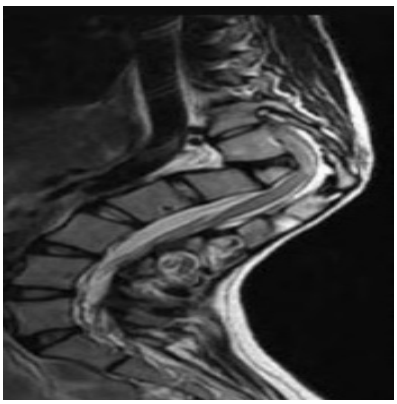
Strongulation of inguinal herni was diagnosed. Urgent surgery planned. The patient had previous history of Von R. Disease. In his history this disease started in childhood with the appearance of multiple hyperpigmented skin macules, at the age of 40 a lot of cutaneous tumors appeared and started to increase in size all over the body surface especially on face (Figüre 1). There was no history of previous any surgeries or drug allergies. He was poorly built systemic examination revealed a normal cardiovascular and respiratory systems. He had protruding upper incissors with inadequate mouth opening, limited neck and temporo mandibular joint movements. Mallampati classification 3 on oropharyngeal examination. He had a moderate kifosis. X-ray chest (PA), blood investigations are within normal limits. Deeper investigations such as neuroimaging couldn't be performed. Regional anaesthesia was planned for the surgical procedures. The anaesthetic procedures were explained to the patient and written informed consent obtained. In the operating room his heart rate was 88/min, blood pressure was 120/80 mmHg, oxygen saturation was 96%. 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 mask at a flow rate 2 lts/min. After 15 min, level of anaesthesia was checked. The patient had bilateral motor block of both legs but minimal sensory block of the L2-3-4 dermatomes on the right. With no improvement in the block by 30 min, we proceeded with general anaesthesia which was induced using a combination of a bolus of Remifentanyl (0.5 µg/kg) and Propofol 2 mg/kg then LMA was inserted. Anaesthesia was maintained with sevoflurane and 0.3 µg/kg/min remifentanil. The surgery lasted for 1½ hour without any problem and the patient was extubated then referred to the ward. Patient was discharged five days later uneventfully from the hospital. Postoperative magnetic resonance imaging of the spinal space demonstrated right intervertebral aracnoid cyst with L2 nerve root compression (figüre 2-3).

Figüre 1. Our patient



Figüre 2. Aracnoid cyst

Figüre 3. Aracnoid cyst

DISCUSSION

Anaesthesia for NF patients can be challenging as it has multisystem involvement and can occur along the airway like tongue, larynx for this reason needs special systemic evaluation. During anaesthetic management this may lead to distortion or difficulty even when ventilating the patient with the mask (Kishan Rao Bagam *et al.,* 2010). The premedication should focus on the airway. Even for painless cervical neurofibromas radiografic examination of neck may be performed as to avoid spinal cord damage. WC Crozier reported a patient with von Recklinghausen's neurofibromatosis who presented severe upper airway obstruction at the induction of anaesthesia and required emergency cricothyroidotomy because of a neurofibroma at the base of the tongue (Crozier, 1987). Mendonça and his friends reported the anaesthetic management of a 15 years old patient with NF1 for the tumor located in the occipitocervical region. Awake intubation was carried out with fiberoptic bronchoscopy (Mendonça *et al.,* 2016). Some authors suggested hypertension should always be investigated for to eliminate the aneurysms so that computerised angiography may be done. Increase in arterial blood pressure during laringoscopy must be prevented (Hirsch *et al.,* 2001). Subarachnoid block is also extremely difficult as neurofibromas close to the needle puncture site. Safety of the procedure can be limited (Hirsch *et al.,* 2001). Because of the neurofibromas involving the spinal cord epidural anaesthesia often considered contraindicated. The presence of kyphoscoliosis, scoliosis, spinal cord tumors, makes regional blocks difficult and sometimes impossible (Calvert, 1989). The tecniques like femoral or sciatic nerve block is usually considered safer. For many conditions, especially when neurological disease is involved, regional anaesthesia has been considered contraindicated because little known about the disease state or the effects of regional anaesthesia on it. Sun Ko and collegues reported patchy sensory blockade in a patient who had undergone three previous spinal operations. Scar tissue often leads to such failures (Sun, 1994). Shahid prefered femoral and sciatic block for lower limb surgery. The patient was 66-year-old with a history of traffic accident following which he developed an open, contaminated wound of size 15×5 cm over the dorsum of the right foot. He was scheduled for wound debridement. He had NF with multiple neurofibromas over the back which made spinal anesthesia impossible and neurofibromas over the face was causing difficulty in mask ventilation (Shahid Mohammed, 2015). MD Esler described a case of epidural analgesia in a parturient with neurofibromatosis (von Recklinghausen's disease) complicated by dural puncture and epidural haematoma was described (Esler *et al.,* 2001). In our case the failure of the spinal block was considered due to the cyct in the arachnoid space. Because of the limited neck movements and the expectations of the difficult ıntubatıon regional anaesthesia seemed much safer, but in the emergency conditions and the neuroimagings absense spinal disorders were missed. For general anaesthesia we didn't use muscle relaxants. Muscle relaxants may be of concern. The neuromusculer monitoring should be instituted when muscle relaxants are used. For this patient there were several important reasons to avoid the general anaesthesia such as the stiffness of the servical spine, protruding teeth, mouth opening 20 mm. Therefore spinal anaesthesia was performed but neurofibroma wasn't eliminated by a CT scan before unfortunately.

Conclusion

It is important to have a knowledge of the clinical features of the disease so that a systematic approach to the pre-operative assessment of these patients can result in succesful perioperative management. If recent radiological examinations can rule out neurofibromas at the planned insertion site, regional anaesthesia can be safely done.

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