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Anesthetic Management in a Patient with Von Recklinghausen Neurofibromatosis

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Abstract

Von Recklinghausen disease or neurofibromatosis Type I (NF1) is a rare genetic disorder with an autosomal dominant transmission with varieties of clinical manifestations. It can manifest as intracranial, spinal, ocular and cutaneous lesions. The lesions can extend to all the systems. We present the anesthetic management of a 46 years old male patient of Von Recklinghausen disease with pseudoarthrosis and bone dysplasia for debulking surgery of right lower limb under general anesthesia.

Keywords : RT fibuka, TLC, Laryngoscopy.

Case Report

46 years old, 60 kg male with known case of von Recklinghausen neurofibromatosis (Fig1) with history of fracture lower end of RT fibula 2 years back and swelling which has progressed gradually leading to difficulty in walking, scheduled for debulking surgery of right lower limb (Fig2). There was no history of seizures, hypertension, diabetes mellitus, stroke or any drug allergies. The patient underwent excisional biopsy of cutaneous Neurofibromas 3-4 years back. The findings of biopsy revealed the swelling to be benign Neurofibromas. He is of average built and systemic examination revealed a normal cardiovascular and respiratory systems. He had adequate mouth opening (>3 Fingers), normal neck and temporo mandibular joint movements & Mallampati class-3 on oropharyngeal examination & Cormack Lehane grade 2 on laryngoscopy.

Figure 1

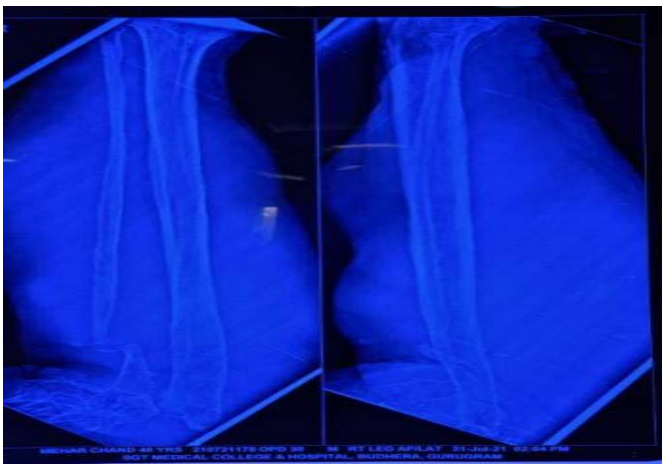

Figure 2

Investigations

ABG

pH (7.46)

PaO2 (84 mmHg)

PaCO2 (25.4 mmHg)

HCO3 (18 mmol/L)

Base excess/deficit (-6)

SaO2 (97%)

Sodium: 129 mmol/L

Potassium: 4.1 mmol/L

Calcium: 083 mmol/L

Hct: 21%

X-ray chest (PA) shows multiple rounds to oval radio opacities noted which was due to mesodermal lesions over the chest wall (Fig-3)

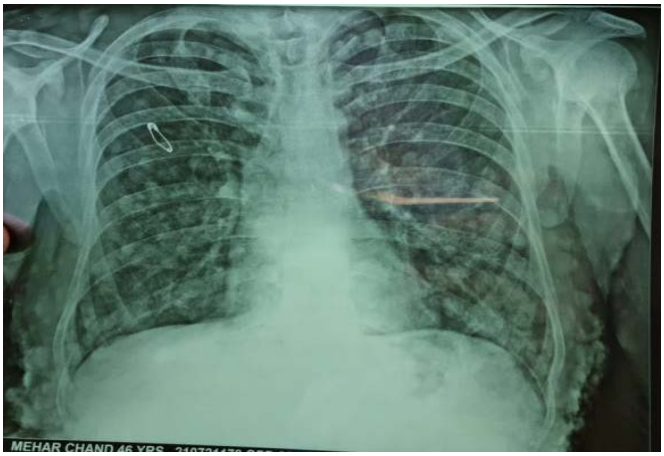

Figure 3

Blood investigations are as:

- HB-10 gm/dl
- TLC-5600 cells/cmm
- DLC :Neutrophils-61Lymphocytes-27 %

Eosinophils-06 % Monocytes-06% Basophils-00%

- PLATELETS COUNT -2,09,000/mm3
- RBS-95mg/dl
- KFT-Urea-38mg/dl,Creatinine-0.7mg/dl,Calcium-9.8mg/dl,

Sodium-140 mmol/L, potassium-5.0 mmol/L, Chloride-107mEq/L

- LFT- S.bilirubin (T)-0.3mg/dl

S.bilirubin Direct -0.1mg/dl

Sgot: 48.0H U/L

Sgpt: 59.0H U/L

Alkaline phosphatase-188.0H IU/L

- Urine-R/M- WNL
- [HBSAG, HIV, ANTIBODIES TO HCV] Non-Reactive
- Covid-19 RT-PCR : Negative

Anesthetic Management

The spinal anaesthesia could not be administered because the patient had not undergone MRI/PET screening for malignant peripheral nerve sheath tumor & multiples lesions over the back. (Fig-4)

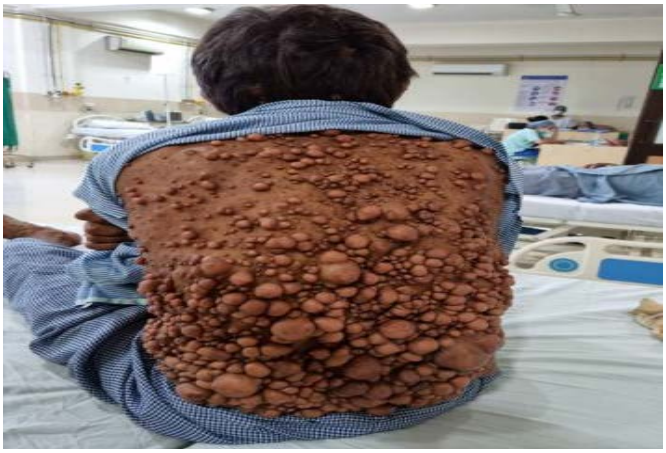

Figure 4

Therefore, General anesthesia was planned for this patient. Procedure was explained to the patient and written informed consent obtained. On the day of surgery after confirmation of NPO status he was shifted to the operating room. The patient was connected to multiparmeter monitor for monitoring of ECG, SPO2, NIBP and ETCO2. The anesthesia was administered as per standard protocol.

In the operating room his heart rate was 77/ min & blood pressure was 109/62 mmHg, oxygen saturation was 100%. Intravenous access with 20G cannula was obtained on left hand. The patient was positioned supine on the operating table.

A head ring was kept under the head to maintain sniffing position (flexion at the lower cervical and extension at the atlanto-occipital joint) He was premedicated with glycopyrrolate (5mcg/kg), ondansetron (0.1mg/kg), midazolam (0.03mg/kg) and fentanyl (1µg/kg) bodyweight intravenously. Then he was preoxygenated with mask for 3 minutes. General anaesthesia was induced with intravenous propofol (2mg/kg) and intravenous atracurium (0.5mg/kg) was given for neuromuscular block. After mask ventilation for 3 minutes with 60% N2O, 40% O2 and isoflurane (0.5%), laryngoscopy was done with McCoy blade. On performing laryngoscopy Cormack Lehane Grading-2 was noted then endotracheal tube size 7.5 was inserted & fixed at 20cm, bilateral air entry was checked after confirming the endotracheal tube then connected to ventilator for mechanical ventilation. Injection Atracurium top up (0.1mg/kg) was given accordingly. The surgery lasted for one and half hour. At the end of surgery throat suctioning was done, isoflurane & nitrous were discontinued. When respiratory efforts were present, residual neuromuscular blockade was reversed with Injection Neostigmine (0.05mgkg-1) & Glycopyrrolate (0.01mgkg-1). Recovery was assessed and extubation was done. Post-operative period was uneventful. Patient was shifted to post anaesthesia care unit (PACU) and observed for 2 hrs for nausea, vomiting, sore throat and Haemodynamic changes.

Discussion

Neurofibromatosis is autosomal dominant diseases 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; may these 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 [1]

There have been many reports suggesting an increased sensitivity of patients with NF1 to non-depolarizing neuromuscular blocking drugs [2, 3]

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 [1] Neurofibromas, the hallmark tumors of NF1, are benign tumors of the peripheral nerve sheath. They exhibit extensive cellular heterogeneity (Schwann cells, perineural cells, mast cells, fibroblasts and axons in an extracellular matrix)

and can be classified into cutaneous, subcutaneous and plexiform neurofibromas. Cutaneous and subcutaneous neurofibromas usually appear in late childhood or early adolescence (57- 99% of patients) while plexiform neurofibromas usually occur from birth to 18 years of age (20-30% of patients) [4]

In adults numerous cutaneous neurofibromas are usually present, but the total number varies from a few to many thousands. Their most typical morphology is one of a raised, sessile lesion of soft or elastic consistency, that is depressible on palpation (buttonholing) (Fig. 3). Solitary neurofibromas can be seen healthy individuals. Plexiform neurofibroma is a benign proliferation of the neural element of peripheral nerve that occurs particularly in the head and neck region and in deep body regions. Regardless of location, they can cause significant morbidity because of pain, disfigurement, local compression, and loss of function of nerves, great vessels, and airways. Although these tumors are usually benign, there is a 2-5% chance of malignant transformation in the setting of NF1 (malignant peripheral nerve sheath tumor; MPNST). The natural history of a plexiform neurofibroma varies, as some lesions remain asymptomatic superficial lesions, while others progress into large invasive disfiguring lesions as seen in our case [5-7]

Neurofibromin is a GTPase-activating protein that down-regulates p21-ras proto-oncogene. The loss of function of neurofibromin in NF1 patients leads to uncontrolled cell growth and increased tumor formation [8]

Conclusion

To summarize, the patient is a case of Neurofibromatosis 1 with bone dysplasia and pseudoarthrosis of right lower limb with no intracranial/optic extension, the asymptomatic patient undergone debulking surgery of right lower limb under general anaesthesia.

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