ORIGINAL RESEARCH PAPER

Anesthesiology

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ANAESTHETIC MANAGEMENT IN A CASE OF GIANT NEUROFIBROMA

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ABSTRACT

Neurofibromatosis is the most common neurocutaneous syndrome characterized by tumours of ectodermal and mesodermal tissues.
Neurofibromatosis type 1 (NF-1) is the most common type, with an incidence of 1 in 3000-4000 individuals.
Neurofibromas are characteristic lesions of the condition which occur mostly in the neuraxis, but maybe found anywhere. Neurofibromatosis can affect any system.
We are discussing a case of 23-year-old male who presented with a giant swelling over the back (plexiform neurofibroma) causing difficulty in walking and was scheduled for resection.
The anaesthetic management aimed at proper pre-operative evaluation to rule out other associated lesions or co-morbidities and airway assessment, proper positioning of the patient during intubation, strict intra-op input output monitoring, neuromuscular monitoring and pain management.

INTRODUCTION-

Neurofibromatosis is the most common neurocutaneous syndrome with autosomal dominant inheritance and has a tendency to form ectodermal and mesodermal tumors. It is of two types; neurofibromatosis type 1 (NF-1) and neurofibromatosis type 2 (NF-2). NF-1 is also known as von Recklinghausen's disease and is the most common type, with an incidence of 1 in 3000-4000 individuals.

Gene for NF1 is located on chromosome 17q11.2 which encodes a protein called neurofibromin which has tumour suppressor role. Mutations of NF1 gene results in decreased levels of neurofibromin causing tumours.

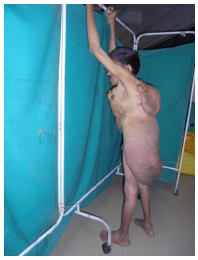
The anaesthetic management in a case of neurofibroma aims at proper pre-operative examination of airway, securing the possible difficult airway along with proper positioning of the patient, accurate assessment of blood loss with required replacement and providing adequate analgesia.

CASE REPORT -

We are discussing a case of a 23 years old male patient who presented with complaint of swelling over back since 5 years. The swelling gradually increased in size over the years causing difficulty in walking. Patient had a protruded chest wall since birth.

On examination patient was conscious & well oriented to time , place and person, moderately built, vitally stable, systemic examination was normal including central nervous system with normal muscle tone and power. Back & Spine examination showed Kyphoscoliosis of dorsal spine with convexity towards right side. Swelling was Soft, 52cm x 28 cm in size.

Airway examination revealed adequate mouth opening with Mallampatti grade 2 and normal neck flexion and extension.



All routine blood investigations along with ECG & 2D Echo were within normal limits. X Ray Chest showed Kyphoscoliotic changes of dorsal spine with convexity towards right side.

PFT showed Restrictive stage COPD as FEV_1/FVC >70% (102%) & FEV_1< 80% (55%) CECT THORAX showed Extreme kyphoscoliosis in thoraco-lumbar spine from D6 to D10 vertebral level with convexity towards right side. (Cobb's angle > 160 degree).

MRI DL SPINE showed Large neurofibroma in back extending from upper thoracic to sacrococcygeal region in subcutaneous plane with multiple small neurofibromas. Extreme kyphoscoliosis in thoraco-lumbar spine.

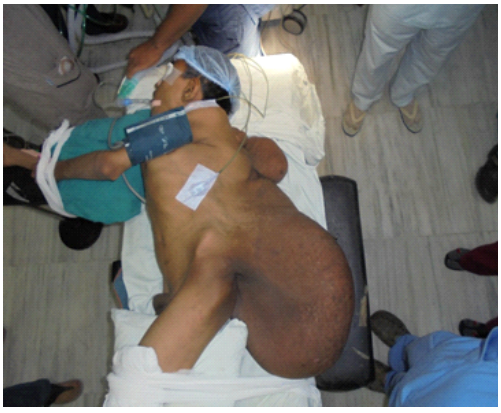
METHOD-

On the day before surgery, patient was kept NBM for 10 hours prior to surgery and explained about anaesthesia risk and Informed written consent of ASA grade IV with ventilatory support was taken, Two 18 G IV lines secured, Urinary catheter inserted, Adequate blood & blood products were kept ready, Difficult airway equipments with emergency drugs cart were kept ready. Routine monitoring including ECG, NIBP, IBP, SPO_2, $ETCO_2$, NMT monitoring, INPUT/OUTPUT, Temperature was done.

Patient was kept in supine position with two rings below the head to bring it in line with the body. The mass was placed in between two bolsters for intubation.

Patient was premediated with- Inj. Glycopyrrolate- 4ug/Kg IV ,Inj. Ondansetron 0.15mg/Kg, IV, Inj. Fentanyl 2 ug/Kg IV. Pre-oxygenation with 100% O_2 via Bains circuit @ 8-10 L/min for 5 mins was started . Induction of anaesthesia was done with Inj. Propofol 3mg/kg IV and check ventilation was done , followed by Inj. Scoline 2mg/kg IV.

Endotracheal tube No. 8.5 portex cuffed inserted, Bilateral air entry checked and confirmed with ETCO2 monitoring ,cuff inflated, tube fixed. Patient was then turned in Right lateral position for surgery. All pressure points were padded, both arms extended and placed on one side, head placed on the ring and ETT secured. All peripheral pulses were checked and the the patient was draped.



Patient was maintained with O2 + N2O+ Sevoflourane with Non depolarising muscle relaxant Inj Atracurium 0.5mg/kg IV loading dose, 0.1mg/kg IV incremental as per the requirement with the help of Train of four (TOF) monitoring. Intra-operatively 500ml DNS, 3000ml RL & 2 Packed red cells were given, Inj Nor – Adrenaline drip @ 100ml/hr (started intra-operatively when BP was 80/60 mmHg and was continued in post-operative period and gradually tapered as per BP control).

Intraoperative Urine output was 700ml and blood loss was 800ml approximately.

Analgesia was provided by Inj Diclofenac 1.5mg/kg IV and Local infiltration of 0.125% bupivacaine in 40ml volume along the margins of the wound before closure.

After the end of surgery, patient was reversed with Inj. Glycopyrrolate 8ug/kg IV and Inj. Neostigmine 0.05-0.07 mg/kg IV. After thorough oral and endotracheal suction, adequate consciousness and when the patient followed verbal command with adequate motor activity as per TOF ratio, patient was extubated. Duration of surgery was 6 hrs and of anesthesia was 6 hrs 45 min. Patient was shifted to post op recovery room and monitored. Nor adrenaline drip continued in post op period and gradually tapered off over next 24 hrs. 1 pint Packed red cell was given post operatively. Post-op histopathological report confirmed Plexiform neurofibroma. Post-operatively, patient was kept in propped up position with Oxygen through simple face mask. Patient was discharged after 7 days post op without complications.

DISCUSSION-

Neurofibromatosis ranges in severity from a benign disease to the one where multiple organ involvement occurs [1].

Diagnostic criteria for NF[2]- The patient should have two or more of the following-

Six or more café-au-lait spots (1.5 cm or larger in post-pubertal individuals, 0.5 cm or larger in pre-pubertal individuals) / Two or more neurofibromas of any type or one or more plexiform neurofibroma / Axillary or groin freckling / Optic glioma / Two or more Lisch nodules (benign melanotic iris hamartomas) / A distinctive bony lesion (Dysplasia of sphenoid bone, Dysplasia or thinning of long bone cortex) / A first degree relative with NF1.

Neurofibromatosis may involve any system therefore the following lesions should be ruled out in individual cases before taking the patient for surgery. Airway involvement due to neurofibromas present on tongue, pharynx or larynx may interfere with tracheal intubation . Patient may present with history of dysphagia, dysarthria, stridor or change of voice.[2]

Above finding require readiness to deal with difficult airway algorithm .

Respiratory system involvement due to Intrapulmonary neurofibroma & pulmonary fibrosis may occur. Patient, in such cases will present with dyspnoea & cough. Scoliosis /kyphosis is a common finding which may compromise lung function. Our patient was having kyphoscoliosis of thoracolumbar region ,we advised for PFT and ABGA preoperatively to assess severity .

Cardio-vascular system involvement in form of essential hypertension / pheochromocytoma/ renal artery thrombosis can occur. There are chances of major blood loss and fluid shifts in cases of huge neurofibroma resection, which require invasive blood pressure monitoring .

Central nervous system involvement include increased incidence of epilepsy, learning difficulties and the possibility of undiagnosed CNS tumours.

Gastro-Intestinal tumors may present with abdominal pain, haematemesis, malena. Carcinoid tumor in duodenum, may result in jaundice & carcinoid syndrome. Genito-urinary system Neurofibromas may cause ureteric / urethral obstruction and hydronephrosis.[3]

Vertebral deformities or spinal cord tumor may make spinal / epidural technique difficult. Neuraxial anaesthesia is contra-indicated in raised intracranial pressure, intraspinal lesions & kyphoscoliosis. It can only be given in cases which have very high risk for general anaesthesia, after ruling out spinal cord neurofibromas and intracranial involvement using CT/MRI.

PHARMACOLOGY-

There is Increased sensitivity to non-depolarising muscle relaxants, which necessitates neuromuscular monitoring. NMT monitoring is best guide for reversal and extubation as per TOF ratio. Sensitivity to depolarising muscle relaxants may be increased / decreased /normal. Inhalational agents and nitrous oxide should be used with caution in patients with large intracranial tumours and raised Intracranial Pressure.

CONCLUSION-

Proper preoperative planning with special investigations like 2D echo , PFT, indirect laryngoscopy, CT & MRI and ABGA along with intraoperative IBP & NMT monitoring , adequate blood transfusion and proper analgesia can lead to successful outcome in such giant neurofibroma.

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