Case Report

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Airway Management in a Case of Neurofibromatosis: Role of I Gel

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Abstract

Neurofibroma is a multi-systemic disorder which may produce considerable challenges in airway management by way of facial asymmetry, macroglossia, parapharyngeal tumors and neurofibromas of pharynx and larynx. Anaesthetic management may further be complicated due to concomitant kyphoscoliosis, sensitivity to neuromuscular blockers and hypertension. We report the case of a 50 year old female, known case of neurofibromatosis, who was posted for open reduction and internal fixation for fracture neck of femur. Patient was planned for General Anesthesia due to multiple neurofibromas over the back and kyphoscoliosis which precluded the use of spinal anesthesia.

Keywords: Neurofibromatosis; Multi system Involvement; I Gel.

Introduction

Neurofibroma is a benign peripheral nerve sheath tumour which arises from schwann cells and perineural fibroblasts. Two clinical types of neurofibromatosis have been described: peripheral-type I; and central- type II. Neurofibromatosis type I, also known as von Recklinghausen's disease, is more common, having widespread effects on ectodermal & mesodermal tissue [1]. NF 2 occurs less frequently than type 1 [2]. The combination of airway involvement in the disease, high likelihood of comorbidities such as hypertension, spinal deformities can make airway management challenging and at the same time necessary, as was observed in this case.

Case Description

A 50y old female, weighing 56kg, and a diagnosed case of Von Recklinghausen's Neurofibromatosis since 15 years, presented with severe pain and swelling in left hip with inability to move after she suffered a fall from height. Upon investigation she was diagnosed to have fracture of the neck of femur left side. Patient was initially managed conservatively and planned to undergo open reduction and internal fixation in elective setting. There was no evidence of abdominal, thoracic or head injury. There was history of surgery for swelling over the left leg 20 yrs back, under spinal anaesthesia, which was uneventful. However since the previous surgery there has been progressive increase in the number of cutaneous neurofibromas as well as the severity of kyphoscoliosis. Patient is a known case of hypertension and was taking 50mg of tablet Atenolol daily since 5 years. Rest of the history was unremarkable. Examination revealed multiple cutaneous neurofibromas present diffusely over her entire body as well as thoracolumbar kyphoscoliosis, Her vitals were stable preoperative with a recorded blood pressure of 150/90 mm Hg and heart rate of 60 bpm with 3-4 missed beats/ min. Patient was able to maintain oxygen saturation

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on room air, and although air entry was equal on both sides the left hemithorax was significantly reduced in volume due to kyphoscoliosis. Airway examination revealed that her upper incisors were protruding with adequate mouth opening, normal neck and temporomandibular joint movements, Mallampati class II. Blood investigations were within normal limits.

CXR revealed cardiomegaly and significant asymmetry with decreased volume of the left hemithorax. X ray spine revealed severe deviation of thoracolumbar spine. ECG showed bradycardia with occasional premature ventricular contractions.

Pt. was advised tablet Ranitidine 150mg at bed time on the night before and on the morning of surgery following an overnight fast and to take her morning dose of antihypertensive on the day of surgery. High risk was explained to the patient as well as the relatives and an informed written consent for likely postoperative ventilator support was also taken.

Patient's vital parameters were stable before anaesthesia. Difficult airway cart was kept ready before induction. Owing to the presence of thoracolumbar kyphoscoliosis and multiple cutaneous neurofibromas over the back of the patient and the unavailability of a spine MRI for undetected intramedullary lesions, general anaesthesia with I- gel as well as surgery in supine position was planned. Securing the airway of the patient was a challenge in itself due to the presence of kyphoscoliosis. Rolls of sheets were kept under the shoulder to support the upper back and head as well as a ring under the head to bring the oropharyngeal and laryngeal axis in one line on assuming the sniffing position. For pre-medication inj. Glycopyrrolate 0.005 mg/kg, inj. Ondansetron 0.08 mg/kg, inj. Morphine 0.6mg/kg were used. Inj.Propofol 2 mg/kg was administered and 2mg/kg succinylcholine was given as the relaxant. I-gel of size 3 was inserted and fixed after confirming equal, adequate ventilation and no leak. Anaesthesia was maintained with O2 + N2O + Sevoflurane +intermittent inj. Vecuronium. The patient was maintained on pressure controlled ventilation in view of the restrictive lung disease. The peak pressures throughout the surgery were maintained within normal limits Intraoperatively, her vitals were stable. Injection dexamethasone was administered in order to reduce airway oedema and inj.paracetamol 15mg/kg slow infusion for supplemental analgesia was given. Duration of surgery was 2 hrs, with blood loss of 450 ml and urine output of 120 ml. 1500 ml of crystalloids were infused throughout the procedure. At the end of surgery, anaesthesia was reversed by giving inj. Neostigmine 0.05 mg/kg and inj. Glycopyrrolate 0.010mg/kg. After awakening and spontaneous breathing, I-gel was taken out keeping difficult airway cart ready. Post-operative period was uneventful and her vitals remained stable throughout the post-operative observation period.

Fig. 1: Picture showing numerous neurofibromas over the back

Fig. 2: Severe thoracolumbar kyphoscoliosis seen in sitting position

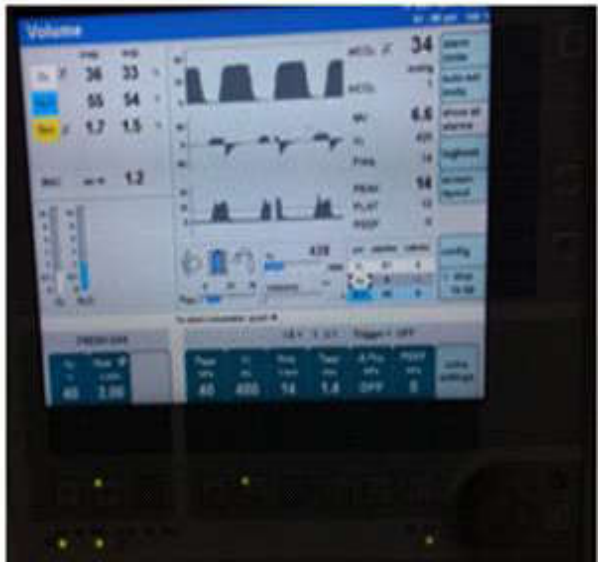

Fig. 3: Intraoperative ventilatory parameters with normal airway pressures

Fig. 4: Patient in supine position

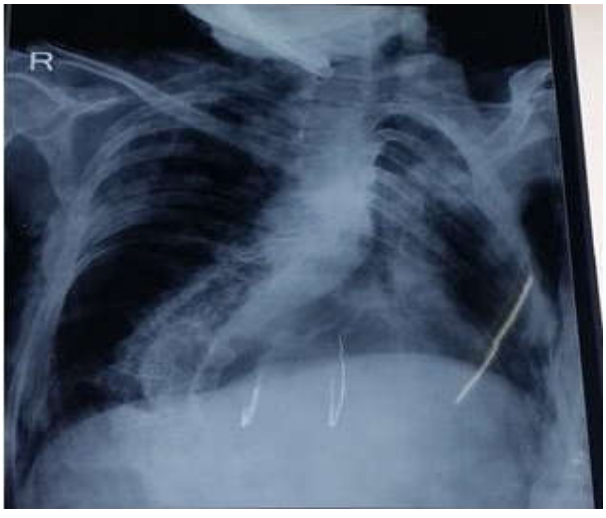

Fig. 5: Xray spine- indicating severe spine deviation

Discussion

Neurofibromatosis is an autosomal dominant disease with widespread effects on ectodermal and mesodermal tissue. The commonest member is Neurofibromatosis type 1 (NF1) which affects all physiological systems. Neurofibromas are the characteristic lesions of this disease and may occur anywhere including dermis, neuraxis, oropharynx and larynx [3]. NF offers a challenge to the anesthesiologist because of the variety of comorbidities in many organs and systems. Some abnormalities of interest for anesthetic-surgical procedures are short stature and bone abnormalities, in addition to cardiovascular abnormalities, such as congenital cardiac malformations, vasculopathy and hypertension. Cognitive disorders, in addition to attention and hyper activity disorders, can be found [4].

Because of the involvement of the CNS, regional anesthesia in NF 2 without careful preoperative examination can be extremely dangerous, and many anesthesiologists prefer general anesthesia. On the other hand, regional anesthesia could be useful in NF 1 because CNS involvement is rare. Additional masses in the tongue, pharynx and larynx in NF 1 may interfere with intubation during general anesthesia, although our patient had no such oral pathology. Endotracheal intubation can be avoided by the use of supraglottic airway devices in surgeries not specifically requiring it, hence minimising hemodynamic stress response to laryngoscopy and intubation as well as post operative pulmonary complications associated with restrictive lung disease.

We secured the airway of the patient with igel. It is a single use supraglottic airway device made of thermoplastic elastomer. It is easy to insert due to the tensile properties of its bowl and the ridge at its proximal end which prevents it from moving upward. I-gel also has the advantage of allowing venting of the air and gastric contents due to its gastric channel and prevent complications like obstruction associated with gastric neurofibromas.

Various Organ System Involvement of Neurofibromatosis is Discussed Briefly Below

Airway- Neurofibroma may occur in tongue, pharynx, larynx and may interfere with laryngoscopy and intubation. In the larynx they usually affect supraglottic structure, and are frequently described in arytenoids, aryepiglottic folds and posterior commissure [3,5]. About 80% of

these arise from false vocal cords and aryepiglottic folds. True vocal cords are rare site of location. It may be suspected by history of dysphagia, dysarthria, stridor or change of voice [5,6].

Respiratory system - Mediastinal neurofibromas may result in tracheobronchial compression with rapidly progressive symptoms. Bilateral upper lobe pulmonary fibrosis resulting in restrictive defect may result in pulmonary hypertension and RVF. There may be an intrapulmonary neurofibroma as well.These may be indicated by history of cough and dyspnea. Kyphosis and scoliosis may compromise pulmonary function [3].

Cardiovascular system – Patient may present with essential hypertension. Young patients are susceptible to renal artery stenosis. Sustained, paradoxical or hypertension resistant to treatment should raise the suspicion of pheochromocytoma [3,7].

During the intraoperative period, fluctuations in blood pressure or cardiac arrhythmia should raise the suspicion of a carcinoid tumor or pheochromocytoma. If not diagnosed before surgery, the patient can develop intraoperative hypertensive crisis after surgical manipulation or usage of triggering drugs such as betablockers and ketamine. Micronodular vascular proliferation may cause aortic, cerebral, coronary aneurysms. Neurofibromas may involve the heart causing both hypertrophy and mediastinal tumours may cause superior vena caval outflow obstruction [8,9].

Central nervous system- It is associated with increased incidence of epilepsy and undiagnosed CNS tumors [3]. Involvement of brain stem structures may result in central hypoventilation syndromes, hence may exhibit delayed weaning from mechanical ventilation post-operatively [7].

Gastrointestinal & genitourinary systems- Gastrointestinal neurofibromas may cause pain, obstruction, perforation, hemorrhage. Retroperitoneal neurofibromas may cause ureteric obstruction and hydronephrosis [3]. Musculoskeletal system – Painless dislocation of cervical vertebrae resulting in spinal cord damage during laryngoscopy and tracheal intubation may occur [10]. Spinal deformities may make spinal and epidural procedures difficult [3].

Conclusion

Neurofibromatosis is a group of conditions which vary in severity but have fundamental implications for anesthesiologists. It is hence essential to have a thorough knowledge of clinical manifestations of this disease, so as to adopt a systemic approach to the pre-operative assessment of these patients, aimed at better perioperative management and favourable outcome.

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