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Role of LMA Fastrach in Airway Management in a Case of Neurofibromatosis

Dr. Shaishabi Bhowmick[1]; Dr. Sarvjeet Kaur[2]; Dr. Rajvir Kaur[3]

[1,2,3] Department of Anaesthesiology, Gobind Singh Medical College, Faridkot 151203, India

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Abstract:- Neurofibromatosis is a recurrent multisystemic disease with a challenging airway management. Failure of the conventional techniques of airway management is seldom a complication but extremely problematic when it occurs. We hereby report a case of neurofibromatosis wherein a failed fiberoptic intubation led to a successful ventilation and subsequent intubation through LMA Fastrach.

Keywords:- *Neurofibromatosis, Difficult Airway, LMA Fastrach.*

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I. INTRODUCTION

Neurofibromatosis (NF) is an inherited autosomal dominant disorder caused by a mutation at neurofibromin gene (NF1). Loss of the normal tumour suppressive role of this gene leads to formation of various types of ectodermal and mesodermal tissue tumours. The incidence of NF1 lies between 1 in every 2500–3300 births.[1] Neurofibromas can cause skin disfiguration, blindness, nerve compressions and airway obstruction along with systemic involvements which poses different challenges to anaesthetist in terms of difficult airway and physiological changes. Role of supraglottic airways over gold standard awake fibreoptic intubation in such patients with difficult airway is epitome of our case report discussion.

II. CASE REPORT

A 45 yr old female patient, weighing 50 kg, previously known case of Neurofibromatosis 1 was posted for elective excision of plexiform neurofibroma of right side of face and right lateral thigh. The patient came with a chief complaint of visual distortion and facial disfigurement due to the mass involving the forehead, bridge of the nose, bilateral medial canthus, right side of face and also mid right lateral thigh(image). Patient's clinical history was unremarkable. On general physical examination, there were neurofibromas (including sizes >1.5cm) and café au lait macules at back.[1] Airway examination revealed Mallampati class 4 and mouth opening of equal to 2 fingers width, rest of the parameters including all routine investigations were within normal limits. Due to anticipated difficulty in both bag mask ventilation and laryngoscopy, the patient was planned for general anaesthesia with awake fiberoptic intubation.

Doing so also excluded any complication arising from an occult intratracheal lesion.[1] After explaining the procedure, written and informed consent was taken. The patient was nebulized with 4 ml of 4% lignocaine and 3 puffs of 10% lignocaine sprayed at posterior pharyngeal wall to anaesthetise oral and laryngeal mucosa. Standard ASA monitors were connected, peripheral venous access was secured and IV fluids started. Pre-operative vitals were within normal limits. Difficult airway cart was ready. Premedication was given with inj. glycopyrrolate 0.005 mg/kg i.v., inj. dexamethasone 0.1 mg/kg i.v. and inj. fentanyl 2mcg/kg. Patient was preoxygenated with 100% oxygen for 5 minutes. But due to poor tolerability of the fiberscope and extreme anxiousness of the patient, the procedure was abandoned. Titrated doses of inj propofol were given for induction. Keeping in mind the possibility for brainstem compression with direct laryngoscopy and succinylcholine induced sympathetic response these were avoided. LMA Fastrach no 3 was inserted and its tracheal tube was reinforced. After confirming its position by capnography, the ETT cuff was inflated and LMA was removed. Intraoperatively, there was an episode of hypotension because of massive blood loss (around 1-1.2 litres) which was managed by rushing 1.5 litres of i.v. crystalloids, 500 ml of colloid and a unit of packed red blood cells after cross match. The whole procedure lasted for 2 hours and 30 minutes. In the end, the patient was successfully reversed with inj. neostigmine 0.05 mg/kg i.v. and inj. glycopyrrolate 0.01 mg/kg i.v. and was extubated upon achieving good muscle tone.

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III. DISCUSSION

Neurofibromatosis 1 is an autosomal dominant disorder caused by heterozygous mutation on chromosome 17q11.2 in NF1 gene.[1] Mutation of this tumour suppressing gene increases the predilection for ectodermal and mesenchymal tumours. The neurofibromas fall into three distinct types: localised, diffuse and plexiform. The condition can have associated bony abnormalities, pheochromocytomas, carcinoid tumours, vertebral deformities.[1-4] As the involvement is multisystemic and widespread, it can give rise to multiple perioperative adverse events. A careful systemic evaluation is not only essential for choosing the appropriate anaesthetic technique but also to predict the anticipated adverse events.

In the presence of hydrocephalus, intracranial and spinal tumours, the use of regional anaesthesia has been discouraged. The tumours can also involve the oro-laryngo-pharyngeal axis making both laryngoscopy and intubation difficult. Even the presence of facial malformations, mandibular abnormalities, vertebral tumours and macrocephaly can complicate facemask ventilation and airway manipulations. [4-5] Thus making the appropriate choice of anaesthesia is debatable. The presence of history of dyspnea, stridor, voice change or dysphagia should alert the anaesthetist about a possible difficult airway.[3] Thus, preoperative indirect laryngoscopy is necessary to predict difficult intubation.

Since difficult airway predictors were affirmative, intubation under fiberoptic guidance was the technique of choice. [4-5] Since the surgical area involved the face securing a definitive airway was imperative. Following the conventional, we decided to opt for awake fiberoptic intubation but due to sheer intolerance of the patient we were pushed into a situation where supraglottic airway came to our rescue. Given the amount of work done on the use of LMA in NF and their favourable outcomes we decided to go ahead with using LMA Fastrach present in our difficult airway cart[6-7]. Intubating LMA have an upper hand over the conventional LMAs due to their preformed anatomically compliant structure along with the provision to secure a definitive airway along with adequate ventilation. In our case, since the distortion of oro pharyngo laryngeal axis was ruled out by IDL, taking a chance with LMA Fastrach was easier. As a backup plan, well lubricated PVC tubes of smaller sizes (6, 6.5mm) were kept handy incase the reusable silicone ETT of LMA Fastrach was difficult to pass through while encountering an intratracheal lesion. With quick decision making and promptness in our actions we were able to do a successful intubation with LMA Fastrach. ILMA can also be used in the first place for fiberoptic intubation in awake state in anticipated difficult airway. LMA Fastrach because of its reusability and better chances of intubation especially in obese pts comes across as a saviour in resource limited areas like ours. Its latex free nature, smoothness of insertion in neutral head position and lesser requirement of muscle relaxation adds to its feasibility.

Thus our case throws light on the utility of supraglottic airway device even when bag mask ventilation is difficult.

The extensive preoperative evaluation is not only recommended for precluding any airway complication but also to predict any perioperative complication.[1] Respiratory compromise can also result from neurofibromas affecting conducting airways, lung parenchyma, mediastinum and thoracic cage. Brainstem tumours can cause hypoventilation syndrome and postoperative protracted weaning from ventilator. Associated scoliosis, restrictive lung disease and cystic fibrosis can further affect effective ventilation.

Cardiovascular complications arising from underlying pheochromocytoma, renal artery stenosis, carcinoid tumours, hypertrophic cardiomyopathy, aortic and cerebral aneurysms and mediastinal tumours causing IVC or SVC compression remain one of the top reasons for unanticipated intra op adverse events.[1-2] Sustained or paradoxical hypertension and cardiac arrhythmia that is resistant to treatment should raise the suspicion of pheochromocytoma and carcinoid tumour.[8]

Gastrointestinal neurofibroma may cause obstruction, pain, perforation or haemorrhage as these tumours are highly vascular.[9]

Some cases have reported altered sensitivity to neuromuscular blockers giving rise to prolonged episodes of apnoea by unexplained mechanism.[10] So in these patients, neuromuscular monitoring should be done.

Hence, a careful preoperative evaluation based on thorough clinical examination and requisite investigations should be done prior to elective surgeries for favourable post-op outcomes.

IV. CONCLUSION

Supraglottic airway devices providing adequate ventilation can be saviour in situations with inadequate bag mask ventilation and later on can serve as a conduit for intubation as well. Our case throws light on the possible use of LMA Fastrach as a primary ventilation tool in difficult cases with difficult bag and mask and anticipated difficult intubation.

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