

Anesthetic Challenges in a Patient having Giant Neurofibromatosis of Thoracic Cage: A Case Report

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ABSTRACT

Neurofibromatosis type 1 (NF1) is an autosomal dominant neurocutaneous disorder with an incidence rate of 1 in 3,500 individuals at birth. The involvement of nasal, sinus or maxillofacial cavities creates airway challenges for anesthesiologists. During the perioperative period, anesthesiologists must be aware of the multisystemic complications of the disorder. Here we report a neglected case of giant NF with deformity of the anterior chest wall (pectus carinatum) and thoracic spine scheduled for tumor resection located at the posterior aspect of the thoracolumbar region. In this case, both anesthesiologists and surgeons faced unique challenges during the perioperative period.

Keywords: Anesthesia, Chest wall, Neurofibromatosis, Patient positioning.

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INTRODUCTION

Neurofibromatosis type 1 (NF1) is an autosomal dominant neurocutaneous disorder with an incidence rate of 1 in 3,500 individuals at birth.¹ The clinical manifestations vary across individuals with the same genotype. During the perioperative period, anesthesiologists must be aware of the multisystemic complications of the disorder. The involvement of nasal, sinus, or maxillofacial cavities creates airway challenges for anesthesiologists.² Thoracic lesions occur in about 10–20% of patients.³ Kyphoscoliosis, which can cause a reduction in lung volume and breathing capacity, may ultimately lead to respiratory compromise or failure.⁴ The preoperative evaluation of NF1 patients should include pulmonary function testing, indirect laryngoscopy, and computed tomography or magnetic resonance imaging to investigate for any respiratory complications and plan proper anesthesia techniques.³ Here, we report a case of giant NF with deformity of the anterior chest wall and thoracic spine scheduled for tumor resection located in the posterior thoracolumbar region. In this case, both anesthesiologists and surgeons faced unique challenges during patient positioning during the perioperative period. Written and informed consent for publication was taken from the patient.

CASE DESCRIPTION

A 42-year-old male patient with single swelling in the upper back since birth was later on diagnosed with a case of NF1. The patient lost follow-up after the age of 14–15 years of age. Secondary numerous swellings of the upper and lower limbs, with obvious bone deformities of the back and chest ribs, that worsen with age up to 25 years. After 30 years of age, swelling

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over the back grew steadily, almost to the size of a football, with a raw region forming over the swelling after 1 month (Fig. 1A). Chest X-ray showed kyphoscoliosis in the thoracic spine and pectus carinatum (pigeon chest deformity of the anterior thoracic wall with prominent sternum). Both site and size of the tumor, along with pectus carinatum deformity, in this case, caused difficulty in positioning the patient supine or prone (Figs 1B and C). Preoperative airway assessment showed a Mallampati score of 3, mouth opening two fingers, large tongue, short neck, thyromental distance >6 cm, normal teeth and temporomandibular joint and normal flexion, and extension cervical spine. Pulmonary function tests showed moderate to severe obstructive patterns [forced expiratory



Figs 1A to C: Neurofibromatosis with giant pectus carinatum and kyphoscoliosis deformity. (A) Erect posture; (B) Prone position; (C) Bag and mask ventilation in lateral position

volume in 1 second (FEV1)—63%, forced vital capacity (FVC)—80%, FEV1/FVC-79%], while other investigations are within normal limits.

After shifting the patient to the operating room, the left lateral position was maintained. A standard American Society of Anesthesiologists monitor was attached, and baseline vitals (heart rate, electrocardiography, pulse oximeter, and noninvasive blood pressure) were taken. All necessary equipment for difficult airway management, including the fibroscope, was kept ready. Two wide bore intravenous lines were secured, and infusion of lactated Ringer's solution started as maintenance fluid. Anesthesia was induced with propofol 2 mg/kg and fentanyl 2 µg/kg in the left lateral position, and after confirmation of bag and mask ventilation, an injection of vecuronium 0.08 mg/kg for paralysis was given. After induction of anesthesia and a minimum of 3 minutes of administering the paralytic agent, the patient underwent direct laryngoscopy in the left lateral position (Cormack Lehane grade 2). This was followed by bougie-assisted endotracheal intubation with a tube size 7.5 mm internal diameter. The operation was uneventful, and the patient's trachea was extubated in the left lateral position. The patient was then shifted to a postoperative ward with oxygen support by a face mask 6 L/minute in a lateral position. The histopathological report shows a malignant peripheral nerve sheath tumor from the neurofibroma in the thoracolumbar region.

DISCUSSION

Surgical conditions requiring anesthesia in NF1 patients include painful neurofibromas, severe kyphoscoliosis, pseudarthrosis, hydrocephalus, intracranial tumors, and other malignancies.³ Anesthesiologists must be aware of intraoral manifestations like discrete neurofibromas of the tongue, larynx, aryepiglottic fold or arytenoids, and of parapharyngeal space which may result in distortion of the airway.² Involvement of recurrent laryngeal nerve results in

vocal cord palsy.⁵ Even if intraoral pathology is recognized preoperatively, elective awake fiber-optic tracheal intubation may fail because of grossly distorted anatomy.⁶ Painless dislocation of cervical vertebrae during laryngoscopy has been reported in a patient with multiple cervical neurofibromas, and it has been suggested that a radiographic examination of the neck should be performed preoperatively.⁷ Response to muscle relaxants is variable, both sensitive and resistant to succinylcholine, but only sensitive to non-depolarizing muscle relaxants.⁸ Dystrophic spinal curvatures are short and sharp and progress throughout life. Severe kyphoscoliosis, although uncommon, may be associated with tumors and a high risk of neurological deficit. Scoliosis with rotation may also occur and produce a reduction in lung volume, which, if severe, may result in respiratory failure. Although pectus excavatum and carinatum occur in up to 30% of patients with NF1,³ they do not contribute to respiratory problems. In our case, there was no intra or extraoral mass. In our case, we used a non-depolarizing agent after confirmation of bag and mask ventilation. Our patients had severe degrees of kyphoscoliosis changes and pectus carinatum, which created difficulty in patient positioning during the perioperative period.

CONCLUSION

Neurofibromatosis type 1 (NF1) is a group of conditions that requires multisystem assessment during the perioperative period. The early corrective measure is required in a patient having the involvement of the cardiorespiratory system and in a patient having increasing progressive kyphoscoliosis and pectus carinatum changes. During the perioperative period, one should always be prepared for a difficult airway.

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