Case Report ISSN 2768-6647

Medical and Clinical Case Reports

Cesarean delivery in a Pregnant Woman with Neurofibromatosis Type 1 and Kyphoscoliosis: Anesthetic Challenges

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Received: 12 Dec 2023; **Accepted:** 15 Jan 2023; **Published:** 22 Jan 2024

Citation: Talita de Freitas Souza, Paul Holzmann Neto, Igor Alcântara Procópio, et al. Cesarean delivery in a Pregnant Woman with Neurofibromatosis Type 1 and Kyphoscoliosis: Anesthetic Challenges. Med Clin Case Rep. 2024; 4(1): 1-3.

ABSTRACT

Neurofibromatosis type 1 (NF1) is associated with the development of tumors in the nervous system, bone abnormalities, respiratory, cardiovascular, airway, and endocrine changes, among others, which interfere with anesthetic management. We present a case of a full-term pregnant woman who underwent elective cesarean section, diagnosed with NF1 and severe scoliosis. The chosen technique was general anesthesia. The management of these patients is highly challenging and requires attention to the possibility of complications, regardless of the type of anesthesia adopted.

This case report highlights the anesthesia management of a cesarian section of a patient with neurofibromatosis and scoliosis.

Keywords
Neurofibromatosis, Pregnancy, Anesthesia.

Introduction
Neurofibromatosis type 1, or Von Recklinghausen's disease, is a genetic disorder characterized by the development of tumors in peripheral nerves and the central nervous system, including the skin, bones, spinal cord, among other areas. Neurofibromatosis type 1 is associated with alterations in the NF1 gene, located on chromosome 17, and accounts for 96% of cases [1]. Additionally, the condition may be linked to structural and neoplastic changes in the spinal cord, including bone abnormalities such as kyphoscoliosis and alterations in vertebral bodies, dural ectasia, spinal tumors, nerve sheath tumors such as neurofibromas, intramedullary tumors, and more. Scoliosis is the most common bone alteration and can be either primary, due to bone dysplasia, or secondary to chronic compression [2].

Neurofibromas can also be found in the oropharynx and larynx, in addition to the possibility of pulmonary and cardiovascular pathologies. This includes cases where there is an association with pheochromocytoma or renal artery stenosis, as well as cases involving the gastrointestinal tract [3].

Case Report
A 22-year-old woman weighing 44 kg and measuring 1.33 m presented for elective cesarean section at 39 weeks of gestation. Her medical history revealed neurofibromatosis type 1 (NF1), severe kyphoscoliosis (Figure 1), and gestational hypothyroidism. The patient had no previous history of general or regional anesthesia. She denied any allergies. There was no reported history of neurological, respiratory, or cardiovascular symptoms. Upon airway assessment, no visible masses were noted in the tongue or pharynx. The patient demonstrated good cervical extension, adequate oral opening, Mallampati class 1, Upper Lip Bite Test class 1, and no airway deviations on physical examination. There were no imaging studies of the spine showing the presence of tumors.

Figure 1

Opted for balanced general anesthesia after monitoring and establishing peripheral venous access. The patient was adequately preoxygenated, and the anesthetic induction included propofol, remifentanil, and rocuronium. Orotracheal intubation was performed under videolaryngoscopy using a size 6.5 endotracheal tube. Neck extension was minimized to avoid cervical spine injury during laryngoscopy. Anesthetic maintenance included remifentanil and titrated sevoflurane to achieve a bispectral index between 40-60. Boluses of vasopressors were administered to maintain hemodynamic stability.

After fetal extraction, a titrated dose of oxytocin was administered through a slow infusion, avoiding hemodynamic adverse effects. At the end of the procedure, a transversus abdominis plane block was performed, and extubation took place in the operating room after neuromuscular blockade reversal with sugammadex.

Discussion

Neurofibromatosis is a complex condition with a wide range of manifestations, bearing significant implications for anesthetic practice, especially in obstetric patients as shown in Table 1. Choosing the anesthetic technique for pregnant individuals with spinal alterations and neurofibromatosis poses a considerable challenge. In such cases, it is crucial to carefully assess the risks and benefits of regional anesthesia techniques.

There are specific concerns related to spinal or epidural anesthesia due to the potential presence of spinal vascular tumors, in addition to reports of epidural hematomas in patients with neurofibromatosis [4]. Ideally, examinations such as magnetic resonance imaging (MRI) should be performed to assess the presence of tumors that could contraindicate spinal anesthesia. These blocks can be technically challenging due to changes in spinal curvature associated with neurofibromatosis.

Table 1: Challenges in anesthesia of patients with neurofibromatosis.

Airway and respiratory sistem	Potential airway changes due to the presence of neurofibromas; thoracic deformities with restrictive lung disease; pulmonary hypertension; tracheobronchial compression.
Cardiovascular	Hypertension; Hypertrophic cardiomyopathy.
Spinal/ Central nervous system	Changes on spinal curvature; Needle path tumors; spinal vascular tumors.
Gastrointestinal	Carcinoid tumours; Intestinal tumours.
Other systems	Kyphoscoliosis; pheochromocytoma; Renal artery stenosis.
Pharmacology	Variable sensitivity to succinylcholine; Exaggerated responses to non-depolarizing neuromuscular blockers.

Regarding general anesthesia, patients with scoliosis and neurofibromatosis present additional challenges, such as potential airway changes due to the presence of neurofibromas. Defects in the cervical spine can influence positioning for direct laryngoscopy and subsequent surgical procedures Additionally, kyphoscoliosis may lead to severe thoracic deformities, resulting in restrictive lung disease, pulmonary hypertension, cardiac or tracheobronchial compression, and heart failure [5]. It is important to note that patients with neurofibromatosis type 1 may exhibit variable sensitivity to succinylcholine, with reports of exaggerated responses to non-depolarizing neuromuscular blockers [6].

In this case, as there was an association of risk factors or difficulties with spinal anesthesia, such as short stature, kyphoscoliosis, presence of a large number of tumors at the puncture site, in a patient without respiratory or cardiovascular symptoms and with the absence of predictors of difficult airway, it was considered safer and more comfortable for the patient to undergo general anesthesia. Despite the lack of predictors of a difficult airway, videolaryngoscopy was deemed a safer option to avoid manipulation of the cervical spine. In the event of failure, recommendations for difficult airway management should be followed [7].

The decision regarding the anesthetic technique in these cases must always be individualized, considering the main risk factors in each patient and the experience of the professionals involved.

Conclusion

The choice of anesthetic technique in patients with neurofibromatosis and severe kyphoscoliosis should be approached with caution. It is important to exercise caution when considering regional analgesia in pregnant women with NF1. In summary, the anesthetic management of these patients requires an individualized approach and careful consideration of potential complications, regardless of the chosen anesthetic technique.

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