Subarachnoid Block a Safe Choice for Cesarean Section in Neurofibromatosis Patient with Severe Kyphoscoliosis

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Abstract

Neurofibromatosis type 1 (NF1) is an autosomal-dominant neuroectodermal hereditary disorder, in which spinal skeletal deformities are one of the manifestations. Pectus carinatum, temporomandibular joint dysfunction, and kyphoscoliosis can be seen with this genetic disorder which can lead to cardiorespiratory system problems and can cause difficulty in airway management. We present the anesthetic management of a NF1 patient with a sharp thoracolumbar dystrophic kyphoscoliosis who have been posted for emergency cesarean section as a result of fetal distress. The operation was successfully performed with subarachnoid block. This case highlights the anesthetic challenges and the significance of early planning anesthetic technique.

Keywords: Dystrophic kyphoscoliosis, neurofibromatosis type 1, subarachnoid block

Case Report

A 32-year-old NF1 primigravida with severe kyphoscoliosis, 36-week pregnant woman with 70 kg diagnosed as a case of cephalopelvic disproportion for emergency cesarean section with fetal distress. She had a full stomach status, taken a meal half an hour before surgery. During the physical examination, airway assessment revealed a mouth opening a fingerbreadth due to the temporomandibular joint dysfunction (TMD); therefore, Mallampati score could not be assessed. Examination of the spine showed a lateral curvature along with thoracolumbar kyphoscoliosis [Figure 1]. According to her complaint, she always had recurrent respiratory tract infection. Laboratory assessments were in normal limits. She was agitated and exhausted in pain. Considering the risks that may be caused by risk of failure in providing airway, we decided to perform subarachnoid block. Metoclopramide 10 mg and ranitidine 50 mg were administered intravenously before the surgery.

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The patient was monitored with electrocardiography, heart rate, noninvasive blood pressure (BP), and pulse oximetry (SpO₂). Spinal anesthesia was performed in the sitting position at the third attempt between lumbar 4 and 5 intervertebral spaces with a 27-gauge atraumatic spinal needle [Figure 2]. 2 ml 0.5% hyperbaric bupivacaine was used. Lactated Ringer’s solution of 1000 ml was administered. The operation was started at the 10th min. Her BP was 100/60 mmHg, SpO₂ was 99%, and pulse rate was 100 b.p.m. Nasale O₂ 5 L/min was administered during the surgery. Surgery lasted for an hour. A live female infant with 2.5 kg with an Apgar of 8/9 was delivered. Her vitals were stable throughout the procedure. After the surgery, the patient was monitored for an hour.

Postoperative period was uneventful. The remainder of her hospital stay was unremarkable and she was discharged home with no change from her preoperative status. The patient was discharged on the 3rd postoperative day.

**DISCUSSION**

The choice of the anesthetic technique and the management of anesthesia in patients with NF1 require careful preoperative evaluation. Factors influencing airway, cardiorespiratory problems, visceral system involvement, and vertebral anomalies make the choice between general and regional anesthesia challenging.

Kyphoscoliosis is associated with restrictive lung disease and hypoxemia which can lead to respiratory failure and cardiovascular compromise. Pregnancy can exacerbate the severity of symptoms.[4,5] One of the problems in this case was the proper position of the patient. The patient could not be placed in supine position because of the kyphosis, the surgery was performed with the pillows positioned beneath the patient.

Subarachnoid block was extremely difficult in this patient due to the NF1 as kyphoscoliosis or neurofibromas close to the puncture site of the needle. Neurofibromas here limited the safety of the procedure. We met technical difficulties. At the third attempt, we achieved subarachnoid anesthesia. Epidural anesthesia is often considered as contraindicated because neurofibromas may involve spinal cord and nerve roots.

NF1 patients have complicated airways. Difficult airway equipment including emergency tracheostomy should be available in the operating room. Regional anesthesia application was more appropriate for this patient, but we did not examine spinal magnetic resonance (MR) because of the fetal distress and the adverse effects of radiation. We did not want to take any risk associated with MR procedures.

In a case in Turkey, anesthetists performed spinal anesthesia in a 20-year-old male patient with NF1 due to the recurrent tumor on the dorsum of his left foot. They ruled out any spinal lesions by craniothoracolumbar MR imaging.[6] Arachnoid cyst can be the reason for the failure of regional anesthesia. Akdemir et al. could not achieve spinal block because of the arachnoid cyst and returned to general anesthesia in urgent hernia. We did not experience such a failure in our case.[7]

As our case, in another study, spinal anesthesia was performed in a critical patient with NF1. The patient had pectus carinatum and TMD. There was a high risk of general anesthesia, and spinal anesthesia was preferable like our patient.[8] TMD is a common symptom in NF1 patients and complicates airway management. For difficult airway and the possibility of the regional anesthesia failure, emergency tracheostomy should be available in the operating room. In consideration of the failure of block and a compulsion to turn back to general anesthesia, ear, nose, and throat specialist was ready in our operating room with the tracheostomy kit and sterile surgical tools.

In another case presentation, spinal anesthesia was applied to a patient who was scheduled dynamic hip screw fixation of the left femur. They did not prefer general anesthesia although the patient’s Mallampati class 1. Involvement of brainstem structures by neurofibroma can cause central hypoventilation syndromes. Such patients may exhibit protracted weaning from mechanical ventilation postoperatively.[9]

In all NF1 patients, complicated airways must be considered. The American Society of Anesthesiologists guidelines for difficult airway are advised.[10] Based on the existing pathology and its severity, preoperative early planning must be done.

As we reviewed the literature, NF1 patients who require surgery general anesthesia have been the preferred method as coexisting cranial and spinal neurofibromas can worsen...
the neurological status of even asymptomatic patients during regional anesthesia. Due to the pregnancy, we could not use MR. We decided to perform subarachnoid block in this NF1 patient with difficult airway.

**Conclusion**

Cesarean section in NF1 patients with severe kyphoscoliosis presents unique challenges. Anesthetic experience in these patients is limited. Careful evaluation preoperatively, best choice and management of anesthesia, and close postoperative monitoring are needed.

**Consent**

Written informed consent was obtained from the patient for the publication of this case report.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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