

## CASE REPORT

# Type-II Neurofibromatosis with Cervical Schwannoma Posted for Emergency Lower Segment Cesarean Section under Sole Epidural Technique

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## ABSTRACT

The prevalence of neurofibromatosis type-II is rare (1 in 33,000 people) worldwide with uniform gender distribution. Patients commonly present with vestibular schwannomas or any of the two among meningiomas, schwannomas, gliomas, cutaneous neurofibromas, posterior subcapsular lenticular cataracts, or cortical wedge cataracts. Pregnancy tends to promote the growth of such preexisting tumors and may reveal an untrodden tumor. This report describes a parturient who had neurofibromatosis type-II presented with cervical schwannoma posted for an emergency lower segment cesarean section (LSCS). General anesthesia is the preferred choice but owing to the restricted neck movements and high risk of autonomic dysreflexia in presence of cervical schwannoma, sole epidural anesthesia was performed successfully.

**Keywords:** Autonomic hyperreflexia, Cervical schwannoma, Epidural anesthesia, Type-II neurofibromatosis.

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## INTRODUCTION

Type-II neurofibromatosis is an autosomal dominant neurogenetic disorder described by the presence of schwannomas, meningiomas, ependymomas, neurofibromatosis lesions, and ocular abnormalities incidence being 1 in 33,000 people worldwide.<sup>1</sup> Fifty percent prevalence of this disorder is found to be hereditary. Cervical schwannomas account for 63% of all spinal schwannomas (1.1%). Pregnancy promotes the growth of preexisting intracranial tumors, and can accidentally reveal an untrodden tumor.<sup>2</sup> The defining feature of NF2 is the presence of bilateral vestibular schwannomas arising from the vestibular branch of the VIII cranial nerve or the presence of two or more meningiomas, gliomas, cutaneous neurofibromas, posterior subcapsular lenticular cataract or cortical wedge cataract.<sup>3</sup> Schwannomas are benign neoplasms with varied neurological phenotypes, including severe, intractable pain. The major concerns for anesthesiologist are the presence of macroglossia, macrocephaly, specific mandibular abnormalities, and cervical spine involvement contributing to difficulties in airway management. It is advised to order a radiographic examination of the neck preoperatively in these patients in order to avoid intraoperative and postoperative complications like spinal cord damage during laryngoscopy and tracheal intubation, aortocaval compression, quadriplegia, autonomic dysreflexia, Horner's syndrome. Aortocaval compression can be prevented with the left tilt of the patient by placing a wedge. Patients presenting with quadriparesis may worsen into quadriplegia due to sudden compression of the intradural tumor during positioning of the patient while intubating or accidental intradural puncture. However, in case of emergency situations, it is mandatory to wisely choose the mode of anesthesia based on the current scenario of the patient. Sudden or extreme neck extension may predispose to autonomic dysreflexia which manifests as an uncontrollable transient increase in blood pressure of 20 mm Hg or more. Onset is generally reported to occur most frequently during the chronic phase after spinal cord injury (3–6 months after injury).

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The choice of anesthesia was sole epidural since the subarachnoid block was contraindicated due to the intradural extension of the cervical lesion and general anesthesia also had its limitation due to restricted range of neck movements, difficult airway, and the possibility of autonomic hyperreflexia.

## CASE REPORT

A 32-year-old patient (weighing 62 kg, height 1.60 m, BMI—24.2 kg/m<sup>2</sup>) G3P2L2 who had previous two normal vaginal deliveries, both of which were uneventful, presented at 36 weeks of pregnancy on active labor with meconium-stained liquor posted for emergency lower segment cesarean section (LSCS). She is a known case of type-II neurofibromatosis diagnosed at 15 years of age. The family history revealed that her mother had a similar illness. The patient had cutaneous neurofibromas all over the body. **Figure 1** represents the cutaneous lesions of the patient. MRI brain and spinal cord revealed cervical schwannoma as an incidental finding. **Figure 2** represents the MRI of the patients' cervical region showing the cervical schwannoma. The patient was advised for surgical excision of cervical schwannoma which was denied by



Fig. 1: Cutaneous lesions of the patient



Fig. 2: MRI showing the cervical lesion

the patient. Preoperative examination revealed that the patient had restricted neck movements. The baseline parameters were found to be SpO<sub>2</sub>—99% at room air, BP—110/60 mm Hg, heart rate—92 bpm, without edema of the lower limbs. The patient had no focal neurological deficit and had muscle power of 4/5 on bilateral upper and lower limbs visual analog scale (VAS) score was 8/10. The obstetric examination revealed meconium-stained liquor with active uterine contractions.

The patient was shifted to the operating room in the left lateral position and all standard monitors were connected. Baseline vitals were recorded and found to be within normal limits. An 18 G intravenous cannula was secured in the left hand. Oxygen was delivered through Hudson mask at 5 L/minute. Sole-epidural was carried out in a sitting position with minimal neck flexion. At T12-L1 space with 18 G Tuohy needle, epidural space was identified at 3.5 cm, catheter was fixed at 9 cm at skin level. After negative aspiration 3 mL of test dose was given with 1% lignocaine, 0.25% bupivacaine, and 10 µg fentanyl followed by 7 mL of bolus dose with 1% lignocaine, 0.25% bupivacaine, and 10 µg fentanyl. Surgery commenced. Intraoperative hemodynamic status was stable throughout the procedure and the VAS score was 1–2. Adequate relaxation of the surgical field was achieved with surgeon satisfaction. A single live, female

baby was delivered which cried at birth with activity, pulse, grimace, appearance, respiration (APGAR) scores of 8/10 and 9/10 at 5 and 10 minutes, respectively. Postoperative analgesia was managed with continuous epidural infusion (0.125% bupivacaine at 5 mL/hour). The postoperative period was uneventful with VAS of 1–2 and hemodynamic parameters were stable.

## DISCUSSION

Type-II neurofibromatosis (NF-2) is a rare condition that involves various organ systems, including the spinal cord which plays a vital role in the choice of anesthetic technique preferred based on the clinical findings in various scenarios. In obstetrics, epidural analgesia is the most effective method of pain relief during labor neuraxial anesthesia in these patients, therefore, carries an increased risk of bleeding, hematoma formation, and increased intracranial pressure. But in our patient, since the tumor was found to be in the cervical region along with other conditions predisposing to difficult airway and also a high risk of autonomic hyperreflexia which may turn fatal to the patient, sole epidural anesthesia was preferred over general anesthesia and subarachnoid block (to prevent the rise in intracranial pressure).

Several cases of post-epidural hematoma have been described. However, neuraxial blocks can be safely performed with expertise and caution in such emergency situations with relative contraindications for general anesthesia. Providing effective analgesia to the parturient is a real challenge for the anesthesiologist in such cases. However, in the absence of spinal imaging, general anesthesia is the technique of choice.

## CONCLUSION

For a parturient with neurofibromatosis performing anesthesia is highly challenging. Due to its rare prevalence, not many such cases have been documented for gaining clear knowledge. General anesthesia is currently the technique of choice if the spinal status is unknown, but in scenarios like the abovementioned case, alternative techniques are to be evaluated precisely based on the need and safety of the patient. Each case should be assessed properly contemplating the uniqueness of each case scenario. Preoperative or recent neuroimaging of the brain and spinal cord is advisable for assessing the neurological involvement of the tumors. Anesthesia of choice for the parturient with neural involvement of neurofibromatosis is still not clear. Taking this case into consideration sole epidural analgesia was the best suitable method of choice in view of the clinical presentation of the patient.

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