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Case Report

Spinal anaesthesia for emergency cesarean section in neurofibromatosis for an antepartum haemorrhage: a case report

Gunmeet Kour[1]*, Shivani Sharma[1], Deeksha Sharma[1], Gurbir Singh[2]

[1]Department of Anesthesiology, Government Medical College, Jammu, Jammu and Kashmir, India
[2]Department of Surgery, Government Medical College, Jammu, Jammu and Kashmir, India

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***Correspondence:**
Dr. Gunmeet Kour,
E-mail: gunmeet8@gmail.com

ABSTRACT

Neurofibromatosis 1 (NF1) is an autosomal dominant disease and is usually diagnosed during childhood. Neurofibromatosis are group of genetic disorders that cause tumors to form, which can develop anywhere in the nervous system including brain, spinal cord and nerves. It poses a great challenge to the anaesthesiologist as it may have difficult airway, fibrosis in lungs and neuromas in spinal cord. Due to the involvement of CNS, regional anaesthesia in type 2 neurofibromatosis without careful preoperative examination can be extremely dangerous. Regional anaesthesia on the other hand could be useful in type 1 neurofibromatosis because CNS involvement is rare. This was a case report of management in a 24-year-old pregnant female with undiagnosed neurofibromatosis for an emergency cesarean section for antepartum haemorrhage and IUD under spinal anaesthesia. Although general anaesthesia is the safer option in neurofibromatosis patients, spinal anaesthesia can be a safe procedure in neurofibromatosis patients but special precautions should be taken to avoid neurological injury and uneventful anaesthesia.

Keywords: Neurofibromatosis, Anaethesia, Antepartum haemorrhage

INTRODUCTION

Neurofibromatosis 1 (NF1) is an autosomal dominant disease and is usually diagnosed during childhood.[1] Neurofibromatosis are group of genetic disorders that cause tumors to form on nerve tissue. These tumors can develop anywhere in the nervous system including the brain, spinal cord, and nerves. There are three types of neurofibromatis: NF1, neurofibromatosis 2 (NF2) and schwannomatosis. Type 1 neurofibromatosis, also known as von Recklinghausen disease is the most common and caused by a mutation in the NF1 gene located on chromosome 17.[2,3] This gene produces a protein called neurofibromin that helps regulate cell growth. It is characterized by café-au-lait spots and benign skin neurofibromas. Type 2 neurofibromatosis affects the central nervous system due to spinal cord tumors and bilateral vestibular schwannomas.[2-4] In NF1, neurofibromas in the tongue, pharynx, and larynx can prevent intubation by making the airway more constricted.[5] It poses a great challenge to the anesthesiologist as it may have a difficult airway, fibrosis in lungs, and neuromas in the spinal cord. Due to the involvement of CNS, regional anesthesia in type 2 neurofibromatosis without careful preoperative examination can be extremely dangerous and that's the reason many prefer general anaesthesia.[2] Regional anaesthesia on the other hand could be useful in type 1 neurofibromatosis because CNS involvement is rare.[5] The choice between general anesthesia or regional anaesthesia is very tricky. This was a case report of management in a 24-year-old pregnant female with undiagnosed

neurofibromatosis for an emergency cesarean section for antepartum haemorrhage and IUD under spinal anaesthesia.

CASE REPORT

A 24- year-old female of primigravida was admitted to our hospital given antepartum hemorrhage with transverse presentation and USG-documented IUD. On preoperative examination, she had had multiple swellings and café-au-lait spots on the body since childhood, a large neurofibroma in the right forearm was present and in the oral cavity there were small flaky lesions.

She was not evaluated previously but based on the history and clinical examination, the diagnosis of neurofibromatosis type 1 was made and taken up for emergency cesarean section given antepartum hemorrhage. Since the patient was not thoroughly evaluated and no investigation has been done, it was decided for the emergency cesarean section to be done under regional anesthesia as the patient was having lesions over the tongue with SpO_2 of 90% on room air and bilateral decreased air entry. On examining the back of the patient, she did not having any lesions and the spine was also apparently normal on palpation. Monitors were attached, and 2 IV cannulas, 18G and 20G were secured.

Blood was started from one IV cannula and ringer lactate was started from another cannula. Spinal anesthesia was given to the patient in a sitting position with 26G quincke needle with 2.4ml of 0.5% hyperbaric bupivacaine between lumber 4 and lumber 5 intervertebral spaces. A sensory level of thoracic level T6 was achieved and the procedure started.

The procedure remained uneventful and the patient shifted to the general ward for further monitoring. The postoperative period was also uneventful and the patient was discharged on 3rd postoperative day. Spinal anesthesia may be extremely difficult in these patients as they may have neurofibromas close to intervertebral space or kyphoscoliosis.

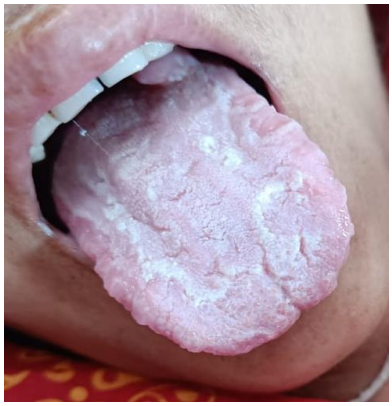

Figure 1: Lesion over dorsum of tongue.

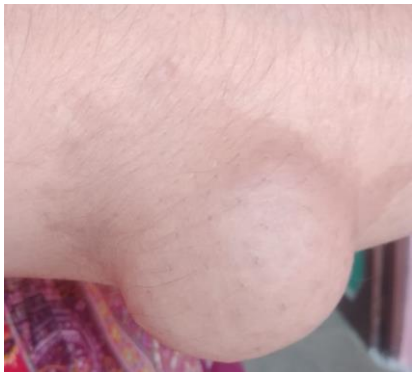

Figure 2: Neurofibroma over forearm.

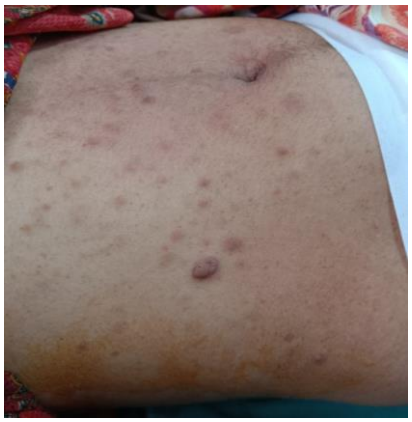

Figure 3: Multiple café au lait spots and neurofibroma over anterior abdominal wall.

DISCUSSION

NF1 also called von Recklinghausen disease is a rare neurocutaneous condition that can have widespread and deleterious effects on various organ systems, including the spine with implications for the choice of anesthetic technique. The multisystem involvement complicates anesthetic management as they may have difficult airway, oral neurofibromas, and out of which tongue has been the most affected part which was also present in our case, supraglottic neurofibromas, phaeochromocytoma and intraoperative hypertension which was also observed in our case, kyphoscoliosis, intrapulmonary fibrosis, decreased cardiopulmonary reserve and occasionally it may develop in the brain, on cranial nerves or the spinal cord.[6,7] Type 2 neurofibromatosis usually invades the central nervous system with the main clinical feature of bilateral vestibular schwannomas leading to gradual hearing loss.[4] Other clinical features like meningioma of the brain, schwannoma of cranial, spinal, or peripheral nerves, and juvenile cortical cataracts can also exist.[2] Pregnant females with neurofibromatosis are also known to be associated with hypertension, HELLP syndrome, spontaneous abortion, and an increase in the number and size of neurofibromas.[4] CNS tumors also rapidly increase in size during pregnancy.[8] Regional anaesthesia in

patients with CNS involvement needs extensive evaluation as the spinal tumor may exist in the injection site or tumor cells can spread into CNS by spinal or epidural needles.

For these reasons, general anaesthesia appears more advisable but other risk factors are also common. Difficult airway management is a known major cause of anaesthesia related maternal deaths. The choice between general anesthesia and regional anesthesia was very difficult as the patient was having antepartum hemorrhage with IUD so it had to be taken in emergency cesarean section. In our case, preoperative radiographic examination was not available. The patient had classical features of type 1 neurofibromatosis and had no neurological symptoms. Keeping in view of her SpO_2 at room air 90% with decreased air entry over bilateral Lung fields and oral neurofibromas, the decision was made to proceed with spinal anaesthesia.

CONCLUSION

This is a case of successful spinal anesthesia during an emergency cesarean section in a patient with newly diagnosed neurofibromatosis without extensive neural evaluation. Although general anesthesia is the safer option in neurofibromatosis patients, spinal anesthesia can be a safe procedure in neurofibromatosis patients but special precautions should be taken to avoid neurological injury and uneventful anesthesia.

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