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Spinal Anaesthesia in an Emergency Lower Segment Cesarean Section in a Patient Diagnosed with Cutaneous Neurofibromatosis: A Case Report

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Abstract

Neurofibromatosis is a neurocutaneous disorder with characteristic skin and nervous system tumours that have implications in anesthesiology. The presence of tumours in the airway and spinal cord can cause difficulty in administering general anaesthesia and spinal anaesthesia respectively. Type 1 neurofibromatosis accounts for 96 % of neurofibromatosis cases which have rare CNS involvements. Prevalence is about 1 in 3000 births. Neurofibromatosis can affect almost all systems. So thorough knowledge is needed to administer safe anaesthesia.

Keywords: Neurofibromatosis, Spinal Anaesthesia, Anaesthesia Management, Pregnancy, Airway Difficulties

Introduction

Neurofibromatosis (NF) is an autosomal dominant genetic disorder which is characterised by the development of multiple neurofibromas and associated multisystem complications. The two primary forms, Neurofibromatosis Type 1 (NF1) and Neurofibromatosis Type 2 (NF2) present unique challenges for anaesthesiologists due to their effects on the airway, nervous system, cardiovascular system, and musculoskeletal structures. Hormonal changes in pregnancy may also accelerate the growth of neurofibromas, exacerbating symptoms. Pregnant women with NF1 may experience worsening neurofibromas, which can affect the airway, spinal cord, and peripheral nerves. Kyphoscoliosis and spinal neurofibromas may complicate epidural or spinal anaesthesia due to anatomical distortion or presence of intrathecal tumours. Hypertension and pheochromocytoma, more common in NF1, necessitate careful hemodynamic monitoring. In NF2, the presence of vestibular schwannomas increases the risk of aspiration, cranial nerve dysfunction, and respiratory compromise. General anaesthesia may be challenging due to difficult intubation and potential respiratory muscle weakness.

Case report

A 25-year-old G2P1L1 at 38weeks plus 2 days of gestational age in labour (height 163 cm, weight 62 kg,) was posted for emergency lower segment caesarean section in view of thick meconium-stained amniotic fluid. She had an uneventful antenatal history. Past medical history revealed neurofibromatosis, which was diagnosed 10 years ago with characteristics of café-au-la-it spots and benign cutaneous neurofibromas. The patient arrived at the operating room with an eight-hour fasting state. There were no neurological symptoms and no mass was observed in the airway. Preoperative laboratory tests were non-specific. After taking consent patient shifted to OT and all ASA monitors were attached. IV fluids started with Ringer's lactate. Preoperative Blood pressure was 130/80 mmHg. Inj.Ondansetron 4 mg IV was given as premedication. Spinal anaesthesia was performed at the L3-4 level with 0.5% heavy bupivacaine 10 mg in sitting position by a paramedian approach as there were lesions in the L3-4 and L4-5 midline. After attaining T4 sensory level operation was started. The intraoperative and post-operative period were uneventful and the patient was discharged on postoperative day five.

Discussion

Neurofibromatosis is an autosomal dominant inherited disease caused by a mutation in a single gene with 100 per cent penetrance and variable expressivity [1]. It can involve multiple systems. Neurofibromatosis is classified into type 1 and type 2. Neurofibromatosis Type 1(von Recklinghausen's disease) can present as cutaneous neurofibroma (benign neurofibromas of the skin) and café-au-lait spots (coffee-coloured spots) [2]. Type 2 neurofibromatosis is more dangerous as it invades the central nervous system and presents as symptoms of bilateral vestibular schwannomas and spinal cord tumours [3]. So careful CNS examination is needed in these patients before attempting the central neuraxial techniques. Pheochromocytoma is known to be associated with type II neurofibromatosis. Because of the hormonal changes in the pregnancy, there can be an increase in the number and size of neurofibromas [4]. The size of CNS tumours in neurofibromatosis patients can also rapidly increase during pregnancy [5]. A spinal tumour may be encountered at the injection level or the tumour cells can spread into the CNS by performing neuraxial techniques. Because of these risks it is advisable to proceed with general anaesthesia, especially in type II neurofibromatosis. Neurofibromatosis in pregnancy can be complicated by pregnancy-induced hypertension, HELLP syndrome, cerebrovascular disease, intrauterine growth retardation, preterm labour, cesarean delivery and spontaneous abortion [6]. The diagnosis of spinal cord neurofibromas pregnancy can be done by magnetic resonance imaging(MRI). Here our case was an emergency case we could not rule out spinal or CNS tumours by doing an MRI. Our patient had characteristic clinical features of type 1 neurofibromatosis and had negative neurological symptoms we performed spinal anaesthesia. Neuraxial anaesthesia could be useful in neurofibromatosis type 1 because of the rare CNS involvement. Difficult airway is a major cause of anaesthesia-related maternal deaths. In neurofibromatosis patients, additional masses in the tongue, pharynx and larynx may interfere with laryngoscopy and intubation. If the airway is difficult, awake fiberoptic bronchoscopy can be done. But this will be challenging during an emergency. Our patient's entire body was covered with neurofibromas, including the needle puncture site (L3-5) in the midline. So here we used a paramedian technique for spinal anaesthesia. Here we successfully managed a case of type 1 neurofibromatosis for an emergency cesarean section under spinal anaesthesia.

Conclusion

Careful preoperative evaluation is needed to rule out CNS and spinal tumours in Neurofibromatosis, especially in type II. Because of rare CNS and Spinal involvement, neuraxial techniques can be done in neurofibromatosis type 1.

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