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Case Report

Anaesthetic Consideration in Patient with Neurofibromatosis Type 1 in Pregnancy

Arshi Taj[1], Mushtaq Ahmad Rather[2], Khurshid Ahmad[2], Shemaiel Maqbool[2]

[1]Consultant, [2]Senior Resident,
Department of Anaesthesia and Critical Care, Government Medical College, Srinagar.

Corresponding Author: Mushtaq Ahmad Rather

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ABSTRACT

Neurofibromatosis type 1, a multisystem relatively common single gene disorder is caused by a mutation of NF-1 gene that results in a loss of activity or in a non-functional neurofibroma protein.

Keywords: Neurofibromatosis, Anaesthetic considerations in pregnancy.

INTRODUCTION

Neurofibromatosis - 1 is associated with wide variety of clinical implications and affects most organ systems presenting as respiratory, cardiovascular, central nervous system, musculoskeletal, gastrointestinal and genitourinary systems and all present various degrees of consideration for anesthesiologist.

Anesthesiologist must perform a thorough preoperative evaluation of every patient and have a well formed and comprehensive anesthetic plan to be prepared for any complication. Anaesthesia for pregnant patient with neurofibromatosis is not well documented.

Pregnancy in patients with neurofibromatosis can result in multiple complications. During pregnancy, hypertension may be exacerbated, neurofibromas may increase in size and large pelvic or genital neurofibromas may cause preterm labour or complicate delivery. [1,2] A study of Segal *et* al showed a significantly higher rate of extrauterine growth restriction, still births and need for caesarean sections in pregnant women with neurofibromatosis type 1. [3] Additionally, the increase in neurofibroma size may cause an increase in intracranial pressure. According to Dounas et al, the presence of increased intracranial pressure and spinal neurofibromas should be evaluated using CT or MRI before spinal or epidural anaesthesia despite the radiation risk to fetus. [4]

Regarding anesthetic management during labour and delivery, the anesthesiologist must consider the possibility of increased intracranial pressure, spinal abnormalities and spinal tumours that can complicate regional anaesthesia.

Neurofibromatosis type 1 is referred to as Von Recklinghausen and characterized by café au lait spots and benign cutaneous neurofibroma. Type 2 neurofibromatosis

affects CNS with spinal cord tumours and bilateral cutaneous neurofibromas.

CASE REPORT

Forty five year old primigravida, 55kg, a known case of Von Recklinghausen neurofibromatosis was scheduled for emergency caesarean section in view of breech presentation. There was no history of previous surgeries and drug allergies. Patient looked poorly built and systemic examination revealed normal cardiovascular and respiratory system. She had protruding upper incisors with adequate mouth opening. Normal neck and temporomandibular joint movement and Mallampati Class I on oropharyngeal examination. There were no neurological symptoms and no mass observed in tongue, pharynx & larynx. As it was an emergency caesarean section in view of fetal distress, regional anaesthesia was not a choice of anaesthesia as no CT scan of head/brain was available. General anaesthesia was planned for caesarean section. The anesthetic procedure was explained to the patient and written informed consent obtained. In the operating room, her heart rate was 118/min, BP 140/80mmHg, oxygen saturation was 95%. Intravenous access with 18G cannula was obtained. Intravenous fluid Ringer lactate 15 ml/kg was started. Difficult intubation cart kept ready. Patient preoxygenated with 100% oxygen for 3 min. General anaesthesia given with Inj. Thiopentone 300mg, Inj. Succinylcholine 75mg and on D/L Cormack Lehane was grade II, hyperextension of the neck was avoided during laryngoscopy & patient intubated with size 7mm cuffed PVC E.T.T. Patient was connected to circuit & ventilated manually. Anesthesia was maintained with Halothane 0.5%-0.77% + N_2O(50%) + O_2(50%)+ inj. Atracurium 25mg. A male infant weighing 2.5kg was delivered with Apgar score 8 at birth followed by 9 & 10 at 1 and 5 min. Intraoperative analgesia was given by injection paracetamol 20mg/Kgbw and after the delivery of baby inj. fentanyl 2µ/kgbw supplemented at the end by intramuscular injection diclofenac 75mg and local anaesthetic ropivacaine wound infiltration. Patient remained hemodynamically stable maintaining vitals and was extubated successfully maintaining SPO_2after the reversal of neuromuscular blockade by injection neostigmine (0.5mg/kgbw) and injection atropine (0.2mg/kgbw). Postoperative course had been uneventful.

DISCUSSION

Neurofibromatosis is an autosomal dominant disease that has widespread effects on ectodermal and mesodermal tissues. The commonest member of the group is neurofibromatosis type 1 which varies in severity but affects all physiological systems. Neurofibromas are the characteristic lesions of the condition and not only occur in the neuraxis, may also be found in the oropharynx and larynx and these may produce difficulties with laryngoscopy and tracheal extubation. Pulmonary pathology includes pulmonary fibrosis and cystic lung disease. The cardiovascular manifestation of NF 1 excludes hypertension, which may be associated with pheochromocytoma or renal artery stenosis. Neurofibromas may also affect the gastrointestinal tract, a cracinoid tumour may be found in the duodenum. [5]

Painless dislocation of cervical vertebrae has been reported in patients with multiple cervical neurofibromas and it has been suggested that a radiographic examination of neck should be performed before administering anaesthesia in these patients in order to avoid spinal cord damage during laryngoscopy and tracheal intubation. [6]

Hypertension presenting in the young NF1 sufferer is usually because of

renal artery stenosis which may be bilateral. [7] The arterial lesions are of variable morphology with fusiform intimal narrowing or nodular or aneurysm formation. [8] Tumours of the central nervous system account for major portion of the morbidity and mortality of patients with neurofibromatosis. Anaesthetic assessment of such patients should be taken into account; there may be increased tendency of epilepsy, learning difficulties and possibility of undiagnosed CNS tumours. Involvement of brain stem structures by neurofibroma or glioma may result in central hypoventilation syndromes. Such patients may exhibit protracted weaning from mechanical ventilation postoperatively. [9]

The genitourinary tract may be involved in NF1 and retroperitoneal neurofibromas result in ureteric obstruction and hydronephrosis. [10]

Increased sensitivity of patients with NF1 to non-depolarizing neuromuscular blocking drugs. [11,12] Special consideration are to be given to patients of NF1 with renal impairment or those on concurrent medication (e.g. anticonvulsant drugs) which may interfere with the normal pharmacokinetics or pharmacodynamics of neuromuscular blocking drugs.

Neurofibromatosis type II NF-2 characteristically has bilateral vestibular schwanomas leading to gradual hearing loss. [13] Other clinical features such as meningioma of the brain, schwanoma of cranial, spinal or peripheral nerve and juvenile cortical cataract can also exist. [5] Neurofibromatosis appears to have no intrinsic effect on fertility, a high rate of spontaneous abortion and still births have been reported. Neurofibromatosis parturients are known to be associated with hypertension, HELLP syndrome, spontaneous abortion and increasing number and size of neurofibromas. [3,14]

Anaesthesia for the pregnant patient with neurofibromatosis is not well documented. Involvement of CNS, in NF-2 has led to choice of anaesthesia to be general rather than regional. We preferred general anaesthesia over regional as patient, risk of CNS involvement was not delineated and no mass in the tongue or pharynx which is normally seen with NF-1 excluded difficult laryngoscopy. Patient was normotensive and keeping in view emergency nature of surgery, we opted for general anaesthesia in this patient. Many studies have been conducted to delineate risks of malignancy in NF1 and it seems overall risk of cancer of cancer was increased by factor of 2.7 and largely owing to marked increase in risk of brain/CNS or connective tissue tumours.

Longitudinal study by Sorensen in 1986, with 212 patients with NF type I were followed up over a period of 40 years, 57 patients were found to have malignant tumours & many of these had increased rate of CNS tumours particularly gliomas.

CONCLUSION

To conclude, manifestations of neurofibromatosis are often mild. Anesthesiologists should consider the complex and the diverse associated factors to provide optimal anaesthesia whether to proceed with general or regional anaesthesia in these cases must be reviewed, within the appropriate clinical context like associated system involved and type of surgery.

REFERENCES

1. Jett K, Friedman JM. Clinical and genetic aspects of neurofibromatosis. Genet Med 2010 Jan; 12(1): 1-11.
2. Hrisch NP, Murphy A, Radcliffe JJ. Neurofibromatosis, clinical presentation and anesthetic implications. Br J Anaesthesia 2001 Apr; 86(4): 555-564.
3. Segal D, Holcberg G, Sapir O, Sheanes E, Mazor M, Katz M. neurofibromatosis in pregnancy. Maternal and perinatal

outcome. Eur J Obstet Gynecol Reprod Biol May 1999; 84(1): 59-61.
4. Dounas M, Mescier FJ, Jhuissier C, Benhamou D. Epidural analgesia for labour in partiturants with neurofibromatosis. Can J Anaest 1995 May; 42 (5Pt 1): 420-422.
5. Hirsch NP, Murphy A, Radcliffe JJ. Neurofibromatosis, clinical presentation and anaesthetic implication.Br J Anaesth. 2001 Apr;86(4):555-64
6. Lovell AT, Alexander EM. Silent, unstable cervical spinal injuries in multiple neurofibromatosis. Anaesthesia 1994; 49: 453-4.
7. Bourk E, Eatenby PBB. Renal artery dysplasia with hypertension in neurofibromatosis. Br Med J 1971; 2: 681-82.
8. Salifer WR, Salyer DL. The vascular lesion of neurofibromatosis. Angiology 1974; 25: 510-9.
9. Sforza E, Colamana V, Lugarin E. Neurofibromatosis associated with central alveolar hypoventilator syndrome during sleep. Acta Paediator 1994; 83: 794-6.
10. Shah S, Murthy PV, Gopal Krishan G, Pandit AP. Neurofibromatosis of the bladder and urethra presenting as obstructive uropathy. Br J Urol 1988; 61: 363-5.
11. Magbagbeola JAO. Abnormal response to muscle relaxant in a patient with Von Rechlinhausen disease. Br J Anaesth 1970; 42: 710.
12. Manser J. Abnormal responses in Von Recklinghausen disease. Br J Anaesth 1970; 42: 183.
13. Ashok R. Asthagiri *et al* Mechanisms of Hearing Loss in Neurofibromatosis Type 2 PLoS One. 2012; 7(9): e46132.
14. Singh T, Hooda S, Anand A, Kaur K, Bala R. Anesthetic consideration in a preeclamptic parturient with Von Recklinghausen's neurofibromatosis. J Obstet Anaesth Crit care 2014; 4: 38-40.

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