Research Paper

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Neurofibromatosis Type 2 for Craniotomy: Anaesthesia Management of a Rare Case

Medical Science

KEYWORDS : Anaesthesia, Bilateral vestibulo-cochlear schwannomas, Meningioma, Neurofibromatosis Type 2

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ABSTRACT

Introduction: Neurofibromatoses are autosomally dominant diseases having tendency to form tumours in ectodermal and mesodermal tissues. Neurofibromatosis type 2 (NF2) is rarer with birth incidence of 1:33,000-40,000.

Case Description: We report the perioperative management of a 22 year old female, known case of NF2, presented with repeated episodes of projectile vomiting and decreased vision. MRI Brain showed bilateral vestibulo-cochlear schwannomas, left parafalcine and bilateral tentorial leaf meningiomas. Patient was posted for craniotomy and was managed successfully. Post operatively patient was shifted to ICU, electively ventilated overnight and extubated next day. After two days patient was shifted to ward and discharged on 10th day.

Conclusion: NF2 is challenging to anaesthesiologists due to possibility of intracranial and intra-spinal canal tumours which is of relevance when patients are posted for other surgeries also. Multidisciplinary management with early diagnosis is mainstay of management.

INTRODUCTION

Neurofibromatoses (NF) are a group of autosomally dominant hereditary diseases [1]. Two distinct forms are neurofibromatosis type 1 (NF 1) and neurofibromatosis type 2 (NF2). NF2 is a rare disease, with incidence of 1 in 33,000-40,000 [2] and a penetrance of almost 100% by the sixth decade of life. The incidence of NF1 is 1 in 3000 births [3]. Disease prevalence is 1 in 5000 for NF1 [4] and 1 in 60,000 for NF2 [5].

NF2 can manifest as intracranial, spinal, ocular and cutaneous lesions. The main tumors occurring in NF2 patients are bilateral vestibular schwannomas, other peripheral, cranial and spinal nerve schwannomas, intracranial and intraspinal meningiomas, ependymomas, and gliomas. As it is rare and can present with various manifestations, it can be challenging for anesthesiologist. We describe here the successful anaesthetic management of a patient with NF2, posted for craniotomy, for Meningioma excision.

CASE REPORT

A 22 year old female weighing 60kg, k/c/o NF2, was posted for craniotomy for her raised intracranial pressure. She came in emergency, with h/o repeated episodes of projectile vomiting, decreased vision since 3 days.

She had a right eye squint, ptosis, multiple subcutaneous nodules, hyperpigmented macules since 10 years. She was operated for squint during childhood but not improved. CT Brain (during that time) showed small size meningioma in anterior frontal pole, which was ignored by the family. She had diminished hearing and tinnitus since 3years, headache and gait imbalance since 3 months. Two months back she was diagnosed as NF2, on CT brain (bilateral vestibulo cochlear schwannomas, multiple left parafalcine and bilateral tentorial leaf meningiomas), which was advised by ENT department after she consulted for her complaints. But patient didn't follow up. Family history of subcutaneous nodules in father and elder brother was present.

MRI Brain on admission showed, same features s/o NF2. Opthalmic examination revealed right complete 3rd nerve palsy and fundoscopy showed bilateral papilloedema. Audiometry revealed left sided severe to profound mixed hearing loss, right sided profound sensorineural hearing loss.

On admission she received, Inj. Eptoin and Mannitol, dexa for raised ICP and craniotomy was planned for decompression for excision of right tentorial meningioma. On examination, patient had hyperpigmented nodules on abdomen, left lower limb and left cheek. Ptosis and right eye squint was present. Her vitals were stable. Her neurological examination including her higher mental functions were normal. Routine blood investigations and chest x-ray was normal.

After appropriate consent she was taken to operation theater. ECG, Spo2, NIBP, ETco2, nasopharyngeal temp,urine output were monitored. Two wide bore IV cannulas were taken under local anaesthesia. After preoxygenation and premedication with IV inj Glycopyrrolate 0.2mg,inj Midazolam 1mg,inj Fentanyl 90mcg,inj Xylocard 60 mg to attenuate laryngoscopy response, pt was induced with inj Thiopentone 300mg and inj Atracurium 35mg and intubated with 7mm endotracheal tube. Balanced Anaesthesia was maintained with O2, Air, Isoflurane and vecuronium and intermittent doses of Fentanyl for analgesia. Right IJV was cannulated for CVP monitoring and fluid management. Park bench position was given.

Intra-operatively vitals were stable and blood pressure was maintained within mean of 60-80 mm of Hg, ETco2 was maintained within 28-30 mm of Hg. Intra-op blood loss was around 2.8 lt. which was replaced with 5 units of packed cells, 5 FFPs and colloids. Operation took around 6 hrs. Post operatively patient was shifted to ICU, electively ventilated overnight and extubated next day. After two days patient was shifted to ward and discharged on 10th day. Histopathology report of tumour showed Meningiothelial Meningioma.

DISCUSSION

NF2 is a autosomal dominant tumour-prone disorder characterised by the development of multiple schwannomas and meningiomas [1], caused by mutation of NF2 gene, a tumour suppressor gene, located on chromosome 22, whose normal protein product is Merlin/Schwannomin [6]. Nearly 50% cases are inherited and little over 50% cases occurs de-novo [7]. The average age of onset

of symptoms is 18 to 24 yrs, and the average age of death is 36 years [8]. The type of mutation in the NF2 gene dictates the severity of disease. Though tumours are not malignant their anatomic location, multiplicity increases morbidity and early mortality.

As NF2 is rare and recently recognised distinctly, not many cases concerning the anaesthetic management are reported. Bilateral vestibular schwannoma (VS) is characteristic of NF2 and other main tumours are schwannomas of the other cranial, spinal and peripheral nerves; meningiomas both intracranial and intraspinal; and some low-grade central nervous system malignancies (ependymomas and gliomas) [7]. Due to bilateral VS, patients usually presents with tinnitus, hearing loss and balance dysfunction, [1] which were present in our patient also.

About 20–30% cases presents with symptoms from an intracranial meningioma (headaches, seizures), spinal tumour (pain, muscle weakness, paraesthesia), or cutaneous tumour [1]. Cutaneous features are more subtle in NF2 compared to NF 1 [2,10].

NF2 is considered as an adult onset disease, it may be underrecognized in children, in whom skin tumours and ocular findings may be the first manifestation [8]. Posterior subcapsular lens opacity is a common ocular finding [8]. Mononeuropathy is increasingly recognised finding in childhood and frequently presents as persistant facial palsy, squint (3rd Nerve palsy), or hand and foot drop [8]. Our patient had squint and cutaneous features in childhood but she was not evaluated for NF2. Hence children with such symptoms should be fully evaluated.

In about 3-5% adults, more generelised polyneuropathy occurs [11]. Approximately half of individuals with NF2 have meningiomas [8]. Most are intracranial and frequently occurs supratentorialy [7,8]. Meningiomas may predate the development of VS and any childhood meningioma should be considered as early sign of NF2 [8]. Our patient had small meningioma on CT Brain during childhood but it was neglected.

Meningiomas are usually vascular tumours hence good IV access is needed for blood transfusion. Our patient had 2.8 lit of blood loss which was replaced adequately. Anaesthesia management during craniotomy was directed towards providing optimal operative conditions, haemodynamic stability and avoiding further neurological damage. We followed these basic neuroanaesthesia principles during the procedure.

Once diagnosis of NF2 is made, its extent should be evaluated by MRI Brain and Spine, hearing evaluation along with brainstem Auditory Evoked Response (BAER), ophthalmologic, neurologic evaluation and cutaneous examination [1,8]. The gold standard for diagnosis is MRI scan of cranial and entire spinal canal [2]. If such individual comes for any surgery, all systems should be evaluated and screened.

MRI screening of the chest and abdomen to look for extensions of the tumour to thorax or abdomen and compression of adjacent structures is vital as multiple intrathoracic schwannomas were previously reported [2]. A cervical spinal scan should be performed before cranial surgery to prevent complications from manipulation under anesthesia [12].

NF2 patients treatment is currently target specific (tumour, cataracts, neuropathy) and mostly surgical i.e. removal of symptomatic cranial and spinal tumours [1,8]. Stereotactic radiosurgery is an alternative [8]. Genetic counselling to first degree relative is also recommended [1].

CONCLUSION

NF2 with multiple tumour manifestation poses challenge to anaesthetist and awareness of its various presentations is required for better management. Pre-operative discussion should involve Anaesthesiologist, Neurosurgeons, Neuroradiologist and ENT surgeons. The proper evaluation helps in better peri-operative management of the cases.

Figure 1. Figure. CT Scan showing Small Childhood Meningioma

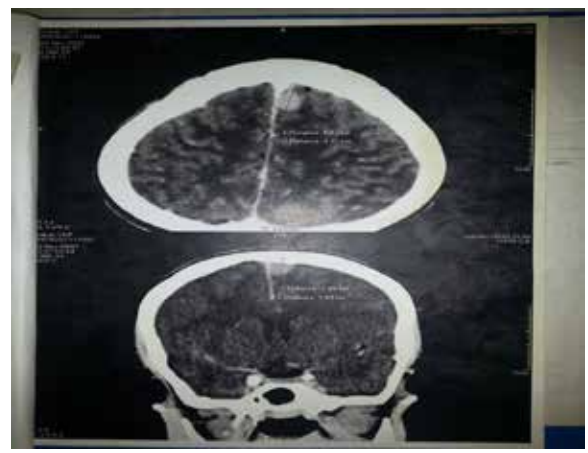
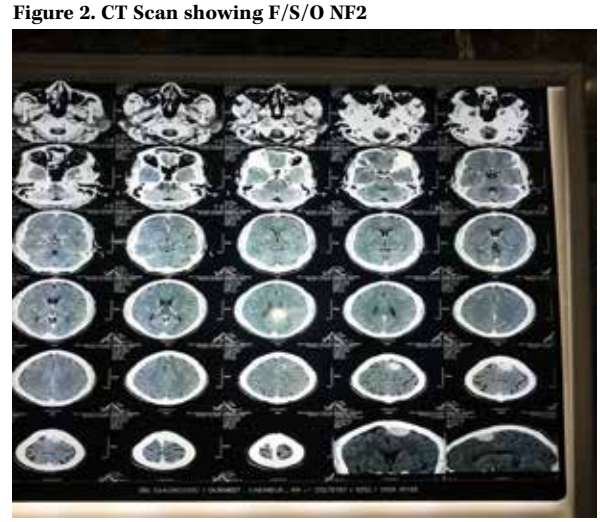


Figure 2. CT Scan showing F/S/O NF2



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