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SPINAL ANAESTHESIA FOR INGUINAL HERNIA REPAIR IN A NEUROFIBROMATOSIS PATIENT WITH KYPHOSCOLIOSIS – A CASE REPORT

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ABSTRACT
Neurofibromatosis type 1 (NF1) is an autosomal dominant neuroectodermal hereditary disorder, in which spinal skeletal deformities are one of the manifestations. Pectus carinatum, temporomandibular joint dysfunction and kyphoscoliosis can be seen with this genetic disorder which can lead to cardiorespiratory system problems and can cause difficulty in airway management. The clinical spectrum of this disorder is quite broad, characterized mainly by skin neurofibromata and café-au-lait spots. We report a case of 61 year old male with neurofibromatosis type 1 with kyphoscoliosis scheduled for left inguinal hernia repair. Patient was administered spinal anaesthesia at L4–L5 intervertebral space. Successful subarachnoid block was obtained and the surgery was performed successfully without any untoward complications.

KEY WORDS: Neurofibromatosis, Spinal Anaesthesia, Kyphoscoliosis

INTRODUCTION
Neurofibromatosis (NF) is a genetic disease, transmitted as an autosomal dominant disorder and characterized by the formation of ectodermal and mesodermal tumours. It can be divided basically into two groups with different clinical manifestations: NF1 and NF2. Von Recklinghausen disease, also known as neurofibromatosis 1 (NF1), is characterized by multiple cafe-au-lait spots in the skin, multiple peripheral nerve tumors, and a variety of other dysplastic abnormalities of the skin, nervous system, bones, endocrine organs and blood vessels.[1,2] It is a genetic disease in humans, inherited by a gene located in chromosome 17, affecting one in 3000-4000 individuals. Pectus carinatum, temporomandibular joint dysfunction and kyphoscoliosis can be seen with this genetic disorder which can lead to cardiorespiratory system problems and can cause difficulty in airway management. NF2 is defined by the presence of bilateral vestibular schwannomas affecting one in 33,000-40,000 individuals.[1,2,3] We present the anaesthetic management of a NF1 patient with thoracolumbar kyphoscoliosis who had been posted for inguinal hernia repair. The operation was

successfully performed with subarachnoid block. This case highlights the anaesthetic challenges, the significance of proper preoperative evaluation & planning anaesthetic technique.

CASE DESCRIPTION

A 61 year old male of 153 cm height, 54 kg weight with past history of neurofibromatosis 1 & severe kyphoscoliosis was scheduled for left inguinal hernia repair. He gave history of right inguinal hernia repair under spinal anaesthesia in the previous year in our hospital, which was uneventful. There was no history of any other associated co-morbidity. He denied any leg weakness, parasthesia, back pain, dizziness or headache. He was observed walking without difficulty & showed no evidence of neurologic compromise on physical examination. His entire body including back was covered with nodules of different sizes. A big tumour, sized 8cmx6cmx5cm was present in the occipital region. Oral examination revealed Mallampatti grade II with no oral lesions. Examination of spine showed a lateral curvature along with thoracolumbar kyphoscoliosis. The rest of the physical examination was normal.

All the routine laboratory investigations were within normal range. Chest X-ray PA view revealed cervicothoracic curvature with tracheal deviation to right. Spine X-ray showed thoracolumbar kyphoscoliosis with Cobb's angle 40^0 [Fig.1]. As he had no neurologic symptoms, MRI spine was not considered for further workup. While being evaluated for PFT, patient did not cooperate.

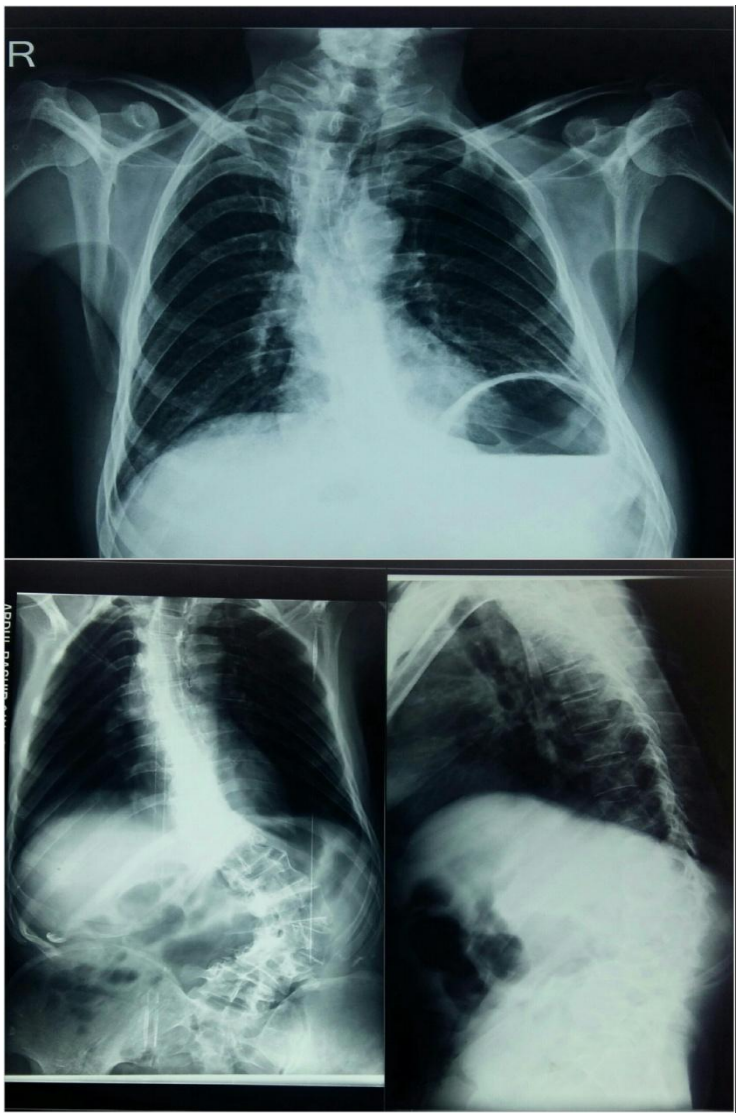

Figure 1: Chest X-ray AP view showing cervical scoliosis with tracheal deviation to right & spine X-Ray with thoracolumbar kyphoscoliosis

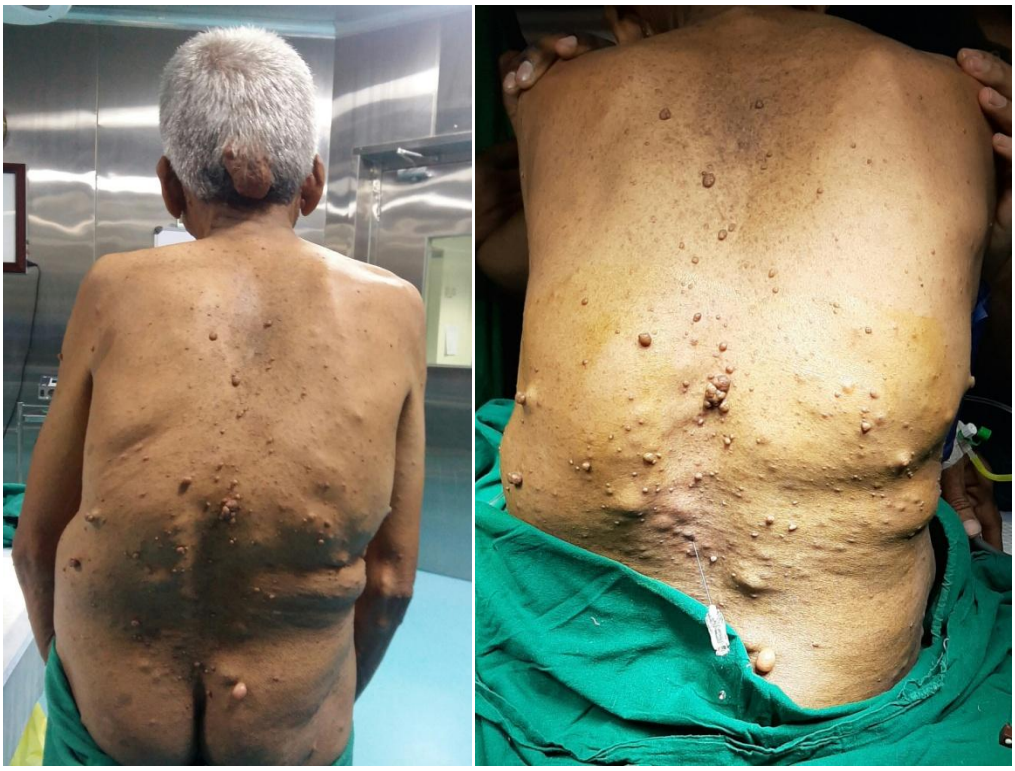

Figure 2: Tumour in the occipital region, back covered with neurofibromata with kyphoscoliosis & spinal needle directed towards convexity of the curve with angulation depicting free, clear flow of CSF

We decided to conduct the case under spinal anaesthesia in view of difficult airway. A written informed consent was obtained. On arrival in the operating room, intravenous line was secured with 18-G cannula & standard monitors were connected. 500ml ringer's lactate solution was given. Under aseptic measures, with the patient in the sitting position, spinal anesthesia was performed between L4-L5 intervertebral space with 26 G Quincke's spinal needle by directing the needle towards the convexity of the curve with significant angulation of the needle & 3ml of 0.5% hyperbaric bupivacaine was administered [Fig. 2]. The patient was immediately placed in supine position. 1mg of midazolam was given intravenously to allay anxiety. Oxygen was supplemented via Hudson's mask at a flow rate of 6L/min.

After 5min, level of anaesthesia was checked to be at T10 level. The surgery was started after 10min & lasted for 1hr. All hemodynamic parameters were stable intraoperatively. The patient was shifted to postoperative recovery room, observed for 1hr & subsequently shifted to the surgical ward, without any complications. He was discharged on the 5th postoperative day.

DISCUSSION

Neurofibromatosis type 1 (NF1) is an autosomal dominant disorder affected by NF1 gene located on chromosome 17. The NF1 gene encodes a protein named neurofibromin which has a role in tumour suppression. Inactivation of the gene leads to loss of function and subsequent development of many different types of tumours seen with the disease.[2,3,4,5,6] Clinical manifestations include cafe-au lait spots, axillary freckling, Lisch nodules, optic gliomas and neurofibromas. Neurofibromas are the most common tumour found in NF1 patients. They can be cutaneous, plexiform or nodular and they can undergo malignant transformation with a risk of 5-13%.[5]

This syndrome has important implications for the anaesthetic practice as the patient may present a difficult airway for both ventilation and intubation depending on the location of the tumours.[7,8,9,10] Neurofibromas on the tongue, larynx, cervical or cranial masses, may hinder the proper positioning of the patient for the best airway approach. Presence of occipital tumour in this case could have

hampered proper positioning to secure airway. In addition, the presence of macroglossia, macrocephaly, mandibular abnormalities and cervical spine involvement may contribute to difficulties of airway management.

Thoracic spinal curvatures are common in NF1 and affect approximately 10% of NF1 patients. Severe kyphoscoliosis, although uncommon, may be associated with tumours and a high risk of neurological deficit. Kyphoscoliosis is associated with restrictive lung disease and hypoxemia which can lead to respiratory failure and cardiovascular compromise.[1,2,11] Although pectus excavatum and carinatum occur in up to 30% of patients with NF1, they do not contribute to respiratory problems.[11,12] NF1 may involve multiple organ systems and therefore raise anaesthetic concerns during surgical procedures. Other anaesthetic considerations include epilepsy, pheochromocytoma, carcinoid tumours and obstructive ureteral stenosis due to neurofibromas.[13] The cardiovascular manifestations of NF1 include hypertension, which may be associated with pheochromocytoma or renal artery stenosis.[14]

The choice of anaesthetic technique in patients with NF1 requires careful systemic evaluation. Factors influencing airway management, respiratory, cardiovascular problems, central nervous system involvement and vertebral anomalies make the choice between general and regional anesthesia more difficult & complex.

In all NF1 patients, complicated airways must be considered. The American Society of Anaesthesiologists provides guidelines for difficult airway.[15] Based on the existing pathology and its severity, preoperative early planning must be done. Patients with NF1 have been reported to be either sensitive or resistant to succinylcholine. Furthermore, all reports indicate that response to nondepolarizing muscle relaxants is exaggerated.[8,16,17] Also prolonged apnea has been reported following both non-depolarizing and depolarizing muscle relaxants for unknown reasons.[18]

Spinal anaesthesia may be extremely difficult & challenging in a patient with NF1 with kyphoscoliosis & neurofibromas close to the needle puncture site, limiting the safety & success of the procedure. We administered spinal anaesthesia in view of difficult airway in the absence of significant neurological manifestations. Due to angulation and rotation of vertebral body, epidural space is deviated toward the convexity of angulation. One method of administering spinal or epidural block is by directing the needle towards the convexity of the curve with significant angulation of the needle. Huang described a modified paramedian approach of spinal anesthesia in such patients.[15] We used same approach in our patient as described above in the procedure at a site that was devoid of neurofibroma lesion.

Anaesthetic experience in patients with neurofibromatosis is limited to few case reports in literature. The anaesthetic challenges in these patients are multiple and anaesthetic management should be designed, based on the existing pathology, its severity and type of surgical procedure. Careful pre-operative evaluation of the case, good preparation, selection and management of effective & safe anaesthesia technique avoid development of fatal airway and other cardiorespiratory complications.

CONCLUSION

The neurofibromatosis, a group of conditions that vary in severity have fundamental, challenging implications for the anaesthesiologists. The choice between general and regional anaesthesia in neurofibromatosis with kyphoscoliosis requires careful consideration. Traditionally, general anaesthesia has been the usual approach as coexisting cranial or spinal neuromas might worsen neurological outcome in patients who have been administered regional anaesthesia. However, associated airway abnormalities, respiratory, cardiovascular, central nervous system and other anomalies make general anaesthesia more risky. Decision between regional and general anaesthesia should also be meticulously weighed. The identification of the spinous process, selection of appropriate intervertebral space & direction of angulation of spinal needle so as to achieve effective spinal anaesthesia, is quite difficult but not impossible by an experienced anaesthesiologist.

Consent
Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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Nil.

Conflict of Interest
There are no conflicts of interest.

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