



Anaesthetic Considerations in Neurofibromatosis Complicated with Kyphoscoliosis and Restrictive Lung Disease

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ABSTRACT

Neurofibromatosis is a genetically transmitted disease arising from the nervous tissue. It is associated with widespread systemic manifestations that pose a challenging situation to the anaesthesiologist. Kyphoscoliosis is one of the musculoskeletal anomalies that present in 10% of patients with neurofibromatosis I. The present case discusses the anaesthetic considerations of a known case of NF I with associated kyphoscoliosis who was posted for surgical removal of mass from the right iliac crest and thigh under spinal anaesthesia. A thorough pre anaesthetic evaluation and workup turn out to be the cornerstone in management of such cases.

Keywords: *neurofibromatosis I, plexiform neurofibroma, scoliosis, respiratory system, anaesthesia, spinal, kyphosis.*

INTRODUCTION

Neurofibromatosis I (NF I) is a hereditary disease with autosomal dominant transmission which is characterised by cafe-au-lait spots, peripheral nerve tumours and dysplasias of skin, nervous system, endocrine glands, blood vessels and bones. Initially described by Freidrich von Recklinghausen in 1882, its incidence ranges from 1 in 3000 to 4000^[1]. It involves the peripheral nerves with long course and plexiform neurofibromas are the hallmark lesions of NF I^[1]. Apart from the characteristic diagnostic factors, NF I can be associated with numerous systemic complications^[1]. These have major implications

on the anaesthetic management for various surgeries.

Kyphoscoliosis which is a deformity of the lateral and posterior curvature of the spine may be associated with NF I, although rarely, affecting 10% of the patients^[2]. Severe Kyphosis is associated with high risk of neurological deficits. Scoliosis may lead to reduced lung volumes and restriction in the lung functions causing hypoventilation, hypercapnia and cor pulmonale^[2]. In addition neurofibromas may lead to erosion of the ribs causing flail chest^[3].

CASE REPORT

A 36 years old male patient, a known case of Neurofibromatosis I with deformity of chest wall and kyphoscoliosis of thoracolumbar spine was posted for debulking surgery of a recurrent mass around the right iliac crest and thigh. During the pre-anaesthetic check-up, patient had a history of multiple nodular lesions all over the body along with deformity of thoracic and lumbar spine since childhood. On examination, patient had numerous neurofibromas over the trunk and extremities. He was moderately built, poorly nourished, had difficulty in lying supine and was unable to walk unaided. The blood pressure (BP) was 120/80 mmHg in the sitting position, heart rate(HR) was 80/ minute and Respiratory rate was 24 per minute. Airway assessment showed normal mouth opening of 3 finger breadth, mallampatti grade III, short neck and restricted neck extension. On chest examination, severe kyphoscoliosis along with deformity of the chest wall was noted. On auscultation the breath sounds were decreased at both lung bases. Bed side pulmonary function tests were done. Cough test and wheeze test were negative. Forced expiratory time was within normal limits while breath holding time was around 20 seconds. Routine investigations including haemogram, renal function test, coagulation profile, blood sugar, liver function tests and ECG were within normal limits. The chest and spine x rays showed crowding of ribs and severe kyphoscoliosis. The pulmonary function tests revealed restrictive lung pattern and patient had mild pulmonary hypertension and an ejection fraction of 60% on echocardiography.



Figure 1



Figure 2

A written informed high risk consent regarding difficult spinal anaesthesia and associated complications like high spinal, failed spinal, partial effect of block and need for general anaesthesia along with post-operative ventilator support was taken from the patient.

Patient was shifted to the operating room. The intravenous access was secured using 18 Gauge peripheral cannula and patient was preloaded with 500 ml ringer lactate solution. Multipara monitor was attached which included monitoring of non-invasive blood pressure (NIBP), HR, electrocardiography (ECG) and pulse oximetry (spO₂). The difficult airway cart was prepared which comprised of different sized face masks, oropharyngeal airways, different laryngoscope blades, endotracheal tubes, laryngeal mask airways, stylet, bougie, fiberoptic bronchoscope and videolaryngoscope. Subarachnoid block in left

lateral position was planned. On palpation, the intervertebral spaces could be barely identified. The spaces were counted using iliac crest as the reference point. Subarachnoid block was administered with great difficulty in space L2-L3 using a 26 gauge quincke point spinal needle. We introduced the spinal needle through the median approach, advancing it towards the convexity of the spinal curvature for achieving a successful placement in the subarachnoid space. Injection bupivacaine 0.5% heavy 2.2 ml was administered after viewing free flow of cerebrospinal fluid.

Patient was made to lie in supine position with using multiple sandbags. Block level of T6 was achieved. The surgery was completed uneventfully in two hours. The haemodynamic parameters and oxygen saturation were monitored and maintained throughout the procedure. No vasopressors had to be used intraoperatively. Patient was shifted to recovery room where monitoring was continued. Post-operative pain was relieved using injection tramadol 50mg intravenously.

DISCUSSION

Neurofibromatosis is one of the most common genetically transmitted diseases. It is associated with multiple complications that present a challenge to the anaesthesiologist. However minor the surgery be, the anaesthetic complications are never minor and need a meticulous pre-operative assessment of all the systems and airway. One of the rare but challenging complications of neurofibromatosis is kyphoscoliosis that progresses with age and presents with impaired lung functions. Being an anatomical deformity, it not only makes regional anaesthesia difficult but also affects the respiratory physiology leading to restrictive lung pattern that hampers the anaesthetic management with routine general anaesthesia as well. Thoracic spine curvatures occur in 10% patients of NF I^[2]. The kyphosis and scoliosis of the spine lead to decrease in vital capacity and functional residual capacity as well as increase in dead space. These patients have a

low tidal volume and high respiratory rate. The first manifestation of restriction of lung functions is reduced vital capacity. Ineffective chest expansion due to distorted anatomy leads to decreased compliance of the respiratory system and ineffective gas exchange in otherwise normal lungs^[4]. All of the above culminate into a high dead space volume, alveolar hypoventilation and increased arterial alveolar gradient leading to ventilation perfusion mismatch and pulmonary shunt^[5]. The end result is chronic hypoxemia leading to right ventricular strain and eventual cor pulmonale^[6]. Our patient had a restrictive lung disease as well as mild pulmonary hypertension as diagnosed by pulmonary function tests and echocardiography.

Apart from respiratory complications due to musculoskeletal abnormalities, neurofibromatosis is associated with cardiovascular, central nervous system, gastrointestinal and genitourinary problems. In a known case of neurofibromatosis, an anaesthesiologist has to prepare for an anticipated difficult airway due to risk of spontaneous cervical dislocation, macroglossia, macrocephaly, recurrent laryngeal nerve involvement and difficult positioning^[1]. Our patient presented with a difficult airway on examination due to short neck, large tongue, restricted neck extension and mallampatti grade III.

Some of the patients with NF I may have hypertension, which could be essential or due to associated pheochromocytoma or renal artery stenosis. Superior vena cava obstruction may also occur due to mediastinal tumours. Associated cardiac pathology like aneurysms and myocardial hypertrophy also affect anaesthetic management. There is also increased incidence of epilepsy, associated cerebrovascular disease, undiagnosed CNS tumours along with cerebral and spinal neurofibromas^[1]. In our case no cardiovascular or cerebrovascular anomaly was diagnosed.

In the present case, we have discussed the management of a known case of neurofibromatosis with kyphoscoliosis posted for excision of mass around the iliac crest. A

subarachnoid block was planned for the same. According to literature, tracheal intubation as well as regional anaesthesia both could be difficult choices for the anaesthesiologists in cases of spinal deformities. However, in a kyphoscoliotic spine, regional anaesthesia is the preferred mode of anaesthesia despite the difficulties associated with it^[7].

The most commonly encountered problems in administering spinal anaesthesia in such patients are failed or multiple attempts and inadequate and unpredictable block. The volume of cerebrospinal fluid is low in these cases leading to a high level of block with low doses of local anaesthetic resulting in more incidence of hypotension^[8]. In addition there could be pooling of hyperbaric solutions in the dependent areas of the spine resulting in an inadequate block. Epidural anaesthesia is also difficult due to improper positioning of the patient and problem in needle and catheter insertion associated with altered epidural space. Combined spinal epidural technique however, offers a faster onset of block along with the chance for improving the level of inadequate block. Also, it helps to prolong the intraoperative analgesia and post-operative pain management^[9].

The spinous processes in scoliosis rotate towards the concave side and the vertebral body rotates towards the opposite side making administration of neuraxial block difficult^[10]. The rotation of spine is directly proportional to the Cobb's angle which determines the degree of cardiovascular compromise and severity of scoliosis^[11]. Cobb's angle is the angle between the perpendicular lines drawn between the most angulated cranial vertebra and caudal vertebrae. The risk of cardiovascular compromise increases with the increase in this angle^[12]. A Cobb's angle > 10 degrees defines scoliosis and it is an independent marker of difficult spinal anaesthesia^[13,14]. Bowens C et al have recommended that in cases with mild scoliosis (Cobb's angle 11-25 degrees) good patient positioning usually results in a successful neuraxial block. Moderate scoliosis

(25-50 degrees) patients may need use of imaging modalities like ultrasound or fluoroscopy and a paramedian approach through the convex side. In cases where the spinous processes are palpable and a median approach is planned the needle should be forwarded in the transverse plane towards the convex side. The authors recommend ultrasound guided access of the subarachnoid space or an alternate modality of pain management for a severely scoliotic spine (Cobb's angle >50 degrees)^[15].

CONCLUSION

Patients of neurofibromatosis with kyphoscoliosis are a major challenge for the anaesthesiologist. Although, no technique is unerring, still regional anaesthesia is a superior choice in these patients. Meticulous preoperative evaluation and planning of anaesthesia is the key to success.

BIBLIOGRAPHY

1. Hirsch NP, Murphy A, Radcliffe JJ. Neurofibromatosis: clinical presentations and anaesthetic implications. *Br J Anaesth* 2001; 86: 555-64.
2. Akbarnia BA, Gabriel KR, Beckman E, Chalk D. Prevalence of scoliosis in neurofibromatosis. *Spine* 1992;17: S244-8.
3. Heriot AG, Wells FC. An unusual case of flail chest: surgical repair using Marlex mesh. *Thorax*. 1997; 52: 488-9.
4. Cooper DM, Rojas JV, Mellins RB. Respiratory mechanics in adolescents with idiopathic scoliosis. *Am Rev Respir Dis* 1984; 130: 16-22.
5. Raw DA, Beattie JK, Hunter JM. Review article: Anaesthesia for spinal surgery in adults. *Br J Anaesth* 2003; 91: 886-904.
6. Kulkarni AH, Ambareesha M. Scoliosis and anaesthetic considerations. *Indian J Anaesth* 2007; 51: 486-95.
7. Ozyurt G, Basagan-Mogol E, Bilgin H, Tokat O. Spinal anaesthesia in a patient with severe thoracolumbar kyphoscoliosis. *Tokuhu J Exp Med*. 2005; 207(3): 239-42.

8. Kleinman W, Mikhail M. Spinal, epidural and caudal blocks in: Morgan GE, Mikhail SM, Murray MJ, eds. Clinical Anaesthesiology .4th ed. New York: Mc Graw Hill Inc; 2006; 289-323.
9. Holmstrom E, Laugaland K, Rawal N, Haliberg S. Combined spinal epidural block versus spinal and epidural block for orthopaedic surgery. Can J Anaesth 1993; 10(7): 601-6.
10. White AA, Panjabi MM. Clinical biomechanics of the spine 2nd ed. Philadelphia:Lippincott, 1990.
11. Peelle MW, Luhmann SJ. Management of adolescent idiopathic scoliosis. Neurosurg Clin N Am 2007; 18: 575-83.
12. Koumbourlis AC. Review: Scoliosis and the respiratory system. Paed Resp Rev 2006; 7:152-160.
13. Atallah MM, Demian AD, Shorrab AA. Development of a difficulty score for spinal anaesthesia, Br J Anaesth 2004;92: 354-60
14. de Filho GR, Gomes HP, da Fonseca MH, Hoffman JC, Pederneiras SG, Garcia JH. Predictors of successful neuraxial block: a prospective study, Eur J Anaesthesiol 2002;19 :447-51.
15. C. Bowens, K. H. Dobie, C. J. Devin, J. M. Corey; An approach to neuraxial anaesthesia for the severely scoliotic spine. Br J Anaesth 2013; 111 (5): 807-11.