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Case Report

Endolaryngeal Neurofibroma: An Unanticipated Anaesthetic Challenge in a Case of Kyphoscoliosis Correction

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ABSTRACT

Laryngeal involvement with neurofibromatosis is rare. Laryngeal neurofibroma can remain symptom free or may commonly present with stridor, hoarseness, dysphonia. We report a case of an asymptomatic 20 years old male k/c/o von Recklinghausen's neurofibromatosis scheduled for kyphoscoliosis correction, with an incidentally discovered aryepiglottic fold neurofibroma during intubation. Patient was intubated successfully and surgery performed in prone position. Intraoperative course was uneventful. But immediately post extubation developed stridor so was reintubated, shifted to ICU and extubated after 24 hours. Although manifestations of neurofibromatoses are often mild, there may be associated pathology of direct relevance and importance to the anaesthetic management.

Keywords: von Recklinghausen's neurofibromatosis, laryngeal neurofibroma, stridor.

INTRODUCTION

Neurofibroma, a benign peripheral nerve sheath tumour arises from schwann cells and perineural fibroblasts. [1] Two clinical forms of neurofibromatosis have been described: peripheral- type I; and central- type II. [1] Neurofibromatosis type I, also known as von Recklinghausen's disease, is a common neurocristopathy, having widespread effects on ectodermal & mesodermal tissue. [1,2] It is associated with a variety of conditions often requiring anaesthesia for surgical treatment. [2] Neurofibromas, the characteristic lesions of the condition occur not only in the neuraxis but may also be found in the oropharynx and larynx; which may produce difficulties with laryngoscopy and tracheal intubation. [2,3] The present paper emphasizes the need for evaluation of airway and larynx of these patients by indirect laryngoscopy before any operation in order to decrease complications of breathing and patient mortality. Also to the best of our knowledge this is the 1st case of an endolaryngeal neurofibroma causing airway difficulty being discussed in anaesthesia till date!

CASE DESCRIPTION

A 17 years male, weighing 45 kgs, known case of Von Recklinghausen's Neurofibromatosis presented with progressively increasing curvature of the back since birth and low backache since 3 months and was scheduled for congenital left sided dorsolumbar kyphoscoliosis correction with posterior instrumentation. He was moderately built and nourished. On physical examination, multiple café-au-lait macules were obvious on his body and extremities. There was no neurological involvement. He had no history of previous surgeries and drug allergies.

Family history was not contributory. Systemic examination revealed normal cardiovascular and respiratory systems. Airway examination showed that the mouth opening was 3 fingers; thyromental distance was 75 mm and a mallampati class-1 on oropharyngeal examination. Neck, temporo-mandibular joint movements and the teeth were normal. Blood investigations as also a screening 2D-echo were within normal limits. Patient's vital signs were stable before anaesthesia. General anaesthesia with endotracheal intubation was planned with induction in supine position and surgery in prone position. For pre-medication inj. Glycopyrrolate 0.004 mg/kg, inj. Ondansetron 0.08 mg/kg, inj. Fentanyl 2 µg/kg and inj. Midazolam 0.01 mg/kg were used. Then, 2 mg/kg inj. Propofol was administered and 1.5 mg/kg succinylcholine was used as the relaxant. On direct laryngoscopy a mass inferior to the epiglottis and coursing posterolaterally over the right aryepiglottic fold was visualized. As the above-mentioned mass did not make any difficulty and vocal cord chink was adequate the patient was intubated successfully with tracheal tube (size 8.5) and a Cormack-Lehane laryngoscopic view of I. Anaesthesia was maintained with O_2 + N_2O + intermittent inj. Vecuronium + inj. Propofol infusion on closed circuit on controlled mechanical ventilation during surgery. Intra-operatively injection dexamethasone was administered in order to reduce airway oedema. Duration of surgery was 6 hrs, with blood loss of 1.2 litres and urine output of 500 ml. 6 crystalloids and 2 colloids were infused throughout the procedure. At the end of surgery, anaesthesia reversal was achieved by injecting inj. Neostigmine 0.05 mg/kg and inj. Glycopyrrolate 0.008 mg/kg. After awakening and spontaneous breathing, the patient was extubated keeping difficult airway cart ready. Immediately post extubation he developed stridor for which was reintubated and shifted to ICU on T-piece. Patient was kept intubated for 24hrs under steroid coverage and intermittent warm saline nebulization and was extubated after 24 hrs with a tracheal tube exchanger in-situ and emergency tracheostomy standby. He was then observed for another 8 hrs in ICU, the course being uneventful. Before shifting him to ward rigid laryngoscopy was performed to map the growth which revealed a submucosal mass involving rt. aryepiglottic fold and rt. arytenoid extending to lt. arytenoid. B/L vocal cords were mobile. Vocal cord chink was adequate.

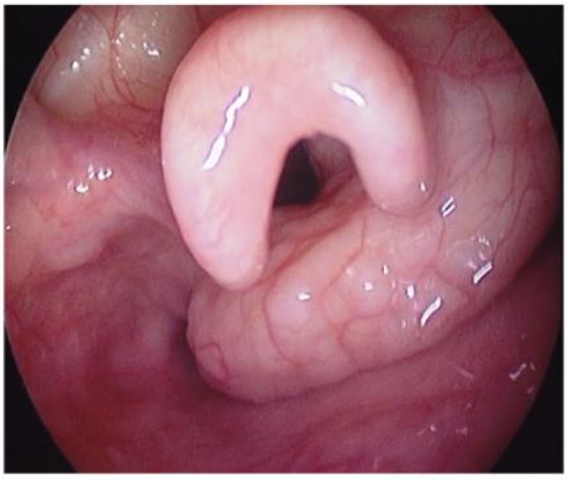

Fig.1: On direct laryngoscopy a mass inferior to the epiglottis and coursing posterolaterally over the right aryepiglottic fold visualized

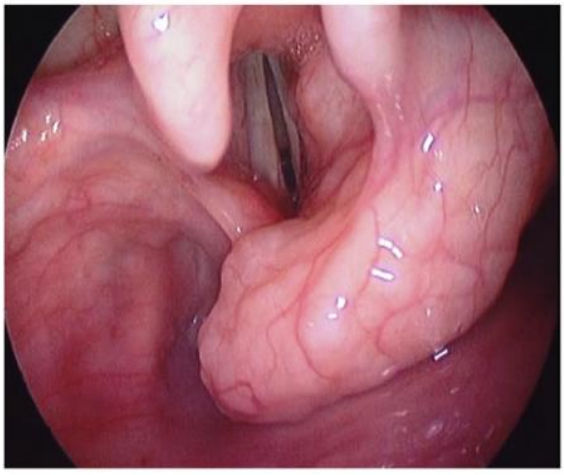

Fig. 2: Vocal cord chink was adequate

DISCUSSION

The term neurofibromatosis (NF) is referred to a group of genetic disorders that primarily affect the cell growth of

neural tissues. [4] There are two forms of NF: Type 1 (NF1) and type 2 (NF2). [1,4] Neurofibromatosis type 1, also known as von Recklinghausen's disease, a neurodermal dysplasia, was first described by Friederich Daniel Von Recklinghausen, the pathologist, in 1882. [4] NF1 is an autosomal dominant disease with complete penetrance but variable expression. [4] It is one of the most frequent human genetic diseases, with a prevalence of one in 3,000 births. [4] There is no sex or racial predilection. The tumour suppressor NF1 gene has been identified on chromosome17q11.2. [1] The pathological alterations behind it begin in the embryonic period, prior to differentiation of the neural crest. Café-au-lait spots, axillary and inguinal freckling, Lisch nodules (pigmented hamartomas of the iris), spinal and peripheral nerve neurofibromas, neurological impairment, scoliosis, abnormalities in the oral and maxillofacial region, malignant tumors of the nerve sheath, pheochromocytoma, vasculopathy are common clinical features of NF1. [4]

Neurofibromas are benign complex tumors that arise from peripheral nerve sheaths and constitute one of the main manifestations of NF1. There are two major types of neurofibroma: 'Discrete' or 'localized' and 'plexiform' neurofibroma. A localized neurofibroma arises from a single site along a peripheral nerve, presents as a focal mass with well-defined margins and is the most common type of neurofibroma occurring in NF1 patients. It can occur superficially or may involve deeper peripheral nerves. [4] They usually appear in late childhood or early adolescence. [1,4] The number of localized neurofibromas tends to increase with age. NF1 patients may have few, hundreds, or even thousands of localized neurofibromas. Neurofibromas are found mostly on the skin. Nevertheless, many organs may be involved, including the stomach, intestines, kidney, bladder, larynx, and heart. [4]

Airway- An estimated 5% of patients with NF1 have an intra-oral manifestation of the disease. [1,6] Discrete neurofibromas may involve the tongue or the larynx. In the larynx they usually affect the supraglottic structure, andare more frequently described in arytenoids, aryepiglottic folds and posterior commissure reflecting areas rich in terminal nerve plexus. [5,8] About 80% of them arise from false vocal cords & aryepiglottic folds. True vocal cords are rare site of location. [5,7] In larynx internal branch of superior laryngeal nerve is involved with preference of sensory rather than motor root. [7] Due to its slow growth rate, variability in location and size, laryngeal neurofibroma can remain symptom free. However symptoms of dyspnoea, stridor, loss or change of voice, dysphonia, or dysphagia should warn of potential airway problems. [3,7,8] Our patient had an incidentally discovered rt. aryepiglottic fold neurofibroma during intubation.

Skin- Pigmented lesions are a common manifestation in NF-1. These lesions usually appear during the first years of life or are present at birth, either as Café-au-lait spots or freckles. Café-au-lait spots are hyperpigmented maculae that may vary in color from light to dark brown. Their borders may be smooth or irregular. They may appear anywhere on the skin, though less common on the face. Inguinal and axillary freckles (Crowe's sign) are frequently present. [4] This patient exhibited more than six Café-au-lait spots and bilateral axillary and inguinal freckles.

Spine- Skeletal involvement is present in almost 40% of the patients with NF-1. Scoliosis is the most common skeletal pathology. [4] Dystrophic spinal curvatures are short, sharp and progress throughout life. [8] Our patient had congenital left sided dorsolumbar kyphoscoliosis which progressed eventually but with no neurological involvement. Painless dislocation of cervical vertebrae resulting in spinal cord

damage during laryngoscopy and tracheal intubation may occur. [3,8] Thoracic spinal curvatures affect approximately 10% which produce a reduction in lung volume and resulting respiratory failure. [8]

Respiratory system-Mediastinal neurofibromas may result in tracheo-bronchial compression with rapidly progressive symptoms. [8] Bilateral upper lobe pulmonary fibrosis resulting in restrictive defect may culminate in pulmonary hypertension and right ventricular failure. [8]

Central nervous system- There is increased incidence of epilepsy & undiagnosed CNS tumors. [4,8] Involvement of brain stem structures resulting in central hypoventilation syndromes, may exhibit protracted weaning from mechanical ventilation post-operatively. [8] Cardiovascular system- Young NF1 are prone to renal artery stenosis. Sustained or paradoxical or resistant to treatment hypertension should raise the suspicion of pheochromocytoma. [4,8] Regular arterial pressure measurement is a vital screen in even young patients. Micronodular vascular proliferation may lead to aortic, cerebral, coronary aneurysms. Neurofibromas may involve the heart causing both hypertrophy and outflow obstruction. [8]

Gastrointestinal & genitourinary systems- Gastrointestinal neurofibromas may result in obstruction, perforation, hemorrhage. Retroperitoneal neurofibromas may lead to ureteric obstruction and hydronephrosis. [8]

Retrospective analysis of stridor in our patient reveals- a vocal cord mass with superimposed factors like surgery in prone position causing dependent edema; long duration of surgery; blood loss and resultant IV fluids infusion causing tissue edema.

In conclusion, the neurofibromatosis is a group of conditions that vary in their severity but which have fundamental implications for the anesthesiologists. Lesson learnt is the importance of advanced airway examination in a case of neurofibromatosis. Although the incidence of airway neurofibroma is low (5 %), a simple non-invasive examination like Indirect laryngoscopy can help us avoid intra and post-operative morbidity. [3] It is therefore important to have a working knowledge of the clinical manifestations of the disease, so that a systemic approach to the pre-operative assessment of these patients can result in rational perioperative management.

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