



Anesthetic Considerations in a Patient With Plexiform Neurofibromatosis: A Case Report

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Abstract

Plexiform neurofibromatosis is an uncommon variant of neurofibromatosis type 1 (NF1) (Von Recklinghausen's disease). There is a greater prevalence of neurofibromatosis 1 in patients with other neoplasms, such as rhabdomyosarcomas, gastrointestinal stromal tumors (GISTs), pheochromocytomas, carcinoid tumors and ganglioneuromas. We report the anesthetic implications of a case of a 33 year old patient with plexiform neurofibromatosis and a history of pheochromocytoma which was operated on previously. He presented with painless swelling on upper eye-lead since childhood and had multiple café-au-lait spots and neurofibromas on the trunk. The surgery was done in two sessions first on the plexiform neurofibroma of the eye followed by the operation on the brain mass.

Keywords: Plexiform neurofibromatosis; Anesthetic implications; Pheochromocytoma.

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Introduction

Neurofibromatosis is an autosomal dominant disorder characterized by neuroectodermal tumors.¹ Neurofibromatosis type 1 (NF1), or Von Recklinghausen's disease is a rare genetic disorder which presents with multiple benign tumors of nerve and skin (neurofibromas) and areas of abnormal coloration of the skin (café-au-lait spots).² NF1 gene has been localized on chromosome 17 and represents 95% of neurofibromatosis cases with the incidence rate of 1 in 3500 newborns and prevalence of 1 in 4500.³

Plexiform neurofibromas represent an uncommon variant (30%) of NF1 in which neurofibromas arise from multiple nerves as deforming masses involving connective tissue and skin, therefore, clinical description of the bag of worms used.⁴

Case Report

A 33 years old male patient presented in our neurosurgical department with the complaint of painless swelling in upper eye-lead since childhood. He had a history of pheochromocytoma which was operated on three months earlier. There was no evidence of pheochromocytoma at the time of the operation and metanephrine, normetanephrine and VMA levels in blood and urine were within the normal range. The diagnosis of NF-1 made according to the presence of two or more criteria of the disease.

Physical Exam

Dermatologic Status

Multiple café-au-lait spots with diameter >1.5 cm on

the chest and back, several neurofibromas on the trunk, axillary and inguinal freckling. The mucous membranes were not affected.

Ophthalmologic Status

Visual acuity was 6/6 in both eyes. A neurofibroma plexiform was present on the upper eye-lead of the right eye.

Head and Neck Examination

He had adequate mouth opening, normal neck and temporomandibular joint movements and a mallampati class 1 on oropharyngeal examination.

Lab and Imaging Studies, Histologic Findings and Consultations

The standard lab test values were within the normal range. CT scan showed soft tissue lesion with widening of orbital canal and mass effect on optic nerve and apex of the orbit. The histological results confirmed the diagnosis of neurofibromatosis (Figure 1).

Anesthetic Implications

General anesthesia was planned for the surgery. After premedication with 20 mg midazolam, 200 µg fentanyl, and 100 mg lidocaine, the induction of anesthesia was done using 100 mg propofol and 50 mg atracurium. The patient experienced transient paroxysmal atrial contractions after intubation which resolved after the administration of the maintenance dose. The maintenance of anesthesia provided with an infusion of 10 mg/kg/h propofol and infusion of 0.05 µg/kg/min remifentanyl. The patient's

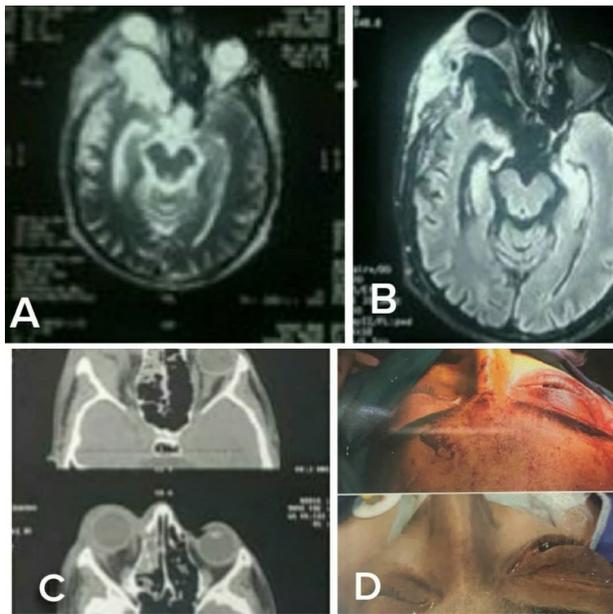


Figure 1. A & B . Magnetic resonance imaging shows hypointense lesions in T1 and hyperintense lesions in T2 posterior to the right orbital cavity. C . In computed tomography a soft tissue lesion with widening of orbital canal and mass effect on optic nerve and apex of the orbit causing proptosis. D . Pre-op and post-op images of the patient with obvious proptosis in right upper eye-lead.

hemodynamic status was normal during the procedure, and the heart rhythm was normal sinus rhythm. Due to the massive amount of bleeding during the evacuation of neurofibroma plexiform, the surgery team decided to postpone the resection of brain mass. The patient received 3 units of isogroup crossmatched packed cells, and the urine output was 400 cc during 5 hours of surgery. General anesthesia administered was uneventful during the second procedure.

Discussion

Neurofibromatosis is an autosomal dominant disease affecting all organs. The diagnosis was made according to the presence of two or more of the following criteria:

- A) 6 or more café-au-lait macules (>0.5 cm in children and >1.5 cm in adults)
- B) 2 or more cutaneous or subcutaneous neurofibromas or one plexiform neurofibroma
- C) Axillary or inguinal freckling
- D) Optic glioma
- E) 2 or more lisch nodules (iris hamartomas visualized on slit lamp examination)
- F) Sphenoid wing dysplasia or bowing of long bone (with or without pseudoarthrosis)
- G) First degree relative with an NF1 diagnosis²

Neurofibromas are the characteristic lesions which can occur anywhere including oropharynx and larynx resulting in difficulty in laryngoscopy.⁵ Painless dislocation

of cervical vertebrae has been reported in a patient with multiple cervical neurofibromas therefore preop radiological exam has been suggested to avoid spinal cord damage during laryngoscopy.⁶ Pulmonary pathologies include pulmonary fibrosis and cystic lung disease. Its cardiovascular manifestations include hypertension which might associate with pheochromocytoma or renal artery stenosis which may be bilateral.⁵ Pheochromocytoma affects between 0.1%-5.7% of patients with NF1 but almost 25% of patients with pheochromocytoma have the disease. The anesthesiologist should also pay attention to the possibility of severe hemorrhage during the surgery on plexiform neurofibromas.⁷ The arterial lesions with different morphologies including fusiform intimal narrowing or nodular or aneurismal formations can be present. CNS tumors account for the major portion of morbidity and mortalities in these patients. Involvement of brainstem structures can cause central hypoventilation syndrome leading to prolonged postop weaning.⁵ The anesthesiologist should take into account the increased risk of epilepsy, learning difficulties and the possibility of undiagnosed CNS tumors during the assessment for patients undergoing surgery due to cranial and spinal masses.⁷

Patient Outcome

The patient discharged without any neurological deficit.

Conflict of Interest Disclosures

The authors declare that they have no conflict of interests.

Ethical Statement

An informed consent form for publication of the study was obtained for the patient.

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