Case Report

Plexiform Neurofibroma of the Face: Challenges Encountered

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Abstract

Plexiform neurofibromatosis is the hallmark lesion of neurofibromatosis Type 1. Facial neurofibromas can be extremely disfiguring and vascular. Surgery for such tumours are challenging and requires multidisciplinary team approach. Inspite of all the efforts surgery may not be successful. Here we present a case of facial neurofibroma and challenges we faced during management of this case.

Key words: Factor VII, massive transfusion, plexiform neurofibroma

Introduction

Neurofibromatosis is a type of benign tumor originating from the nerve sheath cells.[1] It is an autosomal dominant condition affecting about 1:2,500 to 1:3,000 live births.[2] Plexiform neurofibromas are the hallmark lesions of neurofibromatosis type 1. They affect long portions of the nerve, sometimes infiltrating the nerve and the surrounding tissue, thereby giving rise to an extensive disfiguration.[3] Although resection of these highly vascular tumors can be challenging, there are cases where surgeries have been successful. Here, we present a case of plexiform neurofibroma of the face and the challenges we encountered while handling the case.

Case Report

A 23-year-old lady presented to our hospital for reconstruction of her face that was distorted because of plexiform neurofibroma. The lesion involved the entire face [Figures 1 and 2]. She had no other comorbidities. She weighed 42 kg and her height was 152 cm. Her airway examination revealed Mallampati class 3 and an interincisor gap of 3 cm. Neck extension was normal with adequate thyromental distance. Indirect laryngoscopy showed the presence of normal vocal cords but fullness of the left pyriform fossa.

She was accepted for staged reconstruction of the face under American Society of Anesthesiologists (ASA) grade 1. In view of the anticipated difficult mask ventilation and intubation, an awake orotracheal fiberoptic intubation was planned. As part of the preparation, in the preoperative area she received 2% viscous lignocaine mouth gargle and nebulization with 4% lignocaine. An 18-G peripheral cannula was secured and she was briefed about the anesthesia plan. She was premedicated with 0.2 mg of glycopyrrolate intravenously.

In the operation theater, an awake intubation was facilitated by spraying the vocal cords with 2% lignocaine and negotiating the fiberscope through the vocal cords. The awake intubation process was uneventful and once the endotracheal tube was in place, the patient was induced with propofol (100 mg), fentanyl (100 μg), and atracurium (40 mg).

Anesthesia was maintained with oxygen (O_2), air, and sevoflurane. Fentanyl and atracurium were supplemented intermittently. In anticipation of significant blood loss, another 16-G cannula was secured and the radial artery was cannulated.

The first excision was performed on the right side of the cheek. There was a blood loss of about 2 L for the excision of a tissue measuring 15 cm × 3 cm. Two units of packed red blood cells were transfused and her vitals remained stable throughout the surgery that lasted for about 2 h. Multilayered pressure dressing was applied to the surgical site. Extubation was uneventful. She was shifted to the recovery room for observation.

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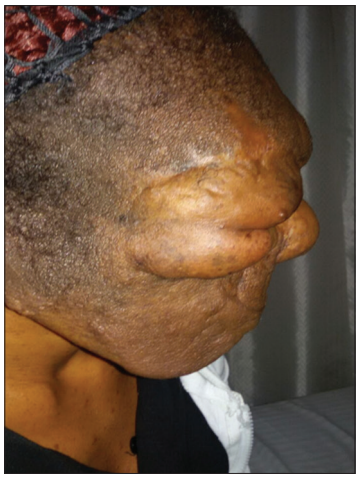

Figure 1: Plesiform Neurofibroma of the face lateral view Figure 1

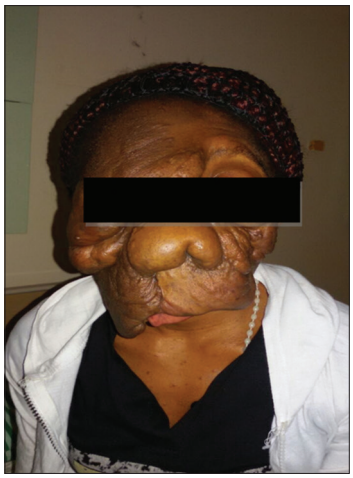

Figure 2: Plexiform Neurofibroma of the face frontal view Figure 2

The first 4 h after the surgery was uneventful. Fentanyl infusion was started at 50 μg/h for pain management. She was receiving supplemental O_2 via Hudson's mask at 5 L/min. There was minimal soakage of the dressing. Her vital parameters were normal except tachycardia (110-120/min). After the 4-h observation period, she was becoming restless. As there was a language barrier, the exact reason for her restlessness was not known but as obvious, it was attributed to pain and bolus fentanyl (50 μg) and midazolam (1 mg) were administered. Tachycardia persisted and there was a slight fall in her blood pressure to 100/70 mmHg. As the patient started to desaturate in spite of supplemental O_2, laryngeal mask airway (LMA) was inserted to facilitate ventilation. Difficult airway cart was mobilized. Intubation was successful by a senior anesthetist requiring McCoy's blade and bougie. There was no increase in the soakage of the dressing.

We shifted the patient to the intensive care unit (ICU) and after about 1 h her blood pressure started to drop, heart rate started to increase, and arterial blood gas (ABG) showed a hemoglobin level of 3.3 g%. The patient was immediately taken up for re-exploration while arrangements were made for additional blood and blood products. A central line was inserted. There was blood loss of about 3.5 L intraoperatively. Nine packed red blood cells (PRBCs), nine platelets, nine fresh frozen plasma (FFP) cells, and six cryoprecipitates were transfused. As there was diffuse ooze, factor VII was considered. It was given in a dose of 1 mg. She required inotropic support throughout the re-exploration. After adequate hemostasis was achieved, she was shifted back to the ICU for further ventilator support and care.

In the ICU, she was weaned of inotropic support over the next 48 h but required ventilator support for 6 days. In view of the difficult airway, tracheostomy was done. Further recovery was uneventful and she was discharged after 1 month of hospital stay.

Discussion

A neurofibroma is a nerve sheath tumor occurring in the peripheral nervous system, inherited as an autosomal dominant trait. They can result in a range of symptoms from physical disfiguration and pain to cognitive disability. Neurofibromas have been subdivided into two broad categories: Dermal and plexiform. Dermal neurofibromas are associated with a single peripheral nerve while plexiform neurofibromas are associated with multiple nerve bundles.[4]

Plexiform neurofibroma is generally considered a hallmark of type 1 neurofibromatosis. Airway assessment is important when it involves the face and the oropharyngeal structures. Intraoral manifestation is seen in around 5% of patients.[5] Plexiform neurofibromas can undergo malignant transformation.

In addition to the careful airway assessment, a thorough systemic examination is equally important. Pheochromocytoma and renal artery stenosis can be associated with plexiform neurofibromatosis. In the presence of preoperative hypertension, these causes should be looked into. It is important to discuss the anesthesia plan, the need for blood transfusion, postoperative ICU care, and prolonged hospitalization.

An awake intubation was done in our case anticipating difficult mask ventilation and intubation. But in the recovery room when airway had to be secured in view of desaturation, initial intubation was not successful and LMA was used to facilitate ventilation. The role of this supraglottic airway device as a part of difficult airway management is commendable. A difficult airway cart should be easily accessible. The cause for postoperative desaturation was attributed to fentanyl infusion.

The need for perioperative transfusion was discussed with our patient but she required massive blood transfusion during the re-exploration. While the vascular surgeon and interventional radiologist were called, factor VII was considered as a desperate measure to control the ongoing hemorrhage. Patients with neurofibromatosis may have associated coagulation abnormality.[6-8] The role of factor VII in surgical trauma is debatable. Whether the patient had some primary coagulation abnormality or massive hemorrhage-induced coagulopathy that got corrected after administration of factor VII is not known.[9] But in our case it was beneficial and helped in reducing further

blood loss. Postoperatively, further investigation to identify any coagulation abnormality was not done as the patient was not willing.

In the postoperative phase, she showed gradual improvement and was discharged about a month later. Prolonged hospitalization and slow recovery led us to think whether it was worth putting this lady at risk.

This case highlights the limitations of ASA grading. It offers inadequate distinction between the presence of a systemic illness and localized manifestation of a systemic illness. It also fails to incorporate the airway grading. A difficult airway may predispose to increase risks for the patient. Also, the grading system does not include the nature surgery, which, by itself, is a predictor of morbidity and mortality.

Conclusion

The treatment of plexiform neurofibromatosis is challenging for the surgical and anesthesia team. In addition to the airway management, intraoperative need for invasive monitoring and transfusion of blood and blood products are also important. Although the role of factor VII cannot be justified, it was beneficial in our case. With the possibility of high recurrence rate and associated perioperative complications, it is questionable whether these patients should be accepted for reconstructive procedures without prior vascular intervention.

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Conflicts of interest

There are no conflicts of interest.

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