Proseal laryngeal mask airway with stylet in drain tube technique in a child with Sturge–Weber syndrome

Sturge-Weber syndrome (SWS), a neurocutaneous disorder, presents with a facial port-wine stain in the ophthalmic distribution of the trigeminal nerve, glaucoma, vascular eye abnormalities, or an ipsilateral occipital leptomeningeal angioma.[1] The varied presentation in the patients, from localized superficial skin lesions to extensive systemic involvement, makes the anesthetic management challenging.[2] Anesthesiologists should plan in advance to avoid trauma to the hemangioma, increase in intraocular and intracranial pressure.[3] The key to safe anesthetic management in such patients is by controlling the convulsive disorder perioperatively, attenuation of hemodynamic responses during airway manipulation and surgery, careful intubation/extubation, avoiding trauma to the angiomatous lesions, and adequate pain relief.[3]

A 2-year-old, male, known case of SWS was posted for syringing and probing for nasolacrimal duct obstruction. Detailed preoperative assessment revealed history of delayed milestones and recurrent episodes of focal motor seizures with last episode 5 months back (on phenytoin). The mouth opening was 1.8 cm with multiple hemangiomas observed in oral cavity [Figure 1]. There were three more hemangiomas on chest, arm, and face. Systemic examination did not reveal anything significant. The computed tomography scan of brain revealed choroidal hemangioma.

We planned to use a supraglottic device (ProSeal laryngeal mask airway [PLMA]) for intraoperative ventilation. After preoxygenation, attaching the achieving inhalational induction of anesthesia with sevoflurane and securing an intravenous line Inj. Glycopyrrolate (5 μg/kg) and inj. Fentanyl (2 μg/kg) were given as premedication. A PLMA size 2, with stylet in situ technique, was used to secure the airway. In this technique, stylet was introduced through drain tube till the distal-most end of PLMA. The PLMA was bent 90° at shaft and adequate lubricant jelly was applied on the dorsal surface [Figure 1]. PLMA was inserted very gently avoiding any vigorous movement. After removal of stylet, PLMA’s position was confirmed by insertion of adult fiberscope. Anesthesia was maintained with sevoflurane (1–1.5 MAC), 66% nitrous oxide in oxygen with spontaneous ventilation. At the end of approximately, 25-minute, the PLMA was gently removed in deeper plane and postextubation oxygenation for 5 minutes was done. The recovery was uneventful and patient was discharged after 6 hours.

Angiomas in SWS may be found on the trunk and extremities and may involve the mucous membrane of nose, gingiva, palate, tongue, larynx, and trachea.[3] Only 8%–20% of...
patients with facial port-wine stain, with and without ocular involvement, develop neurological symptoms. In airway management, there can be difficult mask ventilation, difficult laryngoscopy, difficult supraglottic airway device (SAD) insertion, or difficult intubation. SAD has been successfully used for airway management in these patients if inserted carefully. Furthermore, it prevents increase in the intraocular and intracranial pressures by facilitating smooth insertion and extubation. Endotracheal intubation if required should be done by an expert anesthesiologist with a careful laryngoscopy and using endotracheal tube without stylet to prevent rupture of the airway angiomas. In this patient, the problems were anticipated pre-emptively. There was difficult mask ventilation with angioma over lips, difficult laryngoscopy, difficult supraglottic insertion, and difficult intubation with intraoral angiomas. Mouth opening was 1.8 cm, and thus, gentle PLMA insertion with “stylet in drain tube” approach was used. Pediatric fiberoptic was not available at our setup, and moreover, the passage of endotracheal tube over fiberscope in such patients can lead to severe intraoral bleeding. We just confirmed the position of PLMA with our adult fiberscope and graded it accordingly. A video-laryngoscope-guided intubation might have been another better alternative.

In conclusion, a safe and effective anesthetic management and thorough understanding of SWS are required for management of airway in a patient with SWS. Managing the hemodynamic responses during the airway manipulation and surgery with careful intubation/extubation so as to avoid trauma to the lesions, with adequate pain relief, will contribute to the safe anesthetic management of such patients.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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References
1. Chaudhary SC, Sonkar SK, Kumar V, Golchha S. Sturge Weber syndrome. J Assoc Physicians India 2011;59:327-9.
2. Ramanathan G, Prasanna, Karunanithi P, Venkatraman R. Anaesthetic management of a case of Sturge-Webers syndrome. Indian J Anaesth 2012;56:302-4.
3. Batra RK, Gulaya V, Madan R, Trikha A. Anaesthesia and the Sturge-Weber syndrome. Can J Anaesth 1994;41:133-6.
4. Shirley MD, Tang H, Gallione CJ, Baugher JD, Frelin LP, Cohen B, et al. Sturge-Weber syndrome and port-wine stains caused by somatic mutation in GNAQ. N Engl J Med 2013;368:1971-9.

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