CASE REPORT

Anesthesia and Airway Management of a Patient with Sturge- Weber Syndrome Associated with Extreme Ocular Manifestations and Giant Facial Hemangioma

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Sturge-Weber syndrome (SWS) is a very rare congenital disorder that is manifested by facial capillary malformation (port wine stain) which may be associated with capillary – venous malformations affecting many parts such as the brain, eyes facial skin and mucosa and also airways. In this report we present an unusual 28 years old female case of SWS that presented with signs and symptoms of sudden increased intra ocular pressure (exacerbation of glaucoma) and manifestations of facial and airway involvement. A discussion about anesthetic and airway management of such patient follow the case presentation.
Keywords: sturge–weber syndrome; airway management; hemangioma

Sturge-Weber Syndrome (SWS) is a very rare congenital neuro-occulo coetaneous disorder that is manifested by facial capillary malformation (port wine stain). The disorder may be associated with capillary-venous malformations affecting many parts such as the brain, eyes, facial and airways [1-2].

The cause of SWS is somatic mosaic mutation in the GANQ gene. The GANQ gene encodes a guanine nucleotide binding protein named as G c- alpha – q that regulates intracellular signaling pathways. Somatic mutations in GANQ occurring at a later stage in embryogenesis may affect only precursors of vascular endothelial cells and lead to non- syndromic port wine stains, while those occurring during earlier stages may affect a greater variety of precursor cells and lead to SWS [1].

SWS characterized by a facial capillary malformation (port - wine stain) may be associated with leptomeningeal angioma which involves eyes and the brain. These vascular manifestations are associated with neurologic and ocular abnormalities. The ocular component of the disorder manifests as glaucoma and vascular manifestations of conjunctiva, episclera choroid and retina [2-3]. SWS may involve airways and upper respiratory tract. Oral soft tissues larynx and trachea may involve as a consequence of multiple angiomatous lesions [4].

Case presentation

A 28 years old woman presented at operating room for an acute exacerbation of angle closure glaucoma. She was a well- known case of Sturge-Weber Syndrome (SWS) with neuro–occulo- coetaneous involvement. Neurological involvement consists of a history of intermittent convulsive attacks and mental retardation. Ocular involvement began symptomatic at her adolescence age as increased intra ocular pressure and signs and symptoms of angle closure glaucoma. The disease was under control by medical treatment and by regular ophthalmologic examinations and follow-up. Coetaneous involvement of her SWS consists of a huge facial involvement with extension in to buccal and hard palate mucosa. No other important positive medical or surgical histories were presented. Because of acute uncontrollable exacerbation of glaucoma she was candidate for an emergency trabeculotomy procedure. On arriving at the operating room a fast and thorough examination was done. The patient was considered as a probable difficult ventilation and difficult intubation case due to a Malampati IV Cormac III and ULBT II classification (Figure 1).

Figure 1- General appearance of the patient. Note the altered anatomy of face and oral cavity.

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After about 5 minutes of pre-oxygenation by using a No. 4 face mask, induction of anesthesia wasperformed by using regular dose of fentanyl and midazolam as premedication followed by intubating dose of atracurium (using priming approach) and propofol. As laryngoscopy and airway access seemed to be difficult, the airway was secured with some difficulties in positioning a no.3 laryngeal mask airway. (Figure 2-4)

Figure2- The patient at beginning of anesthesia. LMA secured at proper position.

Figure3- The patient at beginning of anesthesia (lateral view).

Figure 4- The patient at termination of surgical operation and at the time of LMA removal.

All available alternative approaches for managing difficult airway as well as devices for fiberoptic- guided intubation and crico-thyrotomy were present in the operating room for any untoward or unexpected events. After ensuring the airway security, maintenance of anesthesia was continued by total intravenous anesthesia (TIVA) method using propofol and remifentanil. The trabeculectomy procedure continued for about 40 minutes and after terminating the surgical procedure, delivery of intravenous anesthetic agents were discontinued and the laryngeal mask airway removed after ensuring normal and suitable respiration, consciousness and general conditions (Figure 5). The patient was under observation at PACU for about 6 hours and then referred to the ward after ensuring normal and stable respiratory and general conditions.

Figure 5- The patient after removal of LMA. Note the gross discrepancy between No. 4 face mask and altered oral and facial anatomy.

Discussion

Sturge-weber syndrome is a rare congenital disorder which is characterized by facial capillary malformations and capillary– venous manifestations affecting the brain and the eye [1]. Because of the nature of the disease many patients present with signs and symptoms of eye involvement [2-3]. Both kinds of glaucoma can be seen in these patients and surgical interventions may be mandatory especially when uncontrolled intraocular pressure (IOP) rising causes fear of visual complications such as blindness. Our case presented with all typical signs and symptoms of the syndrome and exacerbation of her pre-existing glaucoma mandated an emergent surgical intervention [2-3].

Facial and airway involvement especially in case of giant facial hemangiomas with airway involvement can cause a true anesthesia challenge in the field of airway management especially in urgent and/ or emergency cases. In our case presentation of giant facial hemangioma and airway involvement potentially caused a difficult mask ventilation, difficult intubation and hard to manage anesthesia [4].

Some clinical guidelines as well as algorhythms may help the anesthesiologist for detecting and managing difficult airway. Mallampati classification, Cormack- Lehane grading and upper lip bite test are among the most important and well recognized scoring systems for detecting and managing difficult air way [5-8]. Early detection of probable difficult airway is the most important point for proper management

of it and preparing accessory devices for difficult airway management is the corner stone of any interventional airway management. We made a thorough medical examination before any intervention in our case for insuring the patency of the airway as well as the possibility of safe management of it by preparing all available airway management devices [9-11].

Every effort has to be made in preventing even short periods of hypoxemia during anesthesia management in a suspected difficult airway patient. So accessibility of main and accessory airway devices as well as suitable and safe oxygen supply and accessory oxygen cylinders are very important [5]. Airway trauma during interventional management of airway may cause airway edema, swelling, hemorrhage leading to partial or total obstruction of the airway and their untoward complications. An expert anesthesiologist has to pay attention to gentle but precise airway intervention in preventing any trauma to the airway. In case of any complications during intubation or maintaining the airway, early detection and suitable management have vital roles [12].

In such cases preparing for aggressive and interventional approaches for securing the airway are of prime importance. Preparing for a surgical airway or alternatively an emergent cricothyrotomy or percutaneous dilatational tracheostomy must be considered in mind and all instruments and devices have to be presented at operating room or at bedside [13-14]. Post extubation and post anesthesia periods are very important as respiratory depression and/or obstruction may cause serious complications and everybody must keep in mind that any probable problems in breathing have to be monitored and detected as soon as possible. Precise post anesthesia care unit (PACU) care can reduce such probabilities.

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