Anesthetic Management of a Child with Sturge-Weber Syndrome Undergoing Glaucoma Surgery: A Case Report

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Abstract

Background: Sturge-Weber Syndrome (SWS) is a rare congenital neurocutaneous disorder characterized by leptomeningeal and facial hemangiomas, with consequent neurological and ocular manifestations. The anesthesia of these patients may be challenging and patients with SWS need careful assessment during the perioperative period.

Aim: The aim is to report on a case of successful anesthetic management for glaucoma surgery in a 3-year-old patient with Sturge-Weber Syndrome.

Methods: All the medical procedures on our patient were performed in the Sestre milosrdnice University Hospital Center. The patient first underwent anesthesia to have her eyes examined and then to have glaucoma surgery. General anesthesia was introduced during both procedures.

Results: During both procedures, inhalational anesthesia with 8 Vol% sevoflurane was performed with the intraoperative addition of fentanyl for analgesia. Induction, as well as anesthetic emergence, went smoothly, without blood pressure oscillations. Maintaining blood pressure within normal limits was important, considering the risk of hemangioma rupture. Ibuprofen and paracetamol ensured adequate postoperative analgesia and no rises in blood pressure.

Conclusion: Since clinicians rarely encounter patients with SWS, it is important for all physicians involved in the treatment of such patients to become familiar with the challenges during the perioperative period. Therefore, we should like to emphasize specific anesthetic considerations for patients with SWS.

Keywords: Sturge-Weber Syndrome, anesthesia, glaucoma surgery

Introduction

Sturge-Weber Syndrome (SWS), also known as encephalotrigeminal angiomatosis, is a rare congenital neurocutaneous disorder that affects only one in 50,000 newborns. The etiology is yet unknown, but it is known that SWS is caused by a mutation in a GNAQ gene that occurs randomly in a developing embryo (1). It can be classified as trisymptomatic, bisymptomatic or monosymptomatic, depending on whether the skin, eyes or central nervous system (CNS) is affected (2). The most common types of vascular malformations are leptomeningeal and facial angiomas (3,4). Facial angiomas are often located around the eyes, so SWS frequently includes ocular manifestations, such as glaucoma, buphthalmos, choroidal hemangioma and strabismus. Glaucoma affects about 30–70% of such patients. In about 60%, glaucoma is present at birth, while in other cases it manifests later in life. The first-line treatment for congenital and early-onset glaucoma with associated angle abnormalities is surgical intervention. A goniotomy or trabeculotomy is usually performed. For late-onset glaucoma, surgery is indicated if conventional therapy is ineffective (2).

During the perioperative period, it is mandatory to ensure adequate pre-medication, as well as smooth induction, maintenance and emergence from anesthesia to avoid an increase in blood, intraocular and intracranial pressure in order to prevent hemangioma rupture. Additionally, careful assessment of the airway is necessary due to the possible presence of angiomas in the oral cavity that can compromise the airway and cause difficulty in tracheal intubation.

Case report

We hereby report a case of a 3-year-old girl (weighing 14 kg) with SWS, who first underwent anesthesia to have her eyes examined and then, 2 weeks later, to have glaucoma surgery. All the medical procedures on our patient were performed in the Sestre milosrdnice University Hospital Center. Since our patient was a 3-year-old girl, her mother gave her informed consent for us to report on her syndrome and the anesthetic management for both procedures. The girl had the following signs of SWS: hemiatrophia of the right side of the brain with left-sided hemiparesis, symptomatic epilepsy (seizures having been experienced from birth, with the last one taking place 6 months before the introduction of the anesthesia), left-sided facial hemangioma affecting all three branches of the trigeminal nerve, myopia and congenital glaucoma of the right eye. Magnetic resonance imaging of the brain showed right-sided cerebrocortical atrophy and right-sided frontotemporoparietal calcifications. Additionally, her medical history revealed hypothyreosis. Her mother reported that the girl was allergic to amoxicillin/clavulanic acid. The girl was on the following therapy: valproic acid, clonazepam, levothyroxine, and the topical ocular drugs dorzolamide/timolol and brimonidine/tartrate. Before surgery, routine blood and urine tests and ECG findings were normal, except for elevated thyroid-stimulating hormone (TSH), so the dose of levothyroxine was increased before the introduction of the anesthesia. The mouth opening was adequate (Mallampati grade I), without any visible oropharyngeal hemangiomas. An antiepileptic drug and levothyroxine were continued until the day of surgery.

General anesthesia was introduced during both procedures. The patient was pre-medicated with 4.15 mg oral midazolam 45 min before both anesthesia inductions. Vital parameters were monitored through non-invasive blood pressure monitoring, ECG monitoring, and SpO2 and etCO2 monitoring. The induction was performed by 8 Vol%-sevoflurane inhalation via a face mask, and when the girl was sedated deeply enough, a 24 G-venous cannula was inserted and an infusion of Ringer solution was started. As an induction dose, 25 mcg of fentanyl was administered. In the case of the second anesthesia induction that preceded the glaucoma surgery, 8 mg of rocuronium was administered after fentanyl. On the occasions of both surgeries, a No. 2.5-laryngeal mask was inserted. Before the insertion of the laryngeal mask during the first anesthesia

introduction, a gentle laryngoscopy was performed to evaluate the airway patency; no oropharyngeal hemangiomas were found (Comrack-Lehane grade 1). The anesthesia was maintained with inhalational sevoflurane, oxygen and air, thereby keeping the patient hemodynamically stable. Intraoperative systolic pressure was maintained at around 90 to 95 mm of mercury, and diastolic at around 45 to 55 mm. During the second general anesthesia, fentanyl 50 mcg was re-administered intraoperatively. At the end of the second procedure, a 12.5 mg diclofenac sodium suppository was inserted to ensure initial pain relief. After both procedures, anesthesia emergence went smoothly, without any increases in blood pressure. The surgical procedure went without any complications. Further postoperative pain relief was achieved with 100 mg of ibuprofen syrup, three times per day, and a 120 mg paracetamol suppository, three times per day, as needed. With this therapy, postoperative pain was adequately relieved and no hemodynamic changes were present. The patient was discharged home on the fourth postoperative day.

Discussion

Sturge-Weber Syndrome (SWS) can affect the skin, eyes and/or CNS (2). Neurological manifestations include hemiparesis, hemianopsia, stroke-like episodes, headaches, seizures and mental retardation. Facial angiomas (port-wine stains, PWSs) often involve the eyes, mouth and different parts of the airway. Angiomatous lesions of the skin usually follow the trigeminal nerve distribution pattern (5), so SWS is also called encephalic-trigeminal angiomatosis (6). Ocular manifestations can include glaucoma, buphthalmos, choroidal hemangioma and strabismus.

When it comes to SWS patients, anesthesia may be challenging for several reasons, as discussed below.

Before introducing anesthesia, adequate pre-medication should be administered, in order to avoid increases in blood pressure and possible hemangioma ruptures. Angiomas seated in the upper respiratory tract may cause ventilation difficulties. During the insertion of an airway device, hemangioma rupture involving the airway may occur, making mask ventilation, laryngoscopy and intubation difficult, so the equipment needed for endotracheal intubation must always be ready, should an emergency arise (7). An experienced anesthetist should attempt the intubation using a well-lubricated, cuffed endotracheal tube without stylettes (5). In the case of our patient, the nature of the procedure did not require intubation, so a laryngeal mask was used. An airway examination revealed no visible hemangioma in the oral cavity, but the possibility of the presence of an angioma in the pharynx was also taken into account, so a difficult airway cart was kept ready. The authors think that a laryngeal mask represents a safer and a more suitable airway securing option, provided that the nature of the surgery allows for it and that the patient has no angiomas in the oral cavity, although, in most of the cases reported in the literature, an endotracheal tube was introduced despite the absence of angiomas in the oral cavity (5,8,9). If an airway evaluation reveals the presence of angiomas in the oral cavity, an introduction of an endotracheal tube is a better choice, indeed, since it exerts pressure on a smaller oral cavity area as compared to a laryngeal mask and, therefore, makes oral cavity-seated hemangiomas less prone to rupture.

To prevent angioma ruptures, it is necessary to achieve adequate hemodynamic stability, during both anesthesia induction and maintenance (5). Blood vessels composing hemangiomas have abnormal auto-regulation. Any increase in blood pressure is associated with an outspreading of angiomas that can cause massive bleeding due to hemangioma rupture (6).

Ocular manifestations, primarily the presence of glaucoma, demand smooth anesthesia induction without any rise in the intraocular pressure. During anesthesia, it is necessary to avoid drugs that can raise intraocular pressure, such as succinylcholine, ketamine and atropine, and events that can do the same, such as hypoxia, hypercarbia

and hypertension. Anticholinergics should be avoided in patients with narrow-angle glaucoma (8). During eye surgery, rupture of a choroid hemangioma can be witnessed, which leads to excessive bleeding (6).

SWS patients such as ours often report suffering from epilepsy, so the administration of antiepileptic drugs should be maintained until the day of surgery and continued on that day. Some antiepileptic drugs, such as the valproic acid taken by our patient, affect coagulation so an extensive coagulation workup should be completed prior to major surgery expected to cause substantial bleeding. During anesthesia, hypoxia, hypoglycemia, hypotension and hyperthermia may precipitate an epileptic state and should, therefore, be avoided. Also, ketamine and etomidate should not be used during anesthesia induction, since they may provoke seizures. Concomitant chronic use of antiepileptics can affect the metabolism of some anesthetic agents (8).

Succinylcholine is usually avoided in hemiplegic patients because of its effect on serum potassium levels (6).

Vascular changes may also affect other organs (8). Angiomatous changes have been reported in the pituitary gland, thyroid gland, thymus, lungs, spleen, testicles and lymph nodes (10). In our case, a thorough evaluation ruled out the possibility of the presence of such lesions. The major challenge we had to overcome during both procedures was the placement of the intravenous route. Although the girl was of a normal body weight for her height, her blood vessels were virtually invisible and extremely gracile, so we were left with no other option but to place a 24 G-cannula. During the first anesthesia introduction, we made several attempts to place the cannula and finally succeeded in inserting it into the left radial cutaneous vein. With the second procedure, cannula placement was equally challenging.

Postoperative care should also ensure adequate pain relief, since pain raises blood pressure and might lead to hemangioma rupture.

Conclusion

Patients with SWS need careful assessment prior to anesthesia induction. Adequate pre-medication and perioperative convulsion control are imperative. Anesthesia should be planned in a manner that allows for the avoidance of trauma to eventual hemangiomas seated in the airway. Any drug or occurrence that can raise blood, intraocular or intracranial pressure should be avoided. Throughout induction/intubation and extubation, and the entire course of surgery, hemodynamic and blood pressure stability should be maintained. Moreover, adequate pain management should be provided during the postoperative period. Although the case reports published so far have neglected to mention that intravenous route placement might pose a difficulty, this was seen with our patient.

Declarations

Authors' contributions

All the authors made substantial contributions to the conception or design of the work, were involved in drafting the work or revising it critically for important intellectual content and gave final approval of the version to be published.

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Competing interests

The authors have no conflict of interest to declare.

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