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CASE REPORT

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Anesthetic management of a patient with Sturge–Weber syndrome in sagittal split ramus osteotomy surgery

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Key Clinical Message

Sturge–Weber syndrome (SWS) is a neurocutaneous syndrome characterized by angiomas. This report presents airway management using submental intubation in sagittal split ramus osteotomy under general anesthesia and aimed to explore better anesthetic management for avoiding the rupture of angiomas in a patient with SWS.

KEYWORDS

airway management, general anesthesia, intubation, sagittal split ramus osteotomy, Sturge–Weber syndrome

1 | INTRODUCTION

Sturge–Weber syndrome (SWS) is a neurocutaneous syndrome characterized by facial and leptomeningeal angioma, glaucoma, seizures, and neurological disability.[1] It is a rare nonhereditary condition with an incidence of 1 per 50,000 live births. It has no racial or sex predilection.[2] Cutaneous angiomas called port-wine stains (PWS) are present in 96% of patients with SWS.[3] They are usually distributed unilaterally along dermatomes supplied by the ophthalmic and maxillary parts of the trigeminal nerve.[4] However, they are sometimes bilateral or involve the neck, limbs, or other body parts.[5]

Intraorally, angiomas may involve not only the lips, buccal mucosa, palate, gingiva, and floor of the mouth but also the pharynx, larynx, trachea, and bronchi.[5,6] Anesthetic techniques such as tracheal intubation may cause the rupture of angiomas, which may lead to uncontrolled hemorrhage.

In addition, oral changes have been reported in 40% of patients with SWS because of gingival overgrowth and asymmetric jaw growth.[7] Soft or hard tissue hypertrophy often requires soft tissue correction and bone surgery.[8] Furthermore, these oral changes may make mask ventilation and intubation difficult.

The anesthetic management of patients with SWS has been reported in previous studies.[3,6,9] However, data on the anesthetic management of patients with SWS are limited in oral surgery, even though patients with SWS are predicted to be adapted to orthognathic surgery because of their asymmetric jaw growth. This report presents airway management using submental intubation (SMI) in a patient with SWS in sagittal split ramus osteotomy (SSRO) under general anesthesia. This report aimed to explore better anesthetic management for avoiding the rupture of angiomas in a patient with SWS.

Informed consent for case reporting was obtained from the patient and his parents.

2 | CASE HISTORY

A 20-year-old man (height: 156 cm; weight: 58 kg) was admitted to our institution for SSRO surgery under general anesthesia due to mandibular protrusion. He had a history of SWS and had swelling of the upper lip and maxillary gingiva due to angiomas, bilateral facial PWS, and right hemiparesis (Figure 1). The patient also had a history of glaucoma and retinal detachment.

3 | METHODS

When we saw his face, we realized that mask ventilation might be difficult because his upper lip was markedly swollen and protruded due to angiomas. Therefore, we checked the fitting of a large face mask (Air Cushion Face Mask, Large size for adults: KOO MEDICAL JAPAN, Tokyo, Japan) and total-face mask (PerformaxSE, XL size: Philips Respironics, Tokyo, Japan) for mask ventilation at induction.

In the preoperative evaluation, the patient's blood pressure, pulse rate, and peripheral arterial oxygen saturation (SpO_2) were 121/69 mmHg, 69 bpm, and 97%, respectively. Laboratory tests and chest radiography showed no abnormalities. Electrocardiogram showed sinus rhythm. The brain magnetic resonance imaging scan showed angiomatosis along the cerebral gyrus and brain atrophy. The neurosurgeon instructed us to carefully control blood pressure during the perioperative period because the rupture of angiomas may lead to life-threatening hemorrhage.

We consulted an otolaryngologist to evaluate the nasal cavity and larynx before surgery because airway management in SSRO surgery should be performed by nasal intubation to check the bite during surgery. Computed tomography scans revealed nasal septal curvature and remarkable bony thickening of the inferior nasal dorsum on either side (Figure 2). Fiberscope examination revealed angiomatosis in front of the inferior nasal dorsum. No problems were observed around the larynx. The otolaryngologist suggested that nasal intubation was very severe in this case. Thus, we planned to perform airway management during surgery by SMI, not nasal intubation.

FIGURE 1 The patient's facial appearance. Bilateral port-wine stains and marked swelling of the upper lip.

We also consulted a urologist to evaluate the ureter and bladder using cystoscopy to avoid the rupture of angiomas by urinary drainage. Examination revealed no angiomas or stenosis in the ureter or bladder.

This patient had a history of glaucoma and retinal detachment, for which he underwent ophthalmic surgery under general anesthesia at the ages of 10, 12, and 16 years, and he had been on acetazolamide (250 mg/day). Furthermore, he had a history of epilepsy. However, seizures had not occurred for more than 10 years without medication. His intellectual ability was borderline, and he showed no problems in verbal communication.

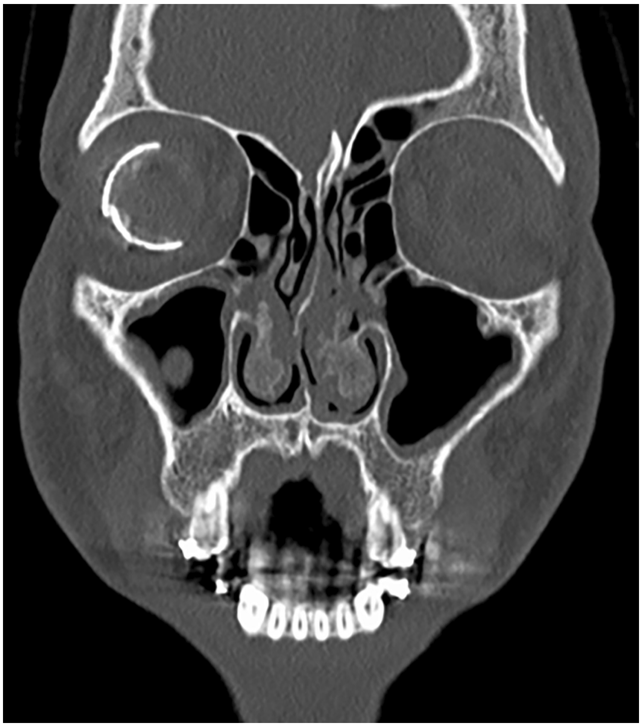

FIGURE 2 Computed tomography image of the patient. The patient's nasal septal curvature and remarkable bony thickening of the inferior nasal dorsum on either side.

No premedication was provided. Noninvasive blood pressure, electrocardiogram results (lead II), oxygen saturation, and muscle relaxation were monitored during general anesthesia. Furthermore, the depth of anesthesia was monitored using an entropy monitor (GE Healthcare, Chicago, IL, USA). The baseline blood pressure, heart rate, and SpO_2 were 123/66 mmHg, 92 bpm, and 99%, respectively, before induction. Oxygen was delivered via a face mask at a flow rate of 6 L/min, and Ringer's acetate solution was infused. Rapid induction was performed with propofol (target concentration 4 μg/mL) and remifentanil (30 μg and 0.35 μg/kg/min). Mask ventilation was possible when a face mask (large size) with an oral airway was used, but a little pressure was required on the upper lip (Figure 3A). Therefore, we changed to a full-face mask (extra-large size) to avoid pressure on the upper lip. The full-face mask allowed us to perform mask ventilation without any pressure on the upper lip (Figure 3B). After we confirmed that we could perform mask ventilation, rocuronium (30 mg) was administered. Oral intubation was performed using a video laryngoscope (McGRATH™ MAC, Aircraft Medical, Edinburgh, UK), and we visually confirmed that there was no angioma in the oro- and laryngopharynx at that time. A wire-reinforced tube (7.0 mm I.D.) was placed into the trachea. After that, general anesthesia was maintained with propofol (target concentration 2.5–2.8 μg/mL) and remifentanil (0.13–0.3 μg/kg/min).

The SMI method was initiated. The surgeon made an incision in the submental skin, and a Kelly forceps was bluntly introduced through the submental incision to the floor of the mouth. Then, the Kelly forceps was replaced with 2.5, 5, and 10 mL syringes to enlarge the tunnel. Ventilation with 100% oxygen was performed to avoid apnea in the following procedures. The endotracheal tube

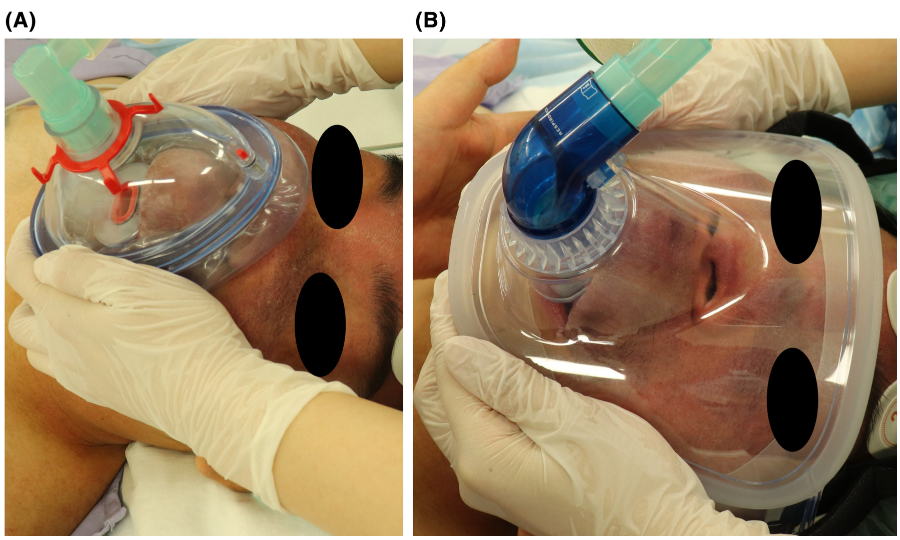

FIGURE 3 Mask ventilation. (A) A face mask (large size) was used with an oral airway. (B) A full-face mask (extra-large size) was used.

(ETT) was removed from the ventilatory circuit. A sterilized echo probe cover was then placed over the ETT and held with a rubber band at the base of the pilot balloon (Figure 4). A Kelly forceps was inserted from the floor of the mouth to grasp the echo probe cover. The echo probe cover placed on the pilot balloon was pulled through the enlarged tunnel of the floor of the mouth using a Kelly forceps, and the ETT was allowed to pass through the tunnel. The ETT was reconnected to the ventilatory circuit. Apnea duration was 60 s, and no decreased SpO_2 was observed. We confirmed with bronchial fibers that the tip of the ETT was properly positioned even when the mouth was opened and closed, and the tube was secured to the skin with a 2–0 silk suture (Figure 5A).

FIGURE 4 Pilot balloon of the endotracheal tube (ETT). The sterilized echo probe cover was placed over the ETT and held with a rubber band at the base of the pilot balloon (white arrow).

After the completion of the SMI, the scheduled SSRO surgery was performed. The tube did not interfere with the operation during surgery, and no accidental extubation occurred. The patient's blood pressure, pulse rate, SpO_2, and $EtCO_2$ were 90–120/40–70 mmHg, 70–90 bpm, 98%–100%, and 35–45 cm H_2O, respectively, and no complications were not observed during surgery.

Immediately after the surgery, ventilation with 100% oxygen was performed to transfer the SMI from submental to oral intubation. In this process, the ETT's connector was stuck in the tunnel because of its wings (Figure 5B). Therefore, the transition took a little bit longer than usual. The apnea duration was 90 s. However, no decreased SpO_2 was observed. After that, the tunnel was closed with sutures.

The anesthetics were stopped after the visual confirmation of hemostasis in the oral cavity. The patient was extubated after confirmation of the recovery of spontaneous respiration, body movement, spontaneous eye-opening, and deglutition reflex. After extubation, the patient was transferred to the ward with stable vital signs. The surgical time, anesthetic time, and bleeding volume were 191 min, 254 min, and 36 mL, respectively. The patient was discharged from the hospital on postoperative Day 11 without any complications.

4 | CONCLUSION AND RESULTS

We experienced anesthetic management of a patient with SWS in SSRO. Anesthetic management with avoiding the rupture of angiomas, controlling epilepsy, and avoiding

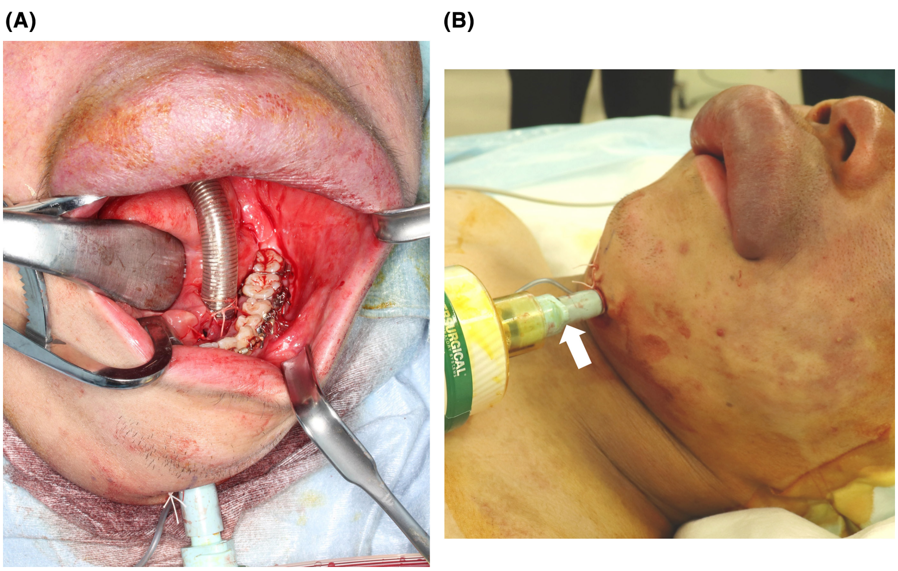

FIGURE 5 Intraoral and extraoral views of the submental intubation (SMI). (A) Intraoral view of the SMI. (B) Extraoral view of the SMI and the wings on the endotracheal tube connector (white arrow).

raising intraocular pressure (IOP) should be performed to achieve safe general anesthesia. Anesthesiologists and surgeons should be consulted to confirm the location of angiomas as far as possible before surgery and apply various strategies to patients to avoid the rupture of angiomas.

5 | DISCUSSION

SWS is characterized by angiomas involving the brain, face, and eye. The rupture of angiomas may result in uncontrolled hemorrhage and may be fatal. Therefore, the rupture of angiomas should be avoided in anesthetic management.

In this case, mask ventilation was difficult because his upper lip was markedly swollen and protruded due to angiomas. Therefore, we checked mask fitting and prepared a large mask and total-face mask, which is often used for noninvasive positive pressure ventilation therapy. We could perform mask ventilation with a large mask. However, we needed to use an oral airway, which could cause pressure on the swollen upper lip. The pressure on the lip might cause the rupture of angiomas. Therefore, we changed to a total-face mask for mask ventilation. The total-face mask allowed us to perform mask ventilation without an oral airway and avoid pressure on the lip.

Generally, airway management is performed with nasal intubation in SSRO because surgeons should check the bite during surgery. However, in this case, the otolaryngologist suggested avoiding nasal intubation because of the angiomatosis in the nasal cavity and the remarkable bony thickening of the inferior nasal dorsum. In such a case, tracheostomy is an alternative option. However, tracheotomy is associated with various complications and should be avoided whenever possible.[10] Thus, SMI is another option. SMI is an alternative to tracheotomy[11] and has been used in patients with severe maxillofacial trauma. Although SMI was applied in orthognathic surgery, the application of SMI to patients with SWS has not been reported. SMI can cause complications such as superficial infection of the submental wound, orocutaneous fistula, abscess formation, and hypertrophic scarring. However, these complications are rare.[12] Fortunately, no angiomas were found in the submental region by magnetic resonance imaging in this case. Therefore, we decided to perform airway management with SMI during surgery. When we pulled the ETT from the floor of the mouth, we placed a sterilized echo probe cover over the ETT to let it go through smoothly because we could not remove the connector of the wire-reinforced tube in our institution. This method helped mitigate bumps on the connector of the ETT to avoid tissue damage and infection and prevent blood from entering the ETT. Goh et al. reported that damage to the tube apparatus is one of the most common complications of SMI.[13] This method also helped to avoid damage to the pilot balloon because the sterilized echo probe cover prevented it from directly gripping.

Blood pressure should be controlled to avoid the rupture of angiomas. Therefore, the depth of anesthesia was monitored using an entropy monitor. This is an electroencephalography-based monitor for determining hypnotic and analgesic levels. In this case, this monitor showed that state entropy (SE) and response entropy (RE) varied from 40 to 60 during anesthesia. The recommended range for adequate depth of anesthesia regarding each parameter (SE or RE) is 40–60.[14] Therefore, anesthetics appeared to be administered adequately, and the blood pressure was stable during surgery.

Maraña Pérez et al. reported that 46% of patients with SWS have mental retardation.[15] Low cognitive understanding and intellectual disability can interfere with perioperative doctor-patient communication, which causes mental stress, elevated blood pressure, and swelling of angiomas.[16] However, the patient's intellectual disability was borderline, and no communication problems were noted. Therefore, we attempted to form a trusting relationship with the patient through several preliminary examinations and detailed explanations. Consequently, we did not prescribe premedication but lidocaine tape to reduce stress as much as possible. These might help maintain stable blood pressure.

Previous studies have reported that propofol can induce seizures in patients with poorly controlled epilepsy. Our patient had a history of epilepsy. However, he had no seizures for more than 10 years without medication. In addition, propofol has significant anticonvulsant effects and has been used to manage status epilepticus.[3] Our patient also had a history of glaucoma. Therefore, we had to avoid raising IOP during surgery because high IOP could damage the optic nerve and cause vision loss. Previous studies have reported that propofol attenuates increased IOP.[17,18] In addition, propofol has been used in patients with SWS. Therefore, we used propofol as an anesthetic in this case.

AUTHOR CONTRIBUTIONS

Aya Oda: Conceptualization; writing – original draft. **Mitsuhiro Yoshida:** Conceptualization; supervision; writing – original draft. **Serika Imamura:** Resources; visualization. **Tamayo Takahashi:** Resources; visualization. **Kana Oue:** Resources; visualization. **Mitsuru Doi:** Writing – review and editing. **Yoshitaka Shimizu:** Writing – review and editing. **Shigehiro Ono:** Conceptualization; writing – review and editing. **Takayuki Nakagawa:** Conceptualization; writing – review and editing. **Koichi Koizumi:** Conceptualization; writing – review and editing. **Tomonao Aikawa:** Conceptualization; supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT

The data supporting the findings of this study are available upon request from the corresponding author.

ETHICS STATEMENT

Permission from the ethics committee was not required for case reports at the institution where the research was carried out.

CONSENT

Written informed consent was obtained from the patient and the patient's parents to publish this report in accordance with the journal's patient consent policy.

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