Anesthetic Management of a Patient With Sturge-Weber Syndrome Undergoing Oral Surgery

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This case involves a possible complication of excessive bleeding or rupture of hemangiomas. Problems and anesthetic management of the patient are discussed. A 35-year-old man with Sturge-Weber syndrome was to undergo teeth extraction and gingivectomy. Hemangiomas covered his face and the inside of the oral cavity. We used intravenous conscious sedation with propofol and N_2O-O_2 to reduce the patient's emotional stress. It was previously determined that stress caused marked expansion of this patient's hemangiomas. Periodontal ligament injection was chosen as the local anesthesia technique. Teeth were extracted without excessive bleeding or rupture of hemangiomas, but the planned gingivectomies were cancelled. Deep sedation requiring airway manipulation should be avoided because there are possible difficulties in airway maintenance. Because this was an outpatient procedure, propofol was selected as the sedative agent primarily because of its rapid onset and equally rapid recovery. Periodontal ligament injection with 2% lidocaine containing 1 : 80,000 epinephrine was chosen for local anesthesia. Gingivectomy was cancelled because hemostasis was challenging. As part of preoperative preparation, equipment for prompt intubation was available in case of rupture of the hemangiomas. The typically seen elevation of blood pressure was suppressed under propofol sedation so that expansion of the hemangiomas and significant intraoperative bleeding was prevented. Periodontal ligament injection as a local anesthetic also prevented bleeding from the injection site.

Key Words: Sturge-Weber syndrome; Hemangioma; Mental retardation; Anesthetic management; Oral surgery.

Sturge-Weber syndrome is a rare congenital disorder characterized by leptomeningeal hemangiomas; a facial port-wine nevus distributed over the trigeminal nerve area, usually unilaterally; and buphthalmos. This syndrome is also called encephalotrigeminal angiomatosis.[1] These hemangiomas cause neurological abnormalities, including epilepsy, mental retardation, and hemiplegia. Resection of gingival tissue and professional oral care are required because of enlargement of the soft tissues as a result of hemangiomas in addition to phenytoin group anticonvulsants. From a perspective of anesthetic management of the patient during dental treatment, Sturge-Weber syndrome involves specific problems that need to be considered.

CASE REPORT

A 35-year-old man with Sturge-Weber syndrome was referred to the university with a complaint of pain and swelling of the gingival tissue. The patient was diagnosed as having Sturge-Weber syndrome shortly after the birth. The port-wine stain extended into the bilateral trigeminal regions and covered both sides of the face, intraoral mucous membrane, and tongue. The patient underwent glaucoma surgery under general anesthesia when he was 3 years old. However, sight from his left eye was lost. The patient also had epilepsy and mental

Received December 6, 2004; accepted for publication December 20, 2005.

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Anesth Prog 53:17–19 2006

ISSN 0003-3006/06/$9.50
SSDI 0003-3006(06)

retardation. Antiepileptic drugs, primidone and ethotoin, had been prescribed. The patient could communicate with other people only by simple questions and answers. He had a tendency to become ill tempered and excited, especially in the morning. The hemangiomas markedly increased in volume during emotional agitation.

The patient had gingival enlargement and tooth stumps. Dental diagnoses were gingival enlargement and periodontitis. The proposed dental treatments were removal of 4 teeth—the right maxillary central incisor, the first and second molars, and the maxillary left first molar—followed by gingivectomies.

There was no contributing familial history. Laboratory reports were within normal range.

PROCEDURES

Informed consent was obtained from the patient's parent before the beginning of intravenous (IV) conscious sedation.

We applied topical anesthesia (Penles, 60% lidocaine tape, Wyeth K. K., Tokyo, Japan) to the venipuncture site. Sedation was started by inhalation of 50% N_2O and 50% O_2 via a nasal cannula. A venous cannula was inserted in the right radial cutaneous vein and an infusion of acetate–Ringer's solution was started. Propofol 35 mg IV was administered as an induction dose followed by an infusion of 3 to 4 mg/kg/h for maintenance of sedation. We monitored blood pressure, pulse rate, Spo_2, and $ETco_2$. After the patient's vital signs became stable, periodontal ligament injection with 2% lidocaine containing 1 : 80,000 epinephrine was administered to the maxillary left first molar area. An absorbable hemostatic agent, oxidized cellulose, was placed into the socket and the wound was sutured tightly because there was a concern with respect to difficulty of obtaining hemostasis. Postoperative bleeding did not occur. The remaining 3 teeth were successfully extracted in succession in a similar manner with periodontal ligament injections and 2% lidocaine with 1 : 80,000 epinephrine. Gingivectomy was cancelled because of the potential for excessive bleeding. The patient was sedated satisfactorily throughout the procedure, and there was no increase in blood pressure throughout the treatment period.

DISCUSSION

Sturge-Weber syndrome is a typical neurocutaneous syndrome defined as capillary or cavernous hemangiomas affecting primarily, but not limited to, cutaneous distribution of the trigeminal nerve. It is sometimes associated with venous malformations involving leptomeninges that may lead to progressive destruction of the adjunct cerebral cortex.[2,3] The patient discussed here had the most common type of Sturge-Weber syndrome, which involves both facial and leptomeningeal hemangiomas as well as glaucoma.[4]

The problems concerning oral surgery and anesthesia in this patient include (a) low cognitive understanding; (b) increased emotional stress, resulting in blood pressure elevation; (c) difficulty with hemostasis; and (d) risk of massive bleeding because of rupture of the hemangiomas covering the surgical sites. We interviewed the patient 3 times before the treatment commenced to establish a rapport with him. As anesthetic strategies to manage these problems, we scheduled the surgery in the afternoon to avoid the most sensitive time for emotional fluctuation for this patient. We also used an IV conscious sedation regimen with periodontal ligament injection with 2% lidocaine containing 1 : 80,000 epinephrine in an attempt to minimize the impact on the hemangiomas. We previously had experienced such a case that required emergency orotracheal intubation because of a rupture of one of the hemangiomas covering the facial region; hence, we prepared the equipment for endotracheal intubation in case of an emergency.

The patient did not resist entering the operation room on the day of the oral surgery. It was essential that we obtund any possible blood pressure rise in this patient. Elevation of blood pressure increases blood flow to and expands the hemangiomas that may lead to difficulty in hemostasis and increases the risk of rupture of the hemangiomas. Deep sedation should be avoided because there are potential difficulties in airway maintenance. We applied topical anesthesia to the venipuncture site and used 50% N_2O inhalation sedation to minimize pain and emotional stress associated with the venipuncture. Propofol was selected as an IV sedative agent because of its rapid onset and short elimination time. Controlling the depth of sedation is easy with propofol, and early discharge is possible. The combination of topical anesthesia and inhalation sedation prevented elevation of blood pressure during venous cannulation. The conscious sedation regimen of propofol 35 mg IV for induction and 3 to 4 mg/kg/h for maintenance produced optimal sedation throughout the operation. For local anesthesia, we used a periodontal ligament injection. This injection technique prevented bleeding from the injection site. There was little bleeding attributed to needle insertion. As expected, hemostasis after tooth extraction was challenging. We placed an absorbable hemostatic agent into the socket and sutured the wound tightly so that postoperative bleeding was prevented. Gingivectomy was cancelled because of difficulty in obtaining hemostasis and increased postoperative bleeding.

CONCLUSIONS

From an anesthetic management perspective of the patient during dental treatment, Sturge-Weber syndrome involves specific problems that need to be considered. We used N_2O inhalation sedation and IV conscious sedation with propofol as the sedative management for a patient with Sturge-Weber syndrome undergoing oral surgery. This sedation regimen was essential to prevent massive bleeding as a result of injury or rupture of the hemangiomas. The depth of sedation was easily controlled with propofol. The recovery from sedation was quick. The elevation of blood pressure was suppressed under sedation so that expansion of the hemangiomas was prevented and intraoperative bleeding was minimized. Periodontal ligament injection as a local anesthesia also prevented bleeding from the injection site.

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