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CASE REPORT

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Anesthetic management of a patient with Freeman-Sheldon syndrome undergoing oral surgery: A case report

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Abstract

In the anesthetic management in this case was how to manage the patient without causing respiratory depression and respiratory muscle fatigue.

KEYWORDS

dexmedetomidine, Freeman-Sheldon syndrome, respiratory depression

1 | INTRODUCTION

Freeman-Sheldon syndrome (FSS) is characterized by multiple joint contractures, characteristic facial features, such as microtia, defects of the hands and feet, such as clubfoot, and skeletal malformations. This report illustrates the case of a patient with FSS who was managed under local anesthesia with intravenous sedation for oral surgery.

Freeman-Sheldon syndrome (FSS), which was first described by Freeman-Sheldon in 1938, is a very rare congenital syndrome with a reported prevalence than 1 per 1 million. FSS is characterized by multiple joint contractures, characteristic facial features, such as microtia, defects of the hands and feet, such as clubfoot, and skeletal malformations.[1] General anesthetic management has been associated with various challenges, such as difficulty in intubation and securing the venous route and malignant hyperthermia. Reports on the general anesthetic management of FSS patients have been disorganized, and most of the reports on the anesthetic management of FSS have been on pediatric patients under general anesthesia, adult general anesthetic cases are rare.[2–6] Although it has been reported that patients with FSS should be treated under regional or local anesthesia if possible, [7] reports on the management of adult patients with FSS under local anesthesia with sedation were lacking. In this report, we describe a case of a patient with FSS undergoing oral surgery that was successfully managed under local anesthesia with sedation using dexmedetomidine. A written consent was obtained from the patient for this case report.

2 | CASE REPORT

The patient was a 42-year-old man, 157 cm in height and 37.5 kg in weight, and was referred to us by his family dentist because he recognized the need for lower right third molar

extraction and cyst removal. Judging from the anatomical position of the third molar and the fact that the mouth opening was only two lateral fingers, surgery under general anesthesia was considered. The patient had a history of FSS and had undergone surgery for scoliosis under general anesthesia at the age of 13 years at another hospital. Compared to the age of 13, he had chronic respiratory insufficiency due to the thoracic deformity caused by FSS, and pulmonary function tests revealed a severe alveolar hypoventilation (Vital Capacity (VC) 1.14 L, %VC 30.0, 1-s forced expiratory volume 1.07 L, 32.8%). Preoperative chest X-ray is shown in Figure 1. We thought that it would be desirable to treat him under local anesthesia if possible because general anesthesia would cause difficulty in extubation, worsening of respiratory insufficiency, and postoperative pneumonia, although the treatment depended on the extent of surgical invasion. In particular, the patient had chronic type 2 respiratory insufficiency, which was likely to be aggravated by general anesthesia.

2.1 | Course of anesthesia

In planning the anesthesia, we considered the following four possibilities:

1. Completely local anesthesia alone. This plan was not possible considering the surgical invasion and the anatomical position of the third molar.
2. Combination of local anesthesia and narcotic analgesics. Considering the respiratory depression and respiratory muscle fatigue caused by narcotic analgesics, this plan was not desirable.

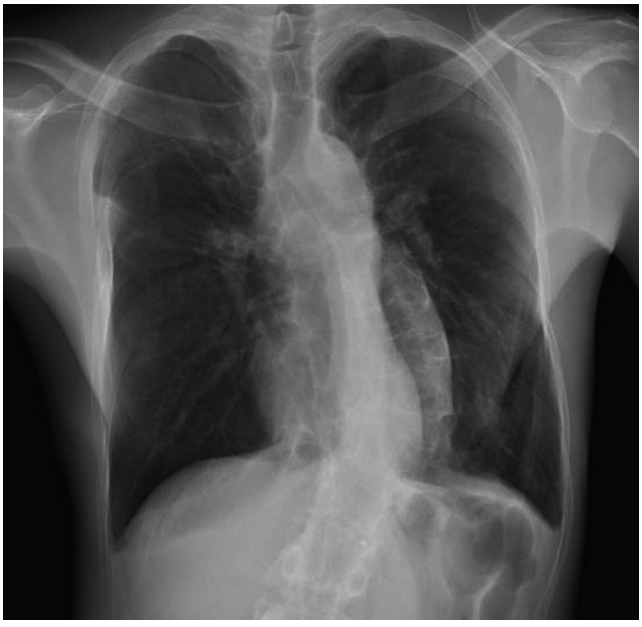

FIGURE 1 Preoperative chest X-ray showed scoliosis

3. Local anesthesia with sedation. In this plan, it is necessary to select a sedative agent that minimizes respiratory depression and respiratory muscle fatigue.
4. General anesthesia with tracheal intubation. If this patient was to undergo general anesthesia, the following parameters should be considered in the light of previous reports:

1) Anticipated difficulty in intubation

In this case, although neck retroflexion was possible, intubation was expected to be challenging because the mouth opening was only two lateral fingers.

2) Respiratory failure due to thoracic deformation

In addition to the restrictive impairment, there was a decrease in the rate of 1-s forced expiratory volume, which was expected to further lead to difficulties in extubation after general anesthesia, worsening of respiratory failure and postoperative pneumonia.

3) Risk of malignant hyperthermia

Several reports [2–6] have postulated that nondepolarizing muscle relaxants could be safely used and there was a slight need to refrain from using rocuronium.

Consequently, general anesthesia was preferred in view of the surgical invasiveness, although in view of the possibility of difficult intubation and postoperative respiratory failure, surgery under local anesthesia with sedation was scheduled first.

After entering the operating room, a standard monitor and a bispectral index (BIS) monitor were attached to the patient, and a venous access was established at the dorsum of the left hand. Together with oxygen administration, end-tidal carbon dioxide level was monitored using Smart CapnoLine® Plus Long, although it was a reference value. For the intravenous sedation, dexmedetomidine, which has a minimal risk of respiratory depression, was used as the sedative of choice. The induction dose was 6 μg/kg/h for 10 minutes continuously, and then sedation was maintained at 0.4 μg/kg/h. The intraoperative BIS value was "60"-"70", oxygen saturation was approximately 98%, and ETCO2 was 40-50 mm Hg without respiratory depression. For intraoperative analgesia, 2% lidocaine containing epinephrine diluted 80 000 times was used as local anesthetic. Intraoperatively, 1000 mg of acetaminophen was administered for postoperative analgesia. One of the side effects of using dexmedetomidine is bradycardia. However, fortunately, it did not occur in this case and intraoperative circulatory dynamics were stable. Although the surgical operation was fairly challenging owing to the small mouth

opening and the protrusion of the buccal mucosa, it was completed as scheduled. The operation time, anesthesia time, and the amount of blood loss were 56 minutes, 1 hour and 30 minutes, and 10 g, respectively. After the surgery, we confirmed no complications observed in the postanesthesia care unit, and the patient was returned to the general ward.

3 | DISCUSSION

Since FSS is thought to cause respiratory function decline in adults, aging is a disadvantage in terms of respiratory complications. The key point in the anesthetic management in this case was how to manage the patient without causing respiratory depression and respiratory muscle fatigue. Since there have been no reports of the challenges encountered in mask ventilation in anesthesia for FSS and there was no facial deformity or limitation of retroflexion in this case, in the event that respiratory depression, such as tongue root subsidence, or respiratory arrest occurred during sedation, mask ventilation was considered to be sufficient. However, in case of intubation, the possibility of a difficult intubation was extremely high; therefore, we chose dexmedetomidine as the sedative drug of choice owing to its minimal risk of respiratory depression. Dexmedetomidine is a central α2-adrenergic sedative and is characterized by a minimal risk of respiratory depression and the ability to maintain cognitive function even under sedation. In Japan, the indication for the use of dexmedetomidine during surgery under local anesthesia was expanded in 2013.[8] One report demonstrated that dexmedetomidine was used during tracheoplasty for tracheal stenosis performed under local anesthesia, reducing the burden on the patient and preventing respiratory depression while enabling high-quality sedation.[8] It was believed to be a good indication for reducing respiratory muscle fatigue and minimizing respiratory depression as in this case. However, respiratory depression is not completely absent when dexmedetomidine is used, and there were cases in which mandibular elevation was temporarily required at a dose of 10 μg/kg/h. [9] Furthermore, it has been reported that mandibular elevation is required in patients with a history of sleep apnea syndrome even when the usual maintenance dose is used. [8] In this case, respiratory depression did not occur even with the prescribed dosage; however, considering the difficulty of intubation, we should have started with a low-dose therapy and increased the dosage after confirming the absence of respiratory depression. Recently, there have been several reports on oxygenation using the nasal high-flow system (NHF). [10–12] Although NHF was not used in this case, it may have been useful to reduce the respiratory muscle fatigue because NHF decreases inspiratory resistance and improves the efficiency of respiratory effort.[13]

4 | CONCLUSIONS

We experienced the case of a patient with FSS who was managed under local anesthesia with intravenous sedation for oral surgery. We used dexmedetomidine, a sedative with a minimal risk of respiratory depression, and were able to manage the patient without intraoperative respiratory depression.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

All authors contributed to the case. TH, YS, MS, YK, and EK: performed preoperative management and anesthetic planning. AS(B) and KT: performed anesthetic management. AS(B): drafted the first manuscript. All authors commented on previous versions of the manuscript. KS: helped with the supervision of the manuscript and development of the overall anesthetic plan. All authors read and approved the final manuscript.

CONSENT FOR PUBLICATION

The authors obtained written consent for publication from the patient.

DATA AVAILABILITY STATEMENT

Not applicable due to patient privacy concerns.

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