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Case report

Negative pressure pulmonary edema after orthognathic surgery for osteogenesis imperfecta

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ABSTRACT

This report describes a case of negative pressure pulmonary edema (NPPE) after orthognathic surgery for osteogenesis imperfecta (OI). An 18-year-old male with a prognathic mandible, earlier diagnosed with OI Type I, was referred by his orthodontist. Presurgical laboratory tests showed his bleeding time and coagulation to be normal, and his chest radiograph was unremarkable. We performed Le Fort I osteotomy and bilateral vertical ramus osteotomy. Continuous bleeding was noted during the operation. After extubation, the patient became agitated and made vigorous inspiratory efforts. His oxygen saturation dropped to 40%, and he was reintubated atraumatically. NPPE was suspected. The patient was transferred to the intensive care unit. Although OI has been linked to several risk factors during and after surgery, such as decreased platelet function causing abnormal bleeding and low-ventilation due to chest wall deformity, follow up periods summing to 5 years showed good bone healing, good occlusion and no further complaints. We predict that prolonged operation time and increased bleeding volume may have influenced the onset of NPPE. Reports of orthognathic surgery for OI and NPPE after orthognathic surgery are currently limited. Our results suggest that a minimally invasive surgery is preferable for OI patients.

Introduction

Negative-pressure pulmonary edema (NPPE) develops in patients with spontaneous respiratory effort who have upper airway obstruction and generate highly negative intrathoracic pressures leading to severe hypoxemia and pulmonary edema. NPPE occurs after intense inspiratory effort against an NPPE-obstructed airway, usually from an upper airway infection, tumor, or oral and neck surgery. NPPE develops in less than 1 in 1000 (0.094%) surgical patients [1].

Osteogenesis imperfecta (OI) is one of the most commonly inherited bone disorders. The most widely used classification of OI was proposed by Sillence et al. [2]. Type I is the most common, occurring in 60% of OI patients or 1 in 30,000 live births [2]. OI is caused by defects in the amount or structure of Type I collagen, an important component of bone matrix. Clinical presentations of OI include bone fragility, short stature, blue sclera, dentinogenesis imperfecta, hearing impairment, kyphoscoliosis, and ligamentous laxity [3]. In addition, OI is associated with risk factors during and after surgery, such as hyperthermia, decreased platelet function causing abnormal bleeding, low-ventilation due to chest wall deformity, and difficult airway [4]. OI is associated with facial deformity, particularly prognathism and Class III malocclusion [5]. Therefore, it is important to understand the perioperative management of surgical orthodontic treatment with OI.

The risk of NPPE with OI is proposed to be much higher after oral and maxillofacial surgery than other types of surgery because of the overlap between the surgical field and airway [6]. Further, OI is associated with risk factors that arise during and after surgery. However, there are currently few reports of NPPE and the management of surgical orthodontic treatment with OI. Therefore, it is necessary to properly examine surgery planning and perioperative management. This case report describes a mandibular prognathic patient with OI who developed NPPE after orthognathic surgery and exhibited marked improvements in occlusion and profile following two-jaw surgery.

Case report

This report was approved by the Kyoto University Graduate School and Faculty of Medicine. Ethics Committee. Written consent for publication was obtained from the patient.

☆ JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JAMI: Japanese Academy of Maxillofacial Implants.

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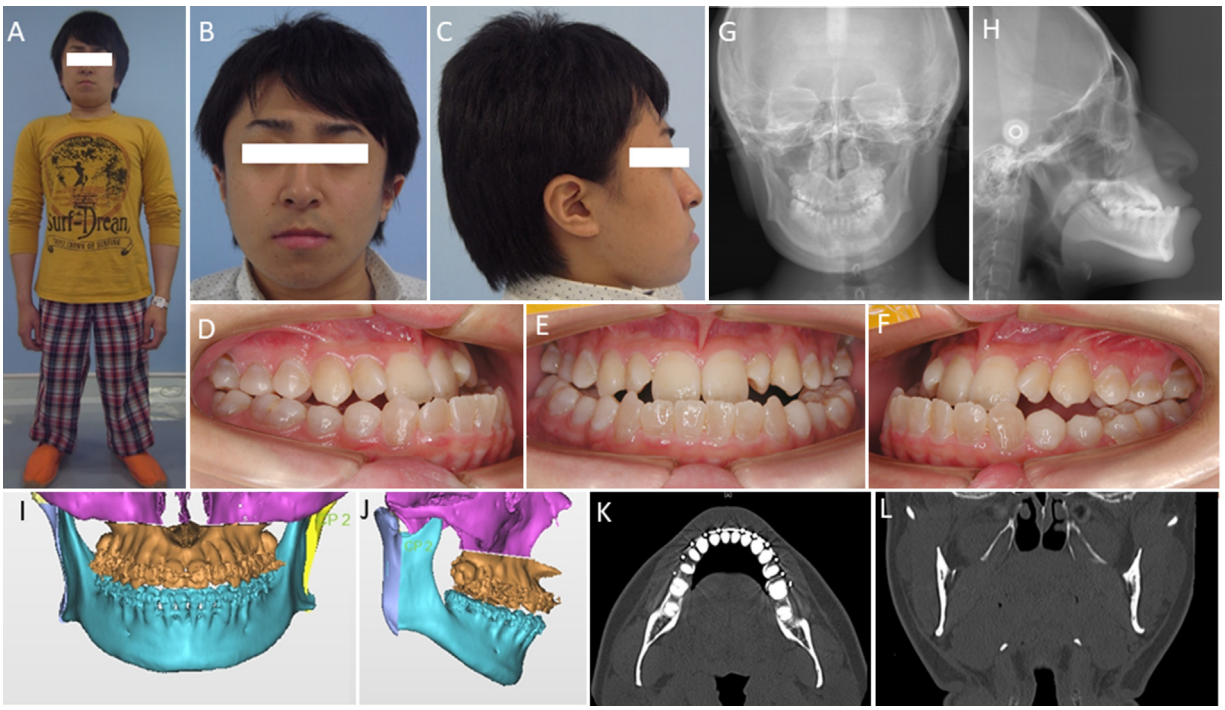

Fig. 1. Preoperative findings and three-dimensional surgical simulation. Whole body image (A), and frontal (B) and lateral photographs (C). Preoperative intraoral photographs (D–F). Lateral (G) and frontal cephalograms (H). Three-dimensional surgical simulation: frontal (I) and lateral images (J). Coronal (K) and axial CT-images (L).

In 2008, an 18-year-old male with a prognathic mandible was referred by his orthodontist to our institute. He had previously been diagnosed with OI Type I and had a family history of OI: his father and younger brother both had OI. He had been treated for about 20 fractures of his limbs and hips since childhood. He had kyphoscoliosis, long bone deformities and was using a wheelchair. Clinical examination revealed that he was 126 cm tall, weighing 37 kg (Fig. 1A). The patient had normal eye sclera. His maxilla was retrognathic and his mandible was prognathic (Fig. 1 B,C). He had a severe Class III occlusion with a negative overjet of 9 mm. The patient had healthy dentition and his teeth had normal enamel and dentin structure (Fig. 1D,E,F). He had intact hard and soft palates. He was taking the bisphosphonates alendronic acid (Fosamax®) and pamidronate (Aredia®). Cephalometric analysis showed SNA and SNB angles of 80.5° and 87.5°, respectively (Fig. 1G,H). The presurgical orthodontic treatment with leveling and alignment lasted for 44 months.

Radiographs and face-bow recordings were taken for final surgical planning 3 weeks prior to surgery. The anesthetic preoperative assessment was normal. We performed presurgical simulation using SimPlant OMS software (Materialise®, Leuven, Belgium) (Fig. 1 I,J). The analysis indicated a maxillary advancement of 3 mm using the Le Fort I osteotomy technique and mandibular set back of 8 mm using the vertical ramus osteotomy technique were suitable. The selected procedure was bilateral mandibular vertical ramus osteotomy because of the patient's very thin mandibular body, which is a bone fracture risk (Fig. 1K,L). We planned to use screws and titanium plates to stabilize the bony fragment. Presurgical laboratory tests showed his bleeding time and coagulation to be normal, and his chest radiograph was unremarkable. To prepare for blood loss during the surgery, 800 g of the patient's autologous blood was preserved before surgery.

The surgery was performed under general anesthesia with nasotracheal intubation. Le Fort I osteotomy was carried out first, and the maxillary bone was found to be thin. Le Fort I osteotomy enabled us to advance the maxilla by 3 mm, which was fixed with screws and titanium plates. The mandible was approached intraorally and set back 8 mm by bilateral vertical ramus osteotomy, and fixed with screws and titanium plates. The bone was thin but no problems were observed regarding the rigid fixation with titanium plates. During the operation, no major bleeding was encountered but continuous bleeding was noted, which was controlled by packing with gauze and external compression. The operative time was 7 h 5 min. The estimated blood loss was 1689 mg; therefore, autologous blood transfusion was conducted.

After surgery and once he was responsive with adequate respiratory and neuromuscular function, the patient was extubated. Immediately afterwards, he became agitated and made vigorous inspiratory efforts. Positive pressure ventilation was manually controlled using a face mask and the oropharyngeal airway, but SpO_2 saturation gradually decreased from 99% to 38%. We therefore performed orotracheal reintubation. At the time of reintubation, we found that the vocal cords were slightly edematous. A small quantity of blood-tinged secretion was suctioned through the endotracheal tube. Arterial blood gas analysis showed pH 7.4, $PaCO_2$ 43.3 mmHg, PaO_2 62.3 mmHg, and SaO_2 91% with FiO_2 0.9. Negative pressure pulmonary edema (NPPE) was suspected. Postoperatively, the patient was transferred to the intensive care unit. A chest radiogram showed the typical pattern of pulmonary edema with symmetric, bilateral middle-lobe infiltrates, and normal heart size and wide vascular pedicle (Fig. 2A). Based on the assumption of NPPE, ventilation was supported by pressure assisted controlled mandatory ventilation with a positive end-expiratory pressure of 5 cmH_2O. A transthoracic echocardiogram demonstrated normal cardiac function. On the third postoperative day, arterial blood gas analysis showed pH 7.5, PCO_2 44.4 mmHg, PaO_2 105.7 mmHg, and SaO_2 98.0% during oxygen supply with a nasal cannula at a flow rate of 4 L/min. The patient's chest X-ray, respiratory parameters, and blood gas parameters had improved, and he was successfully extubated. The patient's postoperative course over the following 3 days was uneventful. The patient had no complaints.

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