Surgical and orthodontic approach for a patient with a severely constricted maxillary arch caused by bilateral cleft lip and palate

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ABSTRACT

Cleft lip and/or palate (CLP) is one of the most frequent craniofacial defects that could happen in 1/500 to 1/1000 live birth depending on different racial background. Among different patterns of facial cleft, complete bilateral cleft lip and palate (BCLP) is one of the most challenging cases for orthodontic and surgeons because of their deformed maxillary dental arch and severe skeletal discrepancy. It is also well known that CLP could occur as part of the phenotype in certain congenital diseases. However, from its extremely diversified phenotypic combination, some of the cases that we encounter remains difficult to diagnose. From these reasons, it is important to continuously report the outcome of orthodontic treatment in such cases which exhibit syndromic phenotypes with CLP.

In the present case report, an 18-year-old man with complete bilateral cleft lip and palate, skeletal Class III and open-bite with maxillary constriction, in addition to hypospadias, bubonocele, opisthotonus, and hypertonia was treated with edgewise appliance therapy combined with orthognathic surgery. The treatment began with surgically assisted rapid palatal expansion (SARPE) in anteroposterior and transverse dimensions with a three-way expander to increase the maxillary anteroposterior length and width. After the expansion, the patient underwent Le Fort I osteotomy and bilateral sagittal split osteotomy to correct skeletal Class III and open bite. At the end of the surgical and orthodontic treatments, functional occlusion

and an improved facial profile were achieved. We also discuss his diverse general phenotype due to his congenital disease.

INTRODUCTION

Cleft lip and/or palate is one of the most frequent craniofacial disorders, exhibiting severe malocclusion and requiring orthodontic treatment. The severity of malocclusion varies depending on the case and complete bilateral cleft lip and palate (BCLP) cases are one of the most challenging situations to manage among all facial cleft pattern [1]. The maxillary dental arch in BCLP cases is frequently collapsed in both the transverse and anteroposterior dimensions [2], [3]. If the skeletal discrepancy remains when the patients are adult, orthodontic treatment combined with various surgical procedure is often required [4].

We herein report the orthodontic treatment of an adult male patient with complete BCLP, a constricted maxilla and skeletal Class III with open-bite. The patient in this report was treated with multiple orthognathic surgeries, including surgically assisted rapid palatal expansion (SARPE) in the anteroposterior and transverse dimensions with a three-way expander and consequent Le Fort I osteotomy and bilateral sagittal split osteotomy (BSSO) in order to correct the severe skeletal deficiency and malocclusion. As a result, his facial profile as well as occlusion showed remarkable improvement at the end of orthodontic treatment. Cases that require both lateral and anteroposterior alveolar distraction of the maxilla are rare and further assessment is required to evaluate the treatment outcome and retention.

Additionally, it is also well known that certain congenital abnormalities exhibit CLP as a part of their phenotypes, such as EEC syndrome, 22q11.2 deletion syndrome and Turner

syndrome [5], [6], [7]. As in this case, there are some patients whose general symptoms do not completely overlap with the existing syndromic phenotype and remain undiagnosed. For these reasons, the continuous reporting of facial cleft cases associated with a general phenotype is important for understanding the etiology and determining possible solutions for improving the facial profile and occlusal relationships of diverse craniofacial disorders.

HISTORY

A 9-year-old boy first visited our hospital with complaints of mandibular protrusion and occlusal disturbance. An examination in infancy had shown a bilateral complete cleft lip and palate, hypospadias, bubonocele, opisthotonus and hypertonia. He also exhibited hypertelorism, short stature, intellectual disabilities and arachnoid cyst at 8 years of age.

He had a history of cheiloplasty at 5 and 8 months of age and pushback palatoplasty at 16 months of age. At 9 years 3 months of age, maxillary expansion and protraction was initiated with a quad-helix and reverse headgear to improve the anterior and posterior cross-bite that resulted from the skeletal Cass III malocclusion and mesial step type terminal plane. The quad-helix could expand the maxillary arch to some extent; however, the orthopedic effect of the reverse headgear was limited (Figure 1A). At 11 years 11 months of age, bilateral alveolar bone grafts from the autogenous iliac bone to the cleft spaces were made to resolve the severe defects in the alveolar bones. At the same time, a tongue flap was utilized to close the palatal fistula.

Growth hormone treatment was performed from 13 to 18 years of age to overcome his short stature, which increased his height from 136 cm to 156.9 cm. Active orthodontic treatment was not performed during this period.

The extraoral examination at 18 years 7 months of age showed severe midfacial deficiency, a concave profile and long face with lip incompetency (Figure 2A). The upper incisor exposure was very small, even in a full smile. Hypertelorism was found. The occlusion was classified as Angle Class III with total cross-bite and anterior open-bite. The maxillary dental arch showed anteroposterior and transverse constriction with moderate crowding, making the width and length of the maxilla narrower than those of the mandible (Figure 2B). The mandibular dental arch exhibited moderate crowding (Figure 2B). The panoramic radiograph showed congenitally missing maxillary lateral incisors and a right mandibular second premolar as well as an upper wisdom tooth (Figure 2C). The presence of periodontal disease was observed in association with horizontal alveolar resorption in both jaws (Figure 2C). Dental compensation was seen as the lingual inclination of the mandibular molars and incisors (Figure 2D). No symptoms or signs of any temporomandibular joint disorder were detected. The result of lateral cephalometric analysis was shown in Table 1. The patient also exhibited borderline velopharyngeal insufficiency and mild hypernasality.

TREATMENT PLAN AND PROGRESS

At 18 years 7 months of age, anteroposterior and transverse expansion of the maxilla using SARPE was performed. The premaxilla and posterior segments were separated to perform maxillary expansion in both the anterior and transverse directions (Figure 3). We fixed the three-way expander with miniscrews to the maxillary posterior alveolar segments to correct both transverse and anteroposterior constriction (Figure 3A). The expansion in the anteroposterior direction and that in the transverse direction were performed for 16 days with 0.5-mm expansion/per day and 1.0-mm expansion/per day, respectively (Figure 3B). All of the wisdom teeth were extracted during SARPE. Preadjusted fixed appliances were then bonded onto the maxillary and mandibular dentition to align and level the teeth, and preoperative orthodontic treatment was started. The dental compensation was corrected in preparation for the orthognathic surgery. Preoperative orthodontic treatment was performed by extracting the lower left second bicuspid to relieve the crowding and maintain the lower incisor inclination.

The patient underwent two jaw orthognathic surgeries after orthodontic preparation at 21 years 3 months of age. The maxilla was set forward 6.5 mm to improve the remaining mid-facial deficiency and low exposure of the upper incisors. The maxilla was also rotated 4.0° clockwise to correct the open-bite. The mandible was set back 2 mm and rotated counter-clockwise for open-bite correction with BSSO. Tongue reduction was also performed as the same time as two-jaw surgery to improve the stability of obtained normal occlusion. Temporary

anchorages were placed in the area of the maxillary and mandibular anterior teeth after the two-jaw surgery for intermaxillary fixation.

After postoperative orthodontic treatment had been continued for 10 months to obtain mutually protected occlusion, all appliances were removed (Figure 4). We tried to make the postoperative orthodontic treatment as short as possible to prevent further alveolar bone resorption. Begg-type retainers were placed on both arches for retention. Speech assessment after the removal of the appliances did not show a significant difference from the start of orthodontic treatment.

RESULTS

SARPE with a three-way expander dramatically corrected the anteroposterior and transverse constricted maxillary arch (Figure 3). As a result, the maxillary width was increased; the intercanine width increased from 20.3 mm to 38.5 mm, and the first molar basal arch width increased from 63.5 mm to 71.6 mm. The anteroposterior length was also increased; the perpendicular distance between the line connecting the upper incisors and the line connecting the distal surfaces of the first molars increased from 29.0 mm to 33.8 mm. Three-dimensional superimposition also revealed an improved maxillary arch form after SARPE (Figure 5).

Subsequent Le Fort I osteotomy improved the midfacial deficiency associated with the skeletal Class III relationship and skeletal open-bite by setting back and counter-clockwise

rotating the mandible through BSSO (table 1). The posttreatment facial photographs showed a preferable straight-type facial profile without lip incompetency (Figure 4). The upper incisor exposure was also notably improved in a full smile. Intraoral photographs showed mutually protected occlusion with proper overjet and overbite (Figure 4). After two-year of retention period, the good occlusion and facial profile were well maintained (Figure 6). Superimposed lateral cephalometric tracings of the each stage were shown in Figure 1. A slightly relapse was shown in that the lower incisors were tipped labially (Figure 1 and table 1).

Panoramic X-ray showed that the horizontal level of the alveolar bone had become lower, especially around the lower premolar and molar region, after the fixed orthodontic treatment (Figure 4).

DISCUSSION

In order to reconstruct their maxillary arch form and correct the intermaxillary discrepancy, various surgical procedures are required for non-growing patients with BCLP [4]. Distraction osteotomy or SARPE have been used for maxillary anterior advancement or lateral expansion in patients with CLP [4], [8], [9]. However, few reports have described the outcomes of SARPE with a three-way expander for maxillary expansion in two directions (anteroposterior and transverse). In this case, the maxilla was constricted transversely and anteroposteriorly in comparison with the mandibular dental arch. We therefore performed

SARPE using a three-way expander to increase the maxillary width as well as the anteroposterior length. Instead of 3-way distraction osteotomy, we could have performed multi-section osteotomy to improve the maxillary constriction. However, in our case, we selected 3-way distraction osteotomy to achieve the amount of expansion required for arch coordination. For these reasons, SARPE with a 3-way expander could be efficiently applied to the patient who exhibited severe dental arch constriction in both the lateral and anteroposterior directions. However, due to the higher risk of damaging the root, cases should be selected after careful assessment.

We corrected the open-bite via Le Fort I osteotomy and BSSO. Through multiple orthognathic surgeries in the present case, including SARPE with a three-way expander and consequent Le Fort I osteotomy and BSSO, the facial profile as well as occlusion were markedly improved at the end of orthodontic treatment. We could have also selected Le Fort I distraction osteotomy to improve the position of the maxilla, as it has been shown to result in good clinical outcomes [10]. Maxillary anterior segmental distraction osteogenesis (MASDO) is another alternative method to move the maxilla forward, especially when severe velopharyngeal insufficiency exists [11], [12].

CLP is also known to occur in the phenotype of certain genetic diseases. Given the present patient's general symptoms of hypertelorism, genitourinary abnormalities, CLP and intellectual disabilities, he was suspected of having Opitz G/BBB syndrome. Opitz G/BBB

syndrome is a rare genetic disorder characterized by multiple anomalies along the midline of the body [13], [14], [15]. The various clinical manifestations of Opitz G/BBB syndrome include facial anomaly, laryngotracheal and esophageal defects, genitourinary abnormalities, CLP and intellectual disabilities [14], [16], [17], [18]. Among those clinical manifestations, hypertelorism, hypospadias and CLP are three major anomalies associated with this syndrome [14], [16], [17], [18]. The clinical manifestation of the present case exhibited symptoms that overlapped with those of patients with Opitz syndrome. Therefore, he was suspected of having Opitz G/BBB syndromes by physicians at Osaka University Medical Hospital in Suita City. Genetic testing of MID1 or SPECCL1 are useful for the definitive diagnosis of Opitz G/BBB syndrome [19], [20]. Some patients have also been reported to exhibit chromosome 22q11.2 deletion [21]. However, he and his family did not wish to undergo genetic testing, so he remained undiagnosed. There have been various reports published on the underlying genetics and medical treatment of patients with Opitz G/BBB syndrome[14,16-18], but reports on orthodontic treatment are limited [22].

Opitz G/BBB syndrome is sometimes associated with intellectual disability and/or congenital heart defects, which sometimes require specific care during orthodontic treatment [23]. The present patient showed poor oral hygiene and periodontal disease before the orthodontic treatment. For this reason, he underwent repetitive professional oral care, including instruction on tooth brushing and his oral hygiene substantially improved. However, he

experienced difficulty in maintaining oral hygiene, especially immediately after orthognathic surgery which possibly led to the worsening of the periodontal situation.

CONCLUSIONS

Surgical-orthodontic treatment with SARPE using a three-way expander followed by Le Fort I osteotomy with BSSO was effective for improving the skeletal disharmony, facial profile and occlusion in a patient with BCLP who exhibited a constricted maxilla and skeletal Class III relationship with open-bite.

ETHICAL APPROVAL

The patient consented to publication of the case in writing.

CONFLICT OF INTEREST

The authors declare that no conflicts of interest exist in association with this study.

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FIGURE LEGENDS

FIGURE 1. Superimposed lateral cephalometric tracings on the SN plane at S: A, initial (black line) and pre-SARPE (gray line); B, pre-SARPE (black line) and before two-jaw surgery (gray line); C, before two-jaw surgery (black line) and post-active treatment (gray line); D, post-active treatment (black line) and post-retention (gray line)

FIGURE 2. Pretreatment records. A, Facial photographs. B, Intraoral photographs. C, Panoramic radiographs. D, Lateral cephalograms.

FIGURE 3. Occlusal photographs of the three-way expander and retainer. A, Before maxillary expansion. B, After maxillary expansion. C, During maxillary retention. D, Three-way expander before activation. E, Three-way expander after activation.

FIGURE 4. Post-active treatment records. A, Facial photographs. B, Intraoral photographs. C, Panoramic radiographs. D, Lateral cephalograms.

FIGURE 5. Three-dimensional models were superimposed using the surfaces of the palate. A, Pretreatment (yellow); B, post-active treatment (green); C, Superimposition of the pretreatment (yellow) model and post-active treatment model (green).

FIGURE 6. Post-retention records. A, Facial photographs. B, Intraoral photographs. C, Panoramic radiographs. D, Lateral cephalograms.