CASE REPORT

Hemifacial Microsomia Caused by First and Second Brachial Arch Syndrome Treated with Orthodontic Approach: A Case Report

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Abstract

Aim and objective: To present a growing patient with unilateral mandibular hypoplasia and microtia involved in the first and second branchial arch syndrome (FSBAS) treated with functional appliance.

Background: The FSBAS comprises several developmental facial hypoplasia in ear and maxillofacial bones, resulting in hemifacial microsomia. Treatment for hemifacial microsomia varies greatly depending on the grade of mandibular deformities. Functional appliance treatment during growth period is available for mild to moderate mandibular deformities. However, there are few reports of hemifacial microsomia treated with functional appliance.

Case description: The patient, an 8-year-and-5-month-old girl, had a chief complaint of mandibular deviation. She had been diagnosed with the FSBAS at birth. Her facial profile was straight and panoramic radiograph indicated that the mandibular ramal height of the affected side was about 60.4% compared to the unaffected side. The occlusal cant was 6°, and the right maxilla and mandible showed severe growth deficiency. At the age of 10 years, functional appliance with expander was used; for 2 years 6 months, the maxillomandibular growth was controlled and from panoramic radiograph, the ramus height of the affected side was increased to 65.0% compared to the unaffected left mandibular ramus. At the age of 12 years and 8 months, multibracket treatment was initiated. After 32 months of active treatment, proper occlusion with functional Class I canine and molar relationships was obtained, although facial asymmetry associated with the difference of ramus heights still existed. The resulting occlusion was stable during 1.5-year retention period.

Conclusion: Our results indicated the importance of orthopedic treatment during growth period in the patient with hemifacial microsomia involving the FSBAS.

Clinical significance: This report proposes an efficacy of conventional orthodontic treatment for growing patients with hemifacial microsomia involved in the FSBAS.

Keywords: Craniofacial anomalies, First and second brachial arch syndrome, Hemifacial microsomia, Orthopedic treatment.

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Introduction

First and second branchial arch syndrome (FSBAS) comprises several developmental facial hypoplasia in external ear, middle ear, and maxillary and mandibular bones. This is a rare, complex disease, and its estimated prevalence is 1 out of every 4,000 births.[1,2] The FSBAS is a multifactorial disease that has not yet been clearly identified. Maternal diabetes and infection caused by rubella and influenza during pregnancy is assumed to involve in the development of this syndrome.[3]

Patients with FSBAS are likely to have unilateral mandibular hypoplasia combined with unilateral or bilateral microtia,[4] resulting in hemifacial microsomia. The severity of the abnormalities and symptoms is variable from one individual to another and ranges from mild to severe forms.[5] The mandibular deformities in hemifacial microsomia was classified by Pruzansky using a radiography.[6] According to his classification, Grade I indicates a minimum mandibular hypoplasia with normal structures; Grade II defines a small and abnormally shaped condyle, ramus, and sigmoid notch with large variation; Grade III is characterized by a lack of the mandibular ramus including the temporomandibular joint. Furthermore, Kaban et al.[7] modified the classification of Pruzansky, and divided Grade II into Grade IIA and IIB: Grade IIA exhibits short mandibular ramus with abnormal shape, and the position of the glenoid cavity is normal, while Grade IIB shows altered position of glenoid cavity.

Treatment for hemifacial microsomia varies greatly depending on the grade of mandibular deformities and the needs of the

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individual. In mild cases of Grade I, no treatment may be needed. The patients with Grade II or III may need surgery including distraction osteogenesis (DO). Indeed, the early DO significantly improves the facial asymmetry and is able to relieve them from psychological disturbances in early days.[8] Functional appliance treatment during growth period is also available for mild to moderate mandibular deformities. The patients with hemifacial microsomia have been applied to early DO and/or orthodontic–orthognathic surgery.[5,8] However, there are few detailed reports of conventional orthodontic treatment with functional appliance regarding hemifacial microsomia. Hence, the prognosis of conventional orthodontic treatment with functional appliance during their pubertal growth period remains unclear.

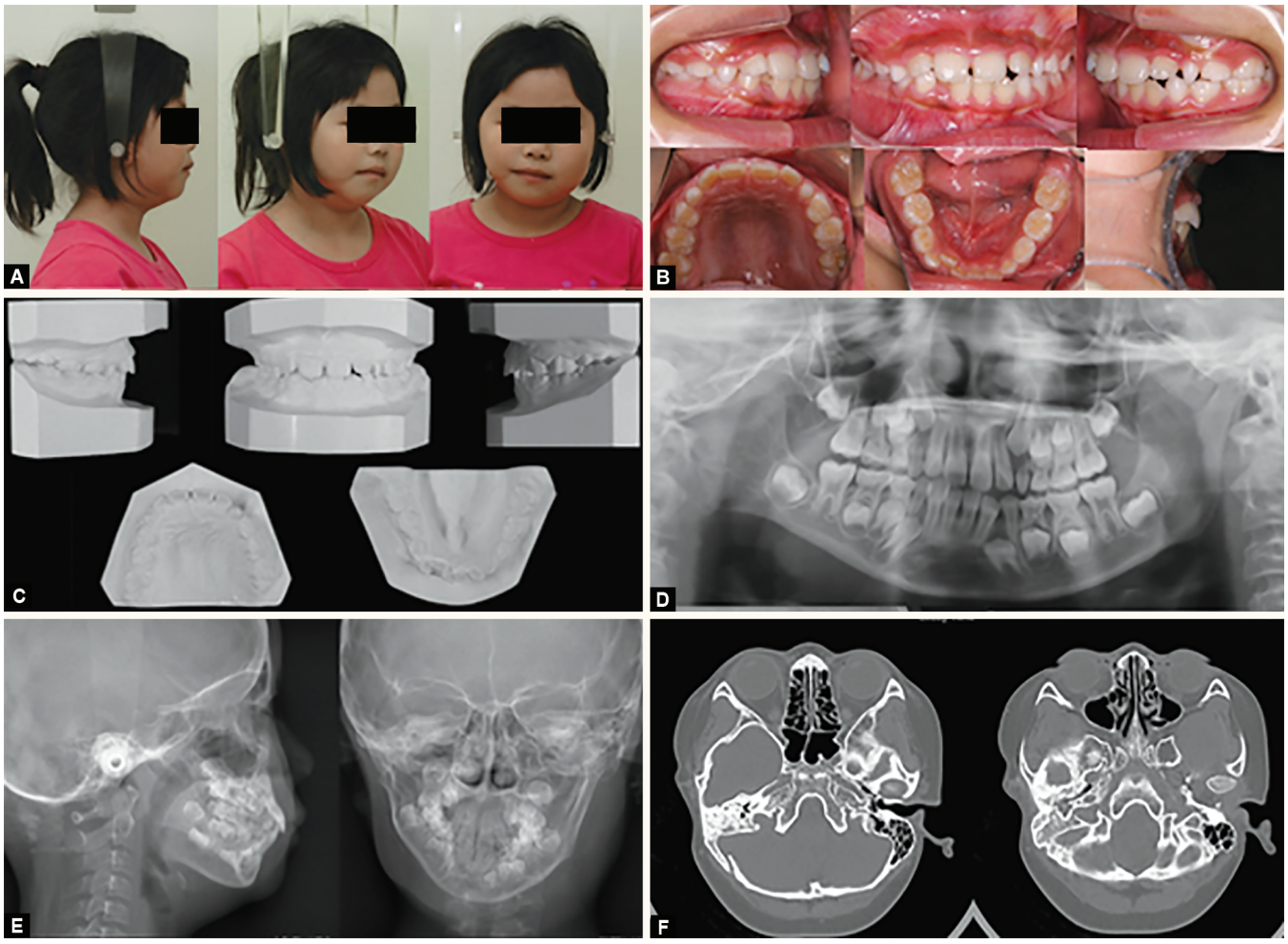
The purpose of this case report is to present a growing patient with unilateral mandibular hypoplasia and microtia involved in the FSBAS treated with functional appliance.

Case Description

A girl, aged 1 year, was introduced by the Department of Plastic Surgery, Tokushima University Hospital. She was diagnosed with FSBAS with a slight mandibular deviation. Once a year she received a follow-up examination from 1-year-old to 8-year-old. The patient, at the age of 8 years and 5 months, had a chief complaint of mandibular deviation. She had no significant medical and dental history except for FSBAS. Facial photographs showed mandibular shift to the right (Fig. 1A). Lateral profile was straight. From computed tomography (CT) images, she had an accessary ear without external auditory canal that had no sense of hearing (Fig. 1F).

The midline of upper dentitions was deviated 2 mm to the right against her facial midline, and the midline of lower dentitions was shifted 0.5 mm to the left against the upper dental midline (Figs 1B and C). The lower right lateral incisor was under eruption with lingual displacement.

Panoramic radiograph showed no congenitally missing permanent teeth. The right condyle had a considerable growth deficiency (Fig. 1D). Compared to the unaffected left mandibular ramus, the ramus height of the affected side was about 60.4%. According to the classification of Pruzansky[6] and Kaban et al.,[7] this case was classified Grade IIB. From cephalometric evaluation, the patient had skeletal Class II jaw-base relationship (ANB, 6.1°) and low mandibular plane angle (FMA, 24.9°) (Fig. 1E, Table 1).[9] The inclination of upper central incisor was almost on average, but the lower central incisor was inclined labially, resulting in a significantly smaller interincisal angle. Frontal cephalogram showed a shorter ramus height of the right side (Fig. 1E). The occlusal cant was 6°, and the right maxilla and mandible showed severe growth deficiency.



Figs 1A to F: Pretreatment facial (A) and intraoral photographs (B), dental cast (C), panoramic radiograph (D), frontal and lateral cephalograms (E), and computed tomography (F) (A: at the age of 7 years and 5 months; B to F: at the age of 8 years and 5 months)

Table 1: Cephalometric summary

	*Mean for Japanese females**			*Pretreatment*	*After phase 1*	*Posttreatment*	*Posttreatment*
Treatment variable	*8 years (SD)*	*12 years (SD)*	*Adult SD*	*8 years 5 months*	*12 years 8 months*	*15 years 5 months*	*16 years 10 months*
Angular measurement (°)							
ANB	3.7 (2.0)	2.8 (2.4)	3.0 (2.2)	6.1	6.5	6.3	6.8
SNA	80.7 (2.8)	80.8 (3.6)	80.7 (3.4)	82.1	82.2	82.0	82.0
SNB	76.9 (2.4)	77.9 (4.5)	77.6 (4.2)	76.0	75.7	75.7	74.2
Mandibular plane/FH	32.2 (3.7)	30.5 (3.6)	29.6 (3.4)	24.9	25.8	25.9	26.5
Gonial angle	128.0 (5.0)	122.1 (5.3)	122.9 (4.4)	121.8	124.7	124.7	126.7
U1-SN	104.0 (6.0)	105.9 (8.8)	105.2 (8.8)	107.8	108.3	103.1	104.1
L1-mandibular plane	92.1 (5.5)	93.4 (6.8)	92.5 (5.4)	111.8	101.9	105.1	106.1
Interincisal angle	124.8 (5.8)	123.6 (10.6)	125.1 (10.1)	109.1	115.2	117.0	114.4
Occlusal plane/SN	21.6 (3.3)	16.9 (4.4)	17.6 (4.2)	19.7	24.8	25.5	25.6
Linear measurement (mm)							
S-N	64.2 (2.4)	67.9 (3.7)	66.9 (3.4)	66.0	71.2	72.9	72.9
N-Me	109.1 (3.9)	125.8 (5.0)	120.9 (4.4)	105.4	120.8	124.5	124.6
Ar-Go	38.5 (2.6)	47.3 (3.3)	44.2 (3.1)	45.9	56.2	56.2	56.3
Ar-Me	91.9 (4.2)	106.6 (5.7)	102.3 (4.8)	89.1	100.1	100.2	101.1
Go-Me	61.7 (3.6)	71.4 (4.1)	69.2 (3.5)	52.7	54.4	54.5	54.3
Overjet	2.6 (1.2)	3.2 (1.0)	3.1 (1.1)	2.0	3.5	2.0	1.7
Overbite	2.4 (1.7)	3.6 (2.0)	3.3 (1.9)	1.5	1.5	1.6	1.5

*Wada et al.[9]

Fig. 2: Functional appliance used in first phase

Treatment Objectives

This patient was diagnosed as FSBAS with mandibular deviation, skeletal Class II, and low mandibular plane angle. The treatment objectives were (1) to control the maxillary and mandibular growth during growing period; (2) to promote the growth of the affected ramus and prevent worsening of facial asymmetry and the occlusal cant; (3) to establish a proper interincisal relationship; and (4) to achieve a functional and reasonable occlusion with a Class I occlusion. The treatment planning was (1) mandibular arch expansion by expansion plate in order to improve scissors' bite on the right molar region, (2) the use of functional appliance to prevent the impairment of mandibular deviation, and (3) surgery after growth if necessary.

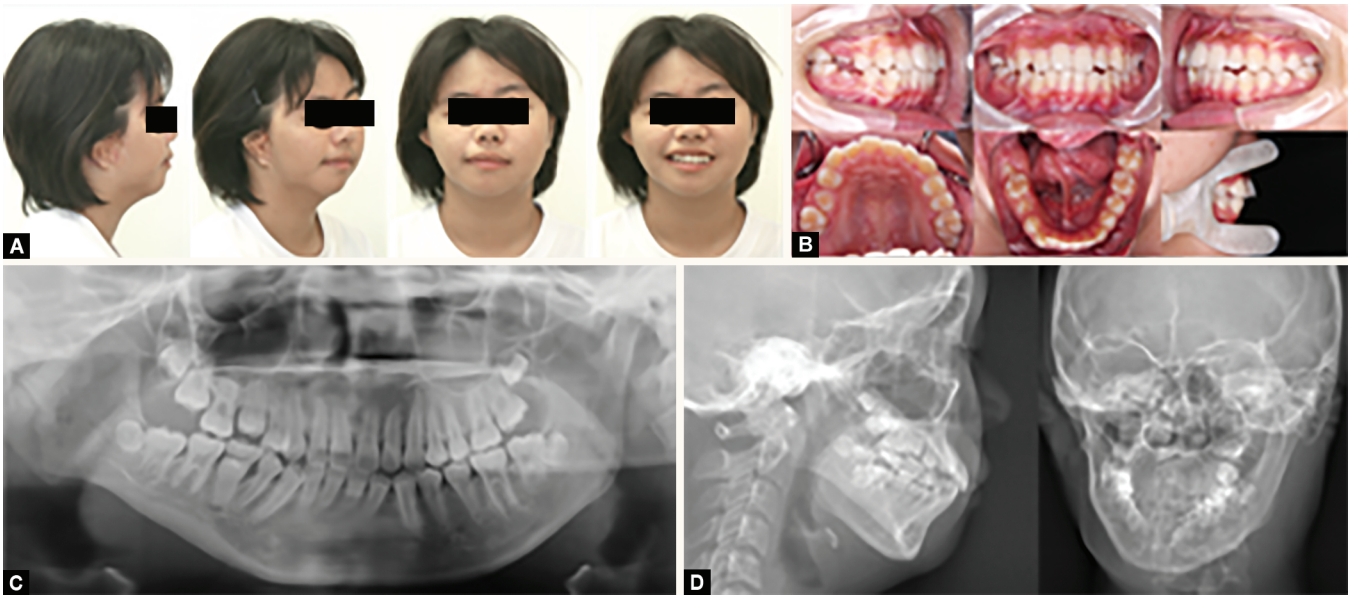
Treatment Alternatives

As patients with hemifacial microsomia Grade I and IIA possess proper function of the temporomandibular joint, mandibular lengthening by DO and/or conventional osteotomy can be commonly applied.[8] In Grade IIB and III patients, the temporomandibular joint is improper in both functional and morphological aspects, and total mandibular reconstruction is often required to achieve the temporomandibular joint function.[10] The present patient was classified Grade IIB, then surgical modalities, such as the DO and conventional osteotomy during or after mandibular growth, were applicable. However, she and her parents desired to avoid surgical procedures because she had received several surgeries for ear reconstruction. Finally, we decided to improve her oral function and facial harmony to use functional appliance as the first phase treatment and conventional multibracket appliance with miniscrews as the second phase.

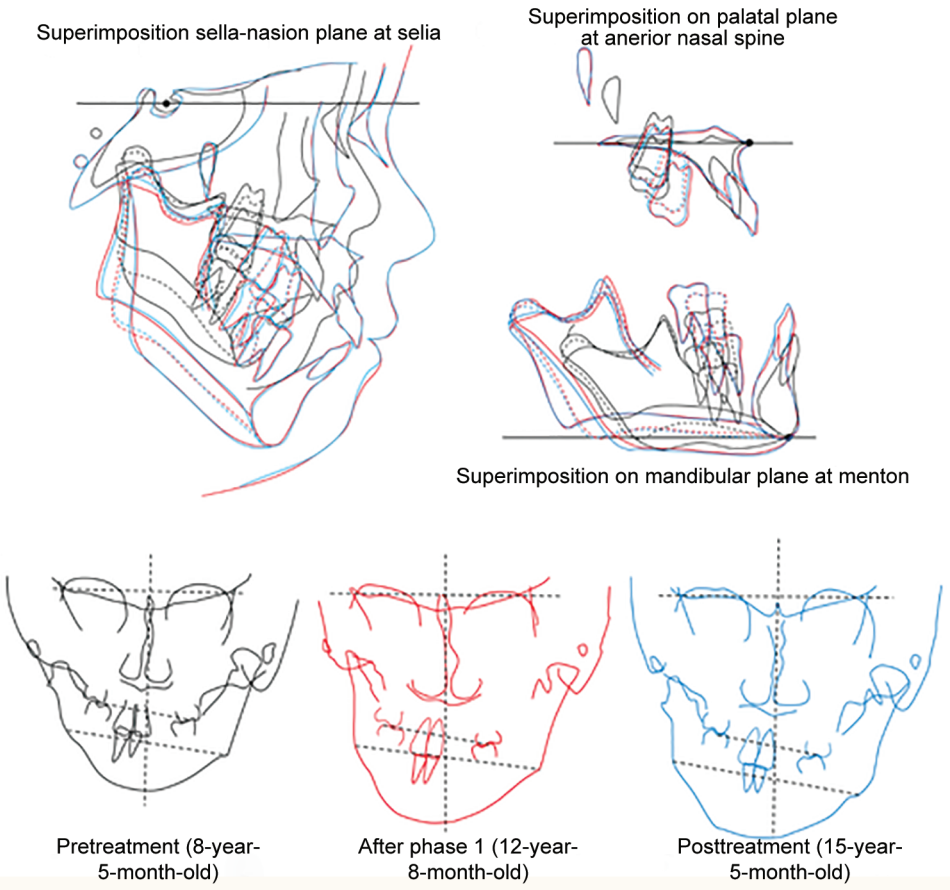
Treatment Progress

At the age of 8 years and 5 months, expansion plate was placed on the lower dentition to improve the scissors' bite at the left molar region. After 1-year treatment, the scissors' bite was improved, then the lingual arch was placed on the upper dentitions to retain the leeway space. At the age of 10 years, functional appliance with expander was placed at least 8 hours per day to correct her mandibular deviation and obtain stable occlusion (Fig. 2). At the age of 10 years and 7 months, sliding plate was additionally used at the daytime. After 2 years and 6 months of orthopedic treatment, the upper dental midline was matched up to the lower one, although the facial asymmetry with right-side mandibular deviation still existed. After the first phase treatment, at the age of 12 years and 8 months, all permanent teeth except right upper second molar had started eruption. The patient had mild crowding around upper and lower anterior teeth, and the overjet and overbite were +3.5 and +1.0 mm, respectively (Figs 3A and B). According to the model analysis, molar relationships were Angle Class I on both sides and arch length discrepancies were calculated to −0.5 mm for the maxillary dentition and ±0.0 mm for the mandibular dentition. Panoramic radiograph showed a considerable growth deficiency

and deformation of right mandibular ramus (Fig. 3C). Compared to the unaffected left mandibular ramus, the ramus height of the affected side was about 65.0%. Lateral cephalometric analysis indicated a skeletal Class II jaw-base relationship (ANB, 6.5°) and low mandibular plane angle (FMA, 25.8°) as it were before treatment (Figs 3D and 4A, Table 1). The upper incisal inclination was within the normal range, but the lower incisor showed labial inclination (IMPA, 101.9°). The frontal cephalogram showed deformation and



Figs 3A to D: Facial (A) and intraoral photographs (B), panoramic radiograph (C), frontal and lateral cephalograms (D) after first phase treatment at the age of 12 years and 8 months



Figs 4A and B: Cephalometric tracings before treatment (black line, 8-year-5-month-old), after first phase treatment (red line, 12-year-8-month-old), and posttreatment (blue line, 15-year-5-month-old). (A) Superimposition of lateral cephalometric tracings. The dotted lines indicate the average of the affected and unaffected sides; (B) Frontal cephalometric tracings

deviation of the mandibular bone to the right (Fig. 4B). The occlusal cant was 6°.

Then, preadjusted edgewise appliances (0.018-in slot) were placed on both upper and lower dentitions to improve mandibular deviation with crowding. After leveling, anchor miniscrews were obliquely placed between the maxillary second premolar and first molar. Since one anchor miniscrew placed on the right maxillary alveolar region was failed, we had to place the miniscrew again after 1 month. To move all upper dentitions distally, elastic chains with 2 N force were applied between the screws and hooks of canine brackets. After 32 months of active orthodontic treatment, a reasonable occlusion was obtained. After removing all appliances, a wraparound retainer and a spring retainer were used on the upper and lower jaws, respectively.

Treatment Results

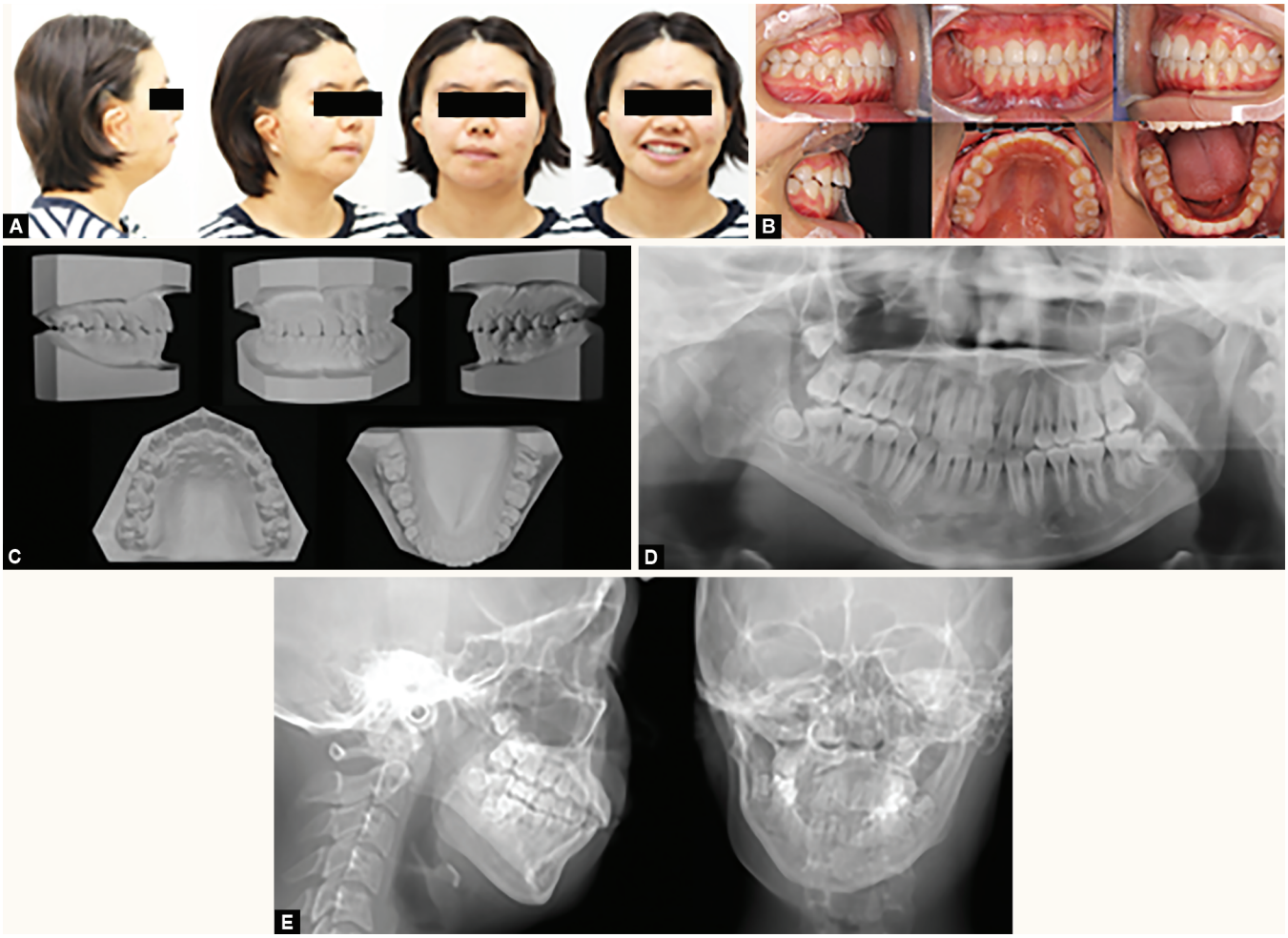
From facial photographs, facial asymmetry involved in the FSBAS was unchanged and her facial profile was maintained straight with a proper position of upper and lower lips (Fig. 5A). The right mouth corner was higher in vertical dimension than the left one even in smiling. The stable intercuspation of the teeth was obtained with a functional Class I relationship (Figs 5B and C).

Panoramic radiograph showed proper root parallelism; the upper and lower bilateral third molars were still impacted (Fig. 5D). Posttreatment cephalometric evaluation indicated a skeletal Class II jaw-base relationship (ANB, 6.3°) and low mandibular plane angle (FMA, 25.9°) (Figs 4A and 5E, Table 1). The maxillary bilateral molars were moved 1.0 mm distally, while the mandibular plane angle was unchanged. Both the maxillary and mandibular incisors were lingually inclined but acceptable interincisal relationship was maintained. The frontal cephalogram revealed that the occlusal cant of 6° remained the same as it was before the multibracket treatment (Figs 4B and 5E). Symptoms and signs of temporomandibular disorders were hardly observed throughout the active treatment.

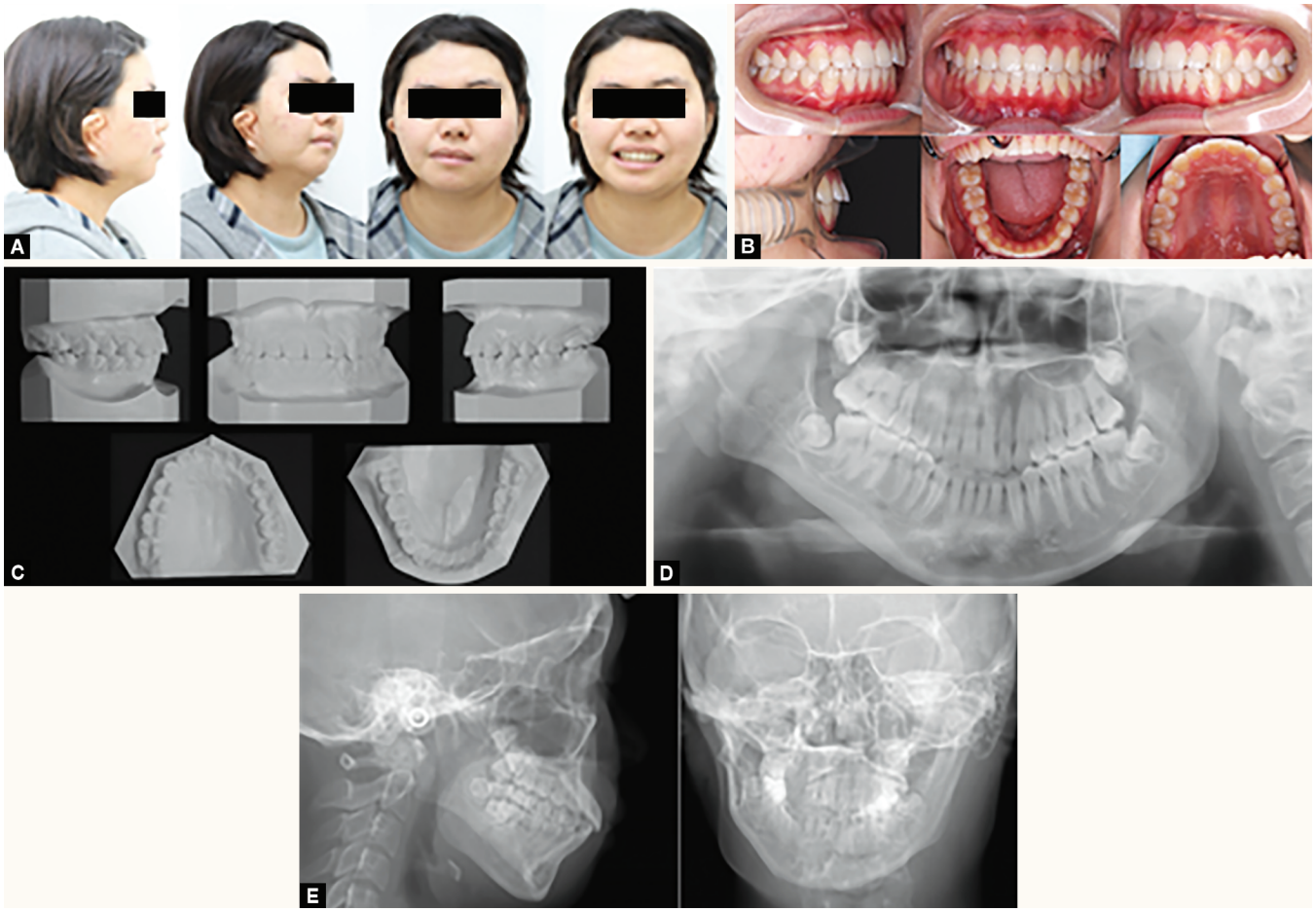
At 1.5-year postretention, the occlusion was stable without any relapse, and good facial profile was maintained (Figs 6A to C). However, facial asymmetry with height difference of the concerns of the mouth still existed. Panoramic radiograph and lateral cephalogram showed no or less changes (Figs 6D and E). From cephalometric analysis, the skeletal Class II jaw-base relationship with low mandibular plane angle was retained and no or minimal relapse of the mandibular deviation was detected (Fig. 6E, Table 1).

Discussion

In this patient, vertical compensation of mandibular ramus and condyle was required to treat the mandibular deviation associated with hemifacial microsomia. We conducted an orthopedic



Figs 5A to E: Posttreatment facial (A) and intraoral photographs (B), dental cast (C), panoramic radiograph (D), frontal and lateral cephalograms (E) at the age of 15 years and 5 months



Figs 6A to E: Postretention facial (A) and intraoral photographs (B), dental cast (C), panoramic radiograph (D), frontal and lateral cephalograms (E) at the age of 16 years and 10 months

treatment by means of functional appliance to minimize the impairment of mandibular asymmetry during growth period, leading to avoidance of mandibular DO and orthognathic surgery.

For patients with hemifacial microsomia involved in the FSBAS, mandibular lengthening by DO during growth period is one of the candidate treatment procedures.[8] Especially in Grade IIB and III patients, since temporomandibular joints are affected in both functional and morphological aspects, mandibular reconstruction is frequently needed to achieve sufficient function.[10] Although the treatment for facial asymmetry involved in hemifacial microsomia remains ongoing, the optimal timing of the surgical intervention has not been identified. Furthermore, little information is available about complications after mandibular DO for patients with hemifacial microsomia.[11] In fact, controversy has existed regarding the treatment for hemifacial microsomia patients with mandibular growth deficiency, especially the treatment outcome of mandibular DO. Zhang et al.[12] investigated the need for orthognathic surgery in adult patients with craniofacial microsomia: Some of them received early mandibular DO, and the other did not undergo early mandibular DO. They demonstrated that no significant difference for the rates of orthognathic surgery was found between the two groups of craniofacial microsomia patients. This implies small benefits of early mandibular DO in the patients with craniofacial hypoplasia. Furthermore, 35% of subjects treated with mandibular DO during growth period underwent repeated distraction or osteotomy after growth. In recent year, virtual three-dimensional surgical planning and guidance was promising tool for accurate treatment planning and sufficient surgical result in the treatment of adult hemifacial macrosomia patients.[13,14] This means that accurate prognosis for hemifacial microsomia treatment may contribute to the avoidance of the repeated distraction and/or osteotomy.

Conventional orthodontic treatment for hemifacial microsomia may initially include functional appliances, such as activator. These devices enable to expand the affected tissue, resulting in taking advantage of the physiological growth. They can often restrain mandibular growth on the healthy side, allowing for vertical compensation of the affected side. In the present case with Grade IIB hemifacial microsomia, the ramus height of the affected side was 60.4% of the healthy side before treatment, and after orthopedic treatment with functional appliance, the ratio of the ramus heights increased by 65.0%, indicating the vertical compensation of the affected condyle and ramus. In addition, the occlusal cant was also well maintained at 6° throughout the multibracket treatment. Although facial asymmetry due to hemifacial macrosomia did not improve completely, the patient was satisfied with the treatment result without early mandibular DO. Recently, Wang et al.[15] developed hybrid treatment strategy for hemifacial microsomia in the combination with early DO and a mandible-guided functional appliance to improve facial asymmetry during growth period, and reported that the hybrid technique proved to

be effective in correcting occlusal cant and mandibular deviation during mandibular elongation, resulting in symmetrical face and balanced occlusion. Therefore, further studies are indispensable to determine the optimal timing and the feasible technique of DO during and after orthopedic treatment, indicating the importance of orthodontic management during and after distraction.

Conclusion

We presented the orthodontic treatment of a patient with hemifacial microsomia associated with the FSBAS. With conventional orthodontic treatment, an acceptable occlusion with functional Class I canine and molar relationships were obtained. Vertical compensation of the mandibular ramus and condyle by functional appliance might be useful in a growing patient with hemifacial microsomia, leading to avoidance of mandibular DO.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Authors' Contributions

Ayami Ito, Kazuna Tachiki, and Ryoko Shioyasono contributed to performing orthodontic treatment and writing the manuscript. Mohannad Ashtar contributed to editing the manuscript. Keiichiro Watanabe and Masahiro Hiasa contributed to managing the orthodontic treatment as senior instructors. Eiji Tanaka contributed to planning of orthodontic treatment and editing the manuscript. All authors read and approved the final manuscript.

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