CASE REPORT

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Orthodontic treatment of a case of Becker muscular dystrophy

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Abstract
Becker muscular dystrophy, similar to Duchenne muscular dystrophy, is a X-chromosomal linked anomaly characterized by progressive muscle wasting and weakness. Duchenne-type is known to have severe openbite with a steep mandibular plane, but there are no studies that describe the occlusal and skeletal patterns of the Becker-type. Here, we report the orthodontic treatment of a Becker muscular dystrophy patient. In the correction of his severe skeletal open bite general anesthesia or orthognathic surgery was not an option. Multiloop edgewise archwires were employed for orthodontic treatment. After 3 years and 8 months the open bite was corrected. During the retention period contact between the anterior teeth was maintained 8 months after active treatment despite a marked relapse tendency.

Key words: Becker muscular dystrophy; myopathy; orthodontic treatment

Introduction

Duchenne (1, 2) and Becker (3, 4) muscular dystrophy patients exhibit malignant and progressive myopathy. The cause of both types is the mutations of the *dystrophin* gene. They are seen only in males as they are both X-chromosome linked conditions (1, 5). The frequency of Duchenne-type is reported to be one in 4000 and the Becker-type is about one tenth of the Duchenne-type. Both types of patients manifest unsteady and clumsy gait in the early stages. The myopathy progressively worsens with age and ultimately effects breathing and circulation (6). Becker type is distinguished from the Duchenne-type by

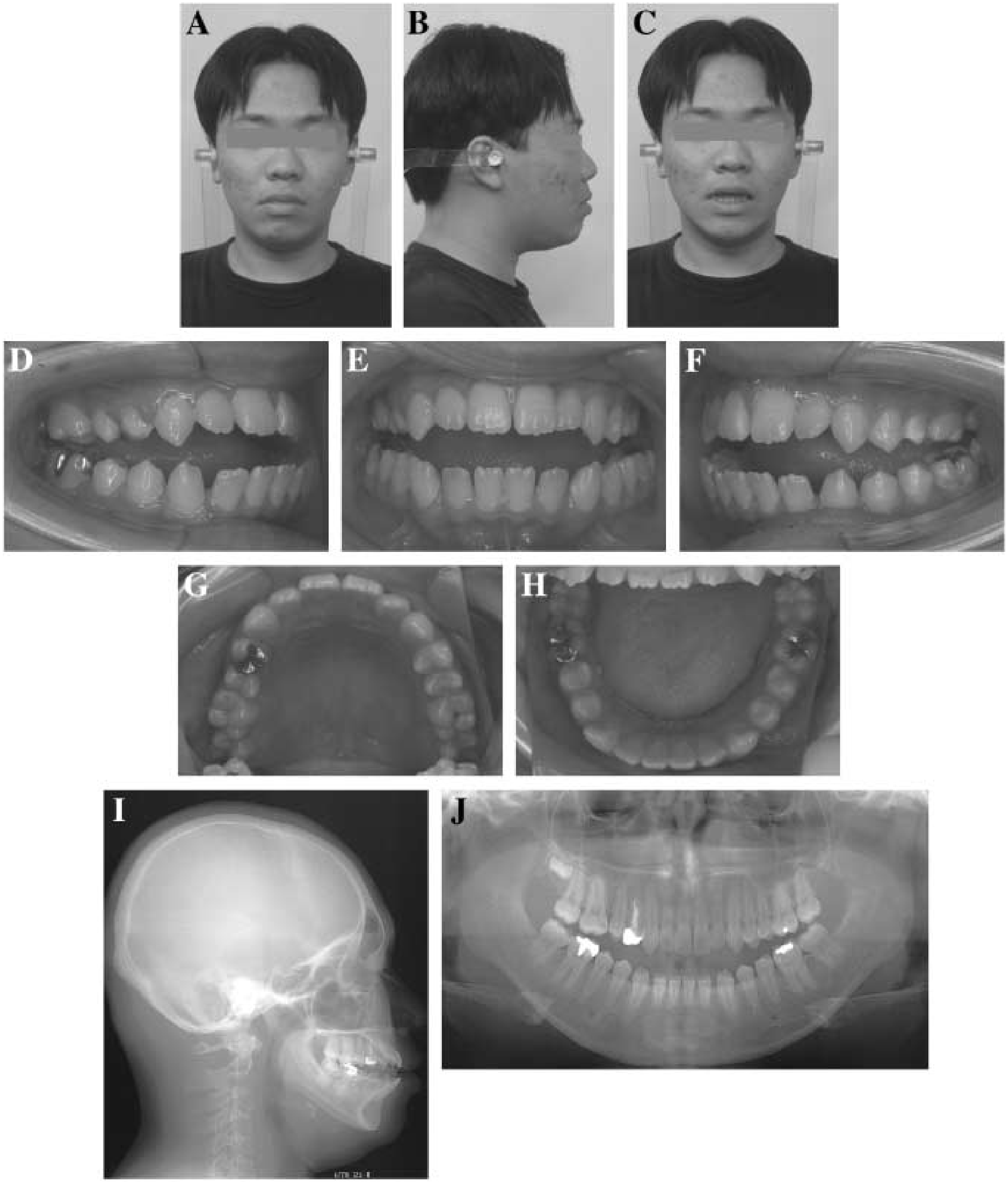

Fig. 1. Pretreatment facial (A–C) and intraoral (D–H) photographs and radiographs (I, J).

partially functional dystrophin peptide. The progression of the Becker-type is slower and the muscles are affected mildly. Also, the life expectancy is longer than that of Duchenne-type (3, 4).

It is reported that patients of Duchenne-type commonly exhibit a severe anterior open bite with a steep mandibular plane (7–9). But there are no reports that describe the occlusal and skeletal characteristics of the Becker-type. Also lacking are reports of orthodontic treatment in these two forms probably because patients suffer from serious masticatory and occlusal problems. Here we report the case of Becker muscular dystrophy patient who was treated for his open bite.

Case report

This patient was born to healthy parents. He has an unaffected older sister and no other members of the family are affected. He started to walk somewhat delayed, at age 14 months. At 10 years of age, he had difficulty going up the stairs; a biopsy of brachialis

Table 1. **Analytic measure before treatment**

	Value	Japanese norm ± SD
SNA (degree)	86.0	81.82 ± 3.09
SNB (degree)	80.0	78.61 ± 3.14
ANB (degree)	6.0	3.28 ± 2.66
U-1 to FH plane (degree)	120.0	108.94 ± 5.62
L-1 to mandibular plane (degree)	108.0	94.67 ± 7.21
Mandibular plane angle (degree)	33.0	26.25 ± 6.34
Gonial angle (degree)	130.0	119.38 ± 5.83
Maxillary inter-first premolar width (mm)*	46.4	44.77 ± 2.61
Mandibular inter-first premolar width (mm)*	44.8	36.26 ± 1.99

*Distance between buccal cusps of the right and left premolars.

muscle and the Western blot analysis were carried out. He was diagnosed as Becker muscular dystrophy. He presented to our orthodontic clinic at 16 years and 10 months of age with the chief complaint of difficulty in mastication. His facial appearance expressed a weak muscular tonus (Fig. 1A–C). The occlusal examination revealed a severe −6.0 mm of overbite with and 0 mm of overjet, and only the distal cusps of the maxillary second molars were in contact with the mandibular second molars (Fig. 1D–F). Widths of the upper and lower dental arches were wide and the tongue appeared to be devoid of tension and was voluminous (Fig. 1G, H). Spaced mandibular arch and proclined maxillary and mandibular incisors were noted. The lateral cephalogram and panoramic radiograph showed mesially tipped molars in both arches and a short

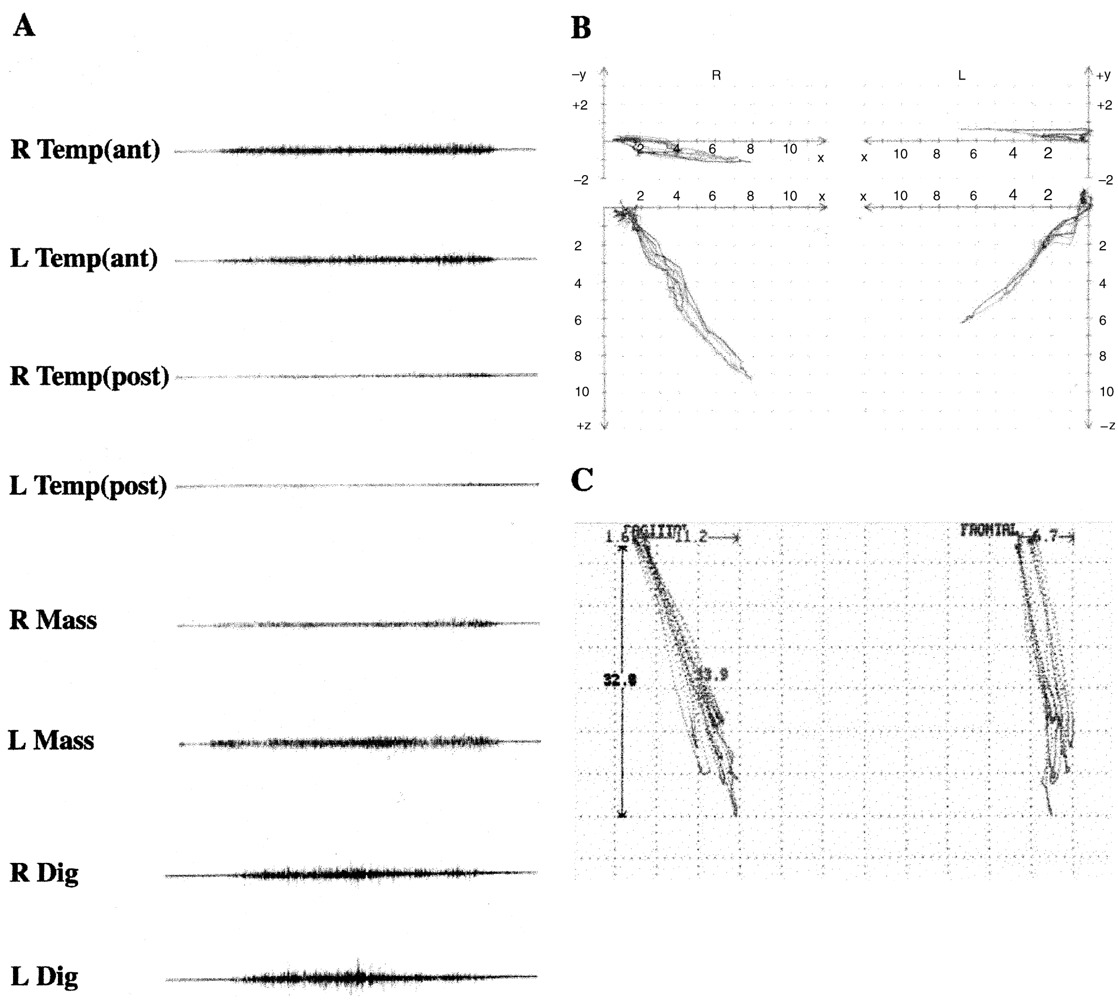

Fig. 2. Pretreatment record of electromyography (EMG) (A), computer-aided diagnostic axiography (CADIAX) (B) and mandibular kinesiography (MKG) (C). R Temp(ant), right anterior temporalis muscle; L Temp(ant), left anterior temporalis muscle; R Temp(post), right posterior temporalis muscle; L Temp(post), left posterior temporalis muscle; R Mass, right masseter muscle; L Mass, left masseter muscle; R Dig, right digastricus muscle; L Dig, left digastricus muscle. EMG was recorded from the temporalis and masseter muscles under the maximum bite force and from the digastricus muscle at wide-open position.

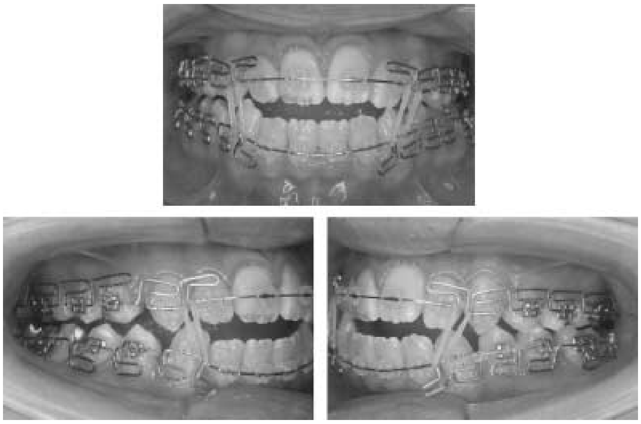

Fig. 3. Intraoral photographs of the active orthodontic treatment.

vertical height of the mandibular body and ramus (Fig. 1I, J). The SNA and ANB angles were larger than the Japanese male norm, and the gonial angle was markedly large resulting with a clockwise rotation of the mandible and a steep mandibular plane (Table 1).

To examine the stomatognathic function, electromyography (EMG) of the masticatory muscles using bipolar surface electrodes (Fig. 2A) and computer-aided diagnostic axiography (CADIAX; Gamma, Klosterneuburg, Austria) (Fig. 2B) and mandibular movements with mandibular kinesiography (MKG; K6-I, Myotronics, Tukwila, WA, USA) (Fig. 2C) were recorded. For the EMG recording, a pair of surface electrodes was attached bilaterally to the masseter and anterior and posterior temporalis muscles at the point of maximum thickening and in line with the muscle fibers. The recordings of EMG exhibited harmonized activity between the right and left sides of temporalis and masseter muscles under maximum bite force, and that of digastric muscle at wide-open position. For the frequency domain analysis, after the EMG record was converted with 12-bit analog-to-digital converter (Powerlab, ADI, Colorado Springs, CO, USA), 1024-point running fast Fourier transforms with a resolution of 8 Hz were computed with a 50% overlap for each data file. The median power frequencies (MPF) were 155.27, 163.07 and 188.72 Hz in the anterior temporalis, masseter and the digastric muscles, respectively. It is

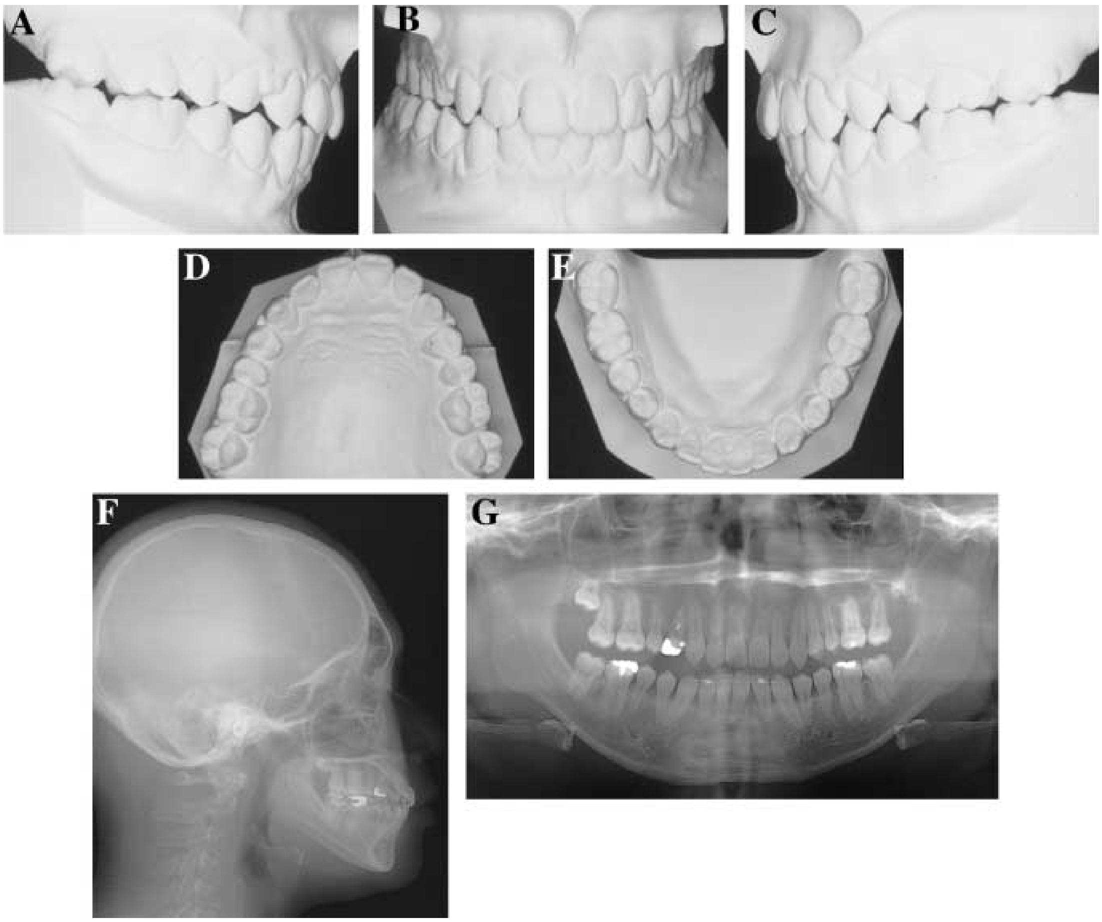

Fig. 4. Cast model (A–E) and radiographs (F and G) immediately after the active treatment.

Table 2. **Analytic measures during the treatment**

	Pretreatment	Immediately after active treatment	Eight months after active treatment
SNA (degree)	86.0	84.0	84.0
SNB (degree)	80.0	80.0	79.5
ANB (degree)	6.0	4.0	4.5
U-1 to FH plane (degree)	120.0	109.5	109.5
L-1 to mandibular plane (degree)	108.0	93.5	93.5
Mandibular plane angle (degree)	33.0	33.0	35.0
Gonial angle (degree)	130.0	130.0	130.0
Maxillary inter-canine width (mm)*	37.4	37.0	37.6
Maxillary inter-first premolar width (mm)*	46.4	46.8	46.4
Maxillary inter-first molar width (mm)*	54.4	56.6	54.7
Mandibular inter-canine width (mm)*	37.1	30.0	30.9
Mandibular inter-first premolar width (mm)*	44.8	42.4	42.9
Mandibular inter-first molar width (mm)*	57.2	54.8	56.7

*Distance between buccal cusps of the right and left teeth.

Fig. 5. Intraoral photographs 3 months after the active treatment.

reported that MPF of each muscle in the healthy Japanese male are 182.80, 183.51 and 186.40 Hz, respectively (9), implying this patient has lower power spectrum of the anterior temporalis and masseter muscles than healthy Japanese male by 11–15%. MKG and CADIAX showed the trace of the mandibular movement and the centric occlusion were both unstable (Fig. 2B, C).

As his open bite was quite severe and tongue voluminous, possibility of orthognathic surgery and glossectomy under general anesthesia were discussed. Unfortunately, because of the high risk that anesthesia might impair his circulation and worsen the muscles, no surgery or general anesthesia could be recommended. Thus, the treatment plan was to treat him by orthodontics alone using multiloop edgewise archwires (MEAW). MEAW was applied with intermaxillary elastics after spaces in the mandibular arch were closed (Fig. 3).

After 3 years and 8 months of active orthodontic treatment, the appliance was removed (Fig. 4A–E). Although the crossbite remained in the right-side molars and the midline was not matched exactly, his open bite was corrected with all teeth in contact. The inter-canine width of the mandibular arch was significantly decreased during treatment (Table 2). The lateral cephalogram and panoramic radiograph showed molars were upright especially in the mandibular arch (Fig. 4F, G). The SNA and ANB angles were slightly decreased (Table 2). The superimposition tracings showed extruded and retroclined incisors in both arches, and upright mandibular molars (Fig. 7).

Retention was initiated with removable and 3–3 fixed retainers with intermaxillary elastics between the incisors. Intraoral photographs taken 3 months after active treatment showed that the return of the open bite started in the premolar and molar regions (Fig. 5). Five months later marked relapse was noted (Fig. 6C–G). In the facial pictures it can be seen that the lips were retracted (Figs 1B and 6B). Also, a difference in the head posture was apparent between these two stages.

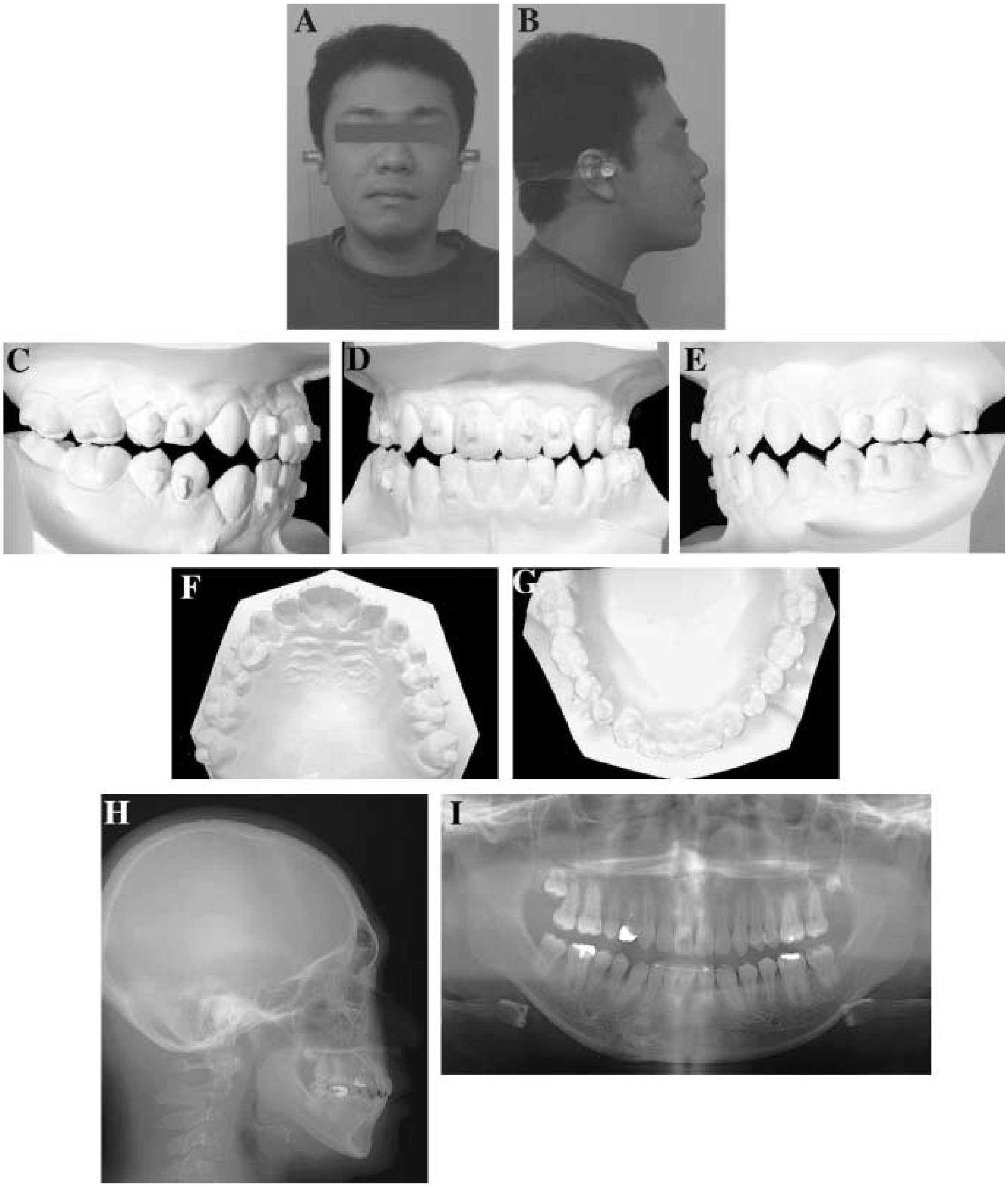

Fig. 6. Facial photographs (A and B), cast model (C–G) and radiographs (H and I) 8 months following the active treatment.

During the retention period, the SNB angle was slightly decreased and the mandibular plane angle increased. Arch width in both jaws returned toward their pre-treatment dimensions, i.e., the maxillary arch contracted and the mandibular arch widened (Table 2). Radiographically, molars maintained their corrected upright position (Fig. 6H and I) but the superimposed tracings showed that the extruded maxillary second molars tended to relapse (Fig. 7).

Discussion

Previous studies report that similar occlusal and skeletal patterns are observed in patients of Duchenne muscle dystrophy (7–9). They commonly exhibit a severe open bite with widened maxillary and mandibular arches. A steep mandibular plane, counterclockwise rotation of mandible and enlarged gonial angle are also invariably observed in this disease. This patient of

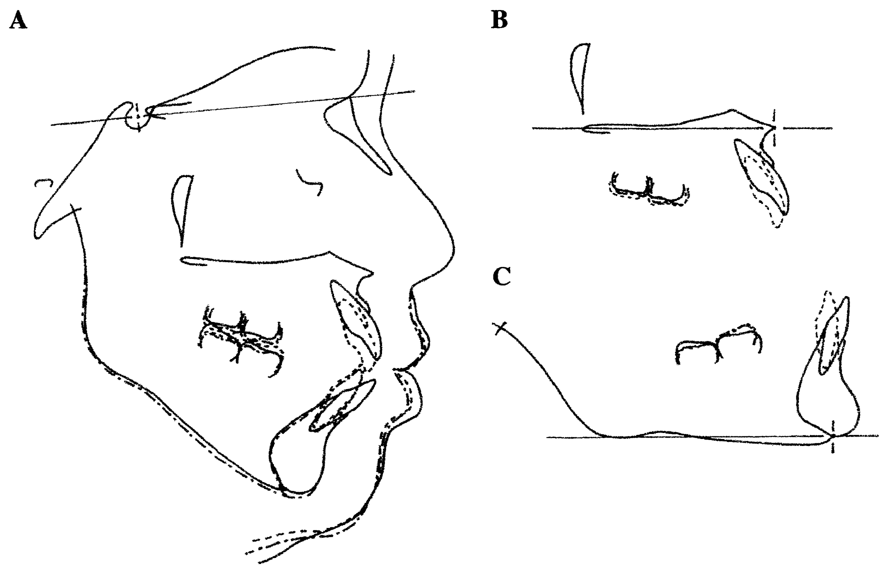

Fig. 7. Superimposed tracings. Pre-treatment (——), immediately after the active treatment, (- - -), 8 months after the active treatment (– · –). Superimposed at Sella turcica (A), ANS (B) and Menton (C).

Becker-type possessed these characteristics. As Becker-type is known to affect the muscles slower and milder than Duchenne-type (3, 4), a particular interest is in the difference of the timing of occurrence of the open bite between these two dystrophies. Unfortunately, neither the patient nor his parents had exact recollection when the open bite began to form.

The result of the frequency domain analysis showed that the present patient had lower MPF in the anterior temporalis and masseter muscles (mandibular elevator muscles), but not in the digastric muscle (mandibular depressor muscle), compared with healthy Japanese male. This was in agreement with the previous studies that the mandibular elevator muscle activity, but not that of the mandibular depressor muscle, tends to impair in patients of Duchenne-type. Mandibular elevator muscles are known to perform isometric contraction during mastication, which is likely to induce excessive loading and damage the muscle fibers (10, 11). This selective impairment of the elevator muscle activity together with the large tongue is speculated to be related to the etiology of the characteristic occlusal and skeletal patterns of Duchenne-type (9). An animal study has shown that the excision of masseter muscle in rats results in small mandibular angle and coronoid process, shortening of mandibular height and ramus, enlarged gonial angle, and extrusion of molars (12), all resembling the skeletal and occlusal patterns reported for Duchenne- and Becker-types.

In this case, incisors of both arches were significantly proclined at the start of treatment. But Eckardt et al. reported that the maxillary and mandibular incisors normally retroclined in patients of Duchenne-type (7). Certainly, the incisor inclination varies among patients depending on the balance of tongue size and tonus in muscles of facial expression and mastication.

In the absence of any surgical options it took 3 years and 8 months to complete the active treatment. Possible factors for this extended treatment time could be the weak closing muscle activity, the large tongue size, and limited cooperation by the patient. MEAW is an effective technique to correct open bites through intrusion of anterior teeth and distal tipping of the molars (13, 14). In the treatment of this young man, however, the intrusion of molars was only seen in the mandibular second molars. This could be due to the weak activity of mandibular elevator muscles. The extrusion of incisors as well as distal-tipping of mandibular molars were more likely to contribute for the correction of open bite (Fig. 7). We expect the long-term retention protocol will involve the continued use of intermaxillary elastics.

The case reported here demonstrates that the open bite caused by Becker-type muscular dystrophy can be corrected orthodontically, but it is difficult to prevent the marked relapse after the active treatment. Intensive care and an informed consent for the potential relapse, together with careful consideration to the systemic condition, would be essential for the orthodontic treatment of this disease.

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