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Case Report

Camouflage Treatment for Skeletal Maxillary Protrusion and Lateral Deviation with Classic-Type Ehlers-Danlos Syndrome

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We herein report the case of a 19-year-old female with a transverse discrepancy, skeletal Class II malocclusion, severe crowding with concerns of classic-type Ehlers-Danlos syndrome (EDS), aesthetics problems and functional problems. The main characteristics of classic EDS are loose-jointedness and fragile, easily bruised skin that heals with peculiar "cigarette-paper" scars. The anteroposterior and transverse skeletal discrepancies can generally be resolved by maxilla repositioning and mandibular advancement surgery following pre-surgical orthodontic treatment. However, this patient was treated with orthodontic camouflage but not orthognathic surgery because of the risks of skin bruising, poor healing and a temporomandibular disorder. A satisfactory dental appearance and occlusion were achieved after camouflage treatment with orthodontic anchor screws and the use of Class II elastics, including the preservation of the stomatognathic functions. Acceptable occlusion and dentition were maintained after a two-year retention period. This treatment strategy of orthodontic camouflage using temporary anchorage, such as anchor screws and Class II elastics, may be a viable treatment option for skeletal malocclusion patients with EDS.

Key words: asymmetry, Class II, camouflage, orthodontic anchor screw, Ehlers-Danlos syndrome

Ehlers-Danlos syndrome (EDS) is a group of congenital connective tissue disorders. Symptoms may include tissue fragility, skin hyperextensibility and articular hypermobility [1]. The incidence is about 1 in 5000 births [2]. EDS is classified into the following six main types: the classic type, hypermobility type, vascular type, kyphoscoliosis type, arthrochalasia type and dermatosparaxis type [3]. The main features of classic EDS are loose-jointedness and fragile, easily bruised skin that heals with peculiar "cigarette-paper" scars [1]. Oral considerations include fragile and sensitive mucosa, early onset of periodontal defects, and poor organization of tooth-supporting tissue collagen. In addition, recurrent subluxation and dislocation of the temporomandibular joint (TMJ) are frequent complaints [4].

In general, patients with severe facial asymmetry and anteroposterior skeletal discrepancy are treated with surgical and orthodontic treatment to improve not only their occlusion but their facial aesthetics [5-7]. Although the most suitable treatment plan generally includes maxillomandibular osteotomy or other orthognathic surgery, surgical orthodontics may be hard for patients to accept because of the risks of bleeding and poor healing due to the invasiveness of such

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procedures. The skin of patients with EDS is stretchable, readily bruised, and slow to heal, and the collagen fibers are thin and form a loose interwoven network. The tissues therefore act like wet blotting paper, surgical sutures hold poorly, and wound dehiscence is a common problem [8]. Subluxation and dislocation of the TMJ are also recurring problems. Accordingly, we propose that it would be better to consider orthodontic camouflage for patients with EDS, even if they have skeletal problems.

This case report presents a relevant and simple camouflage treatment associated with facial asymmetry and skeletal Class II malocclusion in a patient with classic-type EDS. The treatment involves maintaining dental compensation and adjusting the midline position as much as possible using miniscrews and Class II elastics. The treatment was successful, and the patient achieved a satisfactory dental appearance and acceptable occlusion with preserved stomatognathic functions.

Case Report

A 19-year-old female presented at the outpatient department of our hospital with a chief complaint of anterior tooth crowding and a discrepancy of the midlines. According to her patient history, she had been diagnosed with EDS II of the current classic type at the time of her birth. Her mother had the same type of EDS that she had; however, this type of EDS cannot be confirmed by biochemical and molecular tests [9]. The patient underwent umbilical hernia repair at one year of age and had surgery for a groin hernia at five years of age under general anesthesia. She had finger joint hypermobility (Fig. 1A), and easy bruising with "cigarette-paper" scars in areas of trauma (Fig. 1B-D). Considering this information, which was confirmable, the case indicated a general pattern.

In the frontal view, facial photographs showed an asymmetrical face with the mandible deviating to the right. In the lateral view, the facial profile was convex with prominent upper and lower lips. The mandibular dental midline was deviated 5.5 mm toward the right compared with the maxilla, and the occlusal plane was canted (Fig. 2A). Although her occlusal relationships were shifted with functional interference by the upper lateral incisor and the lower first premolar (Fig. 2B),

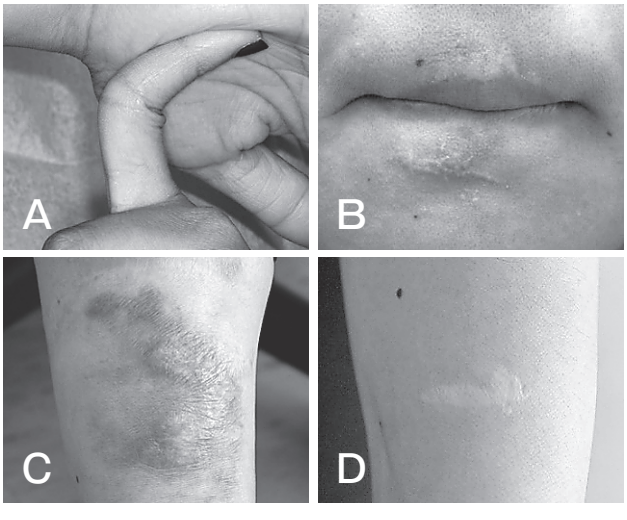

Fig. 1 (A) Hypermobility of finger joints, (B) bruising and atrophic scars, (C and D) "cigarette-paper" scars on the knee.

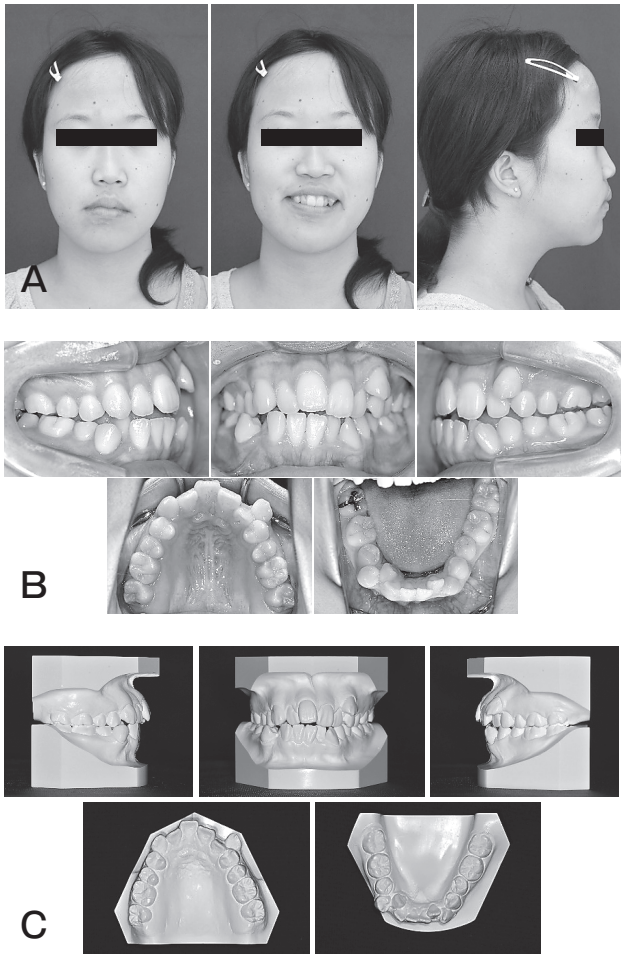

Fig. 2 Pretreatment views. (A) Facial photographs, (B) intraoral photographs (with occlusal interference), (C) dental casts (at centric occlusion).

Class I relationships on the right side and slightly Class II relationships on the left side were observed at the centric occlusion (Fig. 2C). The overjet was 5.5 mm, and the overbite was −0.5 mm. Crossbite or scissors bite was not present in the posterior region except in the right first premolar. There was severe crowding in the maxillary and mandibular arch, and their arches length discrepancies were −13.3 and −11.4 mm, respectively (Figs. 2B and C). Cephalometric analysis showed a skeletal Class II jaw-base relationship, a high mandibular plane angle, and normal inclinations of the maxillary and mandibular anterior teeth (Fig. 3A, Table 1) [10]. The posteroanterior cephalogram showed a maxillary deviation toward the left by 2.0 mm and a mandibular deviation toward the right by 6.0 mm from the facial midline. The difference in height between the maxillary left and right molars was 2.5 mm, and the cant of the occlusal plane was 3.0° (Fig. 3B). On the panoramic view, condylar resorption was observed at the right side, and the condylar neck was posteriorly inclined at the right side compared with the left. In addition, a panoramic radiograph showed that all of the third molars were impacted (Fig. 3C). A six-degrees-of-freedom jaw movement recording system showed that the condylar path length was shorter on the right side than on the left during maximum open-close jaw movements (Fig. 4A).

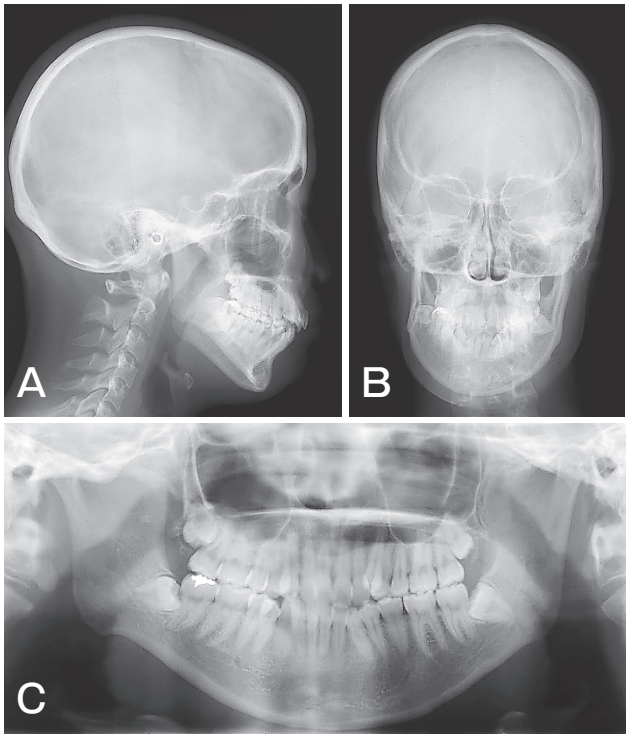

Fig. 3 Pretreatment cephalograms and a panoramic radiograph. (**A**) Lateral cephalogram, (**B**) posteroanterior cephalogram, (**C**) panoramic radiograph.

This patient was diagnosed with severe crowding, midline discrepancy associated with facial asymmetry, and a skeletal Class II jaw-base relationship with classic-type EDS. The treatment alternative was to correct the skeletal deformity, obtain ideal occlusion, and improve the asymmetric facial deformity by a combination of surgery and orthodontic therapy. However, we decided to treat her with orthodontic camouflage because of the risks of skin bruising, poor healing, and a temporomandibular disorder. It is possible to achieve a good treatment outcome using miniscrew anchorage in patients with skeletal discrepancy, as orthodontic camouflage with miniscrew anchorage can correct the anterior teeth crowding and the discrepancy between the midlines, which were the patient's chief complaints. To correct the crowding along with the dental midline in both arches as much as possible, the maxillary right first premolar, left canine and mandibular first premolars on both sides were extracted after making a model setup. Furthermore, the transverse dental compensation of the posterior teeth was to be retained, as it was expected that an optimal overjet and overbite relationship could be obtained by lingual bodily movement of

Table 1 Cephalometric summary

	Japanese norm (adult)		Pretreatment	Posttreatment
	Mean	SD		
Angular (°)				
ANB	2.8	2.4	7.5	7.0
SNA	80.8	3.6	79.0	78.5
SNB	77.9	4.5	71.5	71.5
U1-FH	112.3	8.3	107.5	104.5
L1-FH	56.0	8.1	52.0	47.5
L1-Mp	93.4	6.8	91.5	94.0
Mp-FH (FMA)	30.5	3.6	36.5	38.5
Linear (mm)				
Overjet	3.1	1.1	5.5	3.0
Overbite	3.3	1.9	−0.5	2.0
S-N	67.9	3.7	69.0	69.0
Ar-Go	47.3	3.3	42.0	43.5
Ar-Me	106.6	5.7	108.5	109.5
Go-Me	71.4	4.1	77.0	76.0
A'-Ptm'	47.9	2.8	48.5	47.5

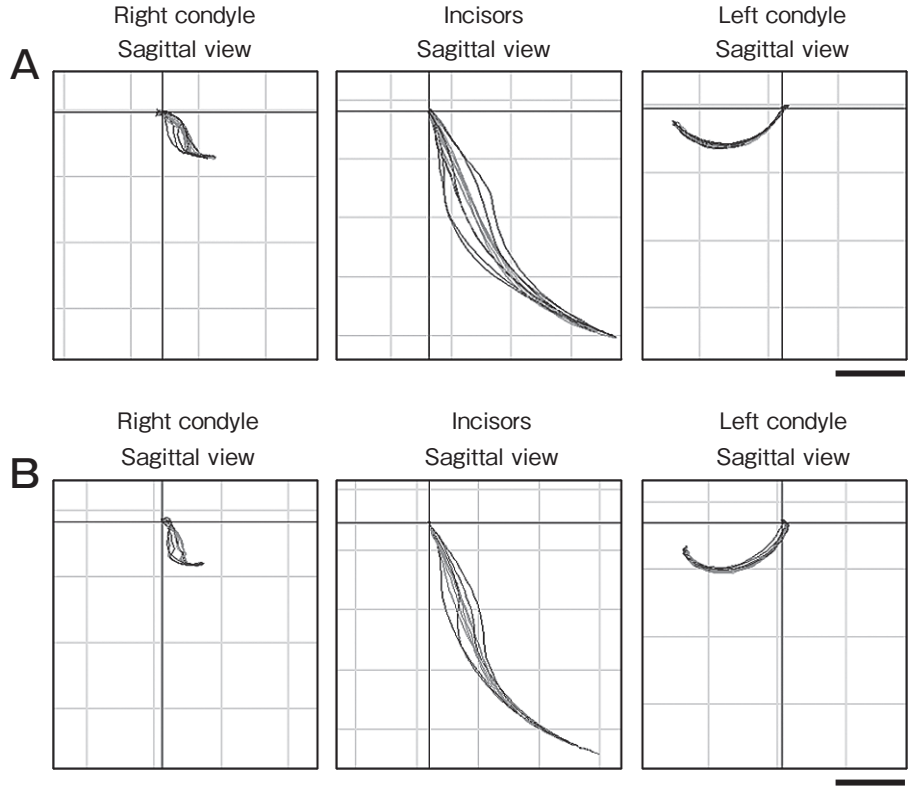

Fig. 4 Condylar movements and the incisal paths, as detected using a 6-degrees-of-freedom jaw movement recording system. The light line indicates the opening phase, and the dark line indicates the closing phase in maximum open-close. (**A**) Pretreatment, (**B**) posttreatment. Scale bar: 10 mm.

the maxillary incisors.

We explained the aims and of the treatment and the treatment alternatives to the patient, and obtained informed consent. After extraction of the maxillary right first premolar and left canine and the mandibular right and left first premolars, preadjusted edgewise appliances with 0.018 inch slots (Ortho-Dentaurum, Tokyo) were bonded to both arches except for the mandibular incisors. The arches were aligned with several replacements of round archwires, and the mandibular canines were retracted for three months. After retracting the canines, 0.018 inch preadjusted edgewise appliances were bonded to the mandibular incisors. The initial alignment was achieved with a 0.016 inch heat-activated nickel-titanium wire in both arches. All third molars were extracted during the leveling and alignment stage.

Under local infiltration anesthesia, miniscrews (diameter, 1.5 mm; length, 6.0 mm; Absoanchor; Dentos, Daegu, South Korea) were inserted into the buccal alveolar bone in the maxillary posterior region on both sides. After the leveling and alignment of both arches, 0.016×0.022 inch stainless steel wires were installed to facilitate midline correction and induce space closure of the extraction spaces intermittently for 5 months using miniscrews and loop mechanics. A lingual crown torque in the left posterior teeth and a buccal crown torque in the right posterior teeth were adequately built into the maxillary archwires. Regarding the mandibular archwires, lingual crown torque in the right posterior teeth and buccal crown torque in the left posterior teeth were also built into the archwires. The buccolingual axial inclination of the posterior teeth was maintained in this way. A Class II elastic was applied on the right side, and a vertical elastic was applied on the left side for 12 months to obtain as close to an Angle Class I molar relationship as possible.

Detailing was initiated with 0.016×0.022 inch stainless steel wires in both arches. The total active treatment period was 33 months. After removing the appli-

ance, the mandibular incisors were stabilized using lingual bonded retainers. Maxillary and mandibular wrap-around retainers were also placed.

Results

The treatment produced retraction of the upper and lower lips, which subsequently improved the patient's facial profile, although the facial asymmetry remained the same. The post-treatment facial and intraoral photographs showed that the midlines of both arches almost coincided with the mesiodistal inclinations of the incisors, and the oral aesthetics had improved. An adequate overjet (3.0 mm) and overbite (2.0 mm) were obtained. A Class I molar relationship was achieved on the left side, although the molar relationship on the right side was slightly Class II (Fig. 5, Table 1).

In our evaluation of jaw movements after the treatment using a jaw-movement recording system with six degrees of freedom, we observed a smooth and stable incisal path during maximum opening and closing movements. Furthermore, the condylar movement was maintained on both sides after orthodontic treatment (Fig. 4B).

The cephalometric analysis showed that the maxillary central incisors were lingually inclined by 3.0°, the mandibular incisors were labially inclined by 2.5°, and both the maxillary and mandibular incisors moved lingually. These changes contributed to a camouflage skeletal Class II jaw relationship. The mandibular plane angle showed clockwise rotation of 2.0° (Figs. 6A and 7A, Table 1). The frontal cephalograms showed that the mandibular deviation did not change after the treatment, and transverse dental compensation of the posterior teeth was maintained during the treatment (Figs. 6B and 7B). Although the posttreatment panoramic radiograph showed the enlarged periodontal space of the upper left central incisor, upper right canine and lower

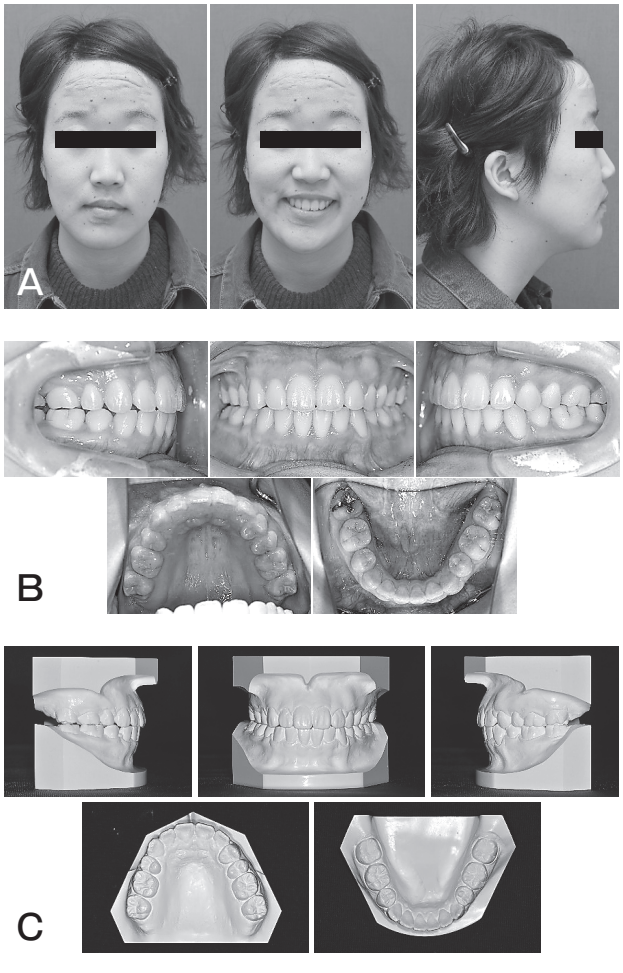

Fig. 5 Posttreatment views. (A) Facial photographs, (B) intraoral photographs, (C) dental casts.

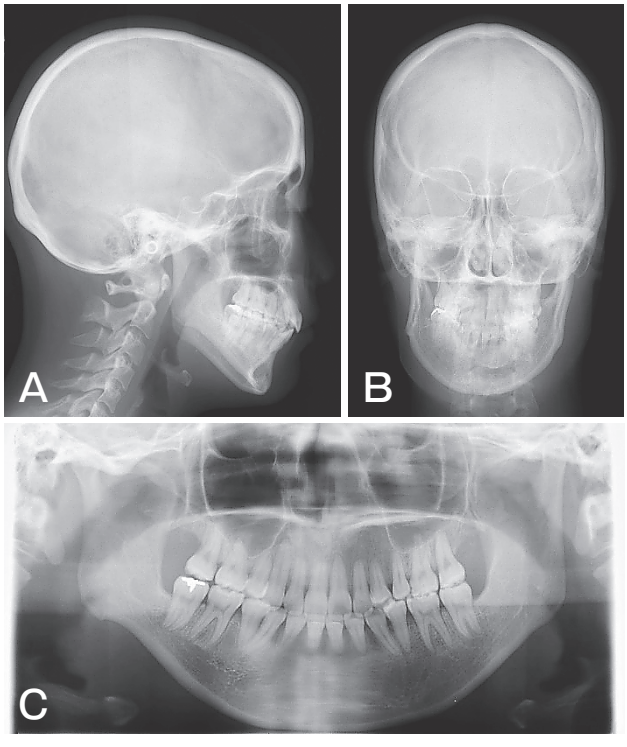

Fig. 6 Posttreatment cephalograms and a panoramic radiograph. (A) Lateral cephalogram, (B) posteroanterior cephalogram, (C) panoramic radiograph.

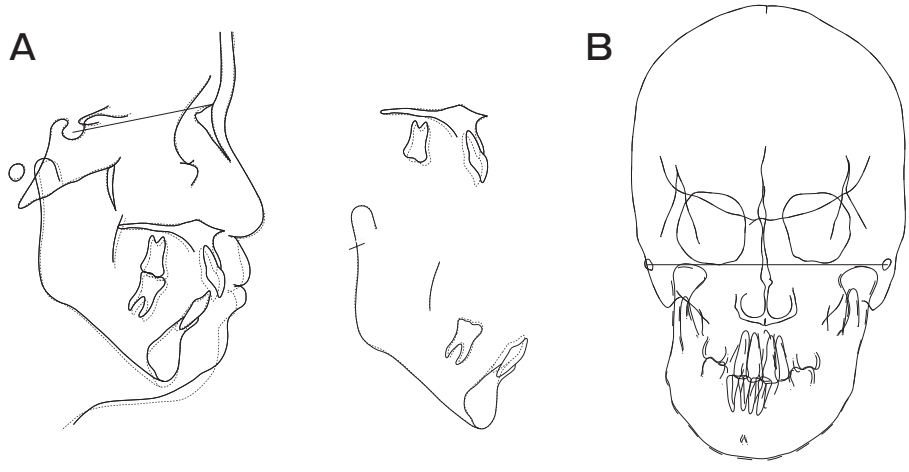

Fig. 7 Superimposed cephalometric tracings showing the changes from the pretreatment (solid line) to posttreatment (dotted line) stages. (A) The sella-nasion plane at the sella, (B) the zygion-zygion plane at the crista galli.

left lateral incisor, there was no marked root resorption (Fig. 6C).

The duration of the active orthodontic treatment was 33 months. After 2 years of retention, acceptable occlusion with a normal overbite and overjet was maintained for stability, and the patient was satisfied with the treatment results.

Discussion

The patient had a transverse discrepancy, skeletal Class II, Angle Class II malocclusion, and severe crowding with concerns about classic-type EDS, aesthetics problems and functional problems. The incisors and posterior teeth were given transverse dental compensation. Subluxation and dislocation of the TMJ were also recurring symptoms. Although defective dentinogenesis, pulp and root deformities, and high susceptibility to the development of temporomandibular disorder were suggested, a relationship between the classic-type EDS within dentofacial disorders and malocclusion has not been recognized [11, 12]. Evaluating of the cast and cephalometric analysis findings helped us make our initial treatment plan, which included orthognathic surgery. The first choice for treatment was orthognathic surgery followed by decompensation of the inclination of the posterior teeth [13, 14]. However, the excessive mechanical stress, stretching and pressure that can occure with orthognathic surgery under general anesthesia can aggravate cutaneous injuries including angulus oris. Orthognathic surgery had not been previously performed in any type of EDS patient. On the other hand, with mandibular third molar extraction under general anesthesia, the post-operative wound including the alveolar socket has previously been reported to heal well [15]. Based on all of the above, in the present case we selected orthodontic camouflage with tooth extraction.

Her camouflage treatment plan included retraction of the upper anterior incisors, correction of the midline discrepancies, and extraction of upper and lower teeth in order to obtain enough space for both arches. Anchorage control in fixed orthodontic treatment is one of the most important factors influencing the treatment plan and outcomes, particularly in adult cases of midline deviation [16]. Therefore, sufficient anteroposterior anchorage was deemed necessary. Miniscrew anchorage was applied to retract the maxillary incisors in order to improve the convex facial profile. In addition to the anteroposterior tooth movement, the correction of the midline deviation also contributed to the improvement of the oral aesthetics [17].

However, miniscrew-assisted maxillary incisor retraction failed to provide a force system that sufficiently maintained the position of her posterior molars, as mobilization of the miniscrews was repeatedly observed in this case. Defects in either the collagen-processing enzymes or collagen structure are ubiquitous in all subtypes of EDS. The miniscrew mobilization may have been caused by fragility of the oral

mucosa and bone throughout the collagen deformity [12, 18, 19]. However, there are no previous reports of the use of miniscrews in an EDS patient. Because our present report describes only a single case, further experience is needed to clarify the cause of the miniscrew mobilization. A previous report demonstrated that EDS patients have reduced bone mineral density and bone quality [20], suggesting that EDS is involved in the bone metabolism. In the present case, the physiological process of healing after the tooth extraction and the biological reaction to tooth movement appeared to be normal. Therefore, Class II interactive elastics were used for maxillary incisor retraction when the miniscrews fell off. Tooth movement in EDS patient is expected to be more rapid when a constant force is applied [21]. Therefore, light forces were used throughout the treatment to avoid any untoward effects. However, interarch elastics can cause molar extrusion, subsequently increasing the mandibular plane angle [22]. To prevent the mandible from rotating clockwise in such cases, the use of additional miniscrews, such as a midpalatal absolute anchorage system, should be considered [23].

In the present case involving camouflage treatment, the buccolingual axial inclinations of the posterior teeth were well maintained by torque control of the rectangular wires. The anterior open bite and crowding were improved by retraction of the maxillary and mandibular incisors with the extraction space. The periodontal spaces in the apical region which were visible on the panoramic radiograph immediately after the debonding of the fixed orthodontic appliances disappeared during the retention. These enlarged spaces may be caused by stress generated in the apical regions of the tooth roots with orthodontic treatment. The position of the mandibular condyle was moved slightly anteriorly in post-treatment. It seems the right side of the mandibular condyle had adaptability because of bone resorption, which is a symptom of EDS. On the other hand, the incisal path and condylar movement during maximum mouth opening and closing were maintained on both sides after the orthodontic treatment. Adequate improvement in both the functional occlusion and aesthetic balance was deemed to have been achieved, and stability was maintained after two years of retention.

With regard to wound healing after tooth extraction, even in EDS patients, surgical repositioning in the alveolar bone as in Wassmund and Köle osteotomy may be able to improve skeletal discrepancies. However, in EDS patients, it is not clear whether these functional disorders are improved with orthodontic treatment as there have been limited reports [8]. This is the first report in which a patient with classic-type EDS was treated by orthodontic camouflage. Further observation of the dental correction is required in order to assess the long-term stability of the patient's dentition.

In conclusion, we herein report the successful camouflage orthodontic treatment of a female patient with Class II malocclusion and facial asymmetry and classic-type EDS. This non-surgical approach may be an efficient alternative for improving the oral aesthetics and promoting a good quality of life.

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