CASE REPORT

Difficulties and treatment outcomes of orthodontic therapy of a patient with Williams Syndrome*

Elif Dilara Şeker(0000-0003-0331-8463)[α], Türker Yücesoy(0000-0002-1356-1574)[β], Ahmet Yağcı(0000-0002-8962-8392)[γ]

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ABSTRACT

Difficulties and treatment outcomes of orthodontic therapy of a patient with Williams Syndrome

Williams Syndrome (WS) is a rare congenital and progressive multisystem disorder which is described with specific systemic disorders, skeletal and dental abnormalities and tongue thrusting. A 13-year-old patient applied to our clinic with complaining dental caries, malocclusion and tongue thrusting. The patient was treated with a full fixed standard edgewise appliance named as Mini Master Series from American Orthodontics (Central Islip, N.Y., USA), 0.018 inch bracket slots) for 2 years and 8 months.

Despite poor oral hygiene, frequent appointments helped to complete the orthodontic treatment of the patient without caries formation. Also extraction of first molars with uncertain prognosis provided social benefits when it was compared to healthy premolar extractions. This case report presents the successful results of the orthodontic therapy and difficulties of an orthodontic treatment of a patient who was diagnosed with WS.

KEYWORDS

Anterior cross-bite, chromosome deletion, orthodontic treatment, Williams Syndrome

ÖZ

Williams Sendromlu hastanın ortodontik tedavisinin sonuçları ve tedavideki zorluklar

Williams Syndrome (WS) nadir görülen ve progresif bir şekilde ilerleyen multi-sistem bir sendrom olup özel sistemik hastalıklarla beraber, iskeletsel ve dental anomalilere ek olarak dil itme durumunun izlenebildiği bir hastalık tipidir. Bu vaka raporunda kliniğimize malokluzyon ve dil itme şikayetiyle başvuran WS sendromlu hastanın tedavisi sırasında karşılaşılan zorluklar ve tedavi planlaması anlatılmaktadır. Hasta standart edgewise braketler(Mini Master Series from American Orthodontics (Central Islip, N.Y., USA),0.018 inch braket slot) ile yaklaşık 2 yıl 8 ay tedavi edilmiştir. Bu vaka raporunda, WS tanısı ile takip edilen hastanın ortodontik tedavisi süresince karşılaşılan zorluklar ve bunlara rağmen elde edilen başarılı sonuçların paylaşılması hedeflenmiştir. Hastanın ortodontik tedavisi kötü oral hijyenine rağmen sık randevu aralıkları sayesinde çürük kaviteleri oluşmadan tamamlanmıştır. Ayrıca hastanın sağlıklı premolar dişlerinin ağızda tutularak prognozu belirsiz molar dişlerinin çekimi sosyal olarak hastaya avantaj sağlamıştır.

ANAHTAR KELİMELER

Anterior çapraz kapanış, kromozom delesyonu, ortodontik tedavi, Williams sendromu

Williams Syndrome (WS) was first defined by Williams *et al*[1] at 1961 and after three years, Beuren *et al*[2] described the phenotype of this syndrome with more dental details such as abnormally shaped teeth, hypodontia, microdontia, short and slender roots, enamel hypoplasia, conical or peg-shaped incisors, high caries rate and malocclusions.[2-6] WS syndrome is an uncommon congenital disorder in which the specific facial characteristics named as “elfin face”, infantile hypercalcemia, mental retardation, growth deficiency, sociable character could be also observed.[2,7]

This syndrome is diagnosed with characteristic facial features, a specific personality, inadequacy of development, short stature, cardiovascular abnormalities and infantile hypercalcemia.[2,8] However, WS occurs depending on submicroscopic deletion in the chromosome 7 (7q11.23) which contains elastin the chrom,osome 7 (7q11.23) which contains elastin gene.9 Although clinical characteristics of the syndrome are well described for diagnosis, fluorescent in situ hybridization (FISH) analysis is recommended as a standard procedure for the diagnosis of WS after the discovery of the microdeletion on chromosome 7 in WS.[9-11]

Because the incidence of WS is 1/20000 among live births and it is rarely reported in the literature, the knowledge about orthodontic or orthognathic treatment of WS patients is very restricted.[7,12,13] The aim of this case report is to describe dento-facial characteristics in a patient with WS before (T0) and after (T1) orthodontic treatment.

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[α] Bezmialem Vakif University Faculty of Dentistry Department of Orthodontics, Istanbul, Turkey

[β] Bezmialem Vakıf University Faculty of Dentistry Department of Oral and Maxillofacial Surgery, İstanbul, Turkey

[γ] Erciyes University Faculty of Dentistry Department of Orthodontics, Kayseri, Turkey

CASE REPORT

A 13-year-old girl was applied to Erciyes University, Faculty of Dentistry, Department of Pediatric Dentistry, Kayseri, Turkey for treatment of dental caries, malocclusion and tongue thrusting. She was consulted for orthodontic examination to the Department of Orthodontics with complaining from crowding in her teeth. When her family history was asked, it is learned that the patient's mother had no complications during the pregnancy and delivery. Her mother had three other healthy children who did not have WS symptoms whereas only one of four children was already diagnosed as WS in Erciyes University, Medical Genetics Department of Medicine Faculty, Kayseri, Turkey.

As another diagnostic symptom of WS; she was quite friendly and talkative and according to extraoral examination, the patient displayed typical "elfin" face and she had convex profile and asymmetrical face (Figure 1). Abnormal breathing pattern (mouth breathing) was also observed. Because of mouth breathing, the patient had a small nose and nostrils. Intraoral findings were poor oral hygiene, gingiva coloring, localized gingivitis, enamel hypoplazia, presence of large restorations in teeth, extracted tooth (mandibular right first molar), anterior cross-bite, open bite tendency and class I malocclusion (Figure 2). Furthermore, the patient had tongue thrusting problem while swallowing. Clinical findings supported the results of genetic test.

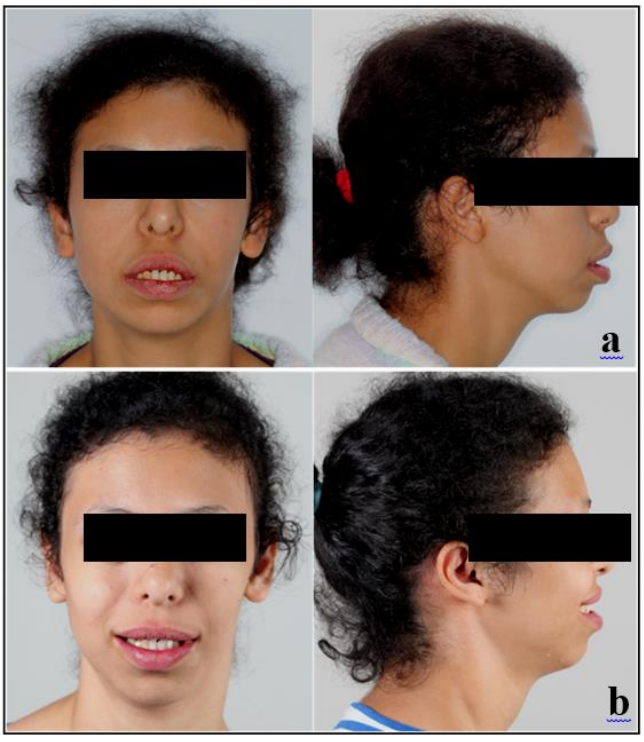

Figure 1.

Extraoral perspective, **a)** T1, **b)** T2

On the other hand, according to orthodontic examination, angle class I molar relationship and an anterior cross-bite were observed due to model analysis of the patient (Figure 3). Overjet and overbite were "-1" and "1" mm, respectively. Panoramic radiograph of the patient revealed that all permanent teeth and all third molar germs were available, lower right first molar was extracted and large restorations were observed in all first molars (Figure 4). The cephalometric radiography findings were shown in Table 1, which showed class I skeletal relationship and normal growth direction. According to the results of the cephalometric radiography, the anterior cross-bite was induced by protruded and proclined lower incisors (Figure 5). Both the upper and lower lips were positioned more anteriorly relative to Ricketts E line.

Due to these findings; the aims of treatment plan were determined as alignment and leveling the teeth, correcting the anterior cross-bite, obtaining a positive overjet and having a satisfactory interdigitation. For these reasons, the patient was consulted to medical genetics department for evaluation of the patient's systemic conditions at the beginning of the treatment to avoid systemic contraindications for our treatment. Fortunately, there were no reported contraindications for our orthodontic treatment plan.

After consultations, the treatment of our patient was decided to start with tooth extractions but however there were three options for the obtaining positive overjet and overbite as follows: extraction all of first premolars; extraction of the maxillary first molars and mandibular left first molar; extraction of the maxillary first premolars and mandibular left first molar. In orthodontic treatments, premolar extraction is generally preferred to provide esthetic, functional occlusion and to solve problems in anterior area. But, in the presented case, right mandibular molar was extracted and left mandibular molar was treated endodontically. Large restorations was observed both on the left mandibular molar and left maxillary first molar of patient. Additionally, primer contact elimination with orthodontic extraction of molar teeth can be more advantageous for patients with increased vertical dimension, So that, we preferred extraction of the maxillary first molars and mandibular left first molar also because of their uncertain prognosis.

Removals of these three teeth were performed to create symmetrical extraction spaces, because the patient's mandibular left first molar had already been extracted. So the patient was consulted to Erciyes University, Oral and Maxillofacial Surgery Department, Kayseri, Turkey for extractions. Three

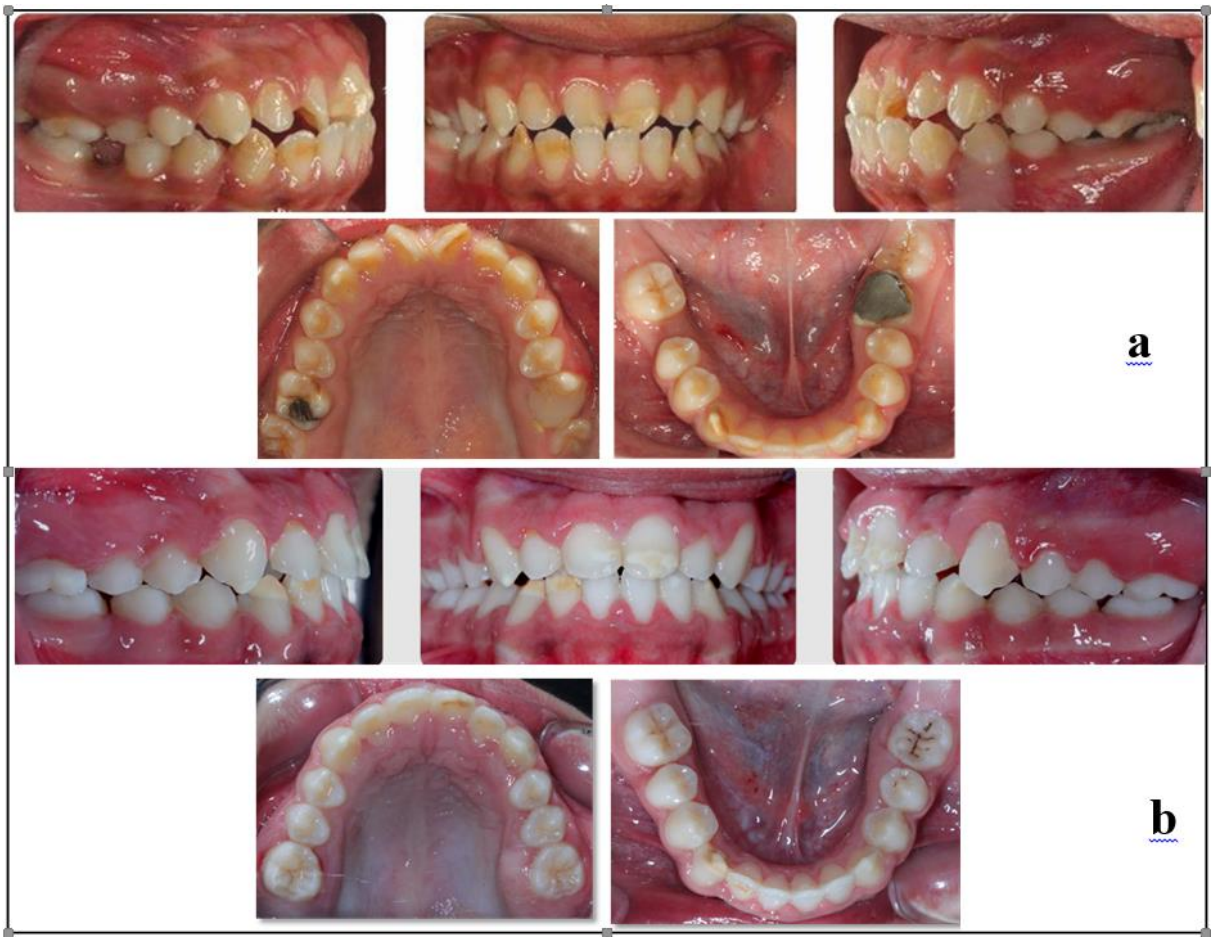

Figure 2.

Intraoral perspective, **a)** T1, **b)** T2

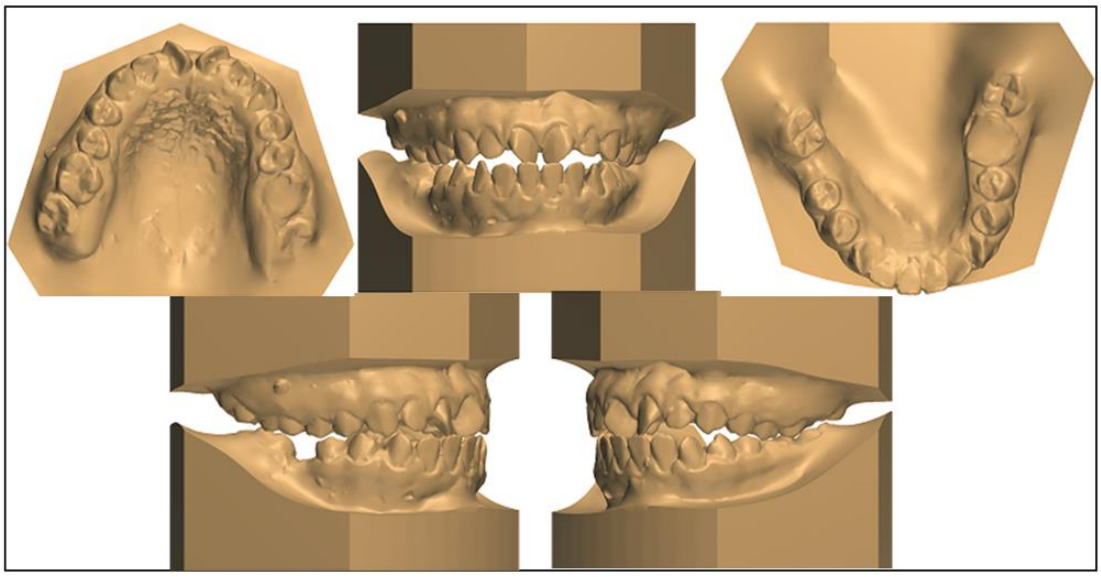

Figure 3.

Dental casts before treatment

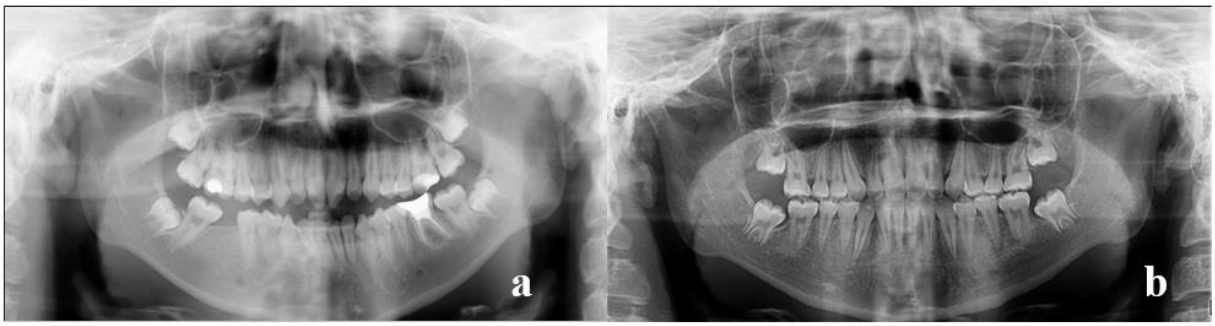

Figure 4.

Panoramic radiographs, **a)** T1, **b)** T2

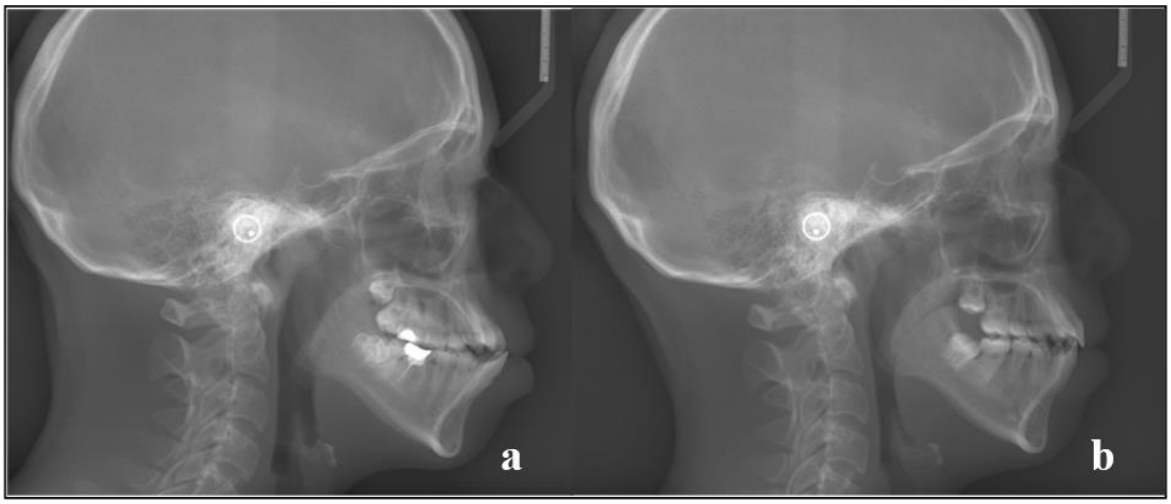

Figure 5.

Cephalometric radiographs, **a)** T1, **b)** T2

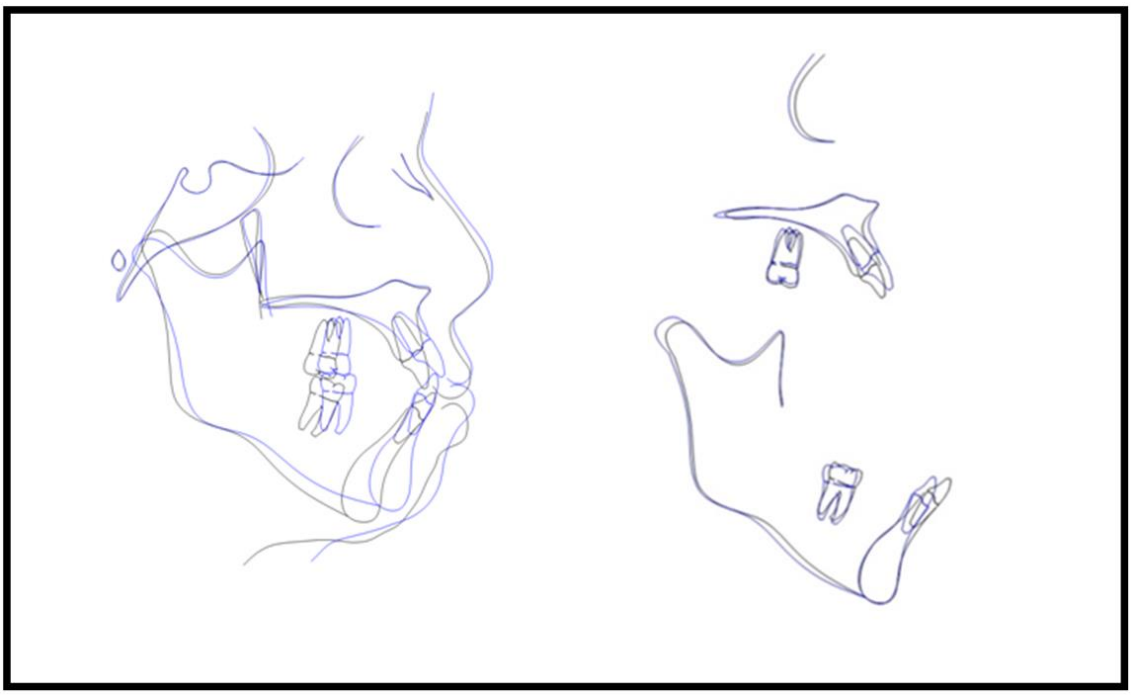

Figure 6.

Superimposed cephalometric tracings, Black line T1, Blue Line T2

Table 1.

Values of cephalometric analysis

	Normal	T1	T2
SAGITTAL			
SNA	82	82,2	82,4
SNB	80	80,7	81
ANB	1,6	1,5	1,3
N-A	0	-1,6	-1,3
N-Pog	-4	-8,5	-7
VERTICAL			
Y Axis Angle	60,5	63,6	63,9
SN-GoGN	38,4	36,2	33,5
SN-PP	7,3	6,8	6,1
Mx-Md Angle	25	31,6	31,3
DENTAL			
U1-SN	102,6	113,1	113,2
U1-PP	110	118,9	115,4
U1-NA	4,3 mm	10,3 mm	8,6
U1/NA	22,8	31	27,8
L1-Apog	2,7 mm	12,8 mm	7,9
IMPA	95	95	85,5
L1-NB	4 mm	12,7 mm	9
L1/NB	25,3	37,2	24
SOFT TISSUE			
Nasolabial Angle	104	108,7	119,7
ULip-E Plane	-4,8	4,3	0,6
LLip-E Plane	-2	9,6	5

teeth were extracted uneventfully under local anesthesia in the same session.

After one week of extractions, the brackets were bonded on the buccal surface of teeth. Straight-wire technique was performed with the purpose of closing extraction spaces. Class III elastics were frequently used during treatment. Also the patient was offered tongue exercises for the abnormal swallowing habit. The patient treated with a full fixed standard edgewise appliance (Mini Master Series from American Orthodontics (Central Islip, N.Y., USA), 0.018 inch bracket slots). After the leveling and aligning of the anterior teeth, the arch wire was changed to stainless steel rectangular arch wires; extraction spaces were closed with chains and elastics. The orthodontic treatment of patient with WS lasted 2

At the end of treatment, the teeth were aligned and leveled, satisfying interdigitation was achieved with the closure of the extraction spaces and positive overjet and overbite was obtained. Figure 6 shows the changes between before (T0) and after (T1) treatment. After debonding process of fixed appliances, the patient was recommended to wear an Essex plan for retention.

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Corresponding Author:

Dr. Elif Dilara ŞEKER
Bezmialem Vakif University Faculty of Dentistry
Department of Orthodontics
34093, İstanbul, Turkey
Tel : +90 543 778 50 93
E-Mail : dilaraarsln@hotmail.com