Aydın Dental Journal

Journal homepage: http://dergipark.ulakbim.gov.tr/adj

LARON SYNDROME: A CASE REPORT

Nil Ceren MUNGAN[1], Serap AKYÜZ[1]

ABSTRACT

Aim: The purpose of this case report is to define oral-dental findings of Laron Syndrome. Laron syndrome, is a very rare familiar disorder with autosomal recessive transition.

Case Report: A 11-year-old patient applied to Marmara University Faculty of Dentistry for dental treatment. She had growth inadequacy, small face and high pitched voice which are among the symptoms of the syndrome.

Materials and Methods: In the intraoral examination, decays and crowded teeth on the upper jaw were observed due to small sized maxilla. The patient also had gingival inflammation due to the poor oral hygiene. The restorations were made and the patient was informed about the oral hygiene behaviors. Clinical Findings: Patients with Laron Syndrome have severe postnatal growth failure and a low level insulin like growth factor- 1(IGF-1). Children have very high-pitched voice, saddle nose, delayed puberty and delayed onset of teething.

Keywords: *: Laron syndrome, growth hormone insensitivity, growth failure*

ÖZET

Amaç: Laron Sendromu otozomal çekinik geçiş gösteren ve nadir görülen ailesel geçişli bir hastalıktır. Bu olgu sunumunun amacı Laron Sendromu tanısı konulmuş hastanın ağız ve diş bulgularının tanımlanmasıdır.

Olgu Sunumu: 11 yaşındaki hasta çürük dişlerinin tedavisi amacıyla Marmara Üniversitesi Diş Hekimliği Fakültesi'ne başvurmuştur. Hasta sendromun bulgularından olan büyüme yetersizliği, küçük yüz ve ince-tiz sese sahiptir.

Materyal ve Metod: Hastanın ağız içi muayenesinde, diş çürükleri ve üst çene gelişiminin yetersizliğine bağlı olarak çapraşık dişler gözlendi. Zayıf ağız hijyenine bağlı olarak dişeti enflamasyonu tespit edildi. Hastanın çürük olan dişleri restore edilip ağız hijyen eğitimi verildi. Klinik Bulgular: Laron Sendromlu hastalar İnsülin Benzeri Hormon (IGF-1) eksikliğine bağlı olarak doğum sonrası gelişim yetersizliğine sahiptir. Bu hastalarda diş gelişiminde gecikme, ince ses, semer burun ve gecikmiş puberteye rastlanır.

Anahtar Kelimeler: *Laron Sendromu, Büyüme Hormonu Duyarsızlığı, Büyüme Yetersizliği*

[1] *Department of Pediatric Dentistry, Marmara University Faculty of Dentistry. Istanbul - TURKEY*

INTRODUCTION

Laron Syndrome (LS) is a rare familial disease with autosomal recessive transition. Growth hormone (GH) receptor gene has mutations or deletions.[1, 2] The syndrome was first reported in 1966 in three siblings of Israeli origin by Zvi Laron.[3,4] The incidence is not exactly known but the cases are reported mostly from Mediterranean or Middle Eastern regions.[1] Syndrome ratio for males and females varies similarly.[1] The growth hormone, which is the main factor of the postnatal growth, leads to inadequacy of generation of insulin-like growth factor 1 (IGF-1) when defects occur.[5, 6]

Patients with LS have a characteristic appearance such as; considerable reduction in adult height, frontal bossing, sparse hair, small maxilla-mandible and orbits, saddle nose, retardation of facial bone growth due to the lack of IGF-1.[4,6,7] Patients also have prominent high-pitched voice due to the narrow oropharynx.[5,8] Clinically progressive dwarfism is manifested with the adult height ranging from 116 to 142 cm in men and 108 to 136 cm in women. In addition to that, the hands and the feet are small that is called acromicria.[6,7] The typical clinical attributes of LS are elevated serum GH and low serum IGF-I.[9]

Patients with LS have smaller maxilla and mandible compared to healthy people therefore the appearance of their faces have smaller pattern and they have dental malocclusion.[10] In most patients the onset of teething is retarded, defective and breaking early with many caries because of small mandible teeth become crowded and irregular.[8] Patients with LS have increased enamel thickness but many of them lose their teeth during their midlife.[8] The roots of the permanent teeth delays development patterns and also the shedding of deciduous teeth are retarded.[10]

CASE REPORT

A 11 years-old patient, has received IGF-1 replacement treatment in Pediatric Department of Istanbul University Faculty of Medicine, for approximately 8 years. Until now, the patient has not caught up with the normal growth ratio. Even though, the patient's 11 year-old height is 110 cm and her weight is 24 kg, the Body Mass Index (BMI) is calculated as healthy. (19.8=healthy weight according to National heart, Lung and Blood Institute) (Figure 1).

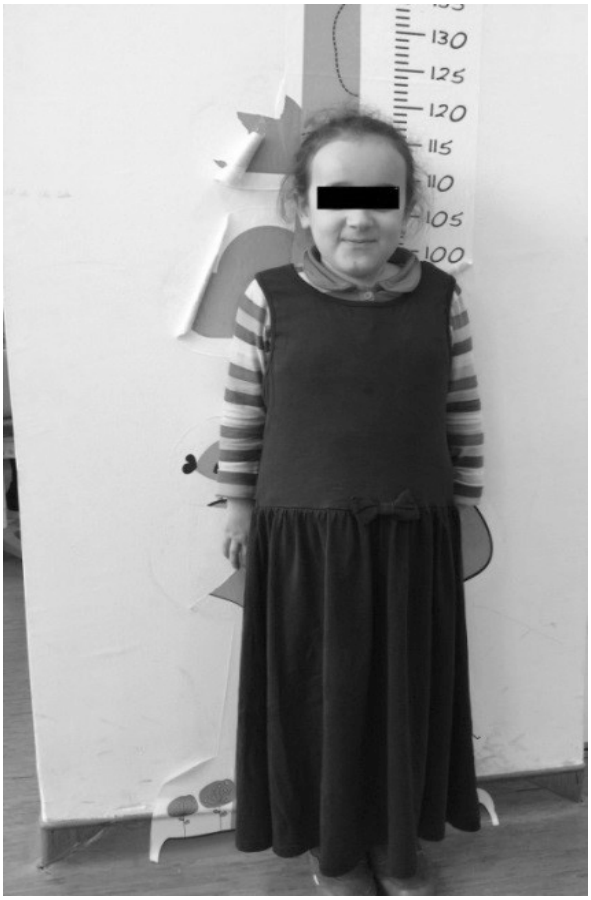

Figure 1: A 11-year-old girl with LS

The patient applied to Marmara University Faculty of Dentistry for the treatments of decayed teeth. She has the growth inadequacy, small face and pitched voice which are among the symptoms of the syndrome. Her parents

have consanguineous marriage and her younger sister also has LS.

In the intraoral examination, crowded teeth on the upper jaw due to small maxilla and decays in teeth with numbers; 16-26-36-55 were observed (Figure 2). The patient also had gingival inflammation due to poor oral hygiene.

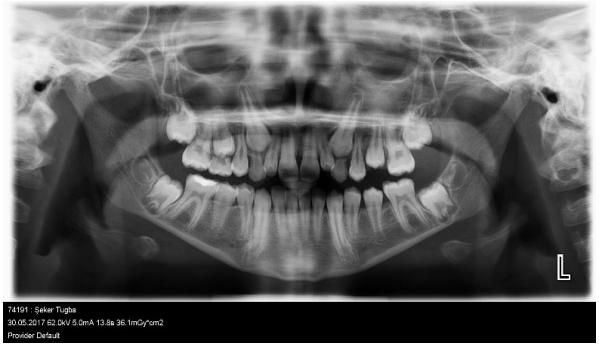

Figure 2 : Panoramic X-ray of the patient

The caries of permanent molars was the enamel caries that composite (Charisma smart) materials were used for the restorations. Additionally, compomer (Dentsply dyract) material was used for the restoration of the superficial enamel decay of the primary teeth. The fluor varnish (duraphat) was applied for the prophylactic treatment in case of potential caries (Figure 3).

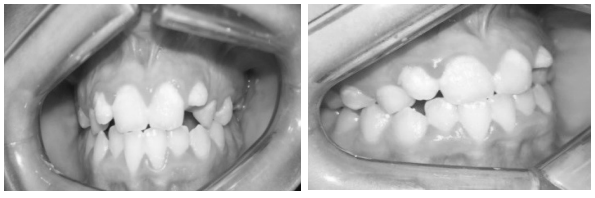

Figure 3: Intraoral clinical views of the patient

DISCUSSION

The only medical treatment option for LS is the injection of recombinant IGF-1 once a day. The injection should be administered with a daily meal so that hypoglycemia could be avoided.[4] If it is not injected with a meal, IGF-I may lead to hypoglycemia[5] Obesity, water retention, intracranial hypertension, tachycardia and arrhythmia are some of the adverse effects of IGF-I treatment.[4] The patient of our study also applies the injection of recombinant IGF-1 once a day.

The characteristic physiognomies of the patients with LS indicate the developmental delays because of the lack of IGF-1.[7] The patients also have oral and dental findings due to LS and treatment procedures.[4] The patient has dental malocclusion due to the small mandible and maxilla however all the dental-oral findings of the LS were not observed.

The patient consumes sucrose containing diets frequently, taking into consideration all high-risk factors, the teeth were expected to be with more caries. However, the patient did not have expected amount of decays despite poor oral hygiene.

The patients with LS have undeveloped body, face and mouth pattern due to the progressive growth failure.[5,6] Because of that; it is hard to administer complex and advanced dental treatments.

Regular dentist visits and prophylactic dental treatments are required to inhibit oral-dental problems when undersized maxilla- mandible and limited mouth opening are considered.

It is very fundamental to contact medical consultation for both medical doctors and dentists in such multiple organ involvement cases since the dentists play a specific role in diagnosing and treatment plans.[10]

Treatments in patients with LS will develop their life quality and also improve their self-confidence in social life.

REFERENCES

[1] Bhatia K, Cockerham R. Anaesthetic management of a parturient with Laron syndrome. International journal of obstetric anesthesia. 2011; 20: 344-46.

[2] Park K. A cell therapy-based cure of the Laron Syndrome. Journal of controlled release: official journal of the Controlled Release Society. 2013; 165: 90.

[3] Goswami S, Ghosh S, Chowdhury S. Growth hormone insensitivity syndrome: A sensitive approach. Indian journal of endocrinology and metabolism. 2012; 16: 817.

[4] Laron Z, Kauli R, Lapkina L, et al. IGF-I deficiency, longevity and cancer protection of patients with Laron syndrome. Mutation Research/Reviews in Mutation Research. 2017; 772: 123-33.

[5] Brooks AJ, Waters MJ. The growth hormone receptor: mechanism of activation and clinical implications. Nature Reviews Endocrinology. 2010; 6: 515-25.

[6] Cotta OR, Santarpia L, Curtò L, et al. Primary growth hormone insensitivity (Laron syndrome) and acquired hypothyroidism: a case report. Journal of medical case reports. 2011; 5: 301.

[7] Kornreich L, Konen O, Lilos P, et al. The globe and orbit in Laron syndrome. American Journal of Neuroradiology. 2011; 32: 1560-62.

[8] Laron Z. Laron syndrome (primary growth hormone resistance or insensitivity): the personal experience 1958–2003. The Journal of Clinical Endocrinology & Metabolism. 2004; 89: 1031-44.

[9] Diniz ET, Jorge AA, Arnhold IJ, et al. Novel nonsense mutation (p. Y113X) in the human growth hormone receptor gene in a Brazilian patient with Laron syndrome. Arquivos Brasileiros de Endocrinologia & Metabologia. 2008; 52: 1264-71.

[10] Atreja G, Atreja SH, Jain N, et al. Oral manifestations in growth hormone disorders. Indian journal of endocrinology and metabolism. 2012; 16: 381.