Patient with Regional Missing Teeth Encountered with Syndactyilia-Clinodactilia and Tongue Anomalies

Sindaktılı-Klinodaktılı ve Dil Anomalisi ile Birlikte Görülen Bölgesel Diş Eksikliği Hastası

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ABSTRACT Dental hypodontia (agenesis) is a common congenital anomaly. One or more missing teeth are more common, but multiple missing teeth are rare. In oligodontia, tooth defect is not usually seen as a segment involvement. The segmental involvement is the first regional odontodisplasia (RO) in mind. However, hand-foot and tongue problems are not characteristic in RO. This suggests that we have a syndrome in our case. In this case report, it is aimed to present and discuss the patient who has sebaceous, palpable adhesions in the hands and feet and missing primary and permanent teeth in a segment of the lower jaw. In this case, which is also likely to be a syndrome, our colleagues were also asked to get ideas.

Keywords: Odontodysplasia; tooth abnormalities; rare diseases


Anahtar Kelimeler: Odontodisplazi; diş anomalileri; nadir hastalıklar

Dental hypodontia (agenesis) is a common congenital anomaly. One or more missing teeth are more common, but multiple missing teeth are rare.

Oligodontia is a relatively rare condition and affects approximately 0.1-1.2% of the population. For these kinds of patients, a personal sequence of treatment through early diagnosis, comprehensive treatment planning, good coordination and timing is determinative to obtain a successful treatment outcome.¹

The defects developing during odontogenesis might occur separately or together with other organs and tissues. Understanding the relation between hereditary dental pathologies and normal odontogenesis will enable an accurate and effective treatment for patients and earlier diagnosis of certain syndromes including dental anomalies.

Regional tooth deficiency is one of the few rare anomalies and can often be seen with syndromes. In this case report, it is aimed to present and discuss the patient who has sebaceous, palpable adhesions in the hands and feet and missing primary and permanent teeth in a segment of the lower jaw. In this case, which is also likely to be a syndrome, our colleagues were also asked to get ideas.

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CASE REPORT

A 13-year-old boy was admitted to Süleyman Demirel University School of Dentistry Department of Pedodontics with the complaint of dental decay. Informed consent form was obtained from the patient. In patient’s anamnesis, it was found out that there is no relationship by affinity between his mother and father, that the patient is the last of three sons in the family, and that he was born mature and normal.

According to the anamnesis of the mother, it was found out that the patient had a tongue-tie and webbed fingers and toes when he was born, and that he had a series of surgeries for three years starting at the age of 10.

During his physical examination, it was observed that he has a depressed nasal bridge, there is color/structural difference above the earlobe level of his ears, and there are pigmented areas on certain parts of his body (Figure 1). Although the webbed fingers and toes (syndactyilia) were separated by surgeries, structural deformation remained (Figure 2). The patient’s height and weight at age 13 are 140 cm and 40 kg.

In the oral examination of the patient; it is seen that the right side of the tongue is not free, the permanent teeth on the lower right side are full, and all primary teeth except the 2nd primary molar teeth are missing and there is excessive crust loss in this region (Figure 3).

He has “V” shaped upper arch and a deep palate. It was determined with the Hayce Nance and Bolton Analysis that there is lack of space for the teeth. The tooth number 13 is located in the vestibule and there are some confusions in the teeth.

It was seen that his center-line was not aligned. Dental caries were detected in the patient’s teeth numbered 16, 21, 26 and 36 and it was found to have poor oral hygiene.

On panoramic radiographs, all premolars are double-rooted and taurodontism is present in pulp chambers. It is seen that the germs of numbers 18, 48 are absent (Figure 4).

Patient’s mother is 36 years old, healthy; father is 41 years old and has learned that there is a problem about eye and perception. In his father’s dental examination; early tooth loss, vertical alveolar crest loss, pulp chamber and linear calcifications in root canals. The mesiodistal dimensions of the teeth were narrow and oral hygiene was poor (Figure 5).

No significant data was found on the examination of the mother. He also learned that his grandmother had a history of early prosthesis use.
DISCUSSION

In oligodontia tooth defect is not usually seen as a segment involvement. Sökücü et al. in their study, incidence of oligodontia was found to be 1.83%. The segmental involvement is the first regional odontodisplasia (RO) in mind. However, hand-foot and tongue problems are not characteristic in RO. This suggests that we have a syndrome in our case.

Possible syndromes include; Oculodentodigital dysplasia (ODDD), a rare congenital autosomal dominant disorder characterized by abnormalities of the eyes, limbs, and teeth, Oro-facial-digital syndrome type 1 syndrome (OFDS) characterized by various degrees of bradydactyly, syndactylic and clinodactyly, and mutation in CXORF5 gene between finger 2 and finger 5 or with digital anomalies and malformed ears, as well as with syndromes such as Lacrimo-au-riculo-dento-digital (LADD).

Regional odontodysplasia is a relatively uncommon condition that affect odontogenic structures. Diagnosis is based on clinical and radiographic findings. In most cases, involvement of the upper jaw is seen, whereas in our case, involvement of the lower jaw is seen.

In addition, RO is more common in female, although our presence is male. The female/male ratio is 1.7/1.

Etiology is not fully known. Heredity does not play a role in etiology, because no familial transition has been reported. In our case, we think that missing teeth is hereditary because the father’s anamnesis showed with his general dental condition that he has many missing teeth though he had a few teeth out and that the father’s mother started to use prosthesis at early ages.

Although both primary and permanent teeth may suggest regional odontodysplasia, hand and foot adhesions are not characteristic of RO.

Be able to think in the diagnosis phase; in cases like Dentinal Dysplasia, Shell tooth, Dentinogenesis Imperfecta, Amelogenesis Imperfecta, Rickets, all dentition is affected unlike ours.

Affected teeth in RO; ghost appearance, abnormal shape, yellow or yellow-brown color, and hypoplasia or hypocalcility, unlike this case, there are no structures and deformities in the clinical crowns of the teeth.

Tervonen et al. reported that 50.4% of patients had inflammation in the gingiva and periodontium in RO patients, and gingivitis is present in our cases. However, this situation may be related to the inadequacy of oral hygiene, so the diagnosis may be insufficient alone.
The most characteristic clinical symptom of RO is the failure of tooth placement of RO-positive teeth in both the primary and permanent teeth. The teeth eruption is delayed and all of the teeth are not erupting. In our presence, number 17 is buried.

There are pigmented areas in certain parts of our body. Walton et al. and Steiman et al. reported RO case related with vascular nevus covering adjacent skin of the face.

Finger malformations are very common in patients with ODDD. Bilateral 4th and 5th fingers are syndactyl or syndactyly type III, the main feature of the syndrome. Camptodactlia, the shortness of the finger and the lack of phalanges of the toes are common. In our case, it is more pronounced in the 4th and 5th fingers and is seen as syndactylia and clinodactyly.

ODDD is due to mutations in the gap junction alpha1 gene (GJA1; OMIM 1210154) on chromosome 6q22-q23. However, since our patient did not accept his or her parents, genetic testing was not performed.

Ophthalmological findings include microphthalmia, microcornea, iris anomalies, glaucoma and hypertelorism. The gray color of our patient is close to eye color and strabismus is present. However, our patient does not have a recognized eye problem, but it is not known whether or not there is an eye problem during the follow-up period. As in our case, dry hair is also seen in this syndrome. Generalized enamel hypoplasia, microdontia, and sporadic cleft lip/palate can be seen, but we do not have these oral findings. Despite the similarity of LADD syndrome to digital anomalies and malformed ear in our patient, lacrimal duct aplasia, deafness, and other characteristics of the syndrome are not available. We also do not have major clinical oral findings of LADD such as microdontia, enamel hypoplasia.

Oro-facial-digital syndrome characterized by mutation in the CXORF5 gene, bradidactylic, syndactylic and clinodactyly, at various degrees between the second finger and the fifth finger. Type 1 syndrome is considered, but the possibility that the syndrome is lethal when seen in men has almost eliminated the possibility.

In such cases, it is important to have a genetic diagnosis. However, genetic testing has not been conducted on whether the parent is not convinced of the necessity of genetic testing or not.

Since our patient had never visited a dentist before and did not use prosthesis, there is atrophy and percent asymmetry due to nonfunctioning. To prevent these problems, a prosthesis was made available to the patient and the controls showed that the patient was satisfied with the chewing function (Figure 6). Patient and their parents were informed about future treatments.

**CONCLUSION**

Bringing many different problems along, this case resembles regional odontodysplasia, and it is considered to constitute a rare syndrome. However, in such cases in which performing a genetic test is not possible as in our case, procedures intended to correct the current oral condition should be followed.

FIGURE 6: Oral views of the patient’s prosthesis.
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Conflict of Interest
No conflicts of interest between the authors and/or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Zuhal Korzioğlu; Design: Zuhal Korzioğlu; Control/Supervision: Zuhal Korzioğlu; Data Collection and/or Processing: Zuhal Korzioğlu, Rüya Alpar; Analysis and/or Interpretation: Zuhal Korzioğlu; Literature Review: Zuhal Korzioğlu, Rüya Alpar; Writing the Article: Zuhal Korzioğlu, Rüya Alpar; Critical Review: Zuhal Korzioğlu, Rüya Alpar; References and Fundings: Zuhal Korzioğlu, Rüya Alpar; Materials: Zuhal Korzioğlu, Rüya Alpar.

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