Ectodermal Dysplasia Treated With One-Step Surgical Rehabilitation: A Case Report

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Abstract

Ectodermal dysplasia (ED) comprises a large heterogeneous group of inherited disorders that are characterized by primary defects in the skin, hair, nails, eccrine glands and teeth. The most characteristic findings are the reduced number of teeth. All rehabilitative programmes involve proper evaluation of skeletal relationships. Prosthetic-implantological treatment at the end of bony growth can be used. In this article a case of ED treated with Le Fort I for maxillary advancement, femur homografts, implants’ insertion and immediate loading is described. In December 2007, a 38-year-old female was referred to the Maxillofacial Department of Galeazzi Hospital (Milan, Italy) who had a diagnosis of ED. Twelve implants were inserted in one-step surgical procedure. No implant was lost and all are stable. The occlusion is stable after 15 months of follow-up. The results indicate that the one-step oral rehabilitation can be performed in adults who are affected by ED. Also, this significantly reduces the time of oral and facial rehabilitation. [Singapore Dent J 2010;31(1):9–14]

Key Words: bone, remodelling, resorption, ridge, alveolar, fixture

Introduction

Ectodermal dysplasia (ED) comprises a large heterogeneous group of inherited disorders that are characterized by primary defects during the development of two or more tissues that are derived from embryonic ectoderm. The tissues primarily involved are the skin, hair, nails, eccrine glands and teeth.1 This condition is classified into two major types: 1. Hypohidrotic, in which the sweat glands are absent or significantly decreased. 2. Hidrotic, in which the sweat glands are normal. Hypohidrotic ED is the more severe form and it affects males more than females. It is associated with sensitivity to heat and frequent high fevers.2 Clinical recognition varies, based on the severity of symptoms and associated complications, from birth to childhood. However, the diagnostic tool is typical clinical physiognomy.3 Men with ED have an easily recognizable face, also referred to as an ‘old man’ face. The forehead appears square, with frontal bossing, and there is a prominent supra-orbital ridge. The nose has a depressed nasal bridge and is called a ‘saddle nose’. The midface is depressed and hypoplastic, giving it a ‘dished-in’ appearance. The cheekbones are high and broad, although they appear flat and depressed as well.

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The chin may be pointed and the lips are exerted and protuberant. Some infants have a premature look with scaling of the skin. This can also form a clue to the diagnosis: the number of sweat glands is reduced and both scalp and body hairs are sparse, with lack of eyebrows and eyelashes. These remarkable variabilities in facial dimensions and harmony found in patients with ED probably correspond to different kinds of dysplasia, with different expression of the associated genes.

The most characteristic finding in man is the reduced number and abnormal shape of the teeth. The dental findings of ED range from complete anodontia to hypodontia of the primary or permanent teeth with or without cleft lip and palate. The delay in teething is often the first step in the diagnosis. However, if teeth are present, they may be tapered, malformed and/or widely spaced. Congenital absence of the teeth is more frequent in the lower jaw and affects the growth of the jawbones, leading to a lack of alveolar bone in both height and width.

Early dental treatment of patients with ED is necessary, because it affords the child the opportunity to develop normal forms of speech, chewing, swallowing and normal facial support. It also improved the temporo-mandibular joint function and self-esteem. The course of the treatment is to restore the function and the aesthetics of the teeth, normalize the vertical dimension and support the facial soft tissues. All rehabilitative programmes involve proper evaluation of skeletal relationships, which will eventually be corrected using the appropriate orthodontic techniques. The use of a total or partial removable prosthesis or overdentures is often the initial treatment of choice. However, prosthetic-implantological treatment at the end of bony growth must be implemented, with the possibility of restoring ad integrum the patient’s masticatory function and aesthetics.

In this article case of ED treated with Le Fort I for maxillary advancement, femur homografts, implants’ insertion and immediate loading is described.

Materials and Methods

A 38-year-old female was referred to the Maxillofacial Department of Galeazzi Hospital (Milan, Italy) who had a diagnosis of ED in December 2007. Informed written consent approved by the local Ethics Committee was obtained from the patient for using her data for research purpose.

The patient was neither a smoker nor a drinker, and she is a nurse. She had no previous major operation in the head or neck region.

Examination of the oral cavity showed the presence of two lateral incisors, two canines, one right premolar and one molar in the mandible (Figure 1). Faces and skin annex were typical for ED, and the clinical history was suggestive for a hidrotic ED (with no high-fever episode).

Ortopantomography and lateral teleradiography (Figures 2 and 3) were performed as along with impressions and custom model splint for maxilla reposition.

An operation was planned to insert implants in the upper and lower jaws (Figure 4): after placement of the surgical guide, a mucotomy was performed, the bone drilled and implants inserted (Neoss S.r.l., Milan, Italy). The implant platform was positioned at alveolar crest level. Then, a maxillary advancement by means a Le Fort I osteotomy (Figure 5), temporary intermaxillary fixation.
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Figure 3. Pre-surgical teleradiography.

Figure 4. Implant inserted by means a customized provisional prosthesis.

Figure 5. Le Fort I osteotomy.

(Figure 6), femur homografts (banked from living donors, Bone Bank of Orthopedic Institute Gaetano Pini, Milan, Italy) inserted in the osteotomic gaps (Figure 7), and internal rigid fixation was performed (Figure 8). The provisional restoration immediately delivered, and after 8 weeks the final restoration was done.

The immediate postoperative clinical (Figure 9) and radiological controls (Figures 10 and 11) demonstrated a successful outcome. The 15 months follow-up demonstrated the stability of the result (Figures 12–14).
Results

There were a total of 12 fixtures. Informed written consent approved by the local Ethics Committee was obtained from patient to use her data for research purposes. The mean follow-up after implant insertion is 15 months: six implants were immediately loaded and six (two upper molars and four inserted in the mandible) are still not prosthetized (because of economic problem of the patient). The mean post-loading follow-up is 7.5 months.

A total of 12 implants (Neoss S.r.l., Milan, Italy) were inserted: eight into the maxilla and four into the mandible. Two measures of fixture length and diameter 11.5 and 13 mm and 3.5 and 4.0 mm, respectively, were used. Implants were inserted to replace four incisors, two cuspids, three premolars and three molars. The gap of maxillary osteotomy was filled with femur graft, whereas no graft was placed in the mandible. Six of the eight fixtures inserted in the upper jaw were immediately loaded.

No implant was lost, and all are stable.

Discussion

Dental implants have become an accepted treatment modality for ageing patients with either completely or partially edentulous arches. However, in partially edentulous children who have ED, multiple implant placement is not possible because of the ongoing development of the jaws and insufficient bone. In addition, the bone height and width will not be sufficient for implant insertion without advanced surgical approaches. In patients not treated with ED, craniofacial deviations from the norm increased with increasing age with a tendency towards a Class III pattern with anterior growth rotation. Cephalometric analysis and anthropometry studies are able to show reduced facial dimensions, decreased lower facial height and variable pattern in facial widths. The maxilla is relatively more retruded than the mandible, and the nasal alar width and mouth width are significantly smaller. In the mandible, sufficient bone may be available only at the mid-symphysial area, where one implant could provide stability for the mandibular denture. However, the maxilla of totally edentulous patients frequently requires bone grafting procedures or Le Fort I and grafting before implant insertion because this may affect their retention and stability.

Orthognatic surgical procedures are the treatment of choice in ED patients who exhibit maxillary and/or mandibular skeletal deficiency. It has
been postulated that the Le Fort I osteotomy that restores normal maxillomandibular relationship might restimulate maxillary growth.\textsuperscript{17,18} First, the jaws must be positioned correctly in relation to each other and the cranium by means of orthognatic surgery; only then implant placement with autogenous or heterologous bone grafts can be attempted.\textsuperscript{19}

Although these surgical procedures can be performed in most ED patients, removable dentures are usually the only viable treatment alternative when the expenses and morbidity of the surgical procedures are taken into account.\textsuperscript{20}

In the present study we describe a case of ED treated with simultaneous implant placement, Le Fort I advancement, grafting of maxillary gaps and provisional immediate rehabilitation of the upper jaw. Although implants were inserted also in the mandible, these lasts are still not prosthetized due to the economic condition of the patient. The mandible is bearing removable dentures.

Conclusion

The one-step oral rehabilitation is stable in terms of occlusion and implant outcome after 15 months follow-up. This procedure can be performed in adults. It significantly reduces the time of oral and facial rehabilitation without compromise with the medium-term clinical outcome.

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Figure 12. The 15 months follow-up panoramic radiograph.

Figure 13. The 15 months follow-up teleradiography.

Figure 14. The 15 months follow-up frontal view of the occlusion.
References


