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Case Report

A Case Report of Ectodermal Dysplasia with Ankyloglossia

[1]Shivani S. Singh, [2]Monika Verma Koul, [3]Faizan Ansari, [4]Azmat Yaqoob, [5]Poonam Gupta

[1,3,4,5]PG student 3rd year, [2]Professor and Head, Pediatric and Preventive Dentistry, Career Post Graduate Institute of Dental Sciences & Hospital (CPGIDSH), Lucknow, Uttar Pradesh, India

ABSTRACT:

Ectodermal dysplasia is a hereditary disorder associated with abnormal development of embryonic ectodermally derived tissues including teeth, nails, hair and sweat glands. Hypodontia of the primary and permanent dentition is the most common oral finding. Therefore, affected patients need dental prosthetic treatments during their early years. This report presents the case of a girl child affected by ectodermal dysplasia with oligodontia and ankyloglossia. Oral rehabilitation was planned with removable acrylic prostheses while ankyloglossia was treated with laser frenectomy. Treatment results aimed at major impacts on self-esteem, masticatory function, speech and facial esthetic.

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Corresponding author: Shivani S. Singh, PG student 3rd year, Pediatric and Preventive Dentistry, Career Post Graduate Institute of Dental Sciences & Hospital (CPGIDSH), Lucknow, Uttar Pradesh, India

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INTRODUCTION

Ectodermal dysplasia (ED) is a heterogeneous group of disorders characterized by development of dystrophies affecting tissues developed from ectodermal origin such as teeth, hair, nails, skin and sweatglandsin varying degrees.[1] The term ectodermal dysplasia was coined by Weechin 1929 but was first used during a medical lecture by Thurmanin 1848. Usually ectodermaldysplasia is caused due to mutation in gene or is inherited in an X-linked recessive pattern disorder. It can also be seen in people without any history in which case new mutations occurs. The incidence of ectodermal dysplasia is around 1-7 cases per 10,000 live births. [2]The unaffected female has 50% chance of transmitting this disorder to her male children and each female offspring has 50% chance of inheriting the defective gene, thereby being a carrier.[3]

Ectodermal dysplasia can be classified according to presence or absence of four primary structures. (Vanessa Ngan, 2005.) [4]

- ED1:Trichodysplasia(hair dysplasia)
- ED2:Dentaldysplasia
- ED3:Onychodysplasia(nail dysplasia)
- ED4:Dyshidrosis(sweat gland dysplasia)

Based on the above, the 150 different types of ectodermal dysplasia's are categorized into one of the following subgroups made up from the primary ED defects:

- Subgroup 1-2-3-4
- Subgroup 1-2-3
- Subgroup 1-2-4
- Subgroup 1-2
- Subgroup 1-3
- Subgroup 1-4
- Subgroup 2-3-4
- Subgroup 2-3
- Subgroup 2-4
- Subgroup 3
- Subgroup 4

The most common ectodermal dysplasia's are hypohidrotic (anhidrotic) ED which falls under subgroup 1-2-3-4 and hydrotic ED which comes under subgroup 1-2-3.

The clinical signs of ectodermal dysplasia includes trichodysplasia (abnormal hair) in 91% cases tooth agenesis in 80% cases onychodysplasia in 75% cases and dyshidrosis in 42% cases. [1] The teeth in ectoodarmal dysplasia patient are markedly less number (oligodontia/hypodontia) and may show abnormality in size and shape of teeth (peg shaped incisor).[5] Although there are phenotypic changes seen in ectodermal dysplasia patient they have normal life span and intelligence.[1]

CASE REPORT

A girl child aged 14 years reported to the department of pediatric and preventive dentistry of CPGIDSH, Lucknow, with the chief complaint of missing lower front teeth since the milk teeth were lost. The patient had syndromic appearance and short strature. On history taking the patient presented no relevant medical or dental history and denied any family history of similar kind or presence of consanguineous marriage. She had normal perspiration.

On extra oral examination there was presence of increased intercanthal width, frontal bossing, flat nasal bridge, mandibular deficiency, maxillary protrusion and sparse eyebrows [Figure 1]. On intraoral examination the mandibular arch showed absence of all six anterior teeth that is both central and lateral incisors and canines root stumps of left first molar, occlusal caries in right andleft second molar, distoproximal caries in right second premolar [Figure 2b]. The lower lip was everted [Figure 1] and there was presence of ankyloglossia [Figure 4 a,b] resulting in unclear speech. The maxillary arch showed the presence of proclinedanterior teeth, high arched palate, lingual pit caries in right and left lateral incisors. [Figure 2a] Reduced vertical height of face was evident. Patient was advised an orthopantomogram, which on observation showed absence of mandibular anterior teeth, radiolucency approaching pulp with respect to mandibular right second premolar, first and second molar with mesially angulated root of mandibular right first molar; grosslycarious teethwith onlyroots stumpspresentwith mandibular leftfirstmolar showingfusion [Figure 3]. Thus, based on history, clinical and radiographic findings, the present case was diagnosed as ectodermal dysplasia (ED 2 Subtype 1-2). The treatment planned was extraction of root stumps with mandibular left first molar followed by removable partial denture for missing lower anteriors and lower left first molar, root canal treatment in mandibular right second premolar, first and second molar followed by porcelain fused to metal (PFM) bridge, composite restoration of carious teeth and lingual frenectomy using soft tissue diode laser [Figure 4 c,d,e].

Figure1: Extra oral photographs showing increased intercanthal width, frontal bossing, flat nasal bridge, mandibular deficiency, maxillary protrusion, everted lower lip and sparse eyebrows

Figure 2: Intra-oral photographs a) maxillary arch showing high palatal vault, lingual pit caries in 12,21 b) mandibular arch showing partial anodontia in anterior region,ankyloglossia, carious 45, 46, 47; 37 and root stump wrt. 36, c) frontal view presenting reduced height of alveolar ridge.

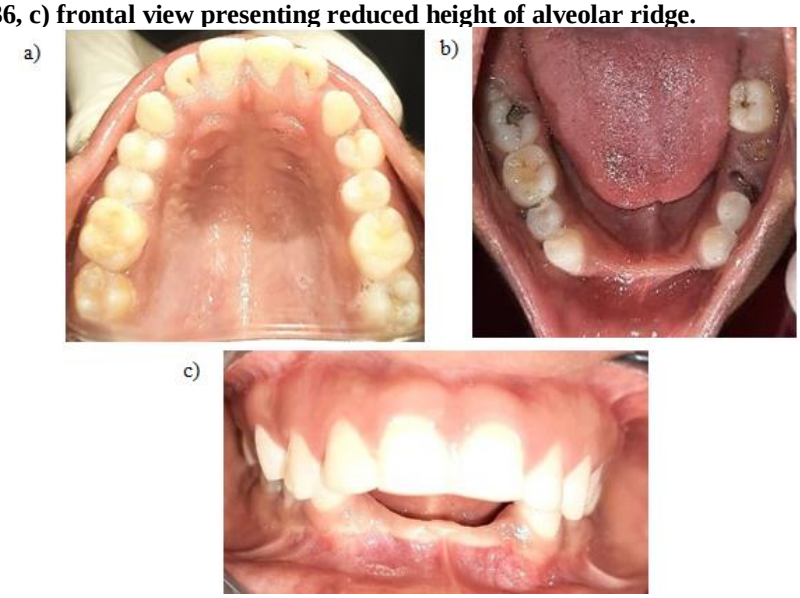


Figure 3: Orthopantomogram, showing congeniitally missingmandibular anterior teeth, carious mandibular right first molar, second molar, second premolar and root stump of mandibular left first molar.

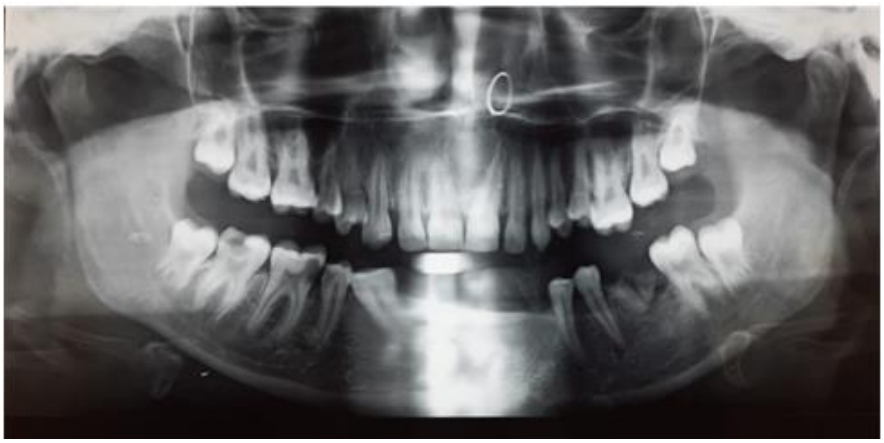
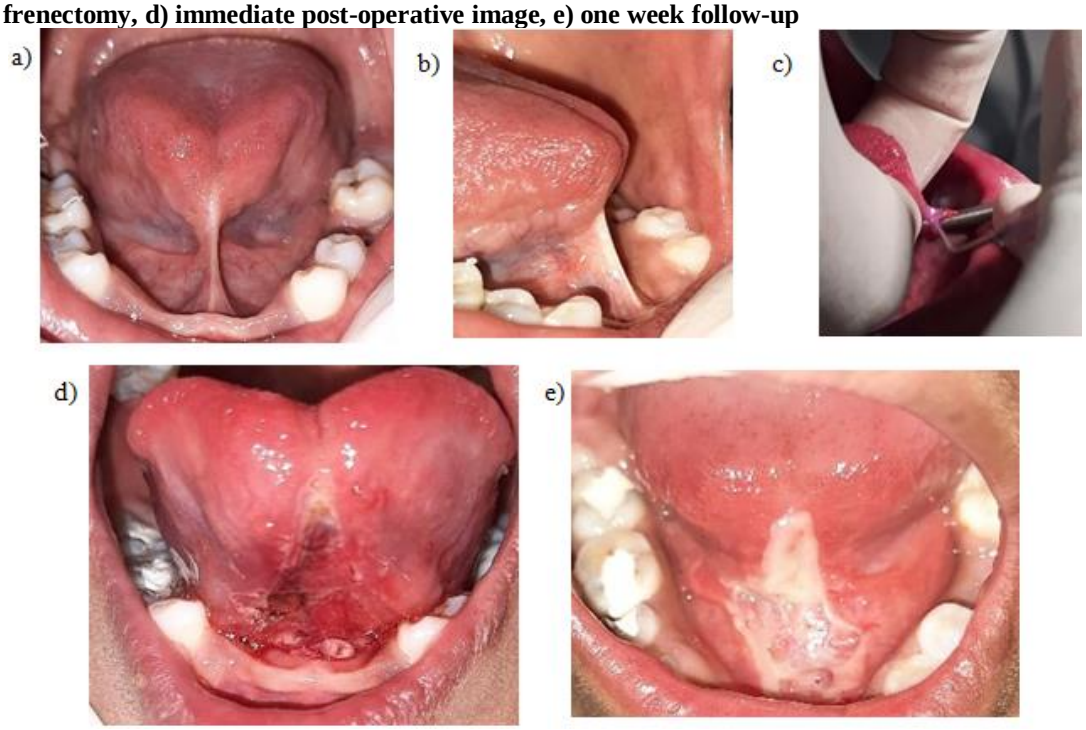


Figure4: a) Front a land b) lateral view of tongue showing high frenum attachment (ankyloglossia) c) laser frenectomy, d) immediate post-operative image, e) one week follow-up



DISCUSSION

Ectodermal Dysplasia is a genetic developmental disorder and chromosome 19 within the region of D19S894 and D19S416 has been postulated as the locus for the abnormalities observed in this syndrome.Nearly 90% cases are caused by mutations of TP63 gene which leads to a reduction of functional levels of normally functioning P63 protein which interferes the proper development of structures derived from ectoderm.[6] The lack of tooth bud formation causes hypoplastic alveolar bone, leading to a reduced vertical dimension of occlusion. Therefore, an old-age appearance is common in affected individuals. To elevate the facial appearance and increase the confidence of children with ectodermal dysplasia rehabilitation of the missing dentition is of utmost importance. Teeth in early ages not only aid in mastication and nutritional gain but also improves the sagittaland vertical skeletal relation, esthetics and speech.[5] In young children using removable prosthesis forreplacementof anodontiais thetreatmentof choicealthough implants can bean alternative. The insertion of dental implants in children or adolescents before completion of craniofacial growth is related to several problems.[7] The mandibular anterior region is most suitable for implants in children owing to least anatomic changes due to growth but thorough evaluation of timing, physical and psychological factors is necessary. Removable partial or complete dentures require regular adjustments and should be replaced when a decreased vertical dimension of occlusion and an

abnormal mandibular posture are detected due to growth. Retention and stability for the prosthesis can pose a problem. When fabricating dentures for these patients, care shouldbeen taken to obtain a wider distribution of occlusal loads by extending the denture base as much as possible. The occlusion of removable partial denture should be in harmony with the patient's occlusion since, oligodontia or anodontia leads to atrophy of the alveolar ridges, reduced vertical dimension and prominent chin. This treatment modality improves the patient's quality of life and it can be regarded as an acceptable treatment modality for functional and esthetic rehabilitation.

Ankyloglossiaor tongue-tie results fromashort, tight, lingual frenulum causing difficulty in speech articulation due to limitation in tongue movement.[8] In the present case lingual frenectomy was performed using a diode Laser after which an improvement was seen in the speech. Lasers are advantageous andhave predictable results overtraditionalscalpelorbladetechnique duetoreduced operating time, minimal amounts of local anesthesia, enhanced haemostasis, no need for sutures and less prescription of post-operative drugs; therefore, this faster treatment option may decreasethe psychological distress of pediatric patients associated with conventional surgical procedures.[9]

CONCLUSION

Patients with ectodermal dysplasia present with various restorative issues.Acomprehensive dental care with multidisciplinary approach should be rendered. Treatment plan depends on patient's needs, wishes and the willingness to undergo treatment and also on the economic possibilities. In young children prosthetic rehabilitation using removable or fixed prosthesis which are cost-effective should be considered and can be the key in their healthy growth both physically and psychologically. Long-term success of such cases depends on regular recall appointments and meticulous maintenance of oral and prosthetic hygiene.

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