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A Palliative Prosthetic Rehabilitation for Improving Quality of Life in Ectodermal Dysplasia Patient: A Case Report

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Abstract: ***Ectodermal dysplasia is a rare inherited disease that affects several ectodermally derived anatomical structures characterized by hypodontia, hypotrichosis and anhidrosis. Early dental intervention can enhance patient's appearance, thereby minimizing the associated emotional and psychological issues in these patients. This clinical report demonstrates prosthetic rehabilitation consisting of lower overdenture to help in psychosocial development and to restore the vertical dimension, esthetics and functioning of the stomatognathic system.***

Keywords: Ectodermal dysplasia, Hypodontia, Hypohidrotic, Tooth-supported Overdenture, Prosthodontic rehabilitation

1. Introduction

Ectodermal dysplasia is an inherited disorder that results in aberrant growth of ectoderm-derived tissues, including nails, teeth, hair and skin as well as occasionally mesoderm-derived tissues.[1] The condition has over 150 documented variations, with hidrotic and hypohidrotic being the two most prevalent types.[2]The key features of the face include a prominent forehead, pronounced lips and a flattened nasal bridge. In addition, convex nails, thin, wrinkled and hyperpigmented periorbital & perioral skin, as well as persistent skin conditions like eczema may be seen.[3]

The oral manifestations include peg-shaped or conical teeth, hypodontia or anodontia, malformation of any teeth that may be present, generalised spacing, underdeveloped alveolar ridges and delayed eruption of permanent teeth. Even when there isanodontia, jaw growth is unaffected which means that except for the alveolar process, the development of the jaws is not dependent on the existence of teeth. Sometimes, the salivary glands, especially the intraoral accessory glands are hypoplasticresuting in Xerostomia.[4]

Ectodermal dysplasia (ED) patients require prosthodontic rehabilitation for a various functional, physiological and emotional reasons.[5]Reduced VDO (Vertical dimension of occlusion)and deficient lip support contributes to the senile appearance of these patients which can be corrected with the prostheses. Most ED patients have permanent teeth, which are frequently present in the incisor and first molar regions which can be used as an abutment for the prosthesis instead of extraction.The final decision to retain a tooth is influenced by its root and crown anatomy, location inside the arch and prosthetic treatment.[6]

2. Case Report

A 16-year-old female patient was referred to the Department of Prosthodontics, KVG Dental College and Hospital with a complaint of unpleasant appearance and inability to eat because of missing lower teeth for 8 years. Patient was diagnosed with Hypohidrotic (anhidrotic) Ectodermal dysplasia past 7 years ago. Family history of Ectodermal dysplasia was negative. Clinical examination revealed that patient was moderately built with hypotrichosis, sparse eyebrows and eyelashes, dry anhidrotic skin, depressed nasal bridge, thick and everted lips, dark pigmented skin around periorbital area and nose along with decreased vertical dimension of face.[Figure 1] Intraoral examination revealed oligodontia with a total of 9 teeth present. Maxillary arch consisted of four peg shaped anterior teeth (11, 13, 21, 23) & malformed posterior teeth (16, 17, 18, 26). Mandibular arch consisted of 33 with severely atrophic mandibular edentulous ridge.[Figure 2] The panoramic radiographic examination revealed the presence of impacted two teeth in the fourth quadrant. [Figure 3]

Figure 1: Extraoral Frontal, right and left lateral profile

Figure 2: Intraoral maxillary and mandibular view

A fixed treatment option was not advised as patient was still in the developmental stage hence a palliative treatment option was preferred. In order to improve the appearance, mastication, speech and quality of life - an overdenture, a functional prosthesis with adequate retention, stability and support was planned for the mandibular arch and removable partial denture with respect to maxillary arch.Patient denied for the maxillary denture due to the palatal coverage which was inconvenient. Other objective of treatment regarding the dental condition was the preservation of bone, the establishment of normal facial characteristics and smile.

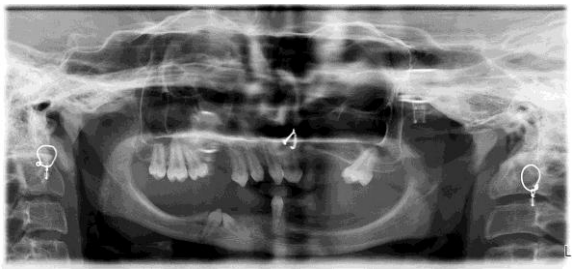

Figure 3: Panoramic radiograph showing impacted teeth in fourth quadrant

2.1 Prosthodontic Management

Informed consent was obtained and treatment procedure was explained to the parents before the procedure. A preliminary maxillary and mandibular impression were made using irreversible hydrocolloid impression material and poured using Type III dental stone. A tentative jaw relation was recorded to evaluate vertical dimension loss and analysis of the available inter-arch space.[Figure4]

Figure 4: Tentative jaw relation

The lower left canine was indicated for the endodontic therapy. After the endodontic therapy the tooth was prepared equigingivallyto receive a dome shaped short coping so as to flush with the remaining residual ridge. Chamfer finish line was made. An anti-rotational groove was given on the mesio-distal wall of post space to prevent the dislodgement of the coping and the canal was prepared with Peesoreamers to the appropriate length.[Figure 5A] An indirect method of coping fabrication was performed; a 22 gauge orthodontic wire with retentive tag at one end was used to make the canal impression with one stage putty wash impression (squash) technique. [Figure 5B]

Figure 5A: Tooth and post space preparation

Figure 5B: Impression of post space and abutment

The metal coping was fabricated and checked for the fit, marginal adaptation and cemented using Type I GIC. A custom tray with uniform 2 mm wax spacer was fabricated using autopolymerizingacrylic resin. Peripheral border seal was recorded using Type 1 low fusing Green stick compound and secondary impression was made with light body impression with the use of tray adhesive. [Figure 6A] Beading and boxing of the impression was done and poured using Type III dental stone and master cast was obtained.[Figure 6B]

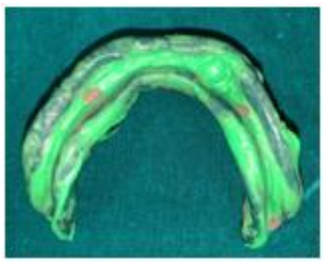

Figure 6A: Secondary Impression

Figure 6B: Master Cast

Denture base was fabricated using auto polymerizing resin and occlusal rim was made. The jaw relation was recorded by manually guiding the mandible to centric relation. [Figure 7] Articulation and teeth arrangement was done on a mean value articulator. The try-in was done and evaluated for the occlusion, phonetics and estheticsthen prosthesis was fabricated using heat cured acrylic resin and delivered. [Figure 8]

Figure 7: Jaw relation recorded

Figure 8: Final Insertion

The patient was instructed on the maintenance of oral hygiene and denture. The patient was followed up for 6 months and was satisfied with the treatment outcome as there was increased masticatory efficiency and speech.

3. Discussion

Ectodermal dysplasia is a group of genetic disorders involving an absence or lack of tissues and structures derived from the embryonic ectoderm. The signs and symptoms include trichodysplasia (abnormal hair) in 91%, tooth agenesis in 80%, onychodysplasia (abnormal nails) in 75% and dyshidrosis (abnormal sweat glands) in 42% of cases.[7]It is diagnosed intraorally by one of its main features that is hypodontia/anodontia.[6] In patients with ectodermal dysplasia, the sagittal and vertical relationship of the somatognathic system are significantly compromised.[8]Patients typically have a loss of vertical relationship, a prominent chin and a class III inter-maxillary relationship.[9] Prosthetic management of ectodermal dysplasia patients requires a challenging approach because of rarity of suchsyndromes which is estimated at incidence of 1 in 10,000 and 1 in 100,000 births.[10]

The prosthetic treatment varies and typically depends on the patient's age, dental agenesis, degree of tooth malformation, growth and development of the stomatognathic system and patient's motivation.[1] The literature describes a wide range of prosthetic treatment options for individuals with ectodermal dysplasia including a removable or fixed partial denture, an overdenture prosthesis (when the vertical dimension of occlusion permits) and an implant retained prosthesis. For the best outcome, these modalities of treatment can be applied individually or in combination.[7]

The lack of fully developed alveolar ridges, xerostomia and under-developed maxillary tuberosity contribute to poor support, retention and stability in conventional complete denture. Anterior conical teeth compromise the stability of a removable partial denture.[9] Fixed prosthodontic management is rarely employed because of limited number of teeth present and often patient are young in which fixed partial denture with rigid connector will impede active growth of dental arches.[4] In this case, the extraction of the conical tooth in the lower arch for the subsequent placement of the prosthetic appliance would reduce the alveolar ridge of the patient. Therefore, maintaining this tooth would promote a larger surface area for distributing masticatory forces and greater prosthesis retention.[7] Thus, it was appropriate to be used as an abutment for overdenture.

Osseo-integrated implants is one of the definitive treatment option in patients with ectodermal dysplasia but adequate bone density is crucial for the effective positioning of implants, which is mainly lacking in these patients.[8] Guckes, Mccarthy, and Brahmin[11] asserted that there is a substantial probability of failure when implants are implanted in ED patients who are under the age of 18. Kearns and Sharma [12] described implant submergence in two young patients (5 and 7 years old), who ultimately needed longer abutments and prosthesis revisions to account for the growth-related change in implant placements. In this case report, implant placement was not considered as treatment of choice because of atrophic under-developed ridges, developing age and economic constraints.

Overdenture was a treatment of choice in this specific case as anterior atypical conical teeth served as an abutment to compensate the drawbacks related to retention, stability and support in the case of conventional removable partial or complete denture.

4. Conclusion

The primary objective of a prosthodontic procedure is to satisfy the young patient's immediate needs. Periodic recall of the young patients with ectodermal dysplasia is also important because prosthetic modification with relining, rebasing or replacement will be needed to accommodate continuous growth and development. The patient's prosthesis restored her vertical dimension, enhanced her physical appearance, speech, and masticatory function, which had a positive impact on her psychological well-being, self-confidence and dietary nutrition.

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