CASE REPORT

Prosthodontic Management of a Patient with Ectodermal Dysplasia— A Clinical Report

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ABSTRACT

Ectodermal Dysplasia is a congenital, diffuse and non-progressive disorder characterized by hypoplasia or aplasia of structures derived from the ectoderm germ layer. Ectodermal dysplasia has three different inheriting patterns with X-linked inheritance being the most common. It is classified into Hypohydrotic and hydrotic based on the condition of sweat glands. Clinical manifestations of this disorder include hypodontia, anodontia, soft and thin enamel, delayed eruption, xerostomia, alveolar ridge atrophy, flat nasal bridge, decreased or no sweat glands, frontal bossing, sunken cheeks, loss of hair, and thin and abnormally shaped nails. Globally, the cases of ectodermal dysplasia are rare, having a prevalence of 1:100,000 with only a few documented and reported in Pakistan. This report presents a female patient, aged 16 years, who came to the OPD with the main complaint of missing teeth. Provisional diagnosis of Ectodermal Dysplasia was made based on examination and history. It aims to contribute to the improvement of treatment planning, promoting documentation and early diagnosis in Pakistan.

Keywords: Complete denture, congenital, ectodermal dysplasia, ectoderm germ layer, hypodontia, prosthetic rehabilitation

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INTRODUCTION

Ectodermal Dysplasia, a genetic disorder, affects structures derived from the ectoderm germ layer. It is characterized by hypoplasia, aplasia and dystrophy of ectodermally derived structures including hair, nails, teeth, salivary, lacrimal, and sweat gland[1]. It is a congenital, diffuse, and non-progressive disorder[2]. Ectodermal Dysplasia has three different inheriting patterns, X-linked, autosomal recessive, and autosomal dominant[3]. The most common inheriting pattern is X-linked therefore, male predominance is seen.

Based on the state of sweat glands, Ectodermal Dysplasia can be classified into Hypohydrotic and Hydrotic. The Hydrotic type has normal sweat glands and is more prevalent. Hypohydrotic or Anhydrotic type has either no sweat glands or reduced in number. Oral manifestations include delayed teeth eruption, conical-shaped teeth, alveolar ridge atrophy, protuberant lips, high palatal arch, etc. These cases are rare with an incidence of 1:100,000[4].

Literature review, done through pubmed and google scholar, found that very few cases of ectodermal dysplasia have been recorded in Pakistan. This might be due to lack of awareness and diagnostic tools. Promoting the documentation of such cases can improve patient care and may lead to early diagnosis, enhancing the treatment plan. Prosthetic rehabilitation is the most commonly considered rehabilitation option for such patients. It includes complete dentures, removable dentures, implant-supported prostheses, and other tooth-supported dentures. Fixed prosthetic treatment options are usually considered but in our case, patient did not have sufficient bone to support an implant and was unwilling to go for procedures like bone grafting. This case report features prosthetic rehabilitation of a patient with ectodermal dysplasia by overdentures.

CLINICAL REPORT

A 16-years old female patient reported to the department of Prosthodontics at the Dr Ishrat-ul-Ebad Khan Institute of Oral Health Sciences (DIKIOHS), DUHS with the

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Fig. 1

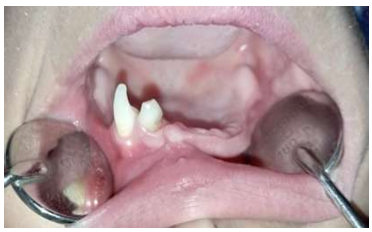
Fig. 2


Fig. 3

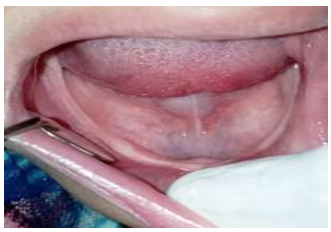
Fig. 4

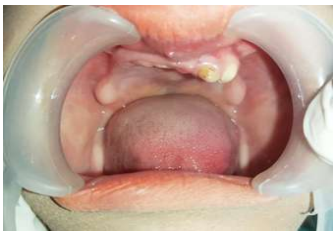
Fig. 5


Fig. 6


Fig. 7


Fig. 8

chief complaint of difficulty in chewing. Extra-oral examination revealed mandibular prognathism and class III skeletal profile along with thin upper lip, depressed cheeks and increased lower face height (Figures 1, 2, and 4).

Intra-oral examination revealed one malformed upper right lateral incisor and a conical shaped upper right canine in the second quadrant with thin mucosa and knife edged residual ridge in maxillary and mandibular arches. General examination revealed lean body structure, low body weight, thin and sparse hair on head and eyebrows.

Considering these findings, a diagnosis of Ectodermal Dysplasia was made. Medical and family history were non-contributory.

Radiographic examination revealed presence of only two teeth (an upper right lateral incisor and a canine) in the second quadrant (Figure 3). No impacted teeth or no other anomalies were found. Treatment options included teeth supported complete overdenture and removable partial denture. Since the alveolar bone was highly resorbed, placement of implants was not possible and the only two malformed teeth were also not suitable as abutments for a removable partial denture. Considering all the factors, a decision was made to provide tooth retained overdentures to the patient. Root canal treatment was performed on the teeth and they were shaped accordingly to be used as abutments for overdentures (Figure 5).

After abutment preparation, impressions were made for fabrication of complete dentures. Primary impression was made by using tissue friendly material (Irreversible hydrocolloid). Secondary impression was recorded using green stick impression compound and impression paste. Jaw registration was done and records were taken to restore lower facial height. Teeth setup was done using semi-anatomic teeth with bilateral balanced occlusion. Teeth were set in cross bite to compensate for arch discrepancy. After processing in the dental lab, complete overdentures were made using heat cure acrylic. Final dentures were inserted and adjustments were made according to the patient (Figure 6, 7 and 8). Since the patient had never had a denture before, she was explained about all the hygiene measure, maintaining periodontal health and denture cleaning protocols. Follow up visit for fluoride applications were also planned.

DISCUSSION

Ectodermal Dysplasia is a congenital disorder that affects ectodermally derived tissues during embryological phases of development. It has different inheriting patterns with X-linked being the most common, therefore it is more common in men with females being carriers. There are two types of ectodermal dysplasia: Hypohydrotic and Hydrotic. Hypohydrotic ectodermal dysplasia (HED) is also known as Chris-Siemen-Tauriane Syndrome[5]. HED is characterized by a triad of symptoms hypotrichosis (sparse hair), hypodontia or anodontia, and inability to sweat or decreased sweating with an X linked inheritance pattern. However, the other type i.e. the hydrotic type is inherited as autosomal dominant (Clouston's syndrome)[2,6].

A report by Chandravanshi stated that mutations in EDA, DARADD, and EDAR genes are responsible for causing HED. These genes mainly code the protein which is responsible for the interaction between the ectoderm and the mesoderm germ layer[5]. Another case

report on Ectodermal Dysplasia states that although the cause of it is still unknown, it is assumed that it may manifest as mutations in thrombospondin-type laminin G domain and epilepsy-associated repeats (TSPEAR) gene affecting hair and tooth development. However, further evaluation is needed on this[7].

Clinical manifestations include flat nasal bridge, everted lips, decreased or no sweat glands, generalized dryness and rash on the skin, frontal bossing, sunken cheeks, low set ears, loss of hair, sparse hair, thin and abnormally shaped nails, and other symptoms may vary depending on the patient's conditions. On the other hand, oral manifestations include hypodontia, anodontia, peg shaped teeth, soft, thin or pitted enamel, delayed eruption, dry oral mucosa, xerostomia which may lead to inadequate oral hygiene conditions, alveolar ridge atrophy, class III malocclusion, upward and forward displacement of cheeks[8,9]. Hypodontia is the most common manifestation in patients with ectodermal dysplasia[3].

Early diagnosis and oral rehabilitation are essential. It helps to reduce unwanted effects caused by the oral manifestations of ectodermal dysplasia. The main goal of the rehabilitation plan is to improve phonetics, aesthetics, occlusion, and mastication, and it also helps patients with self-confidence, while other conditions can be handled with palliative treatment. Xerostomia, which is a feature of hypohydrotic ED, and decreased lacrimation can be relieved by artificial saliva and tears, respectively[10]. The most common treatment considerations are removable partial dentures, implant-supported dentures, complete dentures, bridges and other prosthetic options depending on the manifestations that the patient has.

According to a report, the most acceptable treatment option available for children with hypodontia/anodontia is a removable complete or partial denture[11]. According to another report, the recommended age for early prosthetic treatment is from 5 years, while implant-supported dentures are recommended at the ages of 12 to 15 years keeping in consideration the degree of alveolar bone atrophy[12]. In such patients, denture fabrication must be done to obtain a proper distribution of occlusal loads. In the case of retentive support, the anterior conical teeth might be of little use hence they can be considered as abutments in case of overdentures[4]. Al Nuaimi R et al mentioned that overdentures have also proved to be a great option for rehabilitation purpose but the only drawback is it requires aggressive and complicated procedure for tooth preparation[11]. Removable partial dentures also have some limitations. They are highly considerable but they are short-term rehabilitation options. Severe alveolar ridge atrophy and plaque deposition on teeth might also be the limiting factors[13].

In this case study, we used the treatment options most financially suitable to the patient. The patient opted for tooth supported complete overdenture. The implants could not be placed because of reduced alveolar bone and the unsuitability of the two existing malformed teeth to act as abutments for removable partial denture. To facilitate this, elective root canal treatment was performed on the remaining teeth, and they were shaped to serve as stable abutments for overdenture. This comprehensive plan aimed to address the patient's specific conditions and provide a functional and supportive solution.

CONCLUSION

This report presents a unique case of oligodontia. The patient presented with the complaint of missing teeth in the oral cavity with only a lateral incisor and canine in the second quadrant. Diagnosis of ectodermal dysplasia was made. The treatment plan included teeth retained complete overdentures and lateral incisors and canines were used as abutments.

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