ECTODERMAL DYSPLASIA: A CASE REPORT

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ABSTRACT

Ectodermal dysplasia (ED) is an uncommon genetic disorder that results from abnormalities in the development of tissues, organs, and other structures that originate from the embryonic ectoderm. This condition is rare, occurring in approximately 1 out of every 100,000 births. In this condition anodontia or hypodontia may be associated with other ectodermal abnormalities, such as anhidrosis, hypotrichosis, asteatosis, and defects in salivary glands. Here, we present one such case of ectodermal dysplasia in 13-year-old patient, in which removable partial dentures was inserted for oral rehabilitation. Providing a comprehensive medical care to patients with ED requires a multidisciplinary approach.

KEYWORDS: Hypodontia, Hypohidrosis, Hypohydrotic ectodermal dysplasia (HED), Partial anodontia, multidisciplinary approach, case report.

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INTRODUCTION:

Ectodermal dysplasia (ED) is a rare inherited disorder that is congenital and non-progressive. It is a developmental defect affecting two or more tissues that originate from the ectoderm. The primary tissues affected include the skin, hair, nails, sweat glands, and teeth.3,4 Ectodermal dysplasia is rare, with an incidence of approximately 1 in 100,000 births.1,2 The most prevalent types of ED are hypohidrotic and hidrotic ED.5 Ectodermal dysplasia is associated with various defects, including hypohidrosis, abnormal teeth development, onychodysplasia, and hypotrichosis. The facial appearance is distinctive, featuring prominent forehead (frontal bossing), , a saddle-shaped nose, sunken cheeks and darkened skin around the eyes. Dental features can include conical or peg-shaped teeth, missing teeth (hypodontia or anodontia), and delayed eruption of permanent teeth.6

CASE REPORT

A 13-year-old girl presented to the Department of pediatric dentistry and dental public health at Alexandria university, Egypt. She was accompanied by her father, with the chief complaint of multiple missing teeth in relation to the upper and lower arches and she wants to restore the empty space of teeth to fix the ugly appearance of her smile. There was no history of birth complication during her delivery , and no other live family member presented similar condition. Her father gave history of fine hair growth, dryness of skin, decreased sweating. Her father also gave a history of frequent bouts of fever throughout childhood. It was her first time to seek dental care.On extraoral examination, she had frontal bossing, a prominent supraorbital ridge, sunken cheeks, periorbital skin pigmentation, sparse hair, scanty eyebrows, and dry skin [Figure 1(a-b)].Intraoral examination disclosed that the teeth present include permanent maxillary central incisors, permanent, maxillary first premolars, and primary dentition present include maxillary and mandibular canines, mandibular anterior ,mandibular left and right first primary molar, multiple staining , dense indente of maxillary centrals, deep bite,attrition of lower anteriors ,mobile lower centrals, diastema as well as high labial frenum attatchment was also recorded [Figure 2]. OPG was taken and it revealed multiple missing tooth buds with unerupted tooth including maxillary left canine and mandibular right and left first premolar [Figure 3]. On the basis of history, clinical and radiographic examination, ED was diagnosed.

a

b

Figure 1: extraoral examination of the patient

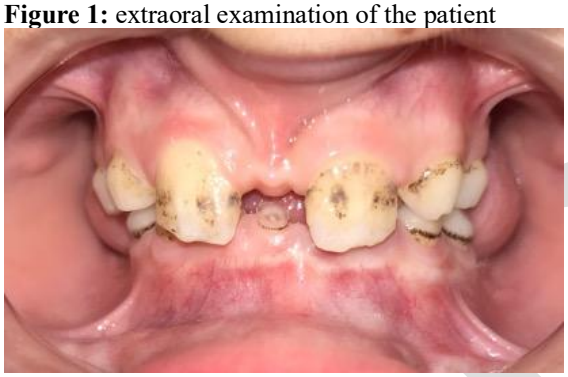

Figure 2: Intraoral photograph

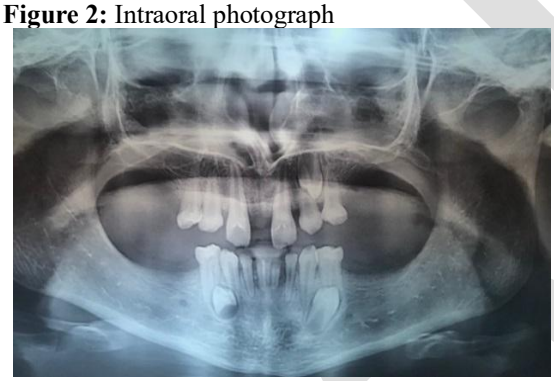

Figure 3: Panoramic radiograph showing multiple missing tooth buds with unerupted tooth buds including including maxillary left canine and mandibular right and left first premolar

The treatment plan proceeded through the regular sequence of consultation (genetic and prosthodontic), preventive, restorative, surgical, rehabilitation and maintenance phases accompanied by basic behavior management techniques .The treatment included oral hygiene instructions(OHI), dietary analysis and counseling, prophylaxis, topical fluoride varnish application, scaling and polishing. Sealing with flowable composite was done to maxillary central incisors, #72, #82 had composite build up restoration.#84 had class I composite restoration. The mobile #71, #81 was extracted.acrylic RPD was inserted Figure 4

Figure 4: Removable partial denture inserted

Treatment options includes; Removable partial denture that has to be frequently relined, rebased or remade to accommodate growth changes, Fixed partial denture, Orthodontic intervention, Complete denture prosthesis (overlaying affected teeth), Implant retained prosthesis (Two implants can be placed in the mandible, intercanine region, Mini implants supported prosthesis.9,10

In this case the adopted treatment modality was removable partial denture to improve the mastication and aesthetics and to put into consideration to raise the bite slightly to avoid the attrition and preserve the teeth available. Since lower ridge tends to be sharp and shallow, which may lead to potential discomfort for the patient, upon planning we chose a flexible RPD, but later we preferred acrylic RPD with metallic clasps, since the remaining teeth are few and the clasp being placed on primary teeth so the rigid clasp of flexible would render the teeth to be at a risk of weakening and mobility so we preferred the resilient metallic clasp of acrylic RPD . the patient was recalled after 1 week from insertion to check any tissue impingement and her satisfaction from the intervention, she was happy and compliant at wearing it and there was no tissue impingement. Continuous follow-ups every six months were

planned for adjustment or replacement of old denture. Long term treatment plan was planned to provide anticipatory guidance, check the removable partial denture, check for eruption of the present tooth buds, relief of labial frenum and closure of diastema. Besides, Patient was advised for dental implants or fixed prothesis in future, after the completion of dentofacial growth.

DISCUSSION

Dental management of a patient with ectodermal dysplasia involves a multidisciplinary team approach that includes expertise in growth, development, and behavior management.7 The focus is primarily on preventing dental disease, with secondary emphasis on oral rehabilitation. This approach is crucial for enhancing the sagittal and vertical skeletal relationships during natural growth and development, and for improving aesthetics, speech, and chewing efficiency. So, the treatment set up for a child with ectodermal dysplasia should embrace a prosthodontics side and a psychological side.8 Additional care should be taken to ensure that the prosthetic and orthodontic appliances used do not interfere with the growth patterns of the maxilla and mandible.

Orthodontic and dentofacial orthopedic treatment has been used in those patients, fundamentally, to facilitate subsequent prosthetic rehabilitation to restore the necessary function and aesthetics.The most frequent orthodontic treatments were the closure of diastemas, the alignment or maintenance of spaces and the traction of impacted teeth. All these measures aim to assist with the placement of implants and/or the subsequent prosthetic treatment.15

Though dentures are poor alternatives to healthy dentition, they produce conditions for maintenance of a normal, satisfactory daily diet. Also, within the absence of occlusal stops (or dentures), the associate rotation of the lower jaw causes an upward and forward displacement of the chin, with a reduction within the height of the lower-third of the face. Dentures facilitate positioning of the chin in situ.10 Thus, 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 during the dentofacial growth phase .

While removable complete or partial dentures remain a common treatment option in many cases, fixed partial dentures (FPDs) and implant-supported prostheses should be considered when feasible.12

Implant placement in a growing child may cause cosmetic problems because implants act similar to ankylosed teeth. As the craniofacial growth continues, the implant may not align properly with the opposite teeth and even the adjacent teeth may tilt into the space. Thus implant supported prosthesis may be less favorable and cannot be adopted in a growing child.13,14

So, removable partial denture was our choice in this case. The success of this treatment modality will depends on regular recall appointments and meticulous maintenance of oral and prosthetic hygiene.11 in addition, our patient was advised to maintain hydration and temperature regulation by frequent intake of water and fluids and wearing light clothing.The patient was also advised to undergo genetic counseling to determine the underlying molecular basis of the disease. This presentation of case is done due to its infrequency .

CONCLUSION

The clinical manifestations of ED lead to a significant social challenges for those affected. A multidisciplinary approach is essential for treating these patients, involving a team that includes pediatricians or physicians for comprehensive medical care, as well as a prosthodontist, pedodontist, and orthodontist to assist with normal functions, aesthetics, and psychosocial well-being. Additionally, consultations with a child psychologist and dermatologist should be considered

Declaration of patient consent

These authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published,but anonymity cannot be guaranteed.

Conflicts of interest

There are no conflict of interest.

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