CASE REPORT

Ectodermal dysplasia and partial anodontia: A case report

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Abstract

Ectodermal dysplasia is a hereditary disorder that occurs as a consequence of disturbances in the ectoderm of the developing embryo. It is usually accompanied by lack of sweat glands and a partial or complete absence of primary and/or permanent dentition. A case report illustrating the prosthetic rehabilitation of a young woman with anhidrotic ectodermal dysplasia associated with partial anodontia and mid facial defect are presented. Since the oral rehabilitation of these cases is often difficult; treatment should be administered by a multidisciplinary team involving orthodontics, prosthodontics and oral-maxillofacial surgery.

Keywords: Ectodermal dysplasia; hereditary disorder; anodontia

Introduction

Ectodermal dysplasia syndrome (EDS) is a large, heterogeneous group of inherited disorders, the manifestations of which could be seen in more than one derivative. These tissues primarily are the skin, hair, eccrine glands, and teeth [1-3]. Ectodermal dysplasia represents a large and complex group of diseases comprising more than 170 different clinical conditions. The incidence of this condition is 1:100,000 when at least 2 types of abnormal ectodermal features occur, such as malformed teeth and extremely sparse hair, the patient is diagnosed with ectodermal dysplasia syndrome [4-8].

The current classification of EDS is based on clinical features. Most commonly EDS is of two types i.e. Hypohidrotic (anhidrotic) ED (christ-siemens-touraine syndrome) and Hidrotic ED (clouston syndrome).

Clinical features

Generally includes reduction of hair follicles varying from sparse scalp hair to complete absence of hair. Eccrine glands may be absent or rudimentary. Mouth
may be dry from hypoplasia of salivary glands, lacrimal glands may be deficient. Nails are often brittle and thin. Oral traits of ectodermal dysplasia (ED) may be expressed as anodontia or hypodontia, with or without a cleft lip and palate. Anodontia also manifests itself by a lack of alveolar ridge development; 7 as a result, the vertical dimension of the lower face is reduced, the vermilion border disappears, existing teeth are malformed, the oral mucosa becomes dry, and the lips become prominent. The face of an affected child usually has the appearance of old age.

Case presentation

A 22-year-old female patient reported to the department of dental care, KIMS hospital, Secunderabad, with the complaint of multiple missing teeth since childhood (Figure 1). She underwent surgery for the cleft palate during childhood. The patient also gave a history of delay in the eruption of deciduous and permanent teeth, intolerance to heat and reportedly less sweat production. There was no history of consanguineous marriage between the parents. On extra oral examination, the patient had dry skin with periocular area being hyper pigmented and wrinkled with sparse hair on the body and scalp. Hairs present were fine in texture & lighter in color. Prominent supraorbital ridges, frontal bossing, small and outwardly placed ears and flattened nasal bridge was also present. Both upper and lower eyelids showed sparse eyelashes. The skin was warm and dry, and absence of tears was also reported.

Intra oral examination revealed multiple missing teeth (Figure 2a,b,c,d) in the maxillary and mandibular arches, malalignment of teeth was present, palatal fistula with narrow maxillary arch seen, and in addition to that swollen gums were present. Based on these findings a diagnosis of ectodermal dysplasia was made. A cone-beam computed tomography (CBCT) scan was made which revealed multiple missing teeth (Figure 3), inadequate bone density in certain areas.

Accordingly her treatment was planned in three phases, which are i) Laser gingivoplasty for inflamed and swollen gums, ii) Orthodontic correction of malaligned teeth followed by closure of the palatal fistula with the intervention of plastic surgeon, iii) Replacement of missing teeth with implants and crowns.

After taking the consent from the patient and her family, laser treatment was done for the gums, orthodontic treatment was done with periodic evaluation to align the teeth. Along with the orthodontic correction 2 stage implants were placed in areas of missing teeth. A period of 6 months was required for proper osseo integration of the implants and proper healing of gums.

After 6 months, a CBCT scan was taken again and proper osseo integration of implants was seen, teeth
were aligned and the fistula was closed. Therefore crowns were placed to replace the missing teeth (Figure 4 a, b). The facial profile and expression improved significantly (Figure 5 a, b).

**Conclusion**

Ectodermal dysplasia is a rare genetic disorder with involvement of various tissues in the body. A careful and a thorough examination will lead to accurate diagnosis. Restoration of normal function should be the main concern in these patients.

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**Conflict of interest**

The authors declare no conflict of interest.

**References**


