

ECTODERMAL DYSPLASIA – A CASE SERIES OF 2 CASES

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Abstract

Ectodermal dysplasia is a disorder associated with the disturbance in ectodermal structures, prominent supraorbital ridges, frontal bossing, nasal bridge depression, conical teeth, anodontia or hypodontia. It can be autosomal dominant, autosomal recessive and X- linked. In ectodermal dysplasia due to deficiency of sweat glands patients may suffer with hyperthermia and dry skin.

Case discussion- This case series presents two cases who reported to the department of oral medicine and radiology, faculty of dental sciences with the complaint of missing teeth and wanted the replacement. On examination multiple teeth were absent and history revealed that teeth are missing since birth and atrophy of alveolar ridge was the evident.

The aim of this case series is to assess self-reported signs of ectodermal dysplasia (problems with hair, nails, skin and sweat glands)

Conclusion-The present case series is based on the usual findings of ectodermal dysplasia which can play important role in proper diagnosis and treatment planning in such patients.

Introduction

Ectodermal dysplasia was first described by Thurman in 1848.^{(1) (2)} These are non progressive, diffuse and congenital.⁽³⁾ Estimated incidence is 1 case per 100,000.⁽⁴⁾ Patients affected with ectodermal dysplasia present with a typical features which includes defective hair follicles and eyebrows, prominent supraorbital ridges with frontal bossing, depression of nasal bridge, conical teeth, anodontia or oligodontia.⁽⁵⁾ Because of the deficiency of sweat glands patient may suffer from hyperthermia, dry skin and fever of unknown origin. Most commonly missing teeth are molars. The teeth which are present in oral cavity are usually malformed.

Case Series

Case 1

An 8-yrs old male patient came to department of oral medicine and radiology IMS BHU Varanasi with complaint of missing teeth and mastication problems.

Extraoral examination revealed dry skin, saddle nose, prominent forehead, very fine scalp hair and eyebrows (fig-1). These are the typical findings of ectodermal dysplasia. On the intraoral examination there was edentulous mandibular arch (fig 2-A) and partially edentulous maxillary arch. Maxillary first molar and central incisors were present. Central incisor was conical in shape. There was atrophy of alveolar ridges (fig-2-B). Parental history revealed febrile convulsions and delayed eruption of tooth and there was no other member of the family with the same condition.



Fig-1
(Facial view of
the patient)



Fig-2A
Intraoral view of
the upper jaw



Fig-2B
Intraoral view of
the lower jaw

Radiographic examination reveals the absence of all the teeth except 11, 16,21,26with two unerupted molar teeth in the upper and lower jaw, alveolar ridges of both maxillary and mandibular jaws were underdeveloped.(Fig-3)

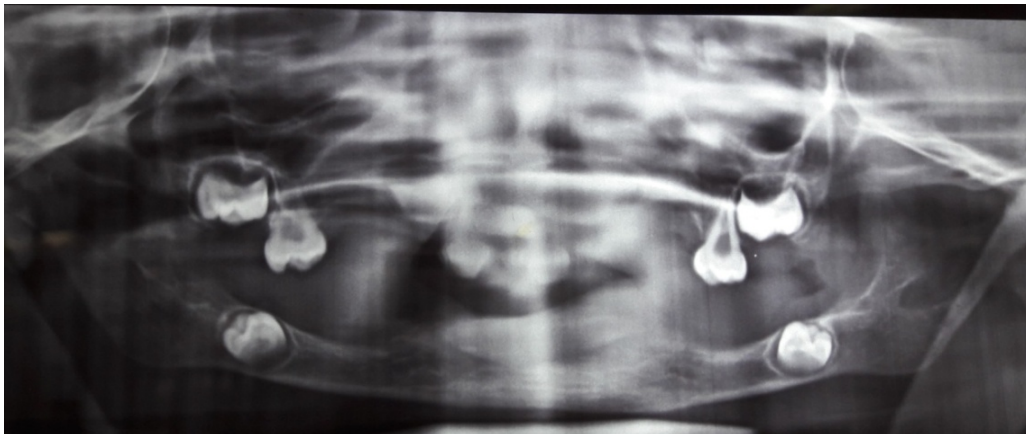


Fig-3
(Panoramic radiograph confirming the absence of teeth)

Case-2

A 24yrs patient came to department with complaint of missing teeth since childhood.

On extra oral examination there was slightly elevation of temperature with dry and scaly skin. Very thin eyebrows was seen and hair examination revealed fine and sparse hairs (fig-4) .Intra orally patient had oligodontia and following teeth were present 16,25,26,36,37,46,47 , retained 55 and 75(fig 5-A& B).

He was also unable to tolerate hot climates and dry weathers.



Fig-4
(Facial view of
the patient)



Fig-5A
(Intraoral view of
the upper jaw)



Fig-5B
(Intraoral view of
the lower jaw)

Radiographic examination reveals presence of few teeth 16, 25, 26, 36, 37, 46, 47, retained 55 and 75. No impacted tooth or tooth buds were present. (Fig. 6)



Fig 6
(Panoramic radiograph confirming complete absence of teeth)

Both patients were prosthetically rehabilitated and are being followed up regularly.

Discussion

Ectodermal dysplasia represents a group of inherited disorders characterized by triad of signs i.e. missing teeth (oligodontia or anodontia), comprising sparse hair (hypotrichosis) and defect in sweat glands (anhidrosis or hypohidrosis).⁽⁶⁾ They are congenital, extremely rare, genetically inherited disorders.

There are 2 major types (1) X-linked anhidrotic or hypohidrotic, where sweat glands are either absent or significantly reduced in number. It is also called as Christ-Siemens Touraine syndrome, (2) Hidrotic, where sweat glands are normal also called as Clouston's syndrome.⁽⁷⁾ In these two major types hypohidrotic ectodermal dysplasia (Christ-Siemens-Touraine syndrome) are more common.⁽⁷⁾

Classification of the ectodermal dysplasias into different subgroups according to the presence or absence defects-⁽⁸⁾

- ED1: Trichodysplasia (hair dysplasia)
- ED2: Oligodontia or anodontia(dental dysplasia)
- ED3: Onychodysplasia (nail dysplasia)
- ED4: Dyshidrosis (sweat gland dysplasia)

Ectodermal dysplasia syndromes can also be associated with midfacial defects such as cleft lip, cleft palate, or both. There are 3 most commonly found syndromes (1) ectodermal dysplasia, ectrodactyly, and clefting (EEC) syndrome (2) Hay-Wells syndrome or ankyloblepharon, ectodermal dysplasia, and cleft lip/palate (AEC) syndrome; and (3) Rapp-Hodgkin syndrome.⁽⁹⁾

In the patients of ectodermal dysplasia because of congenital absence of teeth alveolar ridges may be underdeveloped and it results in decreased vertical facial height.⁽¹⁰⁾

Stability and retention of dentures are difficult to obtain because of dry mouth due to lesser salivary secretions.

Conclusion

Individuals affected by ectodermal dysplasia need special treatment which includes dental management for improvement of aesthetics, function and positive psychological impact. Different treatment modalities are there according to need and growth of the craniofacial structure. Management include fabrication of complete

dentures or implant supported dentures, removable partial dentures, and fixed prosthesis.

References

1. Thurnam J. (1848) Two cases in which the skin, hair and teeth were very imperfectly developed. Proc RM Chir Soc. 31:71-82.
2. Hickey AJ and Vergo TJ Jr (2001) Prosthetic treatments for patients with ectodermal dysplasia. J Prosthet Dent 86:364-8
3. Dhanrajani PJ and Jiffy AO (1998) Management of Ectodermal Dysplasia. A Literature Review. Dental Update 25:73-75.
4. Kupietzky K and Milton H (1995) Hypohidroticb ectodermal dysplasia: Prosthetic managment of hypohidrotic ectodermal dysplasia. Quintessence Int 26:285-291.
5. Fazil . K.A, Subhas Babu. G, Shishir Ram Shetty and Supriya Bhat(2013) hypohidrotic ectodermal dysplasia : a casereport and literature review. Journal of Evolution of Medical and Dental Sciences 10:1551-1554.
6. K.K. Shashibhushan, Revathy Viswanathan, N. SathyajithNaik and V.V. SubbaReddy (2011) Hypohidrotic Ectodermal Dysplasia with Anodontia. J ClinExpDent 3:352-5.
7. Tarjan I, Gabris K andRozsa N. (2005) Early prosthetic treatment of patients withectodermal dysplasia: a clinical report. J Prosthet Dent 93:419-24.
8. Bhadauria R.S., Ranjana Sharma and Gayatri Prajapat (2014) A Review Article on Ectodermal Dysplasia International Journal of Pharmaceutical Erudition: 4 (1), 39-45.
9. Okamura E, Suda N, Baba Y, Fukuoka H, Ogawa T and Ohkuma M(2012), et al. Dental and maxillofacial characteristics in six Japanese individuals with ectrodactyly-ectodermal dysplasia-clefting (EEC)syndrome. *Cleft Palate Craniofac J*.
10. Rashmisingh and Gauri S LELE (2008) Hypohidrotic Ectodermal Dysplasia: A Case Report International Dental and Medical Disorders 1:11-13.

