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Case Report

Ectodermal Dysplasia with Partial Anodontia: A Case Report

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ABSTRACT
Ectodermal dysplasia is an inherited genetic disorder affecting ectodermal structures. It comprises of defects related to hair, nails, eyes, dental and eccrine glands. This leads to unregulated body temperature, oligo or anodontia, dry eyes and dry mouth. Ectodermal dysplasia patients require prompt medical and dental intervention for adequate oral and general health.
Key words: Ectodermal dysplasia, partial anodontia.

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INTRODUCTION

Ectodermal dysplasia is rare hereditary disorder which effects the development and function of ectodermal tissues, mainly hair, nails, teeth and sweat glands .It is a heterogenous group of disorders having a variety of phenotypically distinct entities.[1]

It was first reported by Lanz in 1792.Wedderburn in1838 reported 10 cases of ectodermal dysplasia in male members of a hindu family. First case of ectodermal dysplasia was published in1848 by Thurman. In 1895 Nicolle and Hallipre first reported case of hydrotic ectodermal dysplasia in a french canadian family. Christ in 1913 characterized hypohydrotic ectodermal dysplasia as a congenital defect and its X-linked nature of inheritance was described by Siemens in 1921.[2] The term "Hereditary Ectodermal Dysplasia" was coined by Weech in 1929.[1]

Disorders of this group are diffuse, congenital and non-progressive having prevalence of 1 in 1,00,000 live births. The genes with plausible role in hypohidrotic ectodermal dysplasia are EDA (ectodysplastin A a protein), EDAR (ectodysplastin A receptor) and EDARADD (ectodysplastin A receptor associated death domain).[3]

Among them EDA is associated with X-linked gene, accounting 95% of individuals while EDAR and EDARAAD are associated with autosomal dominant and autosomal recessive forms accounting for only 5%.These genes code for proteins which work in co-ordination during embryonic development effecting the tissues formed.[1]

Clinical features in ectodermal dysplasia include sparse thin hair of scalp, eyebrow and eyelashes, less number of teeth with conical morphology, brittle nails, if eccrine glands are affected dry eyes due to decreased lacrimal activity is observed. [4,5] Other associated features like bossing of frontal bone, very thin alveolar bone, palate with high vault, dry and scaly skin, face skin with hyperpigmentation, hypoplastic maxilla, palmo-plantar keratosis are seen.[2]

CASE REPORT

An 8 years old male visited the department of Oral medicine and Radiology with chief complain of less number of teeth since childhood. There was no history of any live family member presenting with similar condition.

On extraoral examination there were sparse thin hairs on scalp and eyebrows, nails and eyes were normal with adequate flow in eyes. Skin was thin with no apparent dryness.

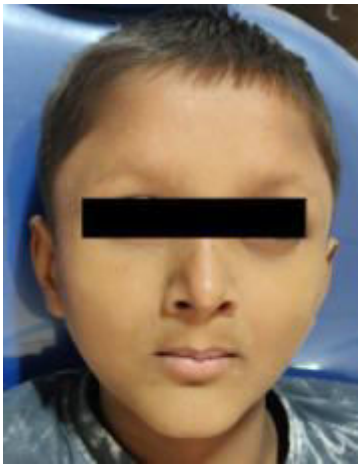

Figure 1: Extra oral picture showing thin sparse hair on eye brows and scalp.

On intraoral examination there were multiple missing teeth. The deciduous maxillary central incisors were retained and maxillary lateral incisors and canine, mandibular central incisors and canine were absent. Salivary flow was adequate.

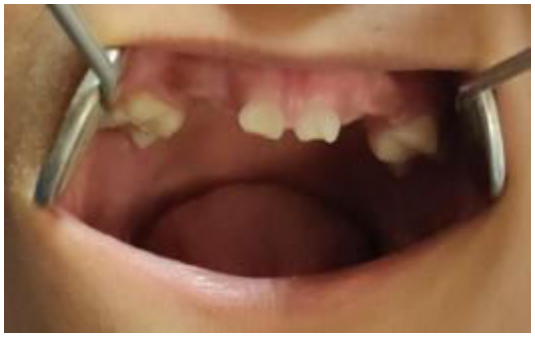

Figure 2: Intra oral picture showing retained deciduous maxillary central incisors and missing lateral incisors and canine

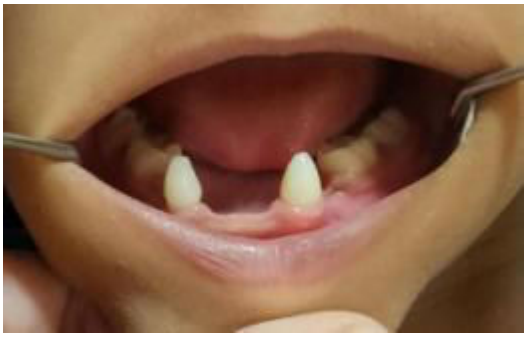

Figure 3: Intraoral picture showing missing mandibular permanent central, lateral incisors.

Orthopantomogram showed mixed dentition with partial anodontia. There were missing deciduous and permanent tooth buds of maxillary lateral incisors, canines, premolars in mandible deciduous and permanent tooth buds of central incisor, canine and premolars were missing.

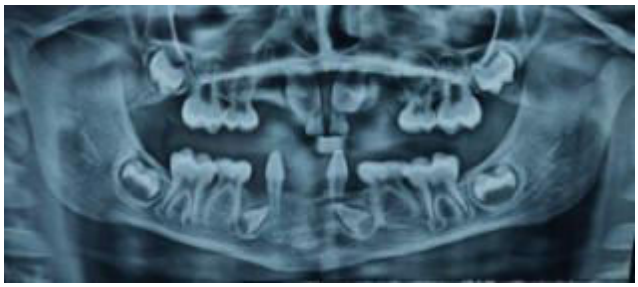

Figure 4: Orthopantomogram showing mixed dentition with partial anodontia.]

Based on above findings diagnosis of ectodermal dysplasia was made. Patient was educated of his condition and need for regular adjustments and reconstruction of dentures at different stages of growth. Treatment was aimed at restoring function and preserving present oral and dental structures. Oral prophylaxis with oral hygiene instructions were given and patient was planned for upper and lower partial dentures and later for dental implants.

DISCUSSION

Freire-Maia and Pinheiro classified Ectodermal Dysplasia into subgroups depending upon the involved ectodermal structures –

1) Hair Anomalies or Trichodysplasia,
2) Dental Anomalies,
3) Nail Abnormalities or Onychodysplasia,
4) Eccrine Gland Dysfunction or Dyshydrosis.

It can be further grouped into:-

Group A- disorders which comprise of defect in at least 2 of the 4 classical ectodermal structure with or without other associated defect

Group B- disorders which comprised of defect in one classical ectodermal structure (1-4 from above) in combination with (5) a defect in any one of the ectodermal structure (ear, lip).

In our patients two of the four classical ectodermal structures were affected hence it was characterized as a Group A disorder.

Ectodermal dysplasia patient needs proper medical and dental care depending upon affected ectodermal structure. In patients with defective sweat glands adequate hydration and thermoregulation is advised. Patient with dental defects require regular dental evaluation and intervention through prosthetic rehabilitation, oral prophylaxis and oral hygiene maintenance. Patients with dry mouth and eyes should be prescribed artificial saliva and tears respectively.[5]

CONCLUSION

Ectodermal dysplasia is a rare genetic disorder, pre-natal genetic analysis is recommended in families which suffer from this genetic disorder. Apart from genetic analysis, patients with ectodermal dysplasia need regular medical and dental monitoring in effort to reduce the effect of anomalies and restoration of function.

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