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Esthetic Rehabilitation of a Patient with Amelogenesis Imperfecta – A Case Report

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Abstract: *Amelogenesis Imperfecta is a disorder which affects the enamel structurally as well as clinically of all or nearly all the teeth in a more or less equal manner.AI is a group of inherited defects of dental enamel formation that shows both clinical and genetic heterogeneity. Enamel findings in AI are highly variable, ranging from deficient enamel formation to defect in the mineral and protein content. AI exists in association or associated with other abnormalities in syndromes. This case report describes the therapeutic planning and rehabilitative procedure experienced by a patient affected by AI.*
Key Words*: Amelogenesis Imperfecta, enamel hypoplasia, tooth discoloration*

I. Introduction

Amelogenesis Imperfecta represents a group of developmental conditions, genomic in origin, which affects the structure and clinical appearance of enamel of an or nearly all the teeth in a more or less equal manner, and which may be associated with morphologic or biochemical changes elsewhere in the body[1]. The prevalence varies from 1:700 to

1:14000 according to the population studied. It is characterized by clinical and genetic heterogenecity in the absence of systemic abnormalities or diseases. AI is also known as by varied names such as hereditary enamel dysplasia, hereditary brown enamel and hereditary brown opalescent teeth. AI pertains to a group of developmental tooth abnormalities which affects the genome of the individual and regard at least of the stages of enamel formation.

AI in general, a hereditary disorder with clinical impact on both deciduous and permanent teeth[2]. AI was first described in 1890, but the substancial separation from dentinogenesis imperfect was not made until 1938, when AI was described as an autonomous entity[3]. Regarding etiology numerous studies have reported a variety in its inheritance pattern including either autosomal, x- linked dominant or recessive modes. More specifically, it was mentioned that enamel hypoplasia is inherited predominantly in a sex- linked, incomplete, dominant trait, whereas the enamel hypo- mineralisation in an autosomally dominant manner[4].

II. Case Report

A 12 year old male patient reported with a chief complaint of yellowish discoloration of his teeth since 5- 6 years. The patient had a non contributory medical history. Familial history reveals the consanguineous marriage. Extra orally patient had straight profile, asymmetrical face competent lips, with mid facial hypoplasia. On clinical examination patient had permanent dentition with yellowish discoloration in upper and lower anteriors, Clinically missing 13 and 23, grossly decayed 46, root stumps of 16, 26, 36 and developing open bite.

Figure 1: Facial view

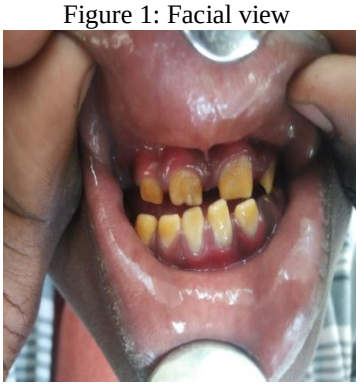

Figure 2: Tooth discoloration in upper and lower anteriors Panoramic radiograph showed the presence of a thin layer of enamel with radiodensity of enamel more than dentin.

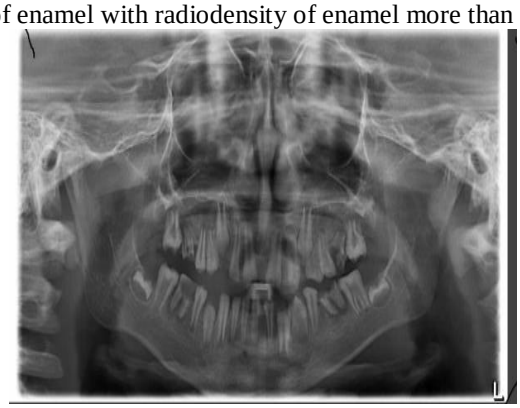

Figure 3: OPG showed thin layer of enamel with radiodensity On the basis of clinical and radiographic features, final diagnosis of hypoplastic AI was made. Parent and patient were counseled and treatment part composite restoration in upper and lower anterior teeth.

Figure 4: composite restoration in upper and lower anteriors.Root canal treatment with gutta percha followed by stainless steel crown in relation to 46 and extraction of 16,26 and 36 under Local anesthesia followed by Removable partial denture were done. Parent and patient were counseled and review done periodically.

III. Discussion

Tooth enamel is the most highly mineralized structure in the human body with 85% of its volume occupied by hydroxyapetite crystals. The physical properties and physiological functions of enamel and directly related to the composition, orientation, the depositions and the morphology of the mineral components within the tissue. During organogenesis, the enamel transition from a soft and pliable tissue to its final form, which is almost entirely devoid of protein. The final composition of enamel is a reflection of the unique molecular and cellular activities that take place during its genesis.AI encomposes a complicated group of conditions that demonstrate developmental alterations of the structure of the enamel in the absence of a systemic disorder[5],[6].

During the secretary stage of enamel formation, enamel matrix proteins, amelogenin, the enamelin and ameloblastin secreted by ameloblasts play a key role in the growth of enamel crystal. Reports have shown that the mutation in the amelogenin gene (AMELX) and enamel gene (ENAM) is implicated in the pathogenesis of the dominant forms of AI [7]. In AI because of poor differentiation of ameloblast it results in poor development or complete absence of enamel of the teeth. Among the three subtypes of AI, Hypoplastic AI had absence of

normal enamel morphology which results in reduced occlusal function and reduced esthetics and on increased chance of caries development[8]. Other dental features associated with AI includes quantitative and qualitative enamel deficiency, pulpal calcifications, taurodontism and root malformation, impaction of permanent teeth, progressive crown and root resorption, congenitally missing teeth, anterior and posterior open bite occlusion[9]. When enamel hypoplasia is the predominant clinical findings, the enamel is reduced in thickness in the histopathological examination. The family history, pedigree plotting, clinical observation and meticulous recording form the backbone of diagnosis[10].

A variety of symptoms can be presented with AI. The most of the substantial findings comprise extensive bone loss, tooth sensitivity, excess attrition, leading to short clinical crowns, spacing in the anterior region of the dentition, normal or light proximal contacts in the posterior region and a general enamel caries resistance[11].

It is important to point out the correlation of AI with two rather rare syndromes named Jalili syndrome and enamel dysplasia with hamartoma atypical follicular hyperplasia [EDHFH] syndrome. Jalili syndrome refers to the co-existence of cone rod dystrophy (CRD) and AI due to mutation of the CNNM4, which is a metal carrier. A variety of symptom including visual deficiency, abnormal dentition, photophobia, nystagmus increasing under photophic condition can also be presented with AI. It may be fully demonstrated either in the infancy or in the childhood. The second syndrome in exclusively reported in black south Africans. Hamartomatous atypical follicular hyperplasia with features similar to central odontogenic fibroma in multiple impacted teeth and also a generalized enamel dysplasia with features of hypoplastic AI are nearly always present. Other conditions often mentioned are open bite, gingival growth, hypodontia, pulpal calcification and there would be aberrant tooth formation of the unerupted tooth[12][13].

IV. Conclusion

Amelogenesis Imperfecta presents with problems of socialization, function and discomfort which may be managed by early vigorous intervention, both preventively and restoratively. A multidisciplinary approach should be planned which focus on early diagnosis, pain management, prevention, sterilization, restoration of defect and regular long term management. The treatment plan should also accommodate factors including the patient's age, socio economic status, disease type and severity and over all oral condition.

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