Case Report

AMELOGENESIS IMPERFECTA- A CASE REPORT

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ABSTRACT:
Amelogenesis imperfecta (AI) is a hereditary heterogenous disorder that causes developmental alterations in the structure of enamel. The Al trait can be transmitted by either autosomal dominant, autosomal recessive, or X-linked modes of inheritance. Genes implicated in autosomal forms are genes encoding enamel matrix proteins, namely: enamelin and ameloblastin, tuftelin, MMP-20 and kallikrein – 4. It is necessary to diagnose the case and provide durable functional and esthetic management of these patients, where the unaesthetic appearance has a definite negative psychological impact. We present a case of AI affecting the dentition of a 10-year-old girl.
Key Words: Amelogenesis imperfect, Enamel Hypoplasia, hydroxyapatite crystals, stainless steel crown.

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INTRODUCTION:

Tooth enamel is the most highly mineralized structure in the human body, with 85% of its volume occupied by hydroxyapatite crystals. The final composition of enamel is a reflection of the unique molecular and cellular activities that take place during its genesis.[1,2] Deviation from this pattern may lead to amelogenesis imperfecta. Amelogenesis Imperfecta (AI) encompasses a complicated group of conditions that demonstrate developmental alterations in the structure of the enamel in the absence of a systemic disorder.[3] AI is also known by varied names such as, Hereditary enamel dysplasia, Hereditary brown enamel, Hereditary brown opalescent teeth. AI follows an autosomal dominant, autosomal recessive or X-linked pattern of inheritance OMIM #301200. These enamel defects are basically a result of gene mutations associated with amelogenin, ameloblastin and enamelin protein. Recent reports involve kallikrein-4 (KLK4), MMP-20 and DLX3 genes in the etiologies of some cases.[1,4] The term "Amelogenesis Imperfecta" was coined by Weinmann et al in the 1940s. The reported prevalence of AI is highly variable, reported to be 1:14.000 in the USA, 1:700 in Europe and 1:8000 in Israel. ICD-9 520.5 is designated for "hereditary disturbances in tooth structure that includes: amelogenesis imperfecta, dentinogenesis imperfecta, odontogenesis imperfecta, dentinal dysplasia, shell teeth". The main classification of AI that is widely adopted is: Hypoplastic type, Hypomineralized type, Hypomaturated type, Hypoplastic and hypomaturated type in addition to taurodontism. Extensive loss of tooth tissue, Poor esthetics, tooth sensitivity are major concerns in such patients. AI has been also associated with additional features of Abnormal eruption pattern, Congenitally missing teeth, Anterior open bite, Pulpal calcifications, Root malformations, Resorption, Hypercementosis, Taurodontism.[1]
This clinical report describes the treatment for a young female patient with mutilated natural dentition caused by amelogenesis imperfecta.

CASE REPORT

10 years old female patient reported to Department of Pedodontics and Preventive Dentistry, with a chief complaint of sensitivity to hot and cold and pain in

relation to all her teeth since last 3-4 years. A detailed case history was recorded which revealed that none of the parents, siblings or relatives suffered from similar dental conditions. Intraoral examination revealed dental fluorosis, extrinsic disorders of tooth formation, chronological disorders of tooth formation, localised disorders of tooth formation. [Figure 1 and 2]

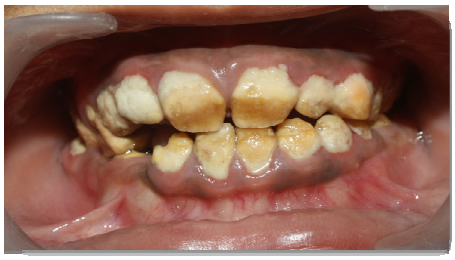

Figure 1: Intra-oral view showing labial surface of teeth with brownish discoloration

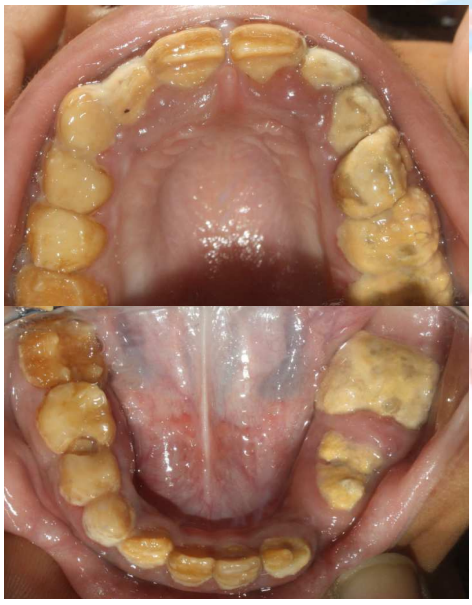

Figure 2: Intra-oral view showing occlusal and lingual surface of teeth with brownish discoloration

Apart from this, her past medical history was noncontributory. Radiographic investigations included an orthopantomogram (OPG) and full mouth intraoral periapical (IOPA) radiographs. The enamel was almost half its expected thickness, but was more radiodense than the dentin. There was normal pulp space, root canal with no obliteration of pulp canal. [Figure 3]

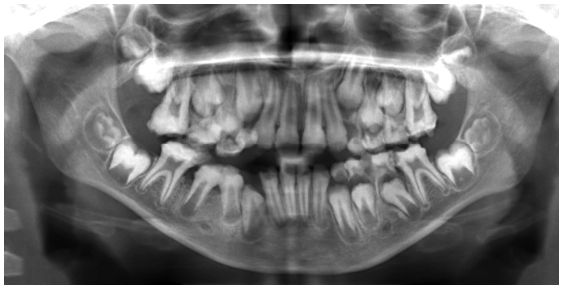

Figure 3: Orthopantomograph

Genetic diagnosis is presently only a research tool that is currently unavailable in India. The dental family history, clinical and radiographic features were suggestive of Autosomal Recessive Hypoplastic Amelogenesis Imperfecta. Treatment plan was developed and discussed with the patient. Complete oral prophylaxis was carried out and oral hygiene instructions were given to patient. Extraction of over-retained deciduous teeth was done. Stainless steel crown for posterior teeth and Polycarbonate crowns for anterior teeth was given. [Figure 4]

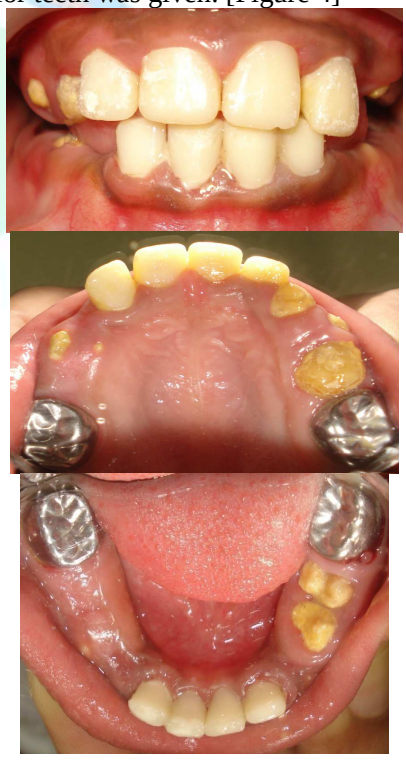

Figure 4: Post-operative photograph

The patient's esthetic and functional expectations were also satisfied. Patient was periodically recalled to maintain oral health.

DISCUSSION

AI is a developmental, often inherited disorder that affects enamel. It occurs in the absence of systemic features and comprises of diverse phenotypic entities.[5] AI can be subdivided at the clinical level into various forms depending on the type of defect and stage at which enamel formation is disturbed, into hypoplastic, hypo mineralized or hypo maturation type.[6] The trait of AI can be transmitted by an autosomal-dominant, autosomal-recessive, or X-linked mode of inheritance.[7]

The distribution of AI types is known to vary among different populations. In a study in Sweden, 63% of the cases were inherited as autosomal-dominant. In contrast, in a study in the Middle East, the most common prevalent type of AI was found to be autosomal-recessive.[8,9] The predominant clinical manifestations of affected individuals are enamel hypoplasia (enamel is seemingly correctly mineralized, but thin), hypomineralization (subdivided into hypomaturation and hyocalcification), or a combined phenotype, which is seen in most cases.[10] Treatment of Amelogenesis Imperfecta presents a challenge for the dentist. The clinical management of an esthetically demanding, complex functional prosthodontic rehabilitation is a clinical challenge. Accurate diagnosis, proper treatment planning, prudent choices of materials, treatment execution are essential for a successful treatment outcome over a long period.[11]

The treatment plan is modified according to the age of the patient and other factors in the present case. PFM or All ceramic crowns were ruled out as the treatment options as the child is in mixed dentition. Laminates/ Veneers were also ruled out due to developing dentition and inability to cover the palatal surface. Composite build-ups were also ruled out because of severe tooth sensitivity complaint of the patient. Stainless Steel crowns for the posterior teeth and Polycarbonate crowns for the anterior teeth were the treatment of choice in present case. Further follow up of such cases should be carried out and as the permanent dentition gets established, patient can be provided with PFM/All ceramic crowns for all teeth.

CONCLUSION:

The pediatric dentist needs to diagnose the condition as early as possible to render proper and prompt treatment to the patient and provide them back with their much deserved beautiful smiles on their innocent faces.

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