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Letter to Editor

Amelogenesis Imperfecta – Report of a rare enamel disorder

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Abstract
In the present article, author reported a case of Amelogenesis Imperfecta which is a rare structural abnormality affecting only enamel of both primary and permanent dentition in a 12-year-old Indian male patient. Documentation and publication of such rare disorders is highly essential for further research pertaining to this dental anomaly to through a more light on the existing literature.

Keywords: Amelogenesis Imperfecta; AI; Enamel defect; Primary dentition; Permanent dentition

Dear Editor-In-Chief,

A 12-year-old male Indian patient reported to a private dental clinic complaining of discoloration in his teeth since birth. His family history was non-contributory with absence of particular signs and symptoms of any systemic, metabolic and syndromic disorders. Patient was apparently normal with normal built and weight. On intraoral examination, patient exhibited mixed dentition period with presence of few primary and erupting permanent teeth. There was generalised yellowish discoloration observed with loss of outer covering of enamel in many teeth. Patient also reported difficulty in chewing hard foods and sensitivity in his teeth. On radiographic examination there was generalised loss of enamel structure affecting both primary and permanent teeth and presence of many impacted and erupting permanent teeth [Figure 1]. Finally, based on clinical characteristics, patient history and radiographic examination features, the present case was diagnosed as Amelogenesis Imperfecta of hypocalcified type (Type III). Detailed treatment options were explained to the parents and scheduled for the treatment.

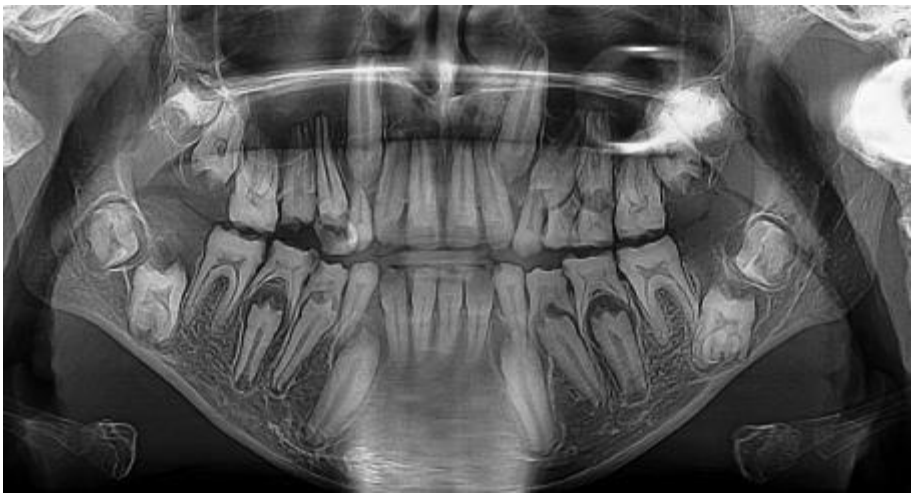

Figure 1: Orthopantomograph showing defective enamel affecting both primary and permanent dentitions

Amelogenesis Imperfecta with a synonym of AI is a hereditary developmental disorder of enamel formation affecting both primary and permanent dentition in the absence of a systemic disorders or syndromes. Clinically, the affected teeth with AI show abnormal color ranging from brown, gray or yellow and are more susceptible to dental caries., tooth attrition, deposition of calculus and generalised gingival or periodontal inflammation [1]. The prevalence of this dental abnormality is reported ranging from 1 in 718 to 1 in 14,000 based on the population studied and depending on the diagnostic criteria used. This condition more commonly shows male predilection however, female distribution is also been reported. Depending on the stage of development with interruption in the development, three types of AI with various 10 subtypes have been mentioned in the dental literature as hypoplastic – 60 – 73% (localized pitted, generalized pitted, generalized thin), hypocalcified (7%) and hypomaturation (20 – 40%) (diffuse creamy, diffuse pigmented, snow-capped) type [2]. One more type has been identified which include hypomaturation combined with hypoplasia and taurodontism. Hypoplastic type is characterized by presence of deficient enamel matrix which is imperfectly formed in quantity but relatively well mineralized. In hypomineralization type, the enamel is present in relatively normal amounts but is insufficiently mineralized. And in the hypomaturation type, the last phases of the mineralization process are anomalous. The subtypes of AI are classified according to the mode of inheritance, clinical, histological, radiological and genetic characteristics [2].

Etiological factor has been associated with mutations in different extracellular matrix (ECM) protein coding genes comprising a total of 10 genes and mainly involving amelogenin, enamelin and kallikrein related peptidase 4 which all help in molecular diagnosis in the majority of the cases [3,4]. The different inheritance patterns associated with AI are autosomal dominant, autosomal recessive and X-linked. Nagaveni NB recently reported an occurrence of AI in Rubinstein-Taybi syndrome patient which was not reported so far [5]. Clinical significance seen with AI are dental sensitivity, loss of vertical dimension, increased tendency for dental caries, anterior open-bite, significant aesthetic problems, gingival and periodontal problems, delayed eruption and tooth impaction. Therefore, meticulous diagnosis is highly essential in these cases to provide appropriate treatment. Various treatment modalities are suggested which include facial veneers for mild type, full coverage crowns for severe form cases. In case of young patients as mentioned in the present report, placement of crown is a major problem because of growth pattern and sensitivity problem associated with conventional crown placement. However, publications have shown positive results following a multisession pattern which includes waiting till for the full eruption of the teeth such as incisors first followed by premolars and cuspids. In some patients reduced sensitivity following crown placement has been reported. Another treatment modality involves use of milled acetal resin overlays until complete eruption of all teeth [4].

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