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Case Report

Amelogenesis imperfecta: A case series

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ABSTRACT

Amelogenesis imperfecta (AI) refers to a group of rare genetic disorders that involve tooth development and that are passed down through families as a dominant trait. This condition is characterized by abnormal enamel formation caused by gene mutations that alter the quality and/or quantity of enamel. This dental problem can impact both primary and permanent dentition, varies among affected individuals, and results in esthetic and functional problems. The treatment planning for patients with AI is related to many factors, including the age of the patient, the type and severity of the disorder, intraoral conditions, and the socioeconomic status of the patient. It is crucial to plan a proper remedy, which requires collaboration among dental specialties to execute comprehensive dental treatment in order to provide a long-term solution with adequate esthetics. This clinical case study looks at three different types of amelogenesis imperfecta patients.

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1. Introduction

Amelogenesis imperfecta (AI) is defined as a group of hereditary developmental defects of the dental enamel affecting both primary and permanent dentition. It affects the structure and appearance of enamel of all teeth. This anomaly exists independent of any related systemic diseases. There are various classification systems proposed for the different types of AI.

AI is classified into

1. Mode of inheritance: autosomal dominant, autosomal recessive, X-linked, isolated case.
2. Molecular basis: chromosomal localisation/locus/mutation.

3. Biochemical outcome: putative result of mutation when known

4. Phenotype: hypoplastic, hypocalcified, hypomaturation, hypomaturationhypoplastic with taurodontism.

2. Case History

2.1. Case 1

17 year old female patient came to the OPD with a chief complaint of discolouration of upper and lower teeth. The discolouration noticed past 10 years back. Intra oral examination revealed generalized yellowish brown discolouration of teeth and rough enamel surface. Chipping of enamel on the buccal surface of 35,36,37,45,46 &47 from the occlusal one third extending to middle one third. Occlusal surface of all teeth is rough and irregular,

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Gingiva is soft and edematous in relation to lower anteriors. (Figure 1 {a-e}) Patient was advised orthopantomogram that showed presence of a thin layer of enamel with radio density of enamel more than dentin, enlarged the pulp chamber (Figure 2). Based on history and clinical examination a provisional diagnosis of AI-hypoplastic. Patient was advised oral prophylaxis and composite restoration or full coverage crown.

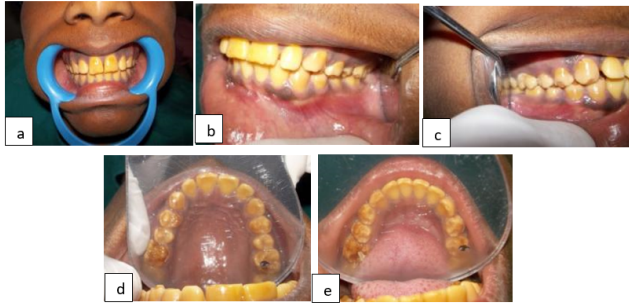


Fig. 1: a: Yellow discoloration of teeth; b: Left buccal segment; c: Right buccal segment; d: Maxillary occlusal view; e: Mandibular occlusal view

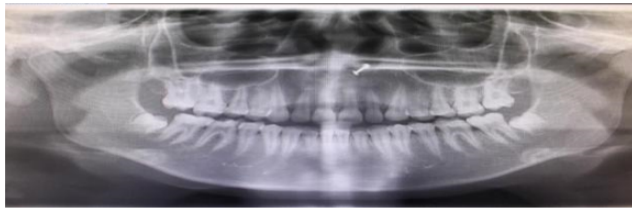


Fig. 2: Panoramicrodiograph showed presence of a thin layer of enamel with radio density of enamel more than dentin, enlarged the pulp chamber

2.2. Case 2

An 8-year-old boy came to the OPD with discoloration of his upper and lower teeth as his main concern. Patient had a history of discoloration on the first tooth since the milk tooth erupted. During an intraoral examination, all of the teeth have a faint brownish discoloration on their labial, lingual, and occlusal surfaces (Figure 3 {a-d}). From the cervical third to the middle third of the crown of the 11, the enamel on the buccal surface has been chipped. The Presence of palatal pit caries in relation to the 11, 21, grossly decayed – 46, presence of gingival abscess -75. Based on history and clinical examination a provisional diagnosis of AI-Hypomaturation. Patient was advised orthopantomogram that showed presence mixed dentition with a thin layer of enamel with radiodensity of enamel was more than dentin and Enlarged pulp chambers (Figure 4) based on the history, clinical findings, Hypomaturational AI. Patient was advised oral prophylaxis and improvement of cosmetic appearance.

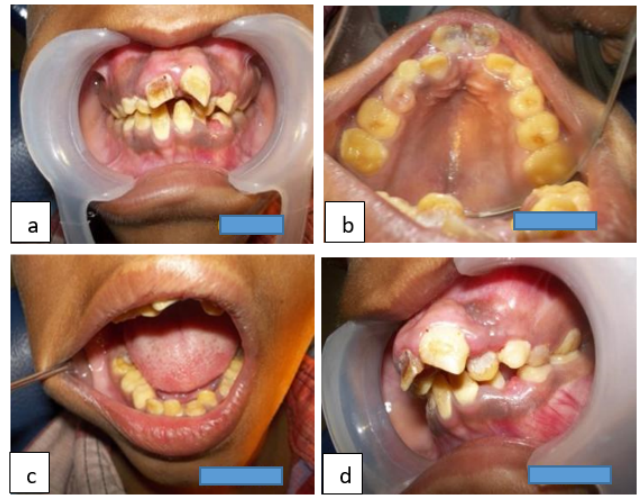


Fig. 3: a: Light brownish discoloration is seen on the labial surface of all teeth; b: Maxillary occlusal view; c: Mandibular occlusal view; d: Left buccal segment

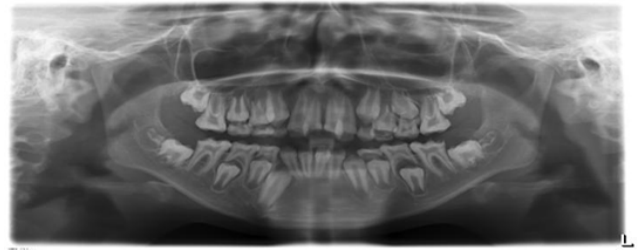


Fig. 4: Panoramic radiograph showed presence mixed dentition present with a thin layer of enamel with radio density of enamel more than dentin, Enlarged pulp chambers

2.3. Case 3

The patient was a 22-year-old man. The patient presented to the OPD with a primary complaint of discoloration of the upper and lower teeth. An intraoral analysis showed yellowish brown discoloration of the teeth and a rough enamel surface. Brownish discoloration on all of the teeth's surfaces, as well as enamel chipping on the labial surfaces of all of the teeth (Figure 6 {a-c}). Based clinical examination a provisional diagnosis of AI-hypocalcified. Patient was advised orthopantomogram that showed Presence of thin radio opaque layer of enamel with normal radio density. Impacted supernumerary 11,33,43 (Figure 7). Based on the clinical findings, radiological report the case was diagnosed as Hypocalcified AI. Patient was advised oral prophylaxis and improvement of cosmetic appearance.

3. Discussion

AI is a group of hereditary diseases affecting the tooth enamel in either quality or quantity, which makes the malformation of the crown due to abnormal enamel

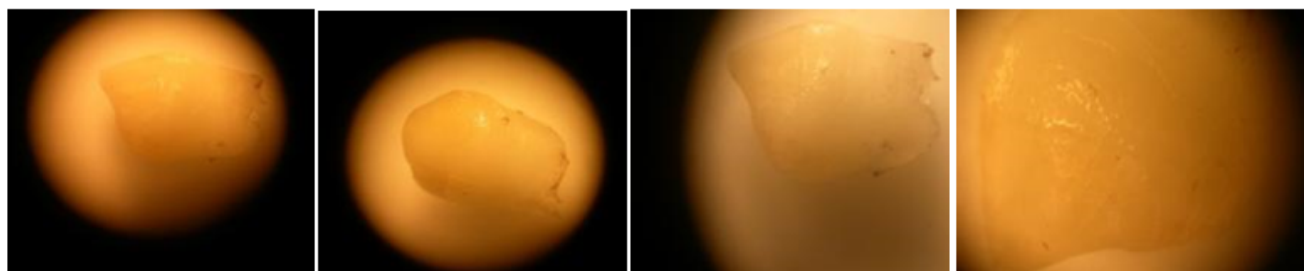


Fig. 5: Tooth Section IRT 74- Section shows reduced enamel thickness with irregular enamel rod patterns and underlying normal dentin. It is consistent with the clinical diagnosis.



Fig. 6: a: Brownish discoloration present on all the surfaces of the teeth; b: Mandibular occlusal view; c: Maxillary occlusal view



Fig. 7: Presence of thin radio opaque layer of enamel with normal radiodensity. Impacted supernumerary 11,3343

density.¹ The prevalence varies from 1:700 to 1:14,000, according to the populations studied.²

The main complaints of patients who are affected by AI are increased sensitivity to hot and cold, reduced aesthetic appearance, and discoloration of mandibular and maxillary anterior teeth and masticatory problems.³ Due to this, dental treatment of patients with AI is a challenge, requiring a detailed interdisciplinary treatment approach based on the correct diagnosis. Prakesh et al. in the year of 2014 investigated the impact of the alterations of AI on the quality of life of patients (61 patients, response rate 61%) suffering from AI. For about 90% of all patients, the main complaint was the discoloration of their teeth, and about 77% asked for an improvement of their smile. For nearly the same percentage (74%), reduction of sensitivity was the most important reason to seek for dental treatment, while improvement of the tooth size was important for 60% of all patients.⁴

Various studies have been done on the different treatment protocols for AI. In 2015, Lundgren et al. published

a split mouth study on the long-term outcome of full crown restoration in patients with AI using different crown materials.⁵ In the year of 2014, Lundgren et al.⁶ concluded that the longevity of dental restorations in patients with AI is considerably reduced and this correlates with the severity of AI.

Hence, extensive coronal restorations of anterior and posterior teeth are often indicated. Besides full metal crowns, especially in the posterior region, also porcelain fused to metal crowns and bridges and recently all-ceramic crowns have been shown to allow sufficient long-term restoration of the affected teeth. At the same time, depending on the age of the patient, other materials like composite crowns, composite veneers, and in younger patients, even stainless steel crowns may be beneficial. Short clinical crowns sometimes require to perform crown lengthening or to perform gingivectomy or gingivoplasty to ensure adequate preparation for fixed crowns and bridges.

Newer, more aesthetic materials for full-crown restorations like lithium disilicate offer a further clinically relevant advantage as these materials require a reduced reduction of dental hard tissues during preparation. Lithium disilicate all-ceramic crowns require only a thickness of approximately 0.8 mm in the cervical region, and an occlusal reduction of only 1.5–2.0 mm is sufficient.

Practice of good oral hygiene and reduction of the degree of gingivitis are essential to ensure optimal longevity of restorations and to improve treatment outcome, because periodontal status of remaining tooth structure has to be good.

Rowley et al. reported that loss of vertical dimension occurred frequently in association with AI, most often in association with the hypo calcified type of AI. These alterations of the vertical dimension were diagnosed more often in female than in male subjects.⁷

As it is a hereditary disorder, it is important to educate the parents and it should be diagnosed in an early stage to minimize the destructions caused by this mineralization disorder. Frequent dental appointments to optimize the gingival and periodontal status, to establish an optimal oral hygiene, and to screen for progressive destructions are

required. Especially for patients with hypo plastic forms of AI, orthodontic treatment to gain the best possible status for further prosthetic treatment is beneficial. As soon as the completion of the skeletal growth is finished, a prosthetic full-mouth reconstruction is indicated for all patients suffering from AI to prevent further attrition and destruction of the teeth and to avoid loss of the vertical dimension.

4. Source of Funding

None.


5. Conflict of Interest

None.

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