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Case Report

Amelogenesis Imperfecta - A Case Report

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Abstract

Amelogenesis imperfecta involves a group of conditions that demonstrate developmental alterations in the structure of the enamel in the absence of a systemic disorder. This condition is characterized by abnormal enamel formation which is compromised both in quality and quantity. This condition can affect both the primary and permanent dentition posing functional and aesthetic problems for the affected individuals. Here we present a case of Amelogenesis imperfecta in a 32 years old male patient.

Keywords: *Amelogenesis Imperfecta; Hypomaturation; Taurodontism*

Introduction

Amelogenesis imperfecta involves a group of conditions that demonstrate developmental alterations in the structure of the enamel in the absence of a systemic disorder.

Case Report

A 32 years male patient came to our department of oral medicine and radiology with the chief complaint of yellowish discolouration of teeth since childhood. He gave no history of adverse or deleterious habits. There was no significant medical history. In the family history patient gave a history of his sister having similar condition since childhood. On general physical examination there was no significant changes seen. On intraoral examination, generalized yellowish discolouration of anterior and posterior teeth were present. Chalky appearance of the enamel was noted. Vertical grooves were present in the labial aspect of maxillary and mandibular incisors. Attrition was present w.r.t posterior teeth. Decay was present w.r.t 46 (Figure 1 and 2). Panoramic radiograph (Figure 3) revealed generalized reduced enamel thickness but radiopacity more than the dentin. Enlarged pulp chambers were seen in the premolars. Taurodontism was present

in relation to maxillary and mandibular molars. Based on the history, clinical examination and radiographic findings, a diagnosis of Amelogenesis imperfecta- Hypomaturation-hypoplastic type with taurodontism was given. Extraction of 46 followed by ground sectioning of the tooth was advised. Since the patient wanted treatment for his teeth discolouration he was not willing for extraction of the teeth.

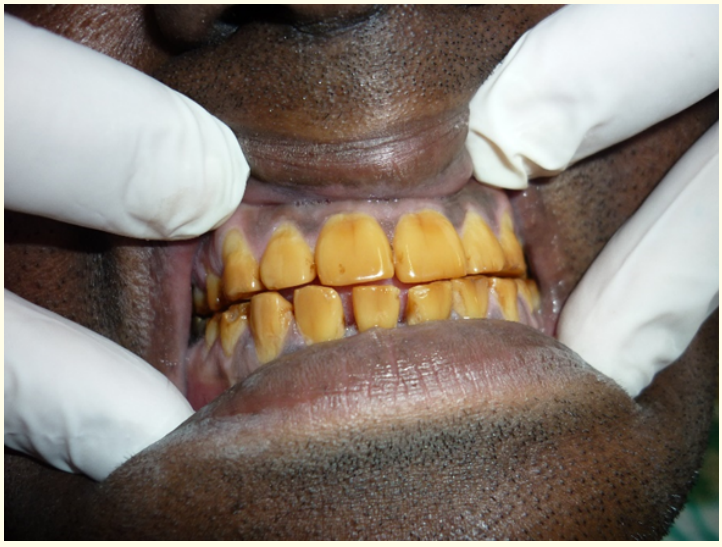

Figure 1: *Yellowish discolouration with vertical grooves on labial surfaces of anterior teeth.*

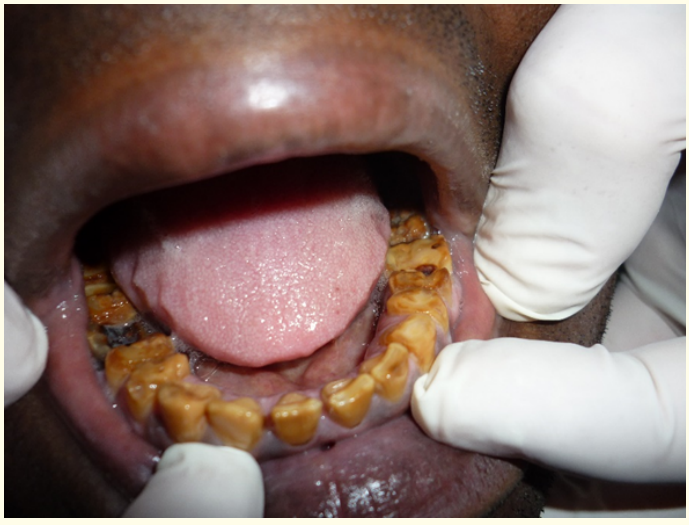

Figure 2: *Mottled and chalky appearance of teeth seen with attrition of posterior teeth.*

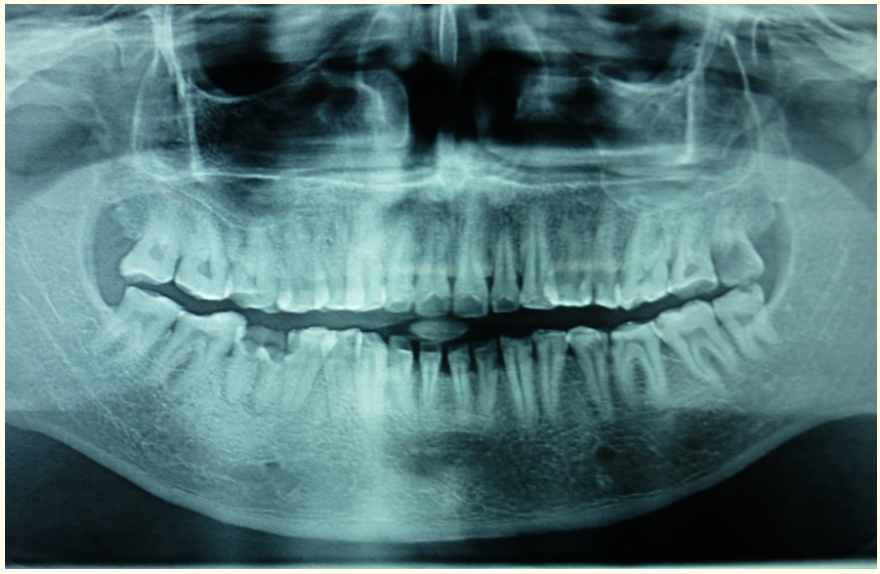

Figure 3: *Generalized reduced thickness of enamel noted with enlarged pulp chambers seen.*

Discussion

Dental enamel is a highly mineralised tissue with over 95% of its volume occupied by unusually large, highly organised, hydroxyapatite crystals. The formation of this highly organised and unusual structure is thought to be rigorously controlled in ameloblasts through the interaction of a number of organic matrix molecules that include enamelin, amelogenin, ameloblastin and other enzymes [1] mutations in these genes can cause Amelogenesis imperfecta.

Amelogenesis imperfecta (AI) is a rare genetically determined defect in enamel mineralization with a prevalence between 1:14,000 and 1:70 [2,3].

AI represents a group of conditions, genomic in origin, which affect the structure and clinical appearance of the enamel of all 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 [4].

There are many classifications of AI-Weinmann., *et al*. [5], 1945, Darling 1956 [6], Witkop 1957 [7]. Based on enamel appearance and hypothesized developmental defects, AI is categorized as 4 patterns: Type I hypoplastic, Type II hypomaturation, Type III hypocalcification and Type IV hypomaturation-hypoplastic with taurodontism.

In patients with hypoplastic amelogenesis imperfecta, the basic alteration centers on inadequate deposition of enamel matrix.

In hypomaturation amelogenesis imperfecta, the enamel matrix is laid down appropriately and begins to mineralize; however, there is a defect in the maturation of the enamel's crystal structure. Affected teeth are normal in shape but exhibit a mottled. opaque white-brown and yellow discoloration.

In the hypomaturation-hypoplastic pattern, the predominant defect is one of enamel hypomaturation in which the enamel appears as mottled yellowish white to yellow-brown. Pits are seen frequently on the buccal surfaces of the teeth. Radiographically, the enamel appears similar to dentin in density, and large pulp chambers may be seen in single rooted teeth in addition to varying degrees of taurodontism [8].

Our case was type IV hypomaturation-hypoplastic with taurodontism.

Diagnosis should be drawn based on the family history, pedigree plotting, meticulous clinical and radiological observation. Dental radiographs of AI teeth provide vital information to the clinician related to the degree of enamel mineralization to design an appropriate treatment plan [9].

According to Seow (1993), the main clinical problems of AI are esthetics, dental sensitivity, and loss of occlusal vertical dimensions. The supportive clinical care needed by these individuals is substantial both in terms of clinical and emotional demands [10]. Management of amelogenesis imperfecta using fixed prosthodontics is reasonable for restoring the function and aesthetic appearance. The long-term efficiency of fixed prosthodontics has advantages to prevent further destructions related to occlusal wear or impairment of the vertical growth [11].

Conclusion

A proper history, clinical evaluation, genetic mapping, radiological interpretation followed by multidisciplinary approach for treatment and adequate follow up is necessary to prevent severe manifestations of Amelogenesis imperfecta.

Bibliography

1. Crawford PJ., *et al*. "Amelogenesis imperfecta". *Orphanet Journal of Rare Diseases* 2 (2017): 17.
2. Witkop CJ Jr., *et al*. "Autosomal recessive pigmented hypomaturation amelogenesis imperfecta". *Oral Surgery, Oral Medicine, Oral Pathology, and Oral Radiology* 36.3 (1973): 367-382.
3. Sundell S and Koch G. "Hereditary amelogenesis imperfecta. I. Epidemiology and clinical classification in a Swedish child population". *The Swedish Dental Journal* 9 (1985): 157-169.
4. Aldred MJ., *et al*. "Amelogenesis imperfecta – a classification and catalogue for the 21st century". *Oral Diseases* 9 (2003): 19-23.
5. Weinmann JP., *et al*. "Hereditary disturbances of enamel formation and calcification". *Journal of the American Dental Association* 32 (1945): 397-418.
6. Darling AI. "Some observations on amelogenesis imperfecta and calcification of the dental enamel". *Journal of the Royal Society of Medicine* 49 (1956): 759-765.
7. Witkop CJ. "Hereditary defects in enamel and dentin". *Acta Genetica et Statistica Medica* 7 (1957): 236-239.
8. Neville BW., *et al*. "Chptr 2: Abnormalities of teeth. In: Oral and Maxillofacial Pathology". 2nd ed. Pub: Saunders (2005): 89-91.
9. Van Heerden WF., *et al*. "Amelogenesis Imperfecta: multiple impactions associated with odontogenic fibromas (WHO) type". *Journal of the Dental Association of South Africa* 45 (1990): 467-471.

10. Seow WK. "Clinical diagnosis and management strategies of amelogenesis imperfecta variants". *International Journal of Paediatric Dentistry* 15 (1993): 384-393.

11. Ergun G and Ataol AS. "An Interdisciplinary Approach for Hypoplastic Amelogenesis Imperfecta: A Case Report". *The Open Dentistry Journal* 12 (2018): 466-475.

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