



The Consequences of Untreated Amelogenesis Imperfecta Type III in a 19-year-old African American Female: A Case Study

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Abstract

Amelogenesis Imperfecta (AI) is a group of hereditary disorders affecting either quality or quantity of enamel formation, resulting in structural and aesthetic defects of teeth. AI is a developmental defect with a complex pattern of inheritance manifesting as various forms of enamel dysplasia often leading to small teeth with discolored rough surface [1]. There are various types of defects associated with Amelogenesis Imperfecta. This case study presents a discovery of AI in a clinical setting during a routine dental examination of a 19-year-old African American female who lacked access to dental care for two years. Long term management of cases such as these can pose challenges to dental practitioners due to variations in the clinical manifestations of AI in the oral cavity, but more importantly from the rapid encroachment of the dental pulp from the breakdown of affected teeth that are a result of this oral disease.

Keywords: Amelogenesis Imperfecta; Enamel Dysplasia; Decalcification Of Enamel

Introduction

AI has several presentations, specifically a Hypoplastic type, also known as Type I, is the most common [2]. Hypoplastic AI is characterized by thin or absent enamel due to deficient matrix formation, leading to yellowish-brown discoloration due to the visibility of underlying dentin, lack of contact between adjacent teeth, flat occlusal surfaces of the posterior teeth due to attrition, and might present with grooves or pits. Hypomaturation, also known as Type II, presents with normal thickness of enamel but improper maturation, resulting in a softened discolored enamel that is prone to wear and pulpal exposure. Hypocalcification, also known as Type III, normal enamel thickness and shape are present, but inadequate mineralization, present as soft, easily damaged enamel with a chalky, yellow, brown appearance. Hypomaturation-hypoplasia with taurodontism, also known as Type IV, is mixed-type AI. Mixed-type AI involves a combination of features from two or more traditional types of AI. Taurodontism is seen on molars with enlarged pulp chambers where the crowns of the teeth are developmentally larger than the roots which can be shortened in

length [3]. In addition to enamel malformation, skeletal changes, particularly in the maxilla and mandible, can also occur. About 24% of individuals with AI exhibit an anterior open bite based on research findings [4,5]. The occurrence of AI varies between 1 in 718 and 1 in 14,000, depending on the population being studied [6,7].

Case Report

A 19-year-old African American female presented with generalized sensitivity in her oral cavity and a severe localized pain in the upper left quadrant in the area of #13. She described a sharp, pulsating pain that was triggered by warm temperatures and the consumption of sweets. Her pain rating from a scale of one to ten was 9 out of 10. She reported the discomfort began the previous afternoon after consuming several pieces of candy and drinking coffee.

The patient had seen a primary care physician one year ago for a medical examination prior to enrollment into university. A comprehensive medical history revealed a diagnosis of Amelogenesis Imperfecta Type III at three years of age, which had been charac-

terized by generalized hypocalcified enamel with a brown-yellow discolored presentation and a pitted, rough texture of her deciduous teeth. The patient reported that as her deciduous teeth exfoliated and were replaced by eruption of her permanent dentition which also exhibited brown-yellow discoloration and the same pitted, rough texture which was seen on her teeth at three years old. Significant to note from the patient's dental history was that her teeth occasionally would slough and crumble necessitating having to expectorate pieces of tooth structure. Additionally, to note is due to financial constraints, the patient had been unable to see a dentist for two years. The patient was a smoker of cannabis for medicinal purposes to control anxiety and the consumption of cannabis had subsequently exacerbated digestive issues and GERD; her physician one year ago had prescribed over-the-counter antacids and recommended reduction of cannabis use as part of GERD management. Due to her daily generalized dental sensitivity, the patient reported that she had transitioned to a pescatarian diet in the hopes of alleviating her sensitivity and issues with mastication of harder foods such as chicken. The patient reported no allergies to any medication and her medical history was non-significant for any systemic diseases.

Two years ago, at her last examination, dental treatment at that time included multiple restorations and an extraction of tooth #3. She had expressed specific dental concerns of the generalized pitting and rough texture, the color and crumbling of her teeth due to AI to the dental provider. As a result, she was given the recommendation to see an oral pathologist and a dentist specializing in AI to receive a comprehensive dental examination to determine the best course of treatment.

Clinical Findings

Extraoral examination yielded a slight lymphadenopathy or swelling of the lymph glands of the left ear, neck and submandibular area and a warm feeling of the patient's facial skin indicative of an elevated temperature. The temporomandibular joint (TMJ) presented non-significantly and had no positive findings of clicking, popping, or deviations of the mandible in either left or right direction. Intraorally, bilateral well-circumscribed 2mm purple papules were noted on the lingual alveolar mucosa apical to canines #22 and #27. The patient presented with acute toothache likely originating from tooth #13 and exacerbated by Amelogenesis Imperfecta-related enamel defects. Additionally, tooth #13 presented with distolingual decalcification that flaked when touched by a Shepard's hook explorer. Additionally present was a periapical radiolucency (PARL) around the apex of tooth #13, accompanied by an existing mesial-occlusal-distal composite restoration with

visible secondary caries that was soft to the touch of the tine of a Shepard-hook dental explorer. This restoration had been placed when the patient was fourteen. The buccal and lingual cervico-gingival areas of #13 presented with erythema, redness and sensitivity to palpation. The patient had been unable to properly brush and floss this tooth since her pain started. The patient also reported feeling her occlusion "was off", and examination revealed that #13 had a mobility grade II, indicating possible enlargement of the periodontal space and inflammation of periodontal fibers in the socket of #13. Percussion proved painful for the patient, and a pulp vitality test was utilized to establish the pulpal health of #13. A Parkell pulp tester device, which uses electricity to send a low current of electricity through a tooth to test pulp vitality was used to test #13 and yielded a value of thirty-nine, indicating pulpal inflammation. Contralateral testing of control teeth #4 and #5 yielded values of three and four respectively, indicating healthy pulpal tissues. Cold testing of teeth #13, #4 and #5 using Endo Ice, an aerosolized refrigerant spray was applied to a cotton roll and applied to these teeth. The patient reported feeling the cold on teeth #13, #4, and #5 for two seconds only.

The lingual papules require further investigation via biopsy and consultation with an oral pathologist to rule out malignancy or varicose vein phenomenon.

At her dental visit at the Howard University dental clinic, clinical examination yielded multiple hypocalcified teeth exhibiting a brown-yellow decalcified discoloration on buccal, lingual and occlusal surfaces. Evident were eroded teeth that presented with flattened cusps and worn surfaces that affected teeth #6 to #11 and #21 to #27. Other observations included an anterior open bite though the patient had not reported thumb-sucking, possibly due to drifting of her teeth since having #3 extracted.

Radiographic findings

Radiographic examination indicated radiolucencies indicative of caries progression on teeth #2, #3, #4, #5, #15, #18 and #32. Tooth #1 was impacted and #16 was partially erupted. Existing full coverage indirect retainer stainless-steel crowns presented on teeth #19 and #30. The cortical bone around the mandibular teeth presented as very trabeculated with an opaque lamina dura lining the sockets of the teeth #23, #24, #25, #26, indicative of periodontal disease [8]. When this area was probed to assess the pocket depths in the areas, reading of 4mm were read interproximally. Trabeculated bone was present on mandibular anterior radiographs and developing vertical bone defects were also apparent. Posterior teeth in

the maxilla and mandible all presented with a horizontal bone loss radiographically, and presented with probing depths of 4mm and above.

Treatment plan

Immediate management included addressing the pain with analgesics and providing dietary counseling to minimize exacerbating factors such as mastication of hard foods in the area of #13. A comprehensive dental examination was recommended and consultations with the Oral Pathology, Endodontic, Periodontic, Prosthodontic and Orthodontic Departments were made to address treatment of #13. It was ascertained that her open bite should be evaluated in the Orthodontic department. Considerations for treatment of #13 included a treatment plan to undergo evaluation for immediate root canal therapy, crown buildup and porcelain-fused-to-metal crown. Another consideration to treat was to perform an occlusal plane evaluation and occlusal adjustment of existing restorations to manage the unbalanced bite the patient had reported. This option was an interim and not a long-term option for the patient. As part of managing this patient’s AI, active caries management was essential, and Prosthodontic consultation revealed the need to restore affected teeth #2,#3,#4,#5,#15,#18 and #32 with full coverage crowns.

The complete occlusal evaluation would serve to establish the correct occlusal plane for the patient, a necessary factor to establish if full coverage crowns are the proposed mode of treatment to establish the correct vertical dimension of occlusion (VDO) of the patient; the occlusal plane evaluation would also effectively establish her antero-posterior and mediolateral planes, given she had lost tooth structure as a result of her condition. The probing depths of 4mm anteriorly and posteriorly in the maxilla and mandible necessitated a complete periodontal examination, where each tooth would be assessed for attachment loss and a plan would be implemented for root planning and scaling to achieve gingival fiber reattachment to aid in improving periodontal health.

Concurrently, the discovery of the bilateral well-circumscribed 2mm purple papules on the lingual alveolar mucosa adjacent to canines #22 and #27 warranted an appointment with the department of Oral Pathology to establish a definitive diagnosis and rule out malignancy.



Figure 1



Figure 2



Figure 3



Figure 4

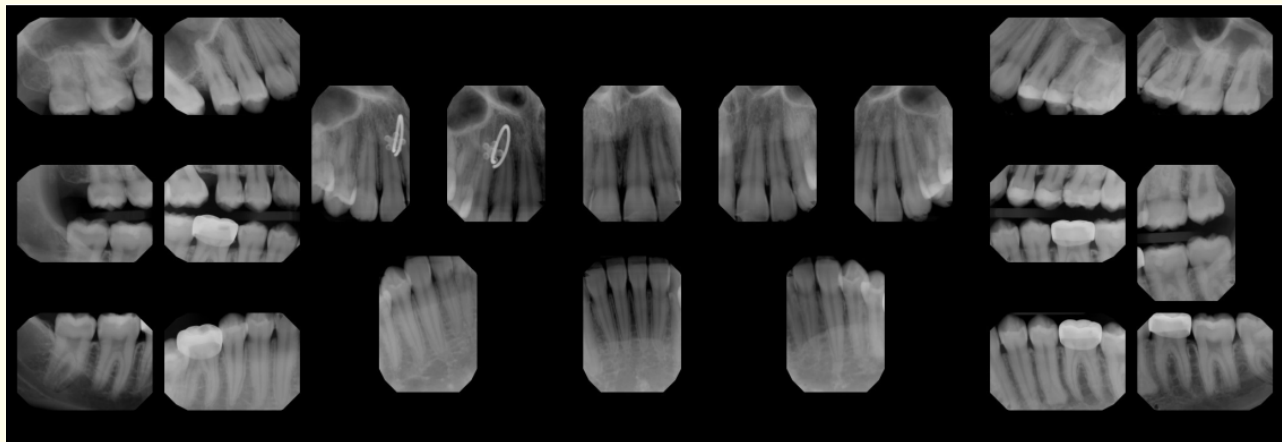


Figure 5: Full Mouth Radiographs

Conclusion

Long-term care for cases such as these must include six-month dental visits for preventive strategies, monitoring of AI-related enamel defects and potential complications such as pulpal encroachment from tooth structure destruction. A recommendation of routine interdepartmental consultation is necessary to monitor the function and integrity of any restorations, the longevity of fixed indirect retainer crowns, or stability of orthodontic brackets to assess periodontal health and tooth function as part of the patient's treatment. Emphasis on fluoride application every six months and meticulous oral hygiene habits such as flossing, using a chlorhexidine 0.12% prescription oral rinse and brushing twice daily are necessary for Amelogenesis Imperfecta cases. This approach seeks to alleviate acute symptoms of sensitivity and pain, preserve dental structure and function, and optimize oral health outcomes in the context of underlying genetic conditions such as AI.

The appearance of teeth and issues related to enamel, such as sensitivity, discoloration, and surface irregularities, can significantly impact both the psychological well-being and functional abilities of the patient [9,10]. The primary goal of any treatment plan is to achieve early detection, prevention, stabilization, restoration of enamel defects, and regular maintenance [11]. When rehabilitating Amelogenesis Imperfecta in adults, it is essential to consider factors such as dental and skeletal alignment, gum health, aesthetic outcomes, tooth vitality, and overall tooth appearance [12].

This case underscores the complex interplay of dental pathology in patients with amelogenesis imperfecta regardless of type, and highlights the importance of multidisciplinary collaboration for effective case management. A multidisciplinary approach in the

management of AI necessitates comprehensive care that encompasses both dental restoration and psychological support [13,14]. Long-term follow-up is critical for monitoring treatment outcomes and addressing evolving dental needs [15]. Regular dental visits allow for ongoing evaluation of enamel integrity, detection of caries or other dental complications, correction of malocclusion through orthodontic treatment, adjustments to prosthetic devices as needed, and reinforcement of oral hygiene practices [16,17].

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