ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 2, Issue – 2, March - April - 2019, Page No. : 460 - 463

Amelogenesis Imperfecta- Hypoplastic Variant

[1]Dr. Abhishek Verma, Oral and Maxillofacial Surgery, Private Practioner, Ghaziabad

[2]Dr.Nitin Kumar Moral, Dept. of Oral And Maxillofacial Surgery, Assistant Professor, IP Dental College, Ghaziabad, Uttar Pradesh

[3]Dr. Nidhi Moral, Dept. of Endodontics, Assistant Professor, Sharda School Of Dental Sciences, Sharda University, Greater Noida, Uttar Pradesh

[4]Dr. Sabourni Sarkar, Post Graduate Student, Department of Paediatrics And Preventive Dentistry, Institute Of Dental Sciences, Bareilly, Uttar Pradesh

[5]Dr. Anmol Bagaria,Private practioner, Mumbai

Corresponding Author: Dr. Sabourni Sarkar, Post Graduate Student, Department of Paediatrics And Preventive Dentistry, Institute Of Dental Sciences, Bareilly, Uttar Pradesh

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Amelogenesis Imperfecta (AI) is a developmental disorder which is hereditary in origin, affecting the formation and mineralization of enamel of one or all the teeth. The mode of inheritance for AI trait may be autosomal dominant, autosomal recessive or X-linked. The variants of amelogenesis imperfecta include hypoplastic, hypomaturative and hypocalcified based on the primary enamel defect. This article intends to present a case of amelogenesis imperfecta in a 8 years old young girl.

Keywords: Amelogenesis Imperfect, Developmental Disorder, Hereditary Disease

Introduction

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.[1] The prevalence of the disease varies from 1:700 to 1:14000, according to the populations studied.[2] Hereditary brown enamel, hereditary enamel dysplasia, hereditary brown opalescent teeth are few other synonyms used for AI. Amelogenesis Imperfecta involves the ectodermal component of teeth and can affect both the primary and permanent dentition.[3] The dentine and roots of the teeth affected by AI appears to be normal with clinical, radiographic, and histopathologic defects seen in the enamel portion of the teeth. The classifications of amelogenesis depend on the mode of inheritance and phenotypic characteristics of the involved tooth. The most commonly used classification of AI was proposed by Witkop in the year 1988, which was later revised by Nusier in 2004.[4] Hypoplastic, hypomaturative, hypocalcified, and hypomaturation-hypoplastic are the four types of AI, depending upon the appearance, structural and developmental defects of enamel.[5] About 60-73% of all the cases of amelogenesis imperfecta are the hypoplastic type, hypomaturation type represents 20-40% and 7% is the hypocalcified variant of amelogenesis imperfecta.[4] No racial predilection is seen in cases of

amelogenesis imperfecta.[6] The treatment aims to relieve any pain or sensitivity in the tooth, so as to prevent any further tooth loss.[7] Here, we report a case of amelogenesis imperfect in an 8 year old girl whose chief concern was the unesthetic appearance of her teeth.

Case report

An 8 year old had reported to the department of oral medicine and radiology with the chief complaint of yellow stains on all her teeth. Also, sensitivity to hot and cold things was present in all the teeth. The parents of the patient revealed that these yellow stains on her teeth are present since early childhood. The medical history and the hereditary involvement of the disease was non-contributory. The extra-oral findings were non-significant. The intra-oral examination revealed generalized yellowish discoloration of teeth along with diffuse pitting, seen more prominently on the labial and buccal aspect of the teeth. The surfaces of the teeth were rough and irregular in shape. Dental caries was probed in the tooth 65 and 85. The probing resistance was felt while palpating the teeth. The tooth material was soft in consistency with mild flaking of residual enamel. (figure 1) Multiple intra-oral periapical radiographs were done to image all the teeth present. The radiographs revealed generalized thinning of enamel surface along with absent enamel at some regions. (figure 2) Based on history, clinical findings, and radiologic examination, the diagnosis was made as Amelogenesis Imperfecta- hypoplastic type. The patient was advised for oral prophylaxis and to wait for all the permanent teeth to erupt.

Discussion

At the time of organogenesis, the transition of enamel takes place from a soft and pliable tissue that is almost devoid of protein to its final form. Various unique molecular and cellular activities are reflected in the final composition of enamel that takes place during its genesis.[8] The specific enamel proteins include Enamelin gene (ENAM), Amelogenin gene (AMELX), Kallikrein 4 gene (KLK4), Matrix Metalloproteinase 20 gene (MMP-20), and Distal-less homeobox 3 gene (DLX3).[9] The aetiologic factors include genetic, febrile illness or vitamin deficiency, local infection or trauma, fluoride ingestion, congenital syphilis, birth defects or idiopathic factors.[10] The pathogenic disease process of AI is described in figure 3.[11] Witkop and Sauk classified amelogenesis imperfect as given in figure 4.[6] All the clinical and radiographic appearances of the three variants of amelogenesis imperfecta, i.e., hypoplastic, hypomaturative, and hypocalcified form are represented in table 1.[10]

Dental care for AI cases can be challenging and protracted.[12] The primary goal for the treatment is to address each concern as it presents with an overall comprehensive plan that outlines anticipated future treatment needs. Clinician treating children and adolescents with AI must understand and solve the clinical and emotional demands of these disorders with sensitivity. It is important to establish good rapport with the child and the family. Timely intervention is critical to spare the patient from psychosocial consequences of these disfiguring conditions. A comprehensive and prompt approach is reassuring to the patient and family and may help decrease their anxiety.[5]

Conclusion: One of the greatest challenges faced by the clinician is the total rehabilitation with amelogenesis imperfecta. Dental practitioners should also consider the social implications for these patients, as it affects the psychology of the patient and intervene to relieve their suffering. The patient should be counselled and motivated to maintain the oral hygiene, which goes a long way in maintaining the dentition.

© 2019 IJDSIR, All Rights Reserved

References

1. Aldred MJ, Crawford PJM, Savarirayan R: Amelogenesis imperfect – a classification and catalogue for the 21st century. *Oral Dis* 2003, 9:19-23.
2. Crawford PJ, Aldred M, Bloch-Zupan A. Amelogenesis imperfecta. Orphanet J Rare Dis. 2007;2:17.
3. Roma M, Shreya H. Amelogenesis Imperfecta: A review of the literature. J.Pharm. Sci. & Res. 2016; 8(9): 1042-44
4. Mayur Chaudary, Shweta Dixit, Asha Singh, Sanket Kunte. "Amelogenesis imperfecta" – Report of a case and review of literature. J Oral Maxillofac Pathol 2009;13(2):70-7.
5. Saraswathi V Naik, Ghousia S, Shashibushan KK, Poornima S. Amelogenesis Imperfecta- A Case Report. J Oral Health Res. 2011;2(4):106-110.
6. Anar Patel et. al. : Amelogenesis Imperfecta. The Journal of Ahmedabad Dental College and Hospital. 2011; 2(1).
7. Nigam et al. Amelogenesis Imperfecta: A case report and review of literature. IJSS. 2015; 2(10)
8. Paine ML, White SN, Luo W, Fong H, Sarikaya M, Snead ML. Regulated expression dictates enamel structure and tooth function (review) Matrix Biol. 2001;20:273–92.
9. Sockalingam S. Dental rehabilitation of amelogenesis imperfecta using thermoformed templates. J Indian Soc Pedod Prev Dent. 2011;29:53–61
10. Witkop C. J. Amelogenesis imperfecta, dentinogenesis imperfect and dentin dysplasia revisited: problems in classification. Jr. J Oral Pathol. 1988 Nov;17(9-10):547-53.
11. Emin Murat Canger, Peruze Celenk, Murat Yenisey, Selcen Zeynep Odyakmaz. Amelogenesis Imperfecta, Hypopalstic Type Associated with some Dental Abnormalities: A Case Report. Braz Dent J. 2010;21(2):170-74.
12. Suchancova B, Holly D, Janska M, Stebel J, Lysy J, Thurzo A et al. Amelogenesis Imperfecta and the treatment plan- interdisciplinary team approach. Bratisl Lek Listy. 2014; 115: 44-8

Legends Figures and Table

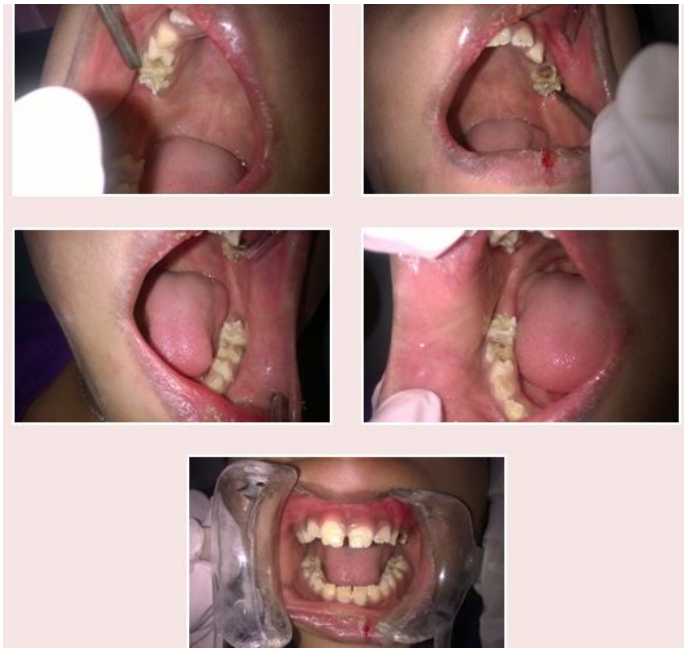

Figure 1: the clinical image of the affected teeth

Figure 2: the intraoral radiographs of the affected teeth

© 2019 IJDSIR, All Rights Reserved

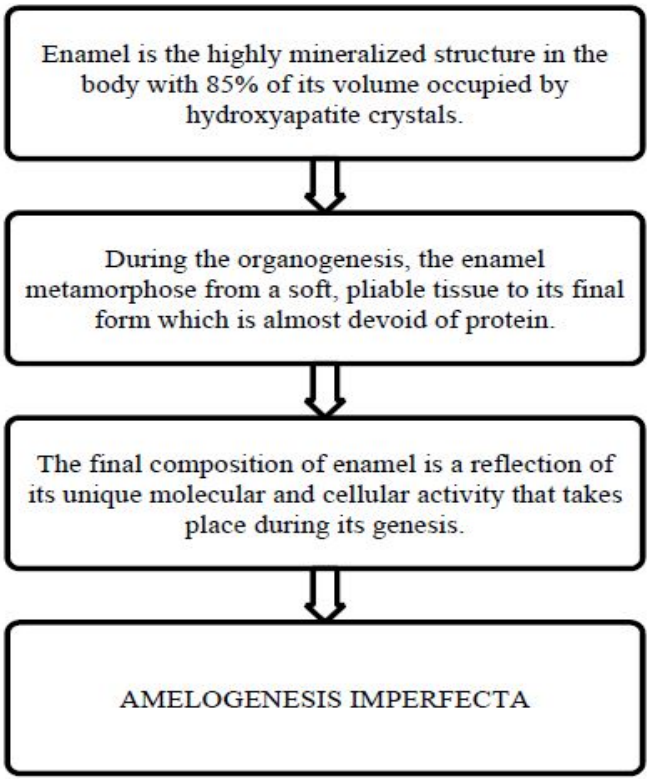

Figure 3: the pathogenesis of amelogenesis imperfecta

Type I hypoplastic	
IA	Hypoplastic, pitted autosomal dominant
IB	Hypoplastic, local autosomal dominant
IC	Hypoplastic, local autosomal recessive
ID	Hypoplastic, smooth autosomal dominant
IE	Hypoplastic, smooth X-linked dominant
IF	Hypoplastic, rough autosomal dominant
IG	Enamel agenesis, autosomal recessive
Type II hypomaturation	
IIA	Hypomaturation, pigmented autosomal recessive
IIB	Hypomaturation
IIC	Snow-capped teeth, X-linked
IID	Autosomal dominant?
Type III hypocalcification	
IIIA	Autosomal dominant
IIIB	Autosomal recessive
Type IV hypomaturation hypoplastic with taurodontism	
IVA	Hypomaturation hypoplastic with taurodontism, autosomal dominant
IVB	Hypoplastic hypomaturation with taurodontism, autosomal dominant

Figure 4: classification of amelogenesis imperfecta

Hypoplastic form Reduction in the thickness of enamel matrix with normal mineralization	**Hypomaturation form** Defect in the mineralization process with normal matrix formation	**Hypocalcified form** Defect in the quality of the mineralization process with normal quantity of matrix formation
Clinical Appearance		
Reduced thickness of enamel	Normal thickness of enamel	Normal thickness of enamel with loss of translucency
Enamel appears normal and less prone to attrition	Enamel is hypomineralized and prone to attrition	Enamel is hypomineralized and exhibits a soft cheesy consistency. Easily broken down.
The color appears normal with translucency to a yellow to dark brown color depending on the thickness of enamel and dentin	Color may be affected by staining from the oral environment. Mottled appearance to yellow-brown or red-brown discoloration	Color may be affected by staining from the oral environment. Teeth appear more dark
Reduction in tooth size		
Rough, irregular or pitted enamel		
Radiographic appearance		
Enamel and dentin appears normal	Enamel has similar radiodensity as dentin	Enamel is less radiopaque than dentin

Table 1: the clinical and radiographic appearance of different forms of AI

© 2019 IJDSIR, All Rights Reserved