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Case Report

Hypoplastic Amelogenesis Imperfecta: Report of Three Cases

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ABSTRACT:
Amelogenesis imperfecta is a genetic linked hereditary disorder affecting enamel. The formation of defective enamel matrix leads to increased tooth discoloration, cracking and loss of tooth structure. Complaints of increased sensitivity, loss of esthetics and function are frequently reported in these cases. A holistic dental treatment comprising of preventive, restorative and prosthetic treatments is required for improved esthetics, function and quality of life in these patients.
Key words: Amelogenesis imperfecta, enamel hypoplasia, tooth discoloration.

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INTRODUCTION

Amelogeneis Imperfecta (AI) is a genetic disorder which leads to formation of defective enamel matrix. The most common inheritance pattern is autosomal dominant, however few cases of autosomal recessive and x-linked have also been reported.[1] AI was first reported in 1938 by Finn as " hereditary opalescent dentin" due to its appearance.[2] Based on the mode of inheritance, clinical, radiographic and histological features AI has been categorized into four types which are further divided into subtypes. Four major types are:

a) Hypoplastic – It results due to defective formation of enamel matrix. It is characterized by thin layer of enamel yellowish-brown in color, square crowns with flat occlussal surfaces and absent contact areas.

b) Hypocalcified – It is due to defect in matrix and mineralization on enamel. In this crown structure is normal but enamel is soft and wears down easily becoming dark-brown in color.

c) Hypomaturation – It is outcome of altered enamel rod and rod sheath structure. Enamel is harder from hypocalcified type but breaks away from crown structure; it can be mottled, yellow, cloudy white or brown in color.

d) Hypoplastic-Hypomaturation – In this enamel is thin, teeth are mottled and show taurodontism with enlarged pulp chambers.[3]

The prevalence of AI ranges between 1 in 718 to 1 in 14,000, depending upon the study population. Hypoplastic AI accounts for 60 - 73% of all AI cases, hypomaturation is 20 - 40%, and hypocalcification is only 7%.[4]

AI can be associated with irregular dental eruption, anodontia, anterior open bite, pulp calcification, dentin dysplasia, resorption of crown and root, hypercementosis, malformation of roots. The main concern in treating cases of AI is restoration of esthetics and function; however differentiating AI from other enamel hypoplastic defects remains a challenge.

The following case reports AI in a female patient which was later traced in her other two siblings.

CASE REPORT

A 16 year old female patient came to the department with complaint of brown stains on her teeth since childhood. She also reported chipping of teeth in posterior region. Her parents gave history of her discolored deciduous teeth and also similar stains in her younger brother and sister. There was no history of systemic disease and patient was in a healthy condition. On examination there was yellowish-brown discoloration of anterior teeth.[fig.1, fig. 2]

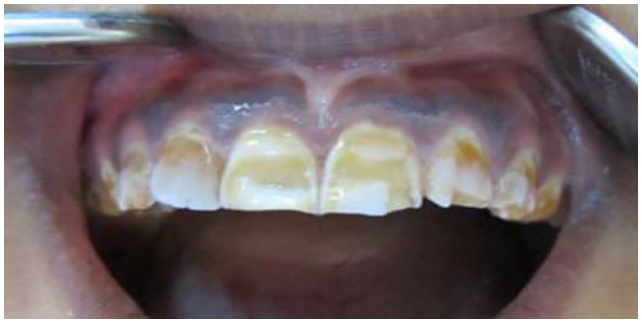

Fig.1 Discolored maxillay anterior teeth

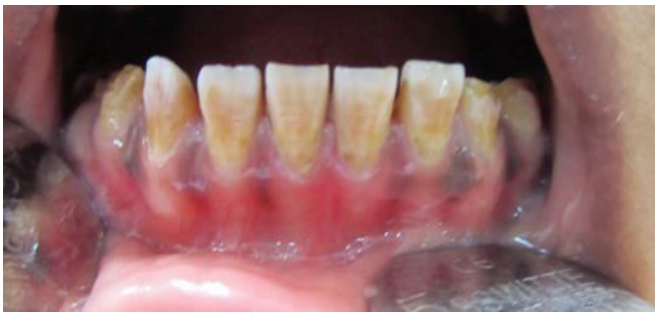

Fig.2 Discolored mandibular anterior teeth

Severe attrition was seen in posterior teeth with 46,36 being carious.[fig.3]

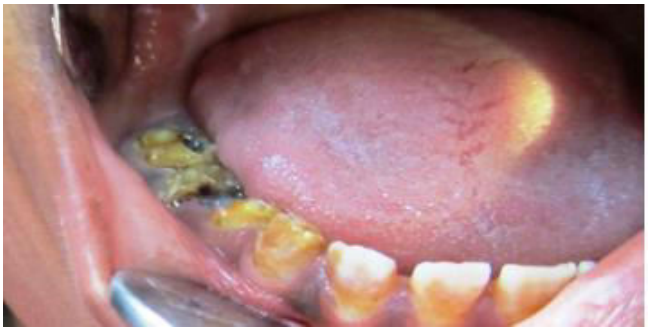

Fig.3 Right mandibular occlussal view

Enamel was hard in consistency and chipping was not present on probing. Orthopantomogram showed thin layer of radiopaque enamel. [fig.4]

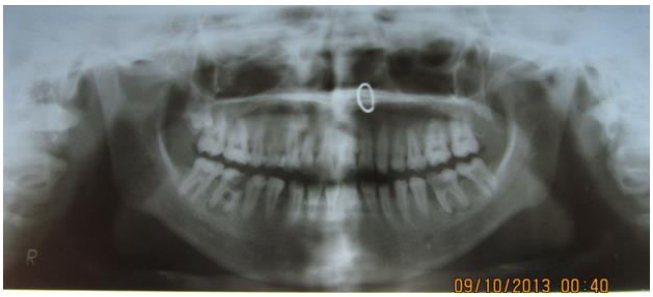

Fig.4. Orthopantomogram showing thin radiopaque enamel layer

On the basis of history, clinical and radiographic findings final diagnosis of hypoplastic AI was made. Patient's younger sister [fig.5, fig.6] and brother were also diagnosed with hypoplastic AI [fig.7, fig.8].

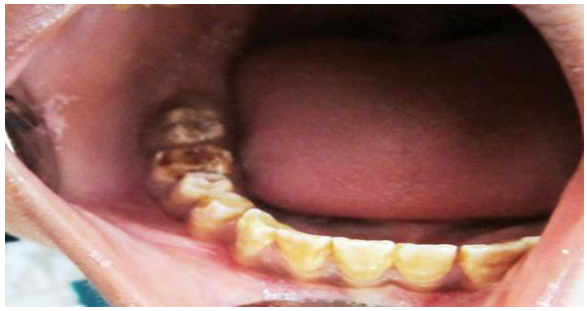

Fig.5 Patient's sister having discolored mandibular anterior teeth and mottled posterior teeth

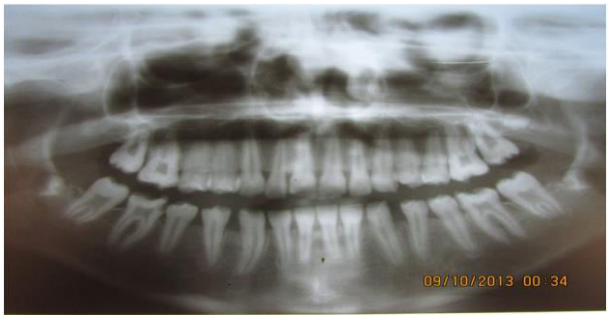

Fig.6 Orthopantomogram of patient's sister showing thin radiopaque enamel layer.

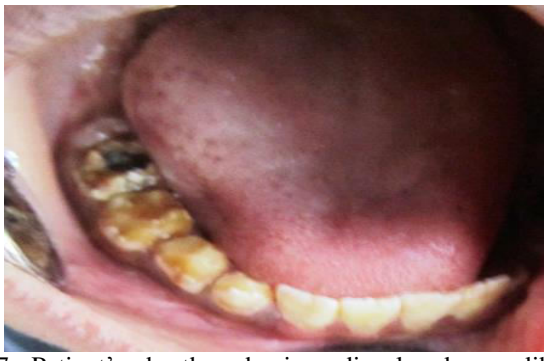

Fig.7 Patient's brother having discolored mandibular anterior teeth and mottled posterior teeth

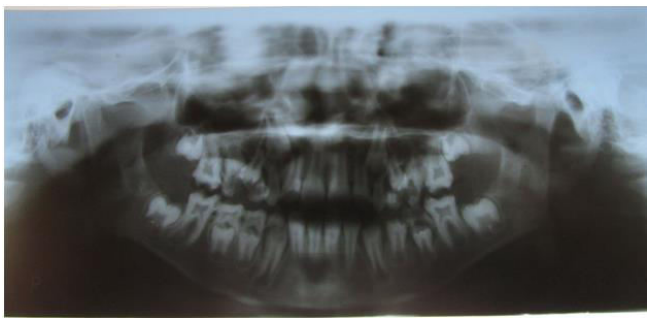

Fig.8 Orthopantomogram of patient's brother showing thin radiopaque enamel layer.

Restoration and prosthetic rehabilitation for correction of esthetic and function was done in all three patients.

DISCUSSION

Amelogenesis Imperfecta refers to a clinical and genetic heterogeneous set of conditions which affect the dental enamel. It might also affect other dental, oral or extra-oral tissues. [5] Clinically enamel hypoplasia due to hypomaturation or hypomineralization of enamel is present in all the types and subtypes of AI. Genetically mutation in coding genes for enamel protein has been implicated in its pathogenesis. Alteration of genes like Enamelin (ENAM), Ameloblastin (AMBN), Amelotin gene (AMELOTIN 4q13),Amelogenin (AMELX), Kallikrein 4 (KLK4), Matrix Metalloproteinase 20 (MMP-20), and Distal-less homeobox 3 (DLX3) have been associated with AI.[1,6]

AI affects both the dentitions primary as well as permanent with permanent incisors and molars more commonly affected than other teeth which was seen in all of our three patients.

Treating AI includes a holistic dental treatment encompassing preventive, restorative and prosthetic procedures. The frequent complaints of patients having AI are unaesthetic appearance, increased sensitivity, loss of tooth structure and vertical dimension. Preventive measures like application of fluoride, oral prophylaxis and dietary changes are advised. Restorative and prosthetic procedures like composite and glass ionomer restorations, fabrication of fixed partial dentures, overdentures, porcelain fused or metal crowns, assist in restoring esthetics and function in these patients.[3]

CONCLUSION

Amelogenesis imperfecta significantly impacts the quality of life in affected patients. Dental surgeons have a vital role in prompt diagnosis, minimizing loss of tooth structure and restoration of lost aesthetics and function in these patients.

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