AMELOGENESIS IMPERFECTA : A CASE REPORT

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ABSTRACT

Amelogenesis imperfecta (AI) is a diverse collection of inherited diseased that exhibit quantitative or qualitative tooth enamel defects in the absence of systemic manifestations. Also known by varied names such as Hereditary of enamel dysplasia, Hereditary brown enamel, Hereditary brown opalescent teeth, this defect is entirely ectodermal, since mesodermal components of the teeth are basically normal. The AI trait can be transmitted by either autosomal dominant, autosomal recessive, or X-linked modes of inheritance. It is necessary to diagnose the case and provide durable functional and esthetic management of these patients, where the unaesthetic appearance has a definite negative psychological impact.

KEY WORDS: Amelogenesis imperfecta, enamel, dental, genetic.

INTRODUCTION

Amelogenesis imperfect (AI) encompasses a heterogeneous group of developmental disorders that demonstrate alterations in the enamel.[1] It is characterized by clinical and genetic heterogeneity in the absence of systemic abnormalities or diseases.[1,2] AI is caused by mutations in genes that control amelogenesis and follows inheritance patterns of autosomal-dominant, autosomal recessive or X-linked modes of transmission.[1-3] There are also patients for whom a family history cannot be identified but where a mutatin is present.[3] The enamel defects of this condition are clinically divided into hypomineralized and hypoplastic forms. Both primary and permanent dentitions are usually affected.

Clinically, AI appears as an alteration of enamel formation resulting in hypoplasia, hypocalcification, and hypomaturation. Enamel hypoplasia results in a decreased quantitative enamel formation. The enamel in hypocalcification appears normal but poorly mineralized while hypomaturation results in an abnormal mineralization in the final stages of tooth formation[2,3]. The most common form, the hypoplastic type, is deficient in normal enamel. The crowns of the teeth appear blanched, snowcapped, yellow-brown, pitted, or grooved. Radiographic examination usually shows a full complement of teeth, but the crowns of the teeth either have very thin enamel or lack enamel completely.[1,3-5]

Review of Literature

Amelogenesis imperfecta (AI) encompasses a complicated group of conditions that demonstrate developmental alterations in the structure of the enamel in the absence of a systemic disorder.[5]

The prevalence of this conditions has been expected to range from 1 in 718 to 1 in 14,000, depending on the population studies. Hypoplastic AI represents 60-73% of all cases, hypomaturation AI represents 20-40%, and hypocalcification AI represents 7%.[6]

Weinmann et al.,1945, subdivided amelogenesis imperfecta into hypoplastic and hypocalcified types.[7] Several classifications have evolved since then, with at least ten subtypes, characterized by clinical features and mode of inheritance. Two X-linked phenotypic variants of amelogenesis imperfecta have been included in these classifications – a hypoplastic form and a hypomaturation form.

Witkop and Sauk listed the varieties of AI, divided according to whether the abnormality lay in a reduced amount of enamel (hypoplasia), deficient calcification (hypocalcification), or impercect maturation of the enamel (hypomaturation), and also recognized the combined defects.[8]

CASE REPORT

A 17 Year-old Female patient presented with the chief complaints of spacing in between teeth. She wanted replacement of her teeth. She had also difficulty in chewing and displeasure with her present dental appearance. A detailed medical, dental, and social history was obtained. The enamel of all teeth was hypoplastic and yellow-brown in color. The surfaces of the teeth were rough, and the enamel was either not visible or very thin over the crowns of all teeth. The dentin, where it was exposed, was brown and hypersensitive.

Past dental history revealed that her deciduous teeth were also similaryly discolored and there was delayed eruption. She had undergone root canal treatment for teeth 11,21 six months back. Apart from this, her past medical history was noncontributory.

On intraoral examination, it was found that she had missing 17, 18, 23, 27, 28, 33, 37, 38, 43, 47, 48. The thickness of enamel was reduced on the teeth and was completely chipped off from some teeth exposing the dentin. The surfaces of the teeth were rough. The teeth, in general, exhibited a yellowish brown discoloration, with diffuse pitting present on the exposed tooth surfaces, more prominent on the labial and buccal aspects. No open bite was present Examination of the periodontium revealed the presence of chronic, generalized, marginal, and papillary gingivitis, with calculus deposition and unsatisfactory oral hygiene. (Fig. 1 and 2)

Radiographic investigations included an orthopantomogram (OPG). The OPG showed the presence of unerupted 17, 18, 23,24,27, 37,38, 43, 47, 48. (Fig. 3)

The diagnosis of hypoplastic, rough, autosomal dominant AI was confirmed on the basis of typical, clinical, radiographic features.

Esthetics along with functional limitations were the reason the patient's parents brought her to the hospital for treatment. The treatment proposed for her ranged from Root canal treatment, Extraction of impacted 23,43, crown lengthening, bonded veneer restorations to tooth preparations for the placement of a full crown.

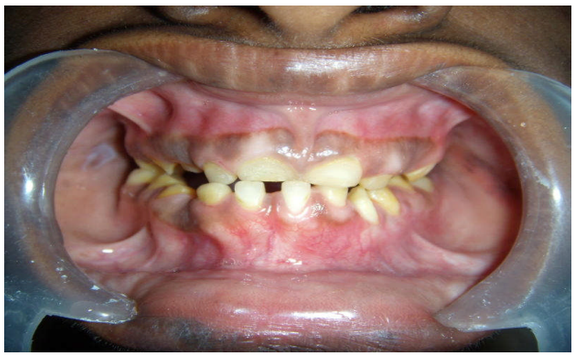

Figure : 1

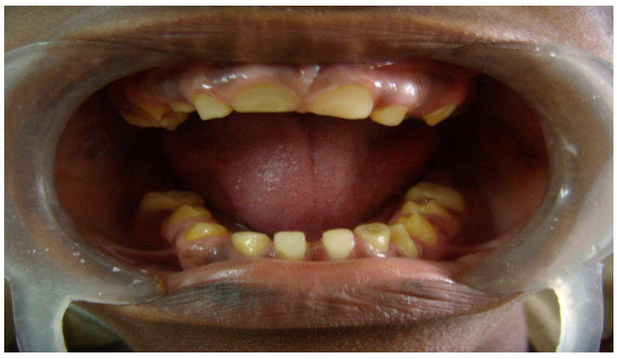

Figure : 2

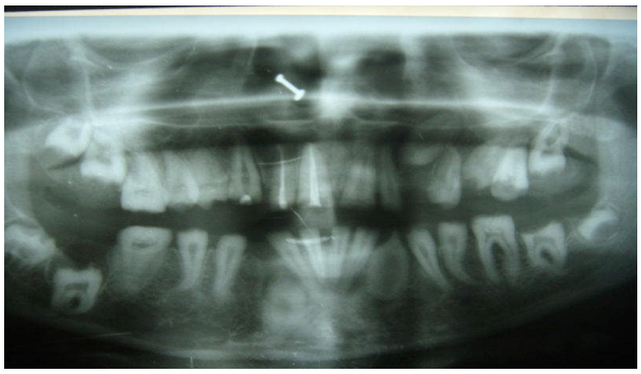

Figure : 3

DISCUSSION

Amelogenesis imperfecta is a developmental, often inherited disorder, affecting dental enamel. It usually occurs in the absence of systemic features and comprises of diverse phenotypic entities.

The predominant clinical manifestations of affected individuals are enamel hypoplasia (enamel is seemingly correctly mineralized, but thin), hypomineralization (subdivided into hypomaturation and hyocalcification), or a combined phenotype, which is seen in most cases. [8,9,10]. The trait of AI can be transmitted by an autosomal-dominant, autosomal-recessive, or X-linked mode of inheritance.[11,12]

CONCLUSION

The objective of esthetic dentistry is to treat diverse problems and achieve natural appearing results. The treatment of patients with amelogenesis imperfecta presents an interesting challenge to the restorative dentist. The main clinical characteristics are extensive loss of tooth tissue, poor aesthetics, and tooth sensitivity.[2,13,14] The treatment plan for patients with AI is related to many factors including the age of the patient, the socio-economic status, the type and severity of the disorder and its intra oral manifestation.[15,16] The complexity of the management of patients with AI should start with early diagnosis to prevent restorative problems at a later stage.

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