Case Report

Restoration of function and esthetics in a patient with amelogenesis imperfecta

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ABSTRACT

Amelogenesis imperfecta (AI) is a rare genetic disorder inherited as either autosomal dominant, autosomal recessive, or X- linked modes. AI is characterized by the defect in enamel formation, resulting in defective mineralization or formation of the matrix. Restoration of the dentition poses great challenges, especially when teeth are severely affected. Treatment aims to relieve pain or sensitivity of teeth and to preserve as much tooth structure as possible while preventing further loss of teeth, to maintain masticatory function, and last but not the least, to improve the aesthetic appearance and quality of life as it has a great psychological impact on the self-esteem of the patient. This case report describes the treatment of a young boy who presented with a severe form of Alimperfect (AI) with a minimally invasive, relatively simple, and cost-effective treatment with composite restorations.

Key words: Amelogenesis imperfecta, hereditary composite restorations, enamel

INTRODUCTION

Amelogenesis imperfecta (AI) is used to describe a number of conditions resulting from defective formation of the enamel. Many cases are inherited, either as autosomal dominant, recessive, or as X-linked traits. There have been a number of classifications, based primarily on the phenotype.[1-3] Tooth enamel is the most highly mineralized structure in the human body, with 85% of its volume occupied by hydroxyapatite crystals.[4,5] The physical properties of enamel are directly related to the composition, orientation, disposition, and morphology of the mineral components within the tissue.[6] During organogenesis, the enamel transitions from a soft and pliable tissue to its final form, which is almost entirely devoid of protein.[4] However, a recent classification based on the molecular basis for the observed enamel phenotype may help clarify the commonly seen overlap in phenotypes. According to this classification, AI is a group of conditions, genetic in origin, which affect the structure and clinical appearance of the enamel of all, or nearly all the teeth, and which may be associated with morphologic or biochemical changes elsewhere in the body.[7] This anomaly of the enamel affects both primary and permanent dentition.[7-10]

The appearance depends on the type of AI, varying from the mild hypo mature 'snow-capped' enamel to the more severe hereditary hypoplasia with thin enamel which has a yellow-brown appearance, resulting in dental problems like: sensitive teeth, poor aesthetic appearance and reduced masticatory function. Patient may develop gingivitis and periodontitis later because of cleaning difficulties arising due to roughness of enamel surface and pulpal involvement may also occur in severe cases. As AI affects both primary and permanent dentition, prevention of future dental problems must start from the beginning by educating the patient or parents regarding oral hygiene and reviewing at regular intervals. Attention must also be paid to the psychological health of the patient.[11]

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CASE REPORT

An 11-year-old boy reported to the Department of Pedodontics and Preventive Dentistry of H.P. Government Dental College and Hospital; Shimla, Himachal Pradesh with the chief complaint of discolored teeth since childhood. Apart from this his past medical and family history was nonsignificant. On intra-oral examination, it was found that he had a normal complement of teeth. The thickness of enamel was reduced to a great extent and was almost completely chipped off from some teeth, exposing the dentin. The surfaces of the teeth had a rough texture exhibiting a

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yellowish brown discoloration, with diffuse pitting present on the exposed surfaces of the teeth, more prominent on the labial and buccal aspects. The pattern of emergence and timing of shedding of teeth seemed to be delayed than for the normal range. An open bite was present. Examination of the periodontium revealed the presence of chronic, generalized, marginal, and papillary gingivitis, with the deposition of calculus and unsatisfactory oral hygiene [Figure 1].

A provisional diagnosis of hypoplastic type ofAIwas made on the basis of typical, clinical, radiographic, and histological features, along with a differential diagnosis from environmental enamel hypoplasia, dentinogenesis imperfecta, dentin dysplasia, regional odontodysplasia, and the tricho-dento-osseous syndrome.

Radiographic investigations included an orthopantomogram (OPG) and full-mouth intraoral periapical (IOPA) radiographs. The OPG showed the presence of all unerupted permanent canines, premolars, and second molars [Figure 2].

DISCUSSION

AI is a rare inherited disorder, ranging from a defect in the formation of the enamel to the coexistence of other medical disorders.[12-14] Both the primary and permanent dentitions are affected. In this case, the patient presented with severe tooth wear due to inherited abnormal development of the enamel. The poor appearance was due not only to the innate color of the teeth, but also to increased staining, chipping, and spacing between the teeth. Treatment of such patients with AI requires special care. They are sometimes psychologically affected due to the poor appearance of their teeth, and esthetic improvement can affect their self-confidence and esteem. In general, little is known about both the etiology of AI and the appropriate dental materials used to restore tooth defects. An appropriate selection of restorative materials is important to preserve function and occlusal harmony and to restore the natural dentition morphology. Resin composites, glass ionomers, modified glass ionomer cements, or a combination of these materials are recommended for restoring developmental dental enamel defects. Besides the esthetic properties of these materials, their adhesion to the structure of the tooth is an advantage that may avoid the unnecessary removal of sound teeth. In this case, it was clearly observed after esthetic restorations with the help of polycarboxylate crowns. Owing to the highly discolored dentine, a more opaque ceramic material had to be used. Although the crowns could have been improved further, as he was contended and were acceptable to him [Figure 3]; thus, no further alteration was done. The treatment required the commitment of a very long period, which was not possible as it would mean visits to the hospital for the patient from a far-flung area of the hilly state.

During the treatment period, oral hygiene and dietary advice were reinforced so that future risk to the periodontium and development of dentalcaries could be prevented to

Figure 1: Periodontium revealed the presence of chronic, generalized, marginal, and papillary gingivitis with calculus deposits and yellowish brown discoloration of teeth with diffuse pitting present on tooth surfaces

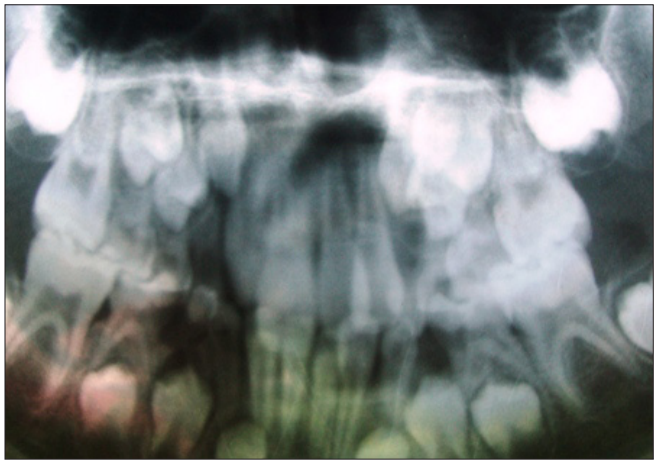

Figure 2: An orthopantomogram (OPG) showing the presence of all unerupted canines, premolars, and second molars

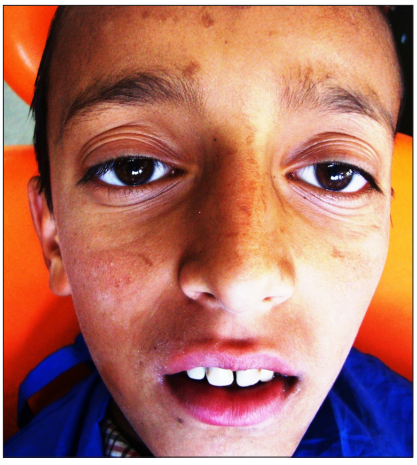

Figure 3: Postoperative photograph, showing markedly improvement in esthetic appearance of the patient

achieve long-term success goals. In AI with the esthetic improvement, psychological health improves to restore quality of life.[15]

CONCLUSION

Unfortunately, some patients seek clinical advice only when they present with problems like: poor esthetics, sensitivity of teeth, and gingivitis. These are the most common problems related to AI patients. Rather I would reiterate that, more complex problems like loss of occlusal vertical dimension and skeletal open bite should also be taken into consideration. In nutshell this case report demonstrates a minimally invasive, relatively simple, and cost-effective option for the correction of hypoplastic AI with composite restorations.

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