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Tackling Unfortunate Legacy: Full Mouth Rehabilitation of Amelogenesis Imperfecta

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ABSTRACT – *Amelogenesis imperfecta (AI) is a developmental disturbance which interferes with the normal enamel formation of both primary and permanent dentitions, leading to functional as well as esthetic inadequacies. Diagnosis is based on the family history, pedigree plotting and meticulous clinical observations. So, early recognition followed by appropriate preventive care and oral rehabilitation is essential in the successful management of AI. This clinical report discusses the oral rehabilitation of 14 year old girl diagnosed with hypocalcified autosomal dominant type of amelogenesis imperfecta. A complex set of problems like decreased occlusal vertical height, caries, tooth sensitivity and psychosocial problems related to poor aesthetics were encountered. The treatment protocol followed was mainly focused on aesthetics, functional rehabilitation and also to develop a positive psychological attitude in the patient. This case report depicts the multidisciplinary approach of full mouth rehabilitation in adolescents with AI.*

Key words-- *Full mouth rehabilitation, Amelogenesis Imperfecta(AI), Hypocalcified Enamel, Stainless steel crowns, Heat cure acrylic crowns, Cross mounting.*

I. INTRODUCTION

Amelogenesis imperfecta (AI) refers to a group of hereditary disorders characterized by defective formation or calcification of enamel. It affects both the primary and permanent dentitions1. Prevalence of AI varies from 1:700 to 1:14,0002. According to Witkop, AI can be classified as hypoplastic, hypomaturation, hypocalcified, and hypomaturation-hypoplastic with taurodontism. When clinical features and inheritance pattern are taken into consideration, 15 subtypes of AI can be distinguished3. AI trait can be either autosomal dominant, autosomal recessive or X- linked mode of inheritance and it may also be transmitted by consanguineous marriages. On the other hand it have been reported that AI may develop when a mutation occurs in the genes4. There are numerous challenges associated with the management of AI patients. Although AI primarily affects enamel formation, but these patients are mainly affected by their poor esthetics, compromised chewing function due to tooth sensitivity, decreased occlusal vertical dimension due to loss of tooth structure and the short clinical crowns caused by attrition and/or incomplete eruption3. Timely and comprehensive intervention is critical to spare the patient from psychological consequences of these disfiguring conditions.

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The treatment plan for AI patients is governed by multiple factors such as patient's age, his socioeconomic status, type and severity of the disorder and the intra-oral condition at the time of presentation5. This clinical report outlines the management of a case of amelogenesis imperfecta with a multidisciplinary approach, to provide aesthetics, function and to avoid vertical dimension loss and hypersensitivity.

Clinical report:

A 14 yr old girl reported to the clinic with a chief complaint of unsatisfactory appearance while smiling and also complaints of sensitivity to hot and cold beverages and reduced chewing ability. A detailed medical, dental and family history was obtained. The family history revealed that similar discoloration of teeth was observed in her mother and also there was a history of consanguineous marriage. No abnormalities were detected on extra oral examination [Figure 1- a,b]. Smile evaluation was done and an average smile line was observed [Figure 2]. On intraoral examination, all the teeth showed yellowish brown discoloration. The incisal edges were thin and attrited and the cuspal structures were also attrited. Tooth surfaces were found to be rough and the enamel layer was either not visible or very thin over the crowns of all the teeth. The dentin was exposed, teeth are brown and premature loss of vertical dimension was noticed. Carious lesions were noticed on the occlusal surfaces of all the upper molars and on the lower second permanent molars. [Figure 3-a,b,c,d,e]. Provisional diagnosis was made as amelogenesis imperfecta.

Radiographic investigations included an orthopantomogram (OPG) and full mouth intraoral periapical (IOPA) radiographs. The OPG revealed the presence of all unerupted third molars with normal pattern and timing of eruption of the teeth [Figure 4]. IOPA's revealed a normal pulp chamber and root canal spaces with no signs of obliteration. The enamel was almost half of its expected thickness. From the obtained history, a pedigree chart was plotted [Figure 5].On the basis of history, clinical and radiographic examinations final diagnosis was made as - hypocalcified autosomal dominant type of Amelogenesis imperfecta.

Treatment planning:

A treatment plan was developed with the aim of restoring masticatory efficiency, improving the aesthetics, reducing the reported tooth sensitivity. The patient was informed about her present condition, treatment procedure and other treatment options including cost differences, levels of tooth structure removal, time period for treatment completion, possible aesthetic results and expected clinical longevity. By considering the patient's age, treatment plan included was stainless steel crowns for molars and heat cure acrylic crowns for anteriors and premolars.

Treatment procedure:

In the initial phase of treatment, preventive measures were taken to improve dental and periodontal health and to modify the girl's attitude and behaviour towards the dental treatment which mainly included the diet analysis. Her oral hygiene was acceptable with no signs of gingivitis, advised extensive oral hygiene instructions. Maxillary and mandibular arch primary impressions were made using irreversible hydrocolloid impression material and two sets of diagnostic casts were made by pouring the impressions with a type III dental stone (Fuji Rock, GC Dental Corp., Tokyo, Japan).

On subsequent appointments, the carious lesions which are present on the molars were restored with the glass ionomer cement. Diagnostic casts were mounted on a semi-adjustable articulator (Hanau wide vue) with a face bow transfer. Centric relation was recorded using polyvinyl siloxane bite registration material. The articulator was programmed using protrusive and lateral records. Diagnostic wax-up on the casts were done to evaluate the amount of tooth reduction for heat cure acrylic crowns for anteriors and premolars. Preformed SSCs (3M ESPE) were placed following minimal slice preparations of the teeth bilaterally on all the molars. The bite has been raised by 1mm to compensate the loss of vertical dimension and also to balance the occlusion [Figure 6- a, b, c]. Patient is kept under follow up for one month. After confirming with the patient that she does not have any discomfort, again impressions were made and with the help of face bow transfer [Figure 7] the fabricated diagnostic casts were mounted [Figure 8] on a semi adjustable articulator. A mutually protected occlusal scheme was developed by doing a diagnostic wax-up [Figure 9]. A minimal tooth preparation with a shoulder margin configuration [Figure 10] was performed to receive heat cure acrylic crowns. Provisional restorations were fabricated using tooth coloured autopolymerising acrylic resin from the diagnostic wax up and they were cemented [Figure 11] with zinc oxide eugenol temporary cement (Temp NE; 3M ESPE, St Paul, USA). Smile analysis [Figure 12] was done placing provisional crowns and necessary corrections were made before cementation. Three weeks follow-up was done with the provisional crowns. Patient has no discomfort with the occlusion established by using the provisional crowns. Final impressions were made for both the arches with polyvinyl siloxane impression material for fabrication of heat cure acrylic crowns. Different occlusal records were made for doing cross mounting to re-establish the occlusion and vertical height same like provisionals. Models were prepared and mounted onto the articulator with the help of face bow transfer [Figure 13]. Final wax pattern was made on the models [Figure 14] and then the cross mounting was done on semi-adjustable articulator using occlusal records [Figure 15]. Flasking [Figure 16] was done for wax patterns along with the die. Later, they were subjected for de-waxing. After making sure that there is no residual wax, packing was done by using tooth coloured heat cure acrylic resin and subjected for acrylization. Then the obtained crowns were removed carefully and excess material was trimmed and the final finishing and polishing were done. These crowns are placed in the oral cavity, highpoints and eccentric contacts were removed. Final cementation was done using type-1 GIC [Figure 17,18].

II. Discussion

AI is a developmental, often inherited disorder that affects the quality and quantity of dental enamel. Transmission of the etiologic gene mutation(s) can occur either by autosomal or X-linked dominant or recessive modes of inheritance. A number of genes critical for enamel formation have been proposed or identified as candidates for AI, including AMBN,27 TUFT1,28 MMP20,29 AMELX30 ENAM,4 KLK4,24 and FAM83H1. In cases of dominant forms of AI, the enamelin gene, ENAM, is implicated in the pathogenesis where as in X-linked form, the disorder results from a mutation in the amelogenin gene, AMELX 6.

Although clinical and radiographic diagnostic methods are frequently used, it is reported that definitive diagnosis can be made with genetic tests because of the lack of objective diagnostic criteria. It has been shown that AI can develop as a result of consanguineous marriages and that similar enamel defects can be found in families of patients with AI4. Wright et al reported, in their study including 71 families with AI patients, that individuals

from the same family have similar genetic mutations7. It suggests that genetic transmission plays an important role in the occurrence of AI.

In AI patients, the radiographic findings of the teeth mainly provide important information for the dentist regarding the degree of the enamel mineralization. These findings help the clinicians to maintain the maximum amount of tooth structure until the patient is old enough for further restorative treatment to rehabilitate the dentition, which is important to allow for maxillary and mandibular growth1. But however it is necessary to intervene, to relieve the patient's symptoms of sensitivity, improve aesthetics and prevent the loss of vertical dimension. So, in this case an interdisciplinary approach is necessary to evaluate, diagnose and resolve the aesthetic problem.

Treatment options for AI include composite resin, acrylic resin crowns, polycarbonate crowns, stainless steel crowns (SSCs), direct composite veneers, ceramic laminates and posterior occlusal table-tops (Ivoclar Vivadent), pressable ceramic crowns, metal ceramic crowns, zirconia crowns. In direct composite restorations, the disadvantages are less longevity and abrasion resistance. Problems in bonding would be observed while using ceramic laminates and occlusal table-top restorations and it would also become difficult to mask the discoloration through pressable ceramic restorations8. Hence full coverage restorations with adequate resistance and retention form were the treatment of choice.

Pediatric dentist and prosthodontist has been participated in the treatment planning. The main goal of treatment was to provide transitional restorations for the protection of remaining tooth structure and to provide considerable aesthetics. Full coverage of all teeth with fixed prosthesis was identified as the ideal treatment; however, this approach requires the removal of a considerable amount of tooth structure. In addition, the patient's incomplete skeletal growth excluded this as an immediate option9. Therefore, stainless steel crowns for molars and heat cure acrylic resin crowns for anteriors and premolars were chosen to provide an excellent prosthetic alternative until the completion of skeletal growth.

In the present case, SSCs were placed on all the permanent molars and the bite has been raised by 1mm to compensate the lost vertical dimension. These SSCs are extremely durable, relatively cheap and are minimal technique sensitive during placement and offer the advantage of full coronal coverage10. A definitive treatment with porcelain fused to metal crowns or all ceramic crowns can be done once facial maturity has been reached. Heat cure acrylic resin crowns were used in this case to restore the anterior teeth and premolars, as these are aesthetically acceptable. When the enamel defects involve proximal surfaces or decreased bonding is expected, these crowns offer reasonable aesthetics and retention due to their full coverage. However, their use in AI teeth has not been systemically evaluated3.

After the treatment, we were able to achieve both functional and aesthetic goals to the satisfaction of the patient. Finally, in the girl a positive psychosocial impact was noticeable.

III. Conclusion

The importance of an interdisciplinary approach to the successful treatment of a patient with hypocalcified AI has been described in this clinical report. The conservativeness and a full coverage treatment for the affected teeth on an adolescent patient, who has not yet completed skeletal growth has also been concluded. Although early identification and preventive interventions poses numerous technical difficulties and challenges, they are

outbalanced by the benefits and it boost up's the self-esteem of the patient. This approach decreased the tooth sensitivity and enhanced aesthetics, masticatory efficiency and it also prevented serious problems in the future occlusion and social adaptation of the patient.

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