Journal of Applied Dental and Medical Sciences
NLM ID: 101671413 ISSN:2454-2288
Volume 2 Issue 1 January - March 2016

Case Report

Amelogenesis Imperfecta: Review of Literature with a case report

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ARTICLE INFO

Keywords:
Amelogenesis imperfecta, Oral rehabilitation, Glass ionomer, Composite laminate veneers, Stainless steel crowns.

ABSTRACT

Amelogenesis Imperfecta (AI) is a hereditary developmental disorder affecting deposition, calcification or maturation of dental enamel in both the primary and permanent dentitions. Also called as hereditary enamel dysplasia, hereditary brown opalescent teeth. This paper describes the review of literature and treatment plan for a young patient affected by pitted hypomaturation type of amelogenesis imperfecta. The objectives of the treatment were to enhance esthetics and restoring masticatory function. Treatment included resin composite laminate veneers on maxillary anterior teeth and stainless steel crowns for posterior teeth.

INTRODUCTION

The term amelogenesis imperfecta is applied to a clinically heterogeneous group of hereditary disorders that interfere with the normal development of dental enamel in primary and permanent dentition. These disorders cause a deficiency in the enamel's quantity and/or the quality that may result in poor dental esthetics.[1,2,3] Witkop and Sauk in 1976 listed the varieties of AI, divided according to whether the abnormality lay in a reduced amount of enamel (hypoplasia), deficient calcification (hypocalcification) or imperfect maturation of the enamel (hypomaturation), and also recognized the combined defects.[3,4,5]

Hypoplastic variants represent 60-73% of all cases. Hypoplastic variants show thin enamel but radiodensity appears normal. Lack of contact points appears to be present in this type. Enamel may be rough, smooth or pitted. Female carriers of x-linked forms manifest vertical banding of normal and abnormal enamel. Unerupted teeth may undergo replacement resorption. It may be associated with anterior open bite. Hypocalicific variants represent 7% of all cases. Radiographically enamel is less opaque than dentin. The enamel thickness is initially normal but enamel is soft and easily removed soon after tooth eruption. Teeth may be light yellow to brown in color. Hypomaturation variants represent 20-40% of all cases. Radio opacity of enamel is similar to that of dentin. Enamel tends to chip away from dentin. Enamel is mottled brown or yellow with localized or diffuse opacities. In hypoplastic – hypomaturation with taurodontism, the enamel is thin, mottled yellow to brown and pitted. Molar teeth exhibit taurodontism and other teeth have enlarged pulp chambers.[3,6,7]

According to Seow, the primary clinical problems of AI are esthetics, dental sensitivity and decreased occlusal vertical dimensions.Restoration of these defects is

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important not only because of esthetic and functional concerns, but also because there may be a positive psychological impact for the patient. [2,8,9] Treatment planning for patients with AI is related to many factors: Age, socioeconomic status of the patient, the type and severity of the disorder and the intraoral situation at the time the treatment is planned. An interdisciplinary approach is necessary to evaluate, diagnose and resolve esthetic problems using a combination of orthodontic, prosthodontic and restorative treatment. This clinical report describes the treatment of a 12-year old boy with AI using resin composite laminate veneers in the anterior region and stainless steel crowns in the posterior region.

CASE REPORT

A 12-year-old boy reported to department of pedodontics and preventive dentistry, Patiala with the chief complaint of discolored anterior teeth since they erupted. Patient also complained of sensitivity while taking cold food in the posterior teeth of lower jaw on both the sides and in the maxillary incisors. Her mother reported that one of the patient's brothers and the father presented similar dental findings. The medical and dental history was noncontributory. Photographs and dental radiographs were made. Clinical examination of patient revealed maxillary anteriors show pit defects and yellowish brown discoloration (Fig. 1). His mandibular permanent first molars were grossly carious [Fig. 2]. Other relevant findings noted were occlusal attrition on posterior teeth, loss of occlusal vertical dimension and compromised esthetics. His oral hygiene was poor, with plaque retention observed on most of the teeth. After thorough examination, the patient was diagnosed as having pitted hypomaturation type of AI because the thickness of enamel was normal and hard in texture but mottled, opaque white to yellow brown in color. Oral hygiene was not satisfactory with evidence of dental plaque (Fig. 1-2). A treatment plan was developed with the following objectives: To improve the esthetics, to reduce the reported sensitivity of the teeth and to restore the masticatory function. Restoration of anterior teeth with composite laminate veneers and posterior teeth with stainless steel crowns was planned. The patient and the parents were informed of the diagnosis and the treatment plan, which they accepted.

Initially, scaling of all teeth was performed. Dental plaque was disclosed, and the patient was taught how to improve her oral hygiene through better brushing techniques. The tooth no. 14, 15, 24 and 25 were restored with resin modified glass ionomer cement (Vitremer, USA) and tooth no. 16 and 26 with hybrid composite resin (Dentsply, USA), after isolating the operating field with cotton rolls. The mandibular second molars were restored with hybrid composite resin (Dentsply, USA).

Pulpal involvement was seen w.r.t 36 and 46 after taking intraoral periapical radiographs(fig. 3), so root canal treatment of 36, 46 was carried out using 5.25% Naocl and EDTA as irrigating and lubricating agent respectively. Obturation was done with 0.2% gutta percha (fig. 4). Post obturation was done with hybrid composite resin followed by stainless steel crowns. The mandibular first permanent molars were prepared to receive stainless steel crowns. Preformed stainless steel crowns (3M ESPE, USA) were fitted on the molars to stabilize the occlusion and to halt attrition (Fig. 5). Minor proximal reduction was undertaken for proper fit of the crowns. Crowns were cemented using luting Glass Ionomer Cement (GC Fuji I, Japan). Occlusal relationship was reevaluated.

A 0.5 mm facial reduction was done on the maxillary anterior teeth for resin composite laminate veneers. Care was taken to confine the tooth preparation within the

enamel to facilitate better bonding of composite to the tooth. The prepared surfaces were acid etched(37% phosphoric acid) and restored with a thin layer of bonding agent (Prime & Bond NT) and Universal Dup-Shade Nano composite (Coltene, Switzerland) (Fig. 6). After the entire restorative treatment, the patient's dental hypersensitivity decreased markedly, and functional chewing was established. Patient was kept on regular recall appointments.

DISCUSSION

Amelogenesis imperfecta encompasses a complicated group of conditions that demonstrate developmental alterations in the structure of the enamel in the absence of the systemic disorder. The incidence of AI has been reported to vary between approximately 1:700 and 1:16,000, depending on the population studied and the diagnostic criteria used.[2,10,11] Four major types of AI were recognized based on the phenotype (hypoplastic, hypomaturation, hypocalcified and hypomaturation-hypoplastic with taurodontism) and subdivided into 14 subtypes based on phenotype and on mode of inheritance. (Table no. 1)

AI may be inherited in an X-linked manner or as an autosomal dominant or recessive trait. The formation of enamel is highly organized and its formation is controlled through the interaction of a number of organic matrix molecules that include enamelin (ENAM; 4q21.), amelogenin (AMELX; Xp22.3-p22.1.), ameloblastin (AMBN; 4q21.), tuftelin (TUFT1; 1q21.), amelotin (AMELOTIN 4q13), dentine sialophosphoprotein (DSPP; 4q21.3.), and a variety of enzymes, such as kallikrein 4 (KLK4; 19q13.3–q13.4.) and matrix metalloproteinase 20 (MMP20; 11q22.3–q23.). Histologically, a ground section of the involved showed very thin enamel composed of lamillations of irregularly arranged enamel prisms.[12,13]

Inherited systemic conditions with enamel defects are amelo-onycho hypohidriotic syndromes, morquio syndrome, kohlschutter syndrome, amelogenesis imperfecta and nephrocalcinosis syndrome, trichodento-osseous syndrome, occulo dento osseous syndrome and epidermolysis bullosa. The commonest differential diagnosis which should be kept in mind during the clinical assessment of AI. Chronological enamel hypoplasia and Rh hypoplasia were ruled out for the patient, since chronological enamel hypoplasia develops when a systemic condition such as high fever during tooth development can produce a pattern of enamel defects (horizontal areas or bands) in the dentition. The timing of tooth development estimates the area of the disturbance. In Rh hypoplasia, hemosiderin is deposited in the forming enamel and enamel is green to bluish in color. Patient's family history ruled out fluorosis and tetracycline staining where tetracycline is deposited in hydroxyapatite crystals and chelation of calcium occurs resulting in yellow discoloration.[9,12,14]

Principles of management for amelogenesis imperfect pose specific challenges to a dentist. Clinical problems associated are poor esthetics, chipping and attrition of enamel, exposure of dentine causing sensitivity, poor oral hygiene, gingivitis and dental caries. Management directed at three aspects of care is prevention, restoration and esthetics. All restorative treatment and materials for AI patients must be carefully considered in the context of the enamel mineral content. Restorations that require the invasion of existing enamel or bonding may be

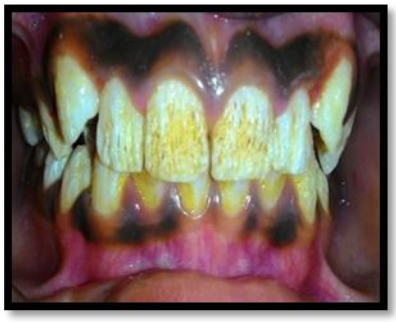

Fig. 1: Pre treatment view of teeth in maximum intercuspation

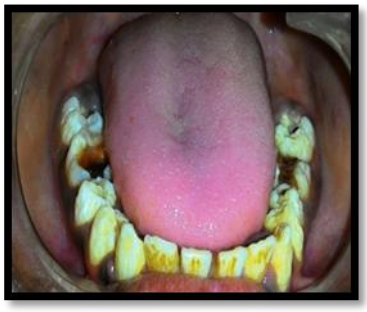

Fig. 2: Pre treatment mandibular occlusal view

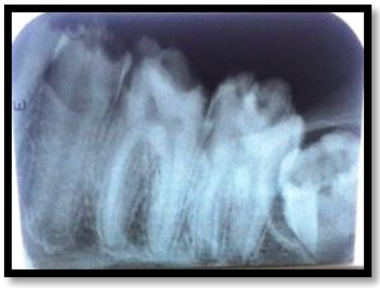

Fig 3: Intra oral periapical view of 36

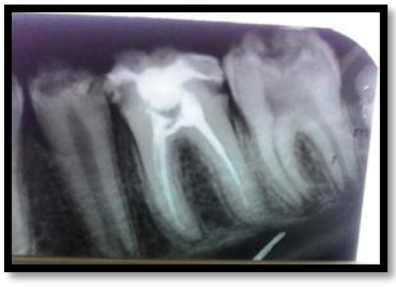

Fig 4: Intra oral periapical view of 36 after obturation

Fig. 5: Post treatment mandibular occlusal view

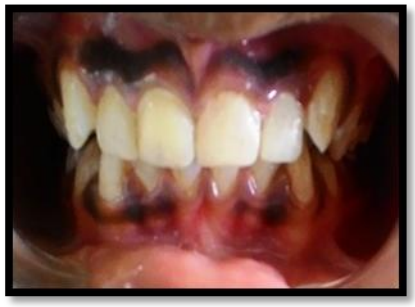

Fig. 6: Post treatment view of teeth in maximum intercuspation

contraindicated for patients with AI who have thin or poorly mineralized enamel. In the present case, composite resin was used, with the duration of the acid etching procedure similar to that indicated for normal teeth.

In the anterior teeth, direct composite veneers were placed to recover esthetics. Stainless steel crowns were placed on the permanent first molars to halt attrition of the occlusal surfaces and decrease the chance of loss of vertical dimension in the future, as well as to protect the dentinal-pulp complex from chemical and thermal attacks.

There are number of alternatives for treatment of anterior teeth affected by AI which includes full coverage coronal restorations (crowns), porcelain laminate veneers and resin composite laminate veneers. Full coverage crowns and porcelain laminate veneers require removal of

Table 1: Classification of amelogenesis imperfecta (Witkop and Sauk)		
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

substantial amounts of tooth structure, thus more invasive and include risk of pulp exposure in young permanent teeth. Similarly for posterior teeth, metal-ceramic or porcelain-fused metal crowns are commonly used. But the relatively high pulp horns in young patients lead to pulp exposure during tooth preparation. The preferred alternative in such cases is preformed stainless steel crowns, which require least tooth preparation.

CONCLUSION

Dentists should be aware of the presentation of amelogenisis imperfecta to assist in early diagnosis and aim to provide the patient with the proper oral rehabilitation treatment. The primary goal of the treatment should address each concern as it presents but with an overall comprehensive plan that outlines the anticipated future treatment needs. In the present case, 12-year old boy with AI were treated using resin composite laminate veneers in the anterior region and stainless steel crowns in the posterior region to alleviate sensitivity, improve esthetics, and restore function.

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