DENTINOGENESIS IMPERFECTA:
Review and Case Report

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INTRODUCTION

Dentinogenesis imperfecta or hereditary opalescent dentine was described in the late 19th century characterized by abnormalities of the enamel and dentin of the condition. In general, both the primary and permanent dentitions are affected[1]. The disease is inherited in a simple autosomal dominant mode with high penertrance and a low mutation rate. It has been reported to occur at a rate of 1 in 6000-8000 births [2].

Dentinogenesis imperfecta has been divided into three types by Shields, Bixler and El-Kawfry[3] (1973):

Type I: In which the dentin abnormality occurs with concurrent osteogenesis imperfecta. In this form primary teeth are more severely affected than the permanent teeth.

Type II: Where patients have only dentin abnormalities and no bone disease.

Type III or Brandywine type: Where only dentin detects occur similar to Type II but some radiographic and clinical differences are present.

Clinically all three types share numerous features. In both dentitions, teeth exhibit an unusual translucent, opalescent appearance with color variations from yellow-brown to gray[4]. The entire crown may appear discolored due to the abnormal underlying dentin.

Inheritance of dentinogenesis imperfecta is said to follow an autosomal path of inheritance, Battagel and Levinkind[5] (1988) reported a case where the pedigree for the condition was traced back to four generations. In cases where there are multiple offspring in a generation the condition has been seen in more than one of the siblings, but it is a rare occurrence. Two affected siblings reported to the Department of Orthodontics, College of Dental Surgery, Manipal.

CASE HISTORY

A brother-sister pair, both middle aged, reported to the department. The male patient complained of discolored, protruding and crowded teeth, whereas the female complained of similar unsightly tooth discoloration and mild protrusion and spacing.

Medical history of both patients was non-contributory with no evidence of associated osteogenesis imperfecta (patient gave no history of bone fractures). The family history revealed that out of five siblings, two had dentinogenesis imperfecta, the patients did not know if any of the previous generations had the condition.

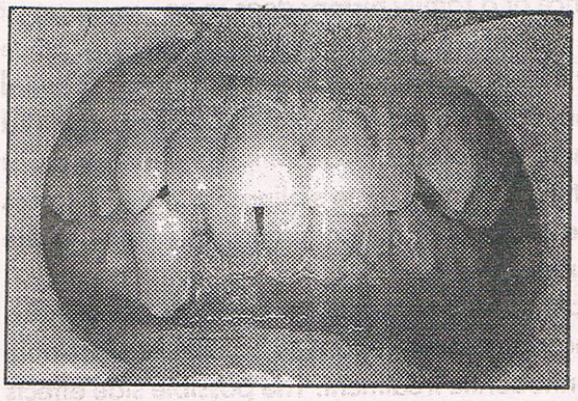

Figure 1: Intra-oral photograph of male patient – frontal view.

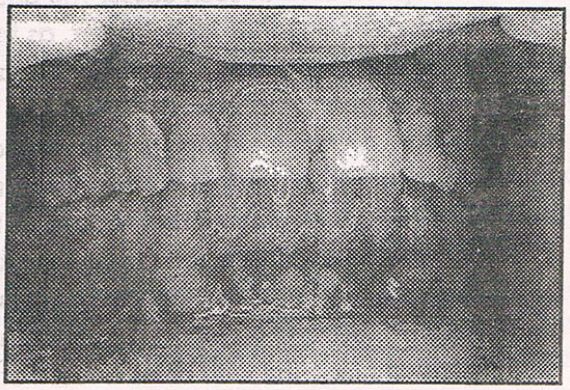

Figure 2: Intra-oral photograph of female patient – frontal view.

Extra oral examination did not reveal anything significant. On oral examination full compliment of teeth were present, the caries incidence was very high, with multiple restored carious lesions in many teeth. There was no history of fracture of any of the teeth.

Radiographic examination revealed that the thickness of the enamel appeared normal, the roots were normal in both cases.

Patient's facial photographs revealed a soft tissue protrusion. The photographs of the teeth in the female patient revealed maxillary spacing with a normal mandibular arch, and a Class II molar relationship.

In the male patient the photographs show mandibular and maxillary crowding, normal overbite, and Class I posterior interdigitation of teeth.

TREATMENT PLAN

It was decided to treat both the brother sister pair orthodontically. The normal condition of the enamel and mild nature of the dentinal defect were encouraging signs, which made us decide in favor of comprehensive fixed appliance orthodontic therapy. Due to the crowding in the male case it was decided to extract the first premolars and retract the anteriors into the residual extraction space. In his sister's case, it was decided to band only the posterior teeth and treat the case using the Strang bypass Technique. This would not only preserve the anterior teeth, from orthodontic banding and debanding; it would greatly shorten the treatment time with optimum treatment results. At present uneventful treatment progress has been recorded in both these cases.

DISCUSSION

The aim of the treatment in dentinogenesis imperfect is to improve the esthetic appearance and maintain the oral masticatory apparatus in a healthy and functional state. Here the patient presented with the dual esthetic complaint of the malocclusion as well as the discoloration. It was decided to first treat the malocclusion and then to mask the discoloration. Porcelain and composite veneers following acid etching of the labial enamel surface is one

treatment option used in such cases. The absence of a family history does in no way suggest to an absence of an inheritance factor, as the patient was not fully aware of the prevalence of the condition among his ancestors. The fact that only two out of total five siblings showed the condition among his ancestors. The fact that only two out of a total five siblings showed the condition, proves a strong genetic component with a possible incomplete penetrance, at the same time the absence of the condition in the remaining three siblings outright indicates that there are no environmental factors in the etiology of the condition, as otherwise the anomaly would have been seen in all the siblings.

Since not many cases are reported on orthodontic treatment of patients with dentinogenesis imperfecta[5,6,7], there is a certain amount of uncertainty with regard to possible undesirable side effects coincidental with or as a result of orthodontic treatment. The possible side effects can be

(1) shearing off or loss of enamel of teeth,
(2) blunting of roots of the teeth,
(3) early loss of teeth,
(4) increased discoloration of teeth, and
(5) fracture of the teeth.

Orthodontic management of patients with dentinogenesis imperfecta involves a certain amount of risk, but treatment can be completed successfully. This case report illustrates the diagnosis, and treatment options for a patient with the condition, and also presents the rare case of siblings presenting with hereditary opalescent dentin.

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