INTRODUCTION

Dentinogenesis imperfecta is an uncommon defect in the collagen formation that is transmitted as an autosomal dominant trait. It was probably first recognized by Barret in 1882. The first published report describing the disorder as an enamel defect was by Talbot as quoted by Witkop. The term ‘hereditary opalescent dentin’ was first used by Skillen, Finn and Hodges to describe the brown translucent teeth that have an opalescent sheen and are lacking in pulp chambers. This condition causes teeth to be discolored (most often a blue-gray or yellow-brown color) and translucent. Enamel may be thinner than normal in this condition. Thus the teeth are also weaker than normal, making them prone to rapid wear, breakage, and loss. In most cases, an affected person has one parent with the condition. Dentinogenesis imperfecta affects an estimated 1 in 6,000 to 8,000 people. This article reports a case of dentinogenesis imperfecta in a 22 year old male with typical clinical and radiographic features.

CASE REPORT

A 22 year old male patient, visited the Department of Oral Medicine and Radiology, Seema Dental College and Hospital, Rishikesh with a chief complaint of pain in the lower right and upper left back tooth region since 1 month. Intraoral examination showed missing 11, 13, 21, rotated 22, and carious 16, 17, 26, 36, 46, 47. Generalized attrition was observed. Gingiva was soft and edematous with loss of stippling. A generalized yellow-brown discoloration of teeth was also observed. (Fig. 1) Based on generalized yellow-brown discoloration and attrition of teeth, a clinical diagnosis of dentinogenesis imperfecta was made.

Orthopantomogram (Fig. 2) showed the following features: Short and slender roots, constricted neck, prominent in posterior teeth, increased contrast between crown and root, and generalized obliteration of pulp canal. (Fig. 3 & 4)
Radiographic features were characteristic and confirmed the clinical diagnosis of dentinogenesis imperfecta.

**DISCUSSION**

Dentinogenesis imperfecta is a genetic disorder of tooth development. It is inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder. In most cases, an affected person has one parent with the condition. Dentinogenesis imperfecta affects an estimated 1 in 6,000 to 8,000 people. It occurs with equal frequency in both sexes. It usually affects whites.

Dentinogenesis imperfecta has been subdivided into three types: Type I occur in people who have osteogenesis imperfecta, a genetic condition in which bones are brittle and easily broken. In type II there is no associated osteogenesis imperfecta; and when the condition is associated with the Brandywine triracial isolate and large pulp chambers, it is classified as type III.

Some researchers believe that dentinogenesis imperfecta type II and type III, along with a condition called dentin dysplasia type II, are actually forms of a single disorder. The signs and symptoms of dentin dysplasia type II are very similar to those of dentinogenesis imperfecta. However, dentin dysplasia type II affects the primary teeth much more than the permanent teeth.

Clinically, the appearance of the teeth with dentinogenesis imperfecta is characteristic. They show a high degree of amber like translucency and a variety of colors from yellow to blue-gray. The colors change according to whether the teeth are observed by transmitted light or reflected light. Affected teeth have broad crowns with constriction of cervical area resulting in tulip shape. The enamel easily fractures from the teeth and the crowns wear readily. In adults they may frequently wear down to the gingiva. The exposed dentin becomes stained. The color of the abraded teeth may change to dark brown or even black. Some patients demonstrate an anterior open bite.

Radiographically the teeth appear solid, lacking pulp chambers and root canals. Radiographs may also reveal slight to marked attrition of the occlusal surface. The roots are usually short and slender.

Early in development, the teeth may appear to have large pulp chambers, but these are quickly obliterated by the formation of dentin. Ultimately the root canals may be absent or threadlike. Occasional periapical radiolucencies are seen in association with sound teeth without evidence of pulpal involvement, which may occur from microscopic communication between residual pulp and the oral cavity. These lesions do not occur as frequently as in dentin dysplasia. The architecture of the bone in the maxilla and mandible is normal.

Histologically the dentin is composed of irregular tubules, often with large areas of uncalcified matrix. The tubules tend to be larger in diameter and less numerous in a given volume of dentin than in normal teeth.

**REFERENCES**

Fig 1: Intra oral examination showing generalised yellow-brown discoloration and attrition of teeth.

Fig 2: Orthopantomogram showing characteristic features of dentinogenesis imperfecta

Fig 3 & 4: Intra oral periapical radiographs showing complete obliteration of pulp canals on all teeth as well as increased contrast between crown & root

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