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CAPDEPONT'S TEETH: A CASE REPORT

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Running title: Dentinogenesis imperfect

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ABSTRACT:

Dentinogenesis imperfecta is an autosomal dominant disorder of tooth development characterized by the presence of opalescent dentine, resulting in a dusky blue to brownish discoloration of the teeth. It is the most common dental genetic disease. This condition is genetically and clinically heterogeneous, it may affect only the teeth or it may be associated with the osteogenesis imperfecta. Diagnosis is based on history, clinical examination and radiographic features. This report describes an 18 year old male patient who showed the characteristic dental features of dentinogenesis imperfecta.

Key words: Dentinogenesis imperfecta, Amelogenesis Imperfecta, Dentine Dysplasia.

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INTRODUCTION:

The teeth which are regarded as the hardest structures of our body are made up of enamel the outermost covering, dentine the middle layer and the pulp which consists of nerves and blood vessels. Tooth development like the development of all epithelial appendages is regulated by inductive tissue interactions between epithelium and mesenchyme. Numerous genes interact, either act in conjunction or antagonize each other in odontogenesis [1, 2]. Certain genes involved in enamel and dentine structures are highly specific for tooth. Mutations in these genes have been identified as causes of Amelogenesis Imperfecta (AI), Dentinogenesis Imperfecta (DGI), Dentine Dysplasia (DD) and anomalies in tooth number [1]. Dentinogenesis

imperfecta is also known as Capdepont's teeth, hereditary opalescent dentin, Brown teeth. It is a genetic disease transmitted as an autosomal dominant trait, and characterized by disturbance in dentin formation [3, 4]. The condition was first described by Barret in 1882 [4]. The term Dentinogenesis imperfecta was coined by Robert and Schour in 1939 [1]. The affected teeth have opalescent, amber color and darken with age and exhibits attrition of incisal and occlusal surfaces [2]. Our present case report is about an 18 year old patient who reported to the department with the chief complaint of discoloration in the anterior teeth.

CASE REPORT:

An 18 year old male patient reported to the department of Oral medicine and radiology, with the chief complaint of discoloration of his anterior teeth (Figure 1). There was a history of similar discoloration of the deciduous teeth which were exfoliated uneventfully. The permanent teeth which erupted were brown in color at the time of eruption. The same type of discoloration was seen in patient's siblings.

On intra oral examination the maxillary and the mandibular anterior teeth were brownish in color with mild upper anterior crowding. The incisors were a darker shade of brown when compared to the other teeth. The incisal and occlusal surfaces of all the teeth were attrited (Figures 2 & 3). Both the upper first maxillary molars were decayed with caries present on the occlusal surface. The mandibular first molars and the premolar on the lower right side were found to be missing. The patient was further subjected to radiographic investigations. Orthopantomograph showed normal anatomical landmarks with full complement of upper teeth and partially edentulous mandible. Generalized cervical constriction of all the teeth was noticed with obliteration of pulp chambers in some suggestive of Dentinogenesis imperfecta. Radiolucency involving enamel and dentin and approximating the pulp was seen in respect to upper first maxillary molars suggestive of dental caries (Figure 4).

DISCUSSION:

The classification of hereditary dentine disorders is currently complicated. The most familiar classification system is that formulated by Shields in 1973 [3]. This categorization discriminates three types of dentinogenesis imperfecta and two types of dentine dysplasia [3].The Shields' system is increasingly out of date as it does not account for the molecular etiologies of the hereditary dentine defect elucidated so far [4]. The genetic defects that have been discovered to date are insufficient to allow for the construction of a comprehensive classification based on the knowledge of the underlying mutations. Shield classified dentinogenesis imperfecta into three types based on clinical and radiographic features [3].

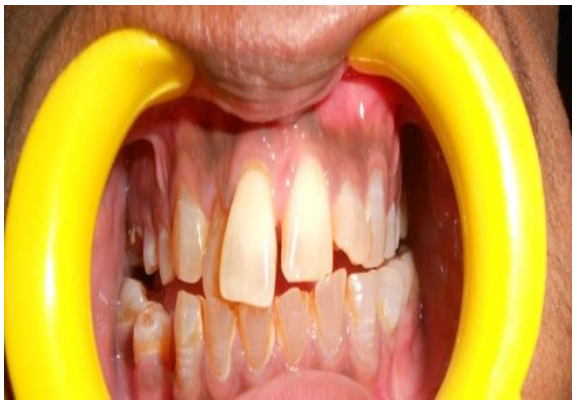

Figure 1: Anterior teeth showing brownish discoloration

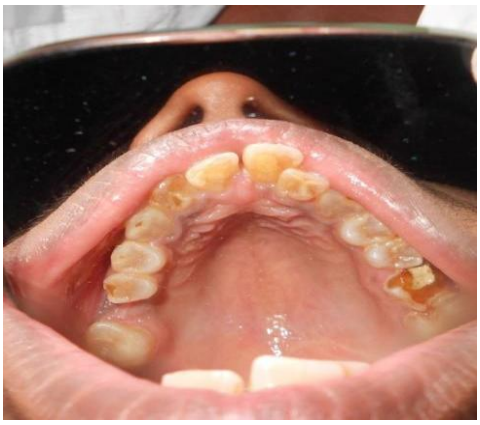

Figure 2: Occlusal view of maxillary teeth showing chipping of enamel and dark brownish pigmentation of posterior teeth.

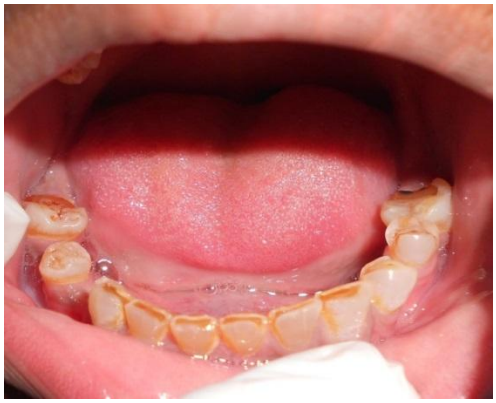

Figure 3: Occlusal view of mandibular teeth showing attrition and dark brownish discoloration

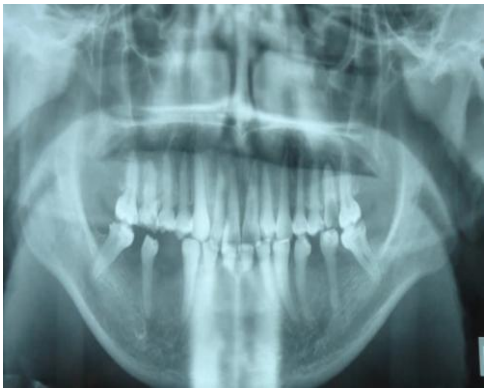

Figure 4: Orthopantomograph showing bulbous crowns and cervical constriction of teeth

Dentinogenesis imperfecta type I: Individuals with DGI-I also have osteogenesis imperfecta. The teeth of both dentitions are typically amber and translucent and show significant attrition. Radiographically, the teeth have short, constricted roots and dentine hypertrophy leading to pulpal obliteration either before or just after eruption. Expressivity is variable even within an individual, with some teeth showing total pulpal obliteration while in others the dentine appears normal [5].

Dentinogenesis imperfecta type II: The dental features of DGI-II are similar to those of DGI-I but penetrance is virtually complete and osteogenesis imperfecta is not a feature. Bulbous crowns are a typical feature with marked cervical constriction. Normal teeth are never found in DGI-II. Short stature and blue sclera are extra oral features which may be seen in individuals affected. Sensorineural hearing loss has also been reported as a rare feature of the condition [6].

Dentinogenesis imperfecta type III: This is a form of DGI found in a tri-racial population from Maryland and Washington DC known as the Brandywine isolate. The clinical features are variable and resemble those seen in DGI-I and -II but the primary teeth show multiple pulp exposures and radiographically, they often manifest "shell" teeth i.e. teeth which appear hollow due to hypotrophy of the dentin.

The present case comes under type II in the Shield's classification and there was absence of blue sclera. The conditions that have similar clinical or radiographic features to DGI need to be considered to give a correct diagnosis. Some of the conditions may mimic the appearance of DGI either clinically or radiographically. Hypo calcified forms of Amelogenesis imperfecta initially develop normal enamel thickness but the poorly calcified enamel is soft and friable and is rapidly lost by attrition leaving dentine cores. But unlike DGI the teeth are usually sensitive and on radiographs enamel is less radio-dense than dentine [7]. Pulp chamber and root canals are usually not sclerosed.

Congenital erythropoietic porphyria is a condition resulting from an inborn error of porphyrin metabolism. This deficiency leads to hemolytic anemia, photosensitivity, blistering of the skin, and deposition of red-brown pigments in the bones and teeth [8]. In case of Rhesus incompatibility, the discoloration ranges from yellow through to green, brown and grey to black is usually found at the necks of teeth and the enamel hypoplasia's are usually located in the coronal third of the teeth [9].

Tetracycline's have the ability to chelate calcium ions and to be incorporated into developing teeth, cartilage and bone, resulting in discoloration of both the primary and permanent dentitions. This permanent discoloration varies from yellow or grey to brown depending on the dose or the type of the drug received in relation to body weight [10].

In the present case discoloration of the anterior teeth was more pronounced than the rest and generalized attrition of teeth was noticed.

Other causes of early loss of teeth as in DGI include hypophosphatemia, immunological deficiencies e.g. severe congenital neutropenia (Kostmann's disease), cyclic neutropenia, Chediak-Hegashi syndrome, neutropenia's, histiocytosis X, Papillon- Lefevre syndrome and leucocyte adhesion deficiency syndrome [11]. With the exception of hypophosphatasia, all of these conditions have an underlying immunological defect which makes those with these conditions susceptible to periodontal breakdown. Mobility of teeth in those with hypophosphatemia however is due to aplasia or marked hypoplasia of cementum.

Vitamin D-dependent rickets and vitamin D-resistant rickets have clinical and radiographic features of DGI. Vitamin D-dependent rickets is characterized by yellowish to brown enamel, chronic periodontal disease, large quadrangular pulp chambers and short roots. Features of vitamin D-resistant rickets include

attrition and exposure of abnormally formed dentine of primary teeth and abscessed non-carious primary or permanent teeth [11].

The aim of treatment provided would be to restore function, aesthetics and protect posterior teeth from wear and maintain the occlusal vertical dimension.

Treatment varies according to the age of the patient, severity of the problem and the presenting complaint. Modern dental technology and materials have promoted new treatment strategies, including the use of an extended provisional phase to better determine the functional and esthetic aspects of a specific case. In the present case the patient was referred to the department of conservative dentistry for the endodontic treatment of the tooth with caries.

CONCLUSION:

Dentinogenesis imperfecta is the most common autosomal dominant disorder causing discoloration of the teeth which in turn affects the quality of life of an individual. Correct diagnosis and carefully planned management would help to restore not only the function but also the aesthetics there by improving the quality of life of the individual.

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