TEETH IN OSTEOGENESIS IMPERFECTA: A LIGHT MICROSCOPIC STUDY

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SUMMARY

The light microscopic features of the dentine in three teeth from two cases of osteogenesis imperfecta (OI) are presented. Results show that teeth in OI distinctively have a more uniform and tubular mantle dentine, and a characteristic laminated circumpulpal dentine formed by distorted incremental bands alternating with parallel rows of interglobular dentine and interrupted by comet-shaped vascular canals. The study indicated that in the absence of overt OI features, the changes in dentine alone are sufficiently characteristic to establish such a diagnosis.

INTRODUCTION

Of all the skeletal disorders, hereditary or otherwise, osteogenesis imperfecta (OI) is the most unique in that it is commonly associated with abnormalities of dentine formation. Based on the classification of Shield *et. al.*, the affected teeth in OI are categorised as dentinogenesis imperfecta (DI) type I to differentiate it from DI type II which occurs as an isolated trait and do not present with any skeletal abnormalities, and from DI type III (Brandywine isolate) which is exclusively found in an inbred population group of mixed white, black and American Indian ancestry in Maryland, USA.[2-5] All three types are transmitted as autosomal dominant traits.[4] Atypical or sporadic cases are rare.[6,7] Probable homozygous cases have also been reported.[3,8]

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The three types of DI share certain clinical and histological characteristics.[4] Both the deciduous and permanent dentitions are affected. The severity of the trait in individual teeth varies with the age at which the particular tooth developed.[9] Accordingly, the deciduous teeth are the most severely affected followed by first permanent molars and incisors and then premolars and cuspids. The least severely affected and best formed teeth are the second and third molars.[10] Clinically, the crowns of the involved teeth have a dark amber opalescence and tend to exhibit severe attrition at an early age.[1-3] The caries index is quite low in these individual teeth and when caries occurs, it tends to spread laterally rather than invade, due apparently to the irregular configuration of the dentinal tubules.[11] The high rate of attrition also tends to retard caries, as well as make the teeth smooth and self-cleansing.[11]

Radiographically, the affected teeth in DI have bulbous crowns with accentuated cervical constrictions, lack of pulp chambers, absent or thread-like root canals and short stunted roots. An extreme

variation that may occur in deciduous teeth giving a "shell tooth" appearance has also been described.[12] Periapical radiolucencies are common and have been associated with exposure of a pulp horn by attrition.[11]

Histologically, the teeth in DI tend to exhibit a normal mantle dentine layer but with the bulk of the circumpulpal dentine exhibiting a spectrum of abnormal features.[1–5] The most striking abnormal feature is the lack of a normal, regular tubular pattern. In many instances there is a relative absence of tubules with a distinct disorganisation in tubule size and direction. Other abnormal findings include frequent occurrence of interglobular dentine, early obliteration of the pulp by similar abnormal dentine and reduced scalloping of the amelodentinal junction.[1-5]

In this paper the histological changes in dentine in three permanent teeth from OI are further delineated and the significance of these findings discussed.

MATERIAL AND METHODS

Three teeth from two cases of OI (DT type I) were used in this study. These were obtained from the files of the Department of Pathology, Eastman Dental Hospital, London. They represented biopsy specimens diagnosed in 1956 and 1964 respectively. Little medical clinical information of these cases were available apart from that provided together with the biopsy forms. The teeth consisted of a permanent first molar from an 11-year-old female, and two premolars from a 22-year-old male. These were fixed in 10% formalsaline, decalcified in 5% trichloroacetic acid and embedded in Stemco wax at 58°C. Longitudinal sections of about 5 microns in thickness were prepared and stained with haematoxylin and eosin (H + E), Schmorl's picrothionin and Gomori's reticulin impregnation technique (modification of Perdrau).

A BHS-2 Olympus microscope was used to study the light microscopic changes in dentine. For the estimation of the mantle dentine width, a 10x eyepiece containing a linear micrometer and a 40x objective were used. This was superimposed on the mantle dentine at 10 randomly chosen sites. The mean value estimated to the nearest 5 microns was obtained.

RESULTS

Amelodentinal Junction (ADJ)

In all the three teeth, the ADJ exhibited a reduced scalloping to almost a smooth union at the cervical third of the crown. In places, part of the ADJ was destroyed by attrition and caries (Fig. 1).

Mantle dentine

Beneath the ADJ, the mantle dentine occurred as a well-defined zone of constant width demarcated from the circumpulpal dentine by a haematoxylinophilic band. Within this zone the dentinal

Fig. 1 Destruction of the mantle dentine in the incisal region of the premolar tooth by caries and attrition. (Schmorl's picrothionin. Original magnification x 33).

tubules were regularly arranged at right angles to the ADJ. These exhibited anastomosing lateral branches (Fig. 2).

Circumpulpal dentine

In all the three teeth, the change in structure from mantle dentine to circumpulpal dentine was sudden and well-defined. Within the circumpulpal dentine the most striking feature was the presence of an abnormal and prominent incremental pattern (Fig. 3). These consisted of distinctive basophilic, distorted bands occurring at irregular frequency and interrupted at fairly regular intervals by comet-shaped vascular canals (Fig. 3). Alternating with these incremental stratifications were rows of interglobular dentine which also extended around the vascular canals (Fig. 3). The vascular canals were usually empty or may be filled with a greyish amorphous material.

In the three teeth a variable degree of abnormality in number, arrangement, size and distribution of the dentinal tubules was also observed. These tubules may occur as short irregular strands running singly (Fig. 4) or in clumps separated by relatively atubular areas. Occasionally they may form loops or terminate in vascular canals.

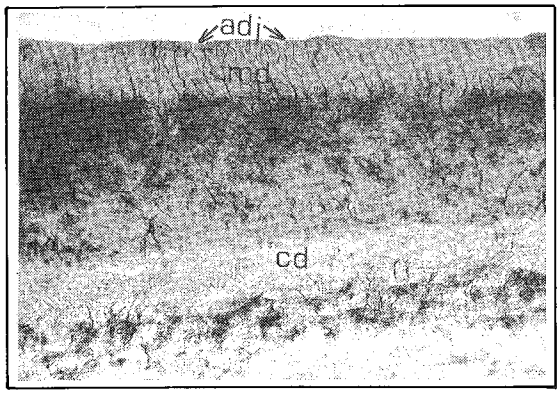

Fig. 2 Higher power view of the mantle dentine zone (md) to show the regular tubular pattern and anastomosing lateral branches. Note the smooth amelodentinal junction (adj) and underlying abnormal circumpulpal dentine (cd). (Schmorl's picrothionin. Original magnification, 132).

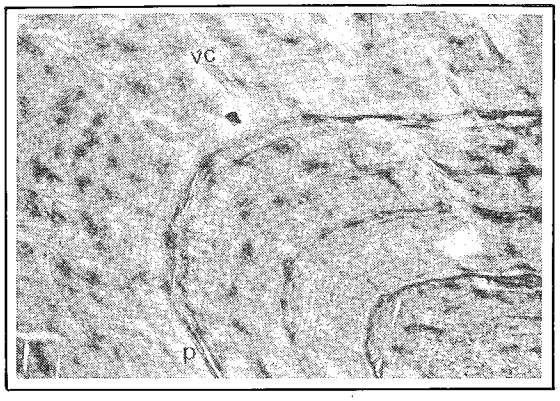

Fig. 3 Photomicrograph of the circumpulpal dentine showing incremental bands alternating with rows of interglobular dentine and interrupted by comet-shaped vascular canals (vc). Note the slit-like pulp. (Schmorl's picrothionin. Original magnification x 33).

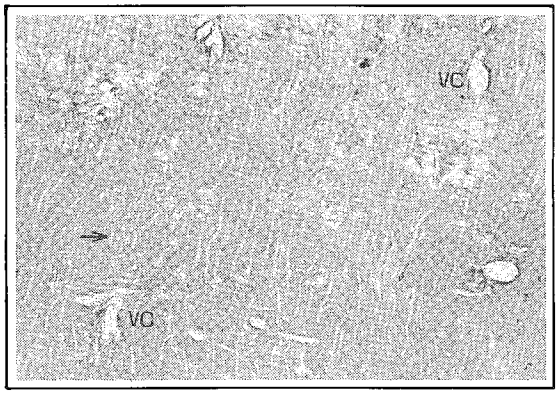

Fig. 4 Circumpulpal dentine with dentinal tubules running singly, forming loops (see arrow) or terminating in vascular canals (vc). (Gomori's reticulin. Original magnification x 200).

Pulp

The pulp chamber and root canals of the molar tooth were reduced to slit-like spaces (Fig. 3). These were largely empty or may be filled with an eosinophilic hyalinised material. The normal pulpal content with its odontoblastic cell layer was lacking. In the other two premolar teeth, the pulp was totally obliterated. The tissue replacing the pulp was similar in structure with the rest of the circumpulpal dentine.

Estimation of the mantle dentine width

The width of the mantle dentine in each tooth represented the average obtained from 10 random sites along the mantle dentine in the crown region. The mean values obtained for the two premolar and molar teeth were 40 and 60 microns respectively.

DISCUSSION

The earliest known case of OI occurring in association with dentinogenesis imperfecta was described in an Egyptian mummy dating back to 1000 B.C.[13] Rushton[9] in his classic paper on "Anomalies of Dentine" discussed the question whether DI occurring as an isolated trait is the same disease as that occurring in association with OI. Based on analysis of published reports and on his personal studies of teeth from OI, he concluded that the two conditions were as regards to the teeth, very similar but there was no substantial evidence that they were in fact the same disease.[14] However current contributors consider the two diseases as distinct, as individuals with DI type II do not exhibit a familial history which included any of the other features of OI.[10,15,16] For these reasons, affected teeth in OI are often designated as DI while that occurring as an isolated trait as hereditary opalescent dentine.[10,15]

In the present study the findings in the mantle dentine were in agreement with those of Sunderland and Smith.[5] However instead of a line of reduced staining intensity demarcating it from the circumpulpal dentine, a thin haematoxyphilic band was identified in the present study. Unlike teeth in OI, the mantle dentine of DI type II has a more amorphous appearance, contain fewer tubules and was often difficult to differentiate from the rest of circumpulpal dentine.[5]

Listgarten[17] and Pindborg[18] observed that the width of the mantle dentine in teeth with DI in general varies between 20 – 100 microns. Godfrey[19] found that in teeth from cases of OI congenita, the mantle dentine width was about 20 microns in paraffin sections and 30 microns in frozen sections. In the present study a much higher value was observed. As with most authors,[19–22] it is also suggested that the normal mantle dentine layer is attributable to normal functioning odontoblasts and subodontoblastic cells.

In 1947, Pindborg[18] observed that teeth in OI often present with a laminated circumpulpal dentine caused by accentuation of haematoxyphilic incremental bands which he likened to as "Northern Lights". Similar structures were also noted by Witkop[10] and Suzuki.[21] According to Sunderland and Smith,[5] this laminated appearance was consistently observed in the dentine in teeth from OI but was absent in DI type II. The present study also confirmed such a finding.

Witkop[10] stated that in teeth from OI, the mildest change in dentine is the presence of vascular canals that resemble a "comet" with the "head" towards the periphery and "tail" towards the pulp. These were generally considered as remnants of vascular channels entrapped within the dentinal matrix during dentinogenesis. In the present study numerous such structures were also encountered in the circumpulpal dentine. Their shape and content also support the concept that they are vascular in origin.

Most authors have noted that the areas of interglobular dentine in DI in general appeared to have both a specific and random distribution within the circumpulpal dentine.[2,10,22] These are indicative of an underlying defective calcification process during dentinogenesis. In the present study the occurrence of rows of interglobular dentine parallel to the incremental lines seems to suggest a periodical retardation of the linear calcification front at each level of dentinogenesis. As only three teeth were studied in the present investigation, it is difficult to ascertain whether this pattern of distribution is specific for teeth in OI.

Sunderland and Smith[5] stated that as most of the teeth in their study on OI were deciduous and those from DI type II were permanent, they could not ascertain whether the differences in the dentine structures observed were due to differences between deciduous and permanent teeth or

to differences between types I or II. The present study confirmed that the observed specific changes in dentine are not due to differences between deciduous and permanent teeth but are distinctive of teeth in OI.

The clinicopathologic significance of the distinct changes in dentine in teeth in OI is that it may provide important medico-legal differentiation between the unexplained fractures of OI and non-accidental injuries as in battered-child syndrome.

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