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STUDIES ON MINERALIZED DENTAL TISSUES

VII. Dental changes occurring in osteopetrosis. Continued studies.

by

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In an earlier publication (*Bergman & Engfeldt,* 1954) a histologic and microradiographic description has been given of dental changes observed in a case of osteopetrosis (Albers-Schönberg's disease). The material was limited to three extracted deciduous teeth of a six-year-old boy in whom skeletal disease had been diagnosed at the age of ten months. The teeth were practically normal in their external shape but examination of ground sections, microradiograms and decalcified sections revealed superficial resorption of the roots, widened Owen's lines in the dentine, slight hypoplasia of the enamel and circumpulpal irregular secondary dentine.

Dental radiographs indicated aplasia of several permanent teeth, impaction of most of the deciduous teeth and malformation of the crowns of the permanent upper incisors. The small number of teeth studied and the non-specific nature of the observed changes called for caution in associating them with the skeletal disease; accordingly, other factors were sought.

The appearance of more substantial material from another case of osteopetrosis — a five-year-old girl — has provided a more reliable basis for a discussion of the connection between

the dental changes and the skeletal disease. The sparse literature on the subject would seem to warrant a fairly detailed account of the findings in this case.

CLINICAL DESCRIPTION

For a general description of the case the reader is referred to the article by *Enell, Melander & Pehrsson* (1956), and for a dental description to the article by *Borggren* (1956). Some clinical details are given here, however, as a background to the following histologic and micro-radiographic examination.

The girl was admitted at the age of four months to the Children's Clinic of the Military Hospital in Boden for examination of a stomach disorder. The patient was pale and had a somewhat prominent forehead and tubera parietalia. The anterior fontanelle was under tension; the spleen was enlarged. Radiographs of the skeleton — the skull, thorax and the extremities — revealed changes typical of osteopetrosis.

The patient is the second cousin to two children (siblings) with the same disorder. The parents of the patient and of these two children are double cousins.

During the following years the patient was admitted to hospital several times. At five years of age she was found to present facial paralysis on the right side, impaired vision and pronounced anaemia with a small number of reticulocytes. Radiographs indicated sclerosis of the whole skeleton and cross striation of the metaphyses of the long bones.

At the age of eighteen months the deciduous central incisors had erupted. No further teeth had appeared by the age of five years. At three it was found that the lips were dry and had sores, the gingivae bled easily and the alveolar crest was swollen. When the patient was five years old the four incisors were loose and the upper ones much discoloured near the gingival margin. The mucosa of the alveolar crest was firm and hard in the edentulous parts of the mouth.

Radiographs showed marginal destruction of the bone in the incisor region, malformation of the crowns and roots of many deciduous and permanent teeth and aplasia of the deciduous upper second molar and of the second premolars in both jaws.

MATERIAL AND METHOD

The following teeth were available for histologic and microradiographic examination: +2, +02, +03, —04, —05.* None of these teeth had erupted. There were also pieces of covering alveolar bone. The teeth and the bone were removed under narcosis (M.B.B.). The material was fixed in 10 per cent formalin. One of the teeth (—05) was cut longitudinally with a diamond saw. One half was decalcified, imbedded in paraffin, sectioned and stained. The other half was used for the preparation of ground sections (see Table 1). The thickness of the decalcified sections was 6 μ and the stains used were Bock, Azan, Mayer's haemalum-eosin (htx-eosin) and Hansen. The teeth for the preparation of ground sections and microradiographic examination were imbedded in plastic (Castolite) after dehydration. Thin slices were obtained with a diamond saw, and these were ground to a thickness of 100—230 μ. Some of the pieces of bone were decalcified for ordinary histologic examination. The remainder were ground to a thickness of 100 μ. In this case, too, Castolite was used as the imbedding material and the ground sections were examined by the microradiographic technique.

The microradiograms were prepared by the procedure described in an earlier article (*Bergman & Engfeldt*, 1954). The ground sections were placed in direct contact with a fine-grained photographic emulsion that was exposed to x-rays generated at 30 kV. As the source of x-rays a Machlett OEG tube with a 0.2 mm beryllium window was used. With the radiation so obtained more than 97.5 per cent of the absorption in the section is due to the inorganic content. The microradiogram thus reveals the quantity and distribution of mineral salts in the tissues. The microradiographic technique as at present applied permits a resolving power of 0.5 to 1 μ which is of the same order of magnitude as that of the optical microscope.

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* In this paper the teeth are designated according to the Haderup nomenclature: + denotes the upper jaw, — the lower jaw. When the + or — is placed to the right of the tooth number, the right side is indicated and vice versa. 0 denotes a deciduous tooth. + 02 thus means the left maxillary deciduous lateral incisor.

RESULTS

Some of the findings are given in Table 1.

Table 1

Data on the teeth studied

	+2	+02	+03	− 04	− 05
MACROSCOPIC EXAMINATION	crown deformed, no root	shape almost normal	crown deformed, no root	crown deformed, root not well developed	crown deformed, suggestion of a root
HISTOLOGIC EXAMINATION	decalcified sections (100)	ground sections (5)	ground sections (3)	ground sections (2)	decalcified and ground sections (70, 3)
ENAMEL HYPOPLASIA (ground sections)		small pits incisally and cervically	deep defects in the enamel	deep defects in the enamel	defects occlusally and buccally
APPEARANCE OF THE ROOTS (ground and decalcified sections)	stunted and deformed	normal	no root	stunted and deformed	stunted and deformed
STRUCTURE OF THE DENTINE (ground and decalcified sections)	regular	regular	regular	regular	regular
MINERALIZATION OF THE DENTINE (Microradiograms)		almost normal (small interglobular spaces near the neonatal line)	almost normal (small interglobular spaces near a hypoplastic defect)	apparently normal	widened Owen's band in crown near the neonatal line; otherwise apparently normal
PULP (decalcified sections)	normal				normal (small cellular infiltration near "root tip")

THE SHAPE OF THE TEETH

Only one of the five teeth — +02 — was of fairly normal external appearance. Fig. 3 shows, however, that the enamel of the tooth was somewhat hypoplastic on the lingual surface and extended further apically on this than on the labial aspect. An adjacent section, not reproduced here, moreover, showed a small hypoplastic pit in the enamel at the incisal edge.

The remaining four teeth were badly deformed (+2, +03, —04, —05), only —05 bearing any resemblance to its normal counterpart. On none of the teeth, however, could a definite root be found with the unaided eye. The crown of —05 was flattened in a labio-lingual direction, especially at the neck. +2 and +03 also had flattened crowns, but it was impossible to decide in which direction the flattening had occurred.

Figures 1 to 4 give some idea of the shape of —05, —04, +02, and of +2. All the sections are cut in a labio-lingual direction through the longitudinal axes of the teeth.

HISTOLOGIC AND MICRORADIOGRAPHIC EXAMINATION

The enamel could be studied on the ground sections of +02, +03, —04 and —05, on the microradiograms of these sections and on the decalcified sections of +2, where remains of the enamel were found near the cervix of the tooth.

Hypoplastic pits were evident in all the teeth, being least pronounced in the case of +02. On the occlusal surface of —05 the enamel was thinner than normal. On the lingual aspect the cemento-enamel junction was nearer the masticatory surface than normal (Figs. 1 a & b). On —04, which bore less resemblance to a tooth than —05, the enamel had a lobate appearance (Figs. 2 a & b), and on +02 it was thin on the lingual surface, extending unusually far towards the "apex" (Fig. 3).

Decalcification had removed much of the enamel from the sections of +2 (Fig. 4). What remained indicated an irregular course of the enamel prisms at the neck, with wavy incremental lines (Fig. 5), suggesting disturbance of the development of the enamel. In some places the ameloblasts were cubic while in others they were low and degenerated. In this region the ameloblastema was separated from the enamel by a band, staining dark

blue in htx and having signs of degenerative calcification (Fig. 5). On other sections of +2 parts of the ameloblastema were entirely calcified (Fig. 6). It was confirmed by the microradiograms that the blue stained bodies observed on the decalcified sections had a high mineral salt content. Round or irregular formations with high x-ray absorption could in many cases be observed near the surface of the enamel (Figs. 11 & 12). In +03 the accumulations of such mineral salt globuli were greatest in the vicinity of the enamel hypoplasia; that is, at the site of the retarded development of the enamel. On decalcified sections inflammatory cells were observed in the neighbourhood of the ameloblastema (Figs. 5, 6 & 9).

Fig. 1 a. Unstained longitudinal ground section of —05; transmitted light; thickness 230 μ.

The enamel presents evidence of hypoplasia occlusally and buccally. On the buccal aspect the enamel extends further apically than on the lingual. Possibly the well-defined line of Retzius corresponds to the neonatal line. The dentine is regular in structure but signs of poor mineralization (interglobular spaces) are evident near the enamel. The tooth tapers considerably towards the stunted root. The section is somewhat to the side of the narrow opening of the pulp at the "root tip". × 15.

Fig. 1 b. Microradiogram of the ground section in Fig. 1 a.

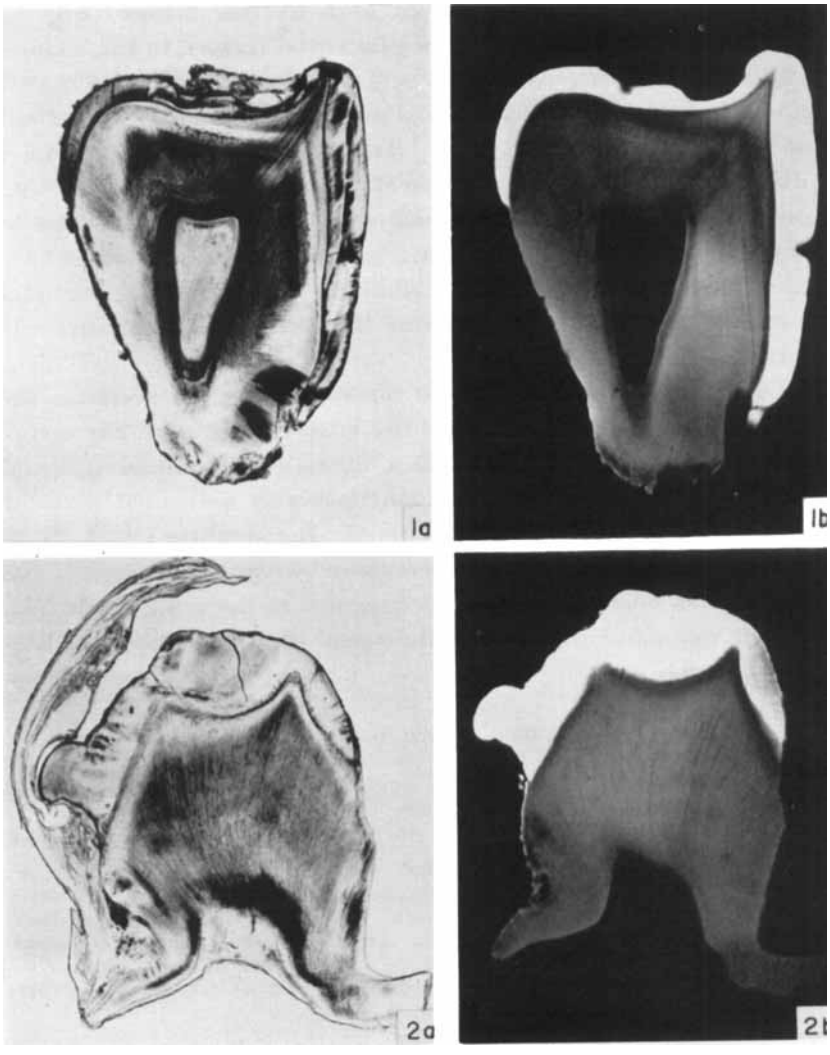
The buccal enamel hypoplasia is clearly visible. In the dentine there are interglobular spaces evidenced as dark "grainy" regions of low x-ray absorption (poor mineralization). Thin bands with alternately high and low mineralization pass near the enamel (incremental lines). The dentine is otherwise evenly mineralized. (The dark area near the "root tip" is due to the fact that the section is thinner there). × 15.

Fig. 2 a. Unstained longitudinal ground section of —04; transmitted light; 100 μ.

The enamel is lobate and does not extend far apically especially not on the right side. Some of the enamel on this side has possibly been lost during the operation or when the section was cut. A large number of enamel spindles are seen in the right cusp. The dentine is regular in structure. The root is deformed and stunted and the pulp widens in an apical direction as the dentine walls diverge (*cf.* —05). The tooth is partly surrounded by soft tissue in which round bodies can be discerned near the surface of the enamel. × 20.

Fig. 2 b. Microradiogram of the ground section of Fig. 2 a.

In one cusp the enamel is darker than elsewhere due to the poor mineral content of the cusp. Thin bands with alternately high and low mineralization occur in the enamel of the cusp. Their course is the same as that of the lines of Retzius. In the region corresponding to the soft tissue in Fig. 2 a there are round bodies of high x-ray absorption, reminiscent of mineral salt globuli. The dentine is evenly mineralized. To the left on the outside of the root there is a small dark area with low x-ray absorption (poor mineral salt content). This corresponds to an area with a cementum-like structure on the section. × 20.



Figs. 1 a, 1 b, 2 a, 2 b.

Spaces in the enamel were found on decalcified sections of +02 (Figs. 7 & 8) and on microradiograms of +03 (Fig. 10). It appeared from the decalcified sections that the spaces were of two types: those partly filled with hyaline bodies (Fig. 7) and those having the appearance of a vessel formed in the enamel with red blood corpuscles in a state of decomposition (Fig. 8). It was not possible to ascertain the type of the spaces observed on the microradiogram. The irregular course of the enamel prisms was observed not only on the decalcified sections of +02 but also clearly on the microradiograms of +03. This kind of wavy arrangement of the prisms occurred only at places where the enamel was hypoplastic or lobate. In the region of the enamel with normal surface configuration the course of the prisms was normal.

The dentine could be studied on all the ground sections, decalcified histologic sections and the microradiograms. The structure was regular with straight, uniformly distributed dentinal tubules (Fig. 13). Only in the most severely deformed parts of the dentine were the tubules curved. The dentine could, however, nowhere be described as irregular in type. The shape of the dentine was normal in +02 but irregular in the other teeth. The dentine was more deformed in the apical than the coronal regions (Figs. 1—4).

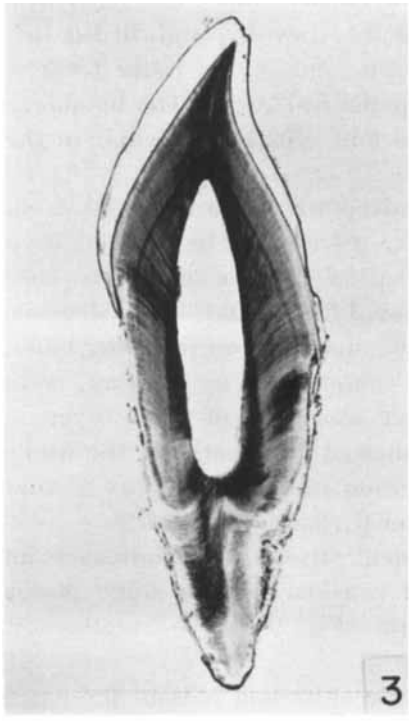
Fig. 3. Unstained longitudinal ground section of +02; transmitted light; thickness 100 μ.

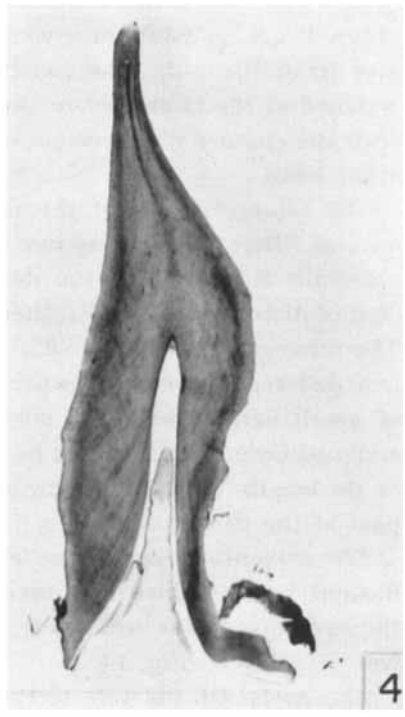
The enamel is thin on the lingual aspect, has a shallow hypoplastic area and extends further apically than the enamel on the labial aspect. The dentine is regular in structure. The tooth is fairly normal in shape. × 9.

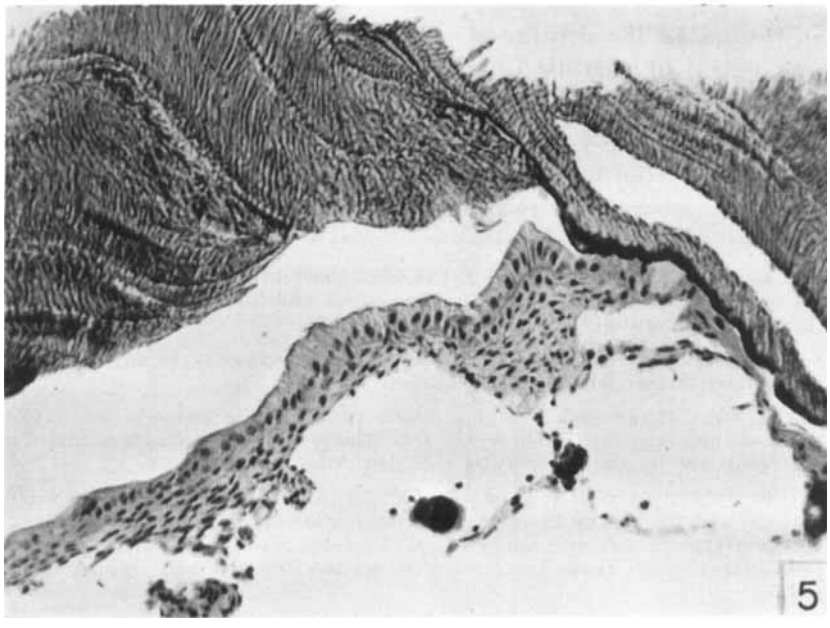
Fig. 4. Decalcified section through + 2.

Large parts of the enamel are absent, the remains being visible as a dark U-shaped area at the "tooth neck" low to the right. The upper part of the dentine is normal in shape, the lower part (the "root") is deformed and stunted. There is a similar tendency to divergence of the apical part of the pulp chamber walls as in —04 (*cf.* Figs 2 a & b). The pulp has come away from the walls. Stain: Bock. × 15.

Fig. 5. Decalcified enamel from another section of the same tooth as in Fig. 4. The enamel prisms are irregular in their arrangement and they are crossed by wavy incremental lines. The ameloblastema that has partly come away from the enamel, consists to some extent of cubic cells. In the lower part of the figure the ameloblastema has degenerated and is now separated from the enamel by a thin dark layer, poor in structural detail (degenerative calcification?). The dark, rounded regions on the left are reminiscent of mineral salt globuli (*cf.* microradiogram in Fig. 2 a). Near these globuli occasional leucocytes are seen. Stain: Bock. × 325.







Figs. 3—5.

In —05 the dentine walls converged toward the "root tip" (Figs. 1 a & b), while in —04 and 2+ they diverged. In the former tooth the pulp was constricted, and in the latter teeth it widened at the boundary to the periapical tissue. The boundary with the enamel was even in —05 and +02 but irregular in the other teeth.

The mineralization of the dentitne was little different from normal. The microradiogram of —04 (Fig. 2 b) showed even mineralization, and on the decalcified sections of +2 no sure sign of disturbed mineralization could be observed in the dentine. The microradiograms of +02, +03 and —05, on the other hand, revealed regions of low mineral content. These, however, were of small extent, with the possible exception of —05, where a widened Owen's band could be followed for practically the whole of its length (Fig. 1 b). The position of this band was at that part of the dentine normally formed shortly after birth.

The cementum showed no evident structural deviations from normal but its thickness varied considerably. In some places the cementum was unusually thin (Fig. 9) while in others it was very thick (Fig. 14).

The roots. Of the five teeth only +02 had a well developed root. The roots of the other teeth were malformed and abnormally short. In the dentine of —05 there was near the "root tip" a fragment of necrotic immature bone (Fig. 15).

The pulp was normal in structure and the odontoblastema regular. In —05 a few inflammatory cells were found at the transition of the pulp and the "periapical" tissue. Similar findings were recorded in respect of +2, and in addition there were

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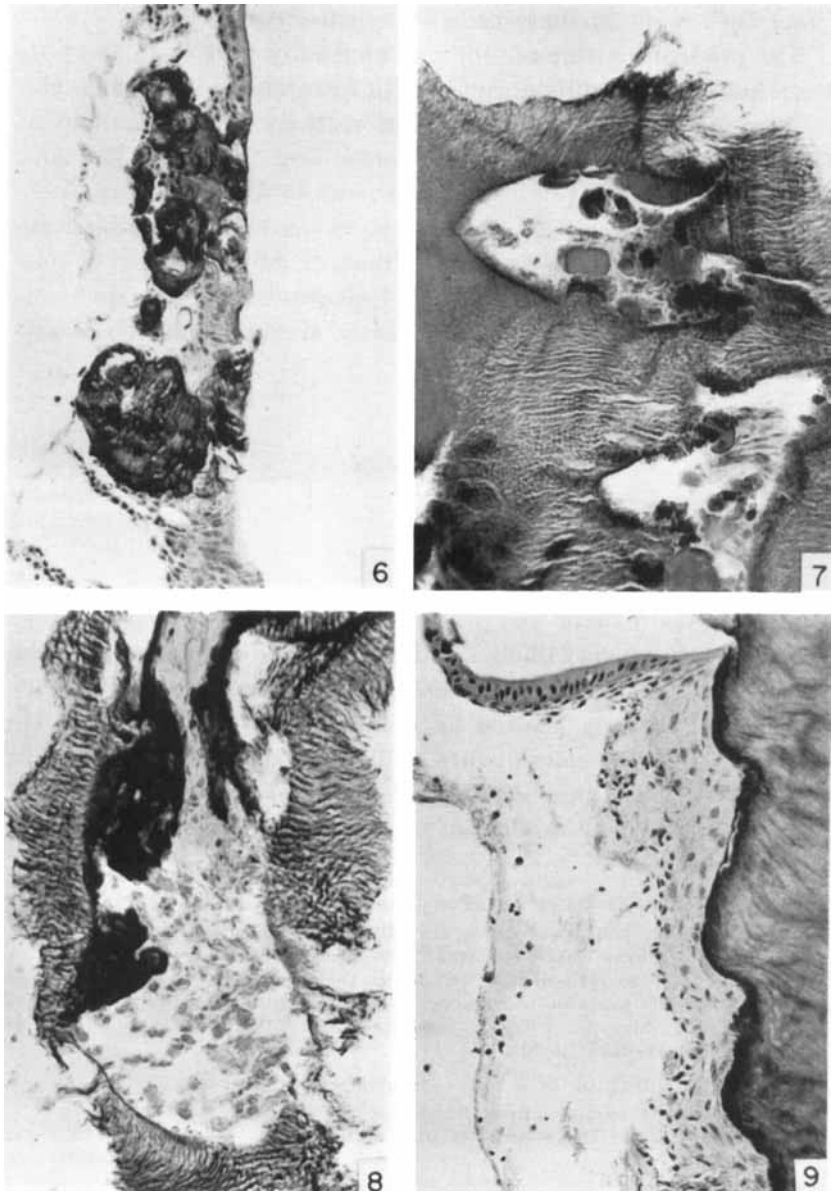
Fig. 6. Decalcified section of +2. Calcified ameloblastema. The dark areas are accumulations of calcium salts. Uppermost and lowermost on the figure there are exudate cells. Stain: Htx-eosin. × 325.

Fig. 7. Decalcified enamel of +2. Irregular spaces, partly filled with hyaline bodies. Stain: Htx-eosin. × 325.

Fig. 8. Decalcified enamel of +2. Blood vessel in the enamel. The vessel contains decomposing erythrocytes and poorly defined necrotic substance. The walls are in places markedly calcified. Stain: Bock. × 450.

Fig. 9. Decalcified section from the neck of +2. The root surface is wavy and covered by a thin layer of cementum and cementoblastema. High up on the figure is seen the union of the ameloblastema and the neck of the tooth. On the left there is necrotic tissue and exudate cells. Stain: Bock. × 325.



Figs. 6—9.

exudate cells near the odontoblastema some millimetres from the "root tip". Some of these cells were leucocytes.

The predentine zone of +2 was unusually narrow in the pulp horn but it assumed its normal width towards the apex (Fig. 13).

The jaw bone. The decalcified sections showed immature, fibrillar bone with little sign of remodelling (Fig. 16). The bone fragments were in places necrotic and in their vicinity there were accumulations of inflammatory cells with marked infiltration of leucocytes (osteomyelitis). A detail worthy of note was the occurrence of stratified epithelium between the bone trabeculae. The epithelium was partly necrotic and at one site manifestly keratinized (Fig. 16).

DISCUSSION

In this case, as in that previously reported, the radiographs revealed that most of the deciduous teeth were impacted and that many were much deformed. In the vicinity of the few erupted deciduous teeth there was profound marginal alveolar osteitis in both cases. It appears likely that he abovementioned changes were associated with the systemic disease but the question is whether they were directly caused by it or whether they occurred secondarily, e.g. as a consequence of the disturbance in the development of the bone that takes place in osteopetrosis.

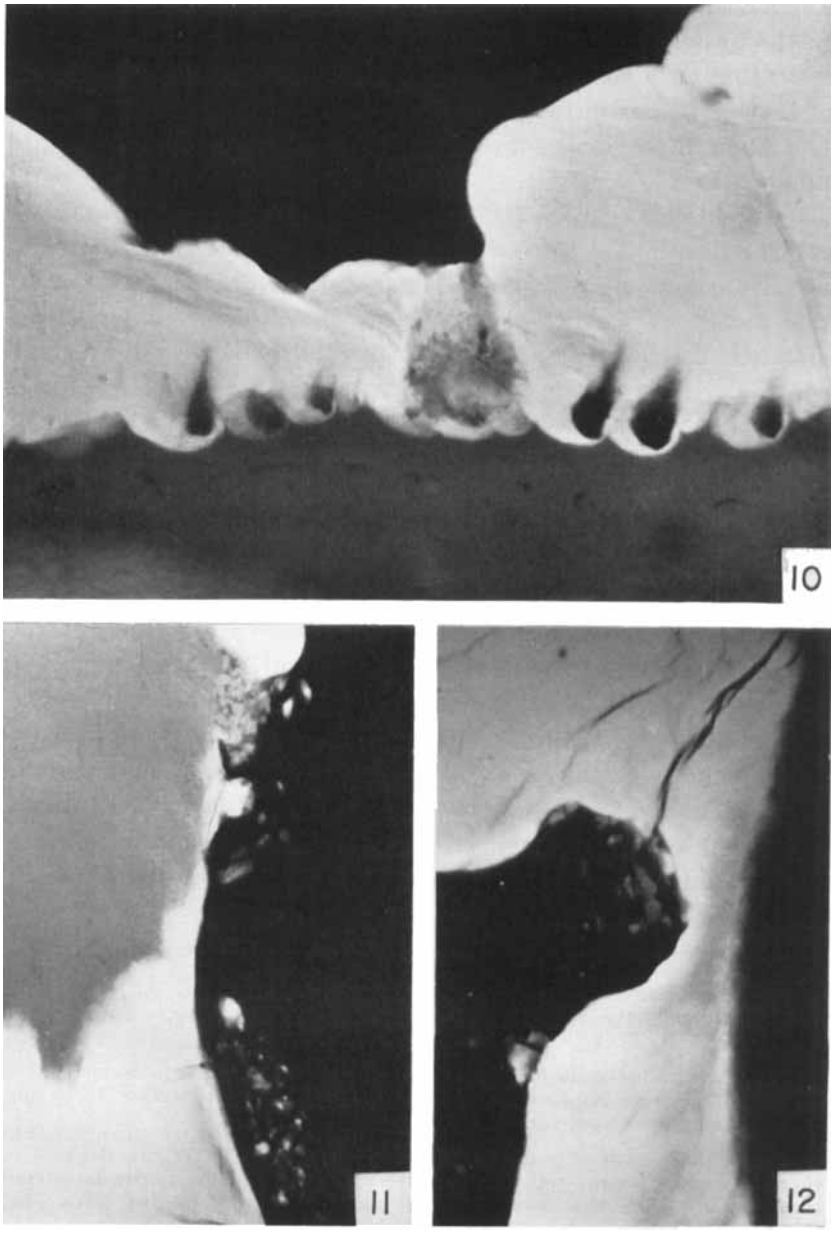
In the previously described case there was definitely enamel

Fig. 10. Microradiogram of a ground section of +03, 100 μ in thickness. In one part of the microradiogram corresponding to the dentine (lowermost in the figure) there are small interglobular spaces with low x-ray absorption. The surface of the enamel is pitted and the x-ray absorption varies in various parts of the enamel (irregular mineralization). Elsewhere in the enamel there are wavy incremental lines with alternately large and small mineral content. × 90.

Fig. 11. Microradiogram of a 100 μ ground section of —04.

The dentine is evenly mineralized and its junction with the enamel is in places irregular. The enamel is pitted and at one place very thin. Near the centre of the picture there is no enamel at all. Instead, the dentine passes directly into a hard substance with a "grainy" structure and irregular mineral salt content. Near the surface of the enamel there are irregularly shaped bodies with high mineral salt content. × 80.

Fig. 12. Microradiogram of a 100 μ ground section of +03. There is marked hypoplasia of the enamel, the groove being partly filled with round or irregularly shaped bodies with high mineral salt content. × 80.



Figs. 10--12.

hypoplasia of one of the three deciduous teeth studied (+02). In the present case, the enamel was so deformed in three of the teeth that they were impossible to identify other than by virtue of their positions in the jaws (+2, +03, —04), and in the other two teeth there were pits in the enamel suggestive of enamel hypoplasia (+02, —05).

Two possible reasons for the enamel defects may be considered. The first is that the disturbance in the enamel formation was directly connected with the skeletal disease, or, in other words, the factor or factors giving rise to the skeletal changes also affected the amelogenesis. This possibility is to some extent contradicted by the fact that the enamel formation of +02 was practically normal, while that of +03 and —04 was extremely irregular (*cf.* rickets where teeth developing simultaneously present similar defects). It cannot be entirely excluded, however, that different teeth, or groups of them, have different sensitivity to the general factors suggested above; this might explain the dissimilarities evident between the teeth.

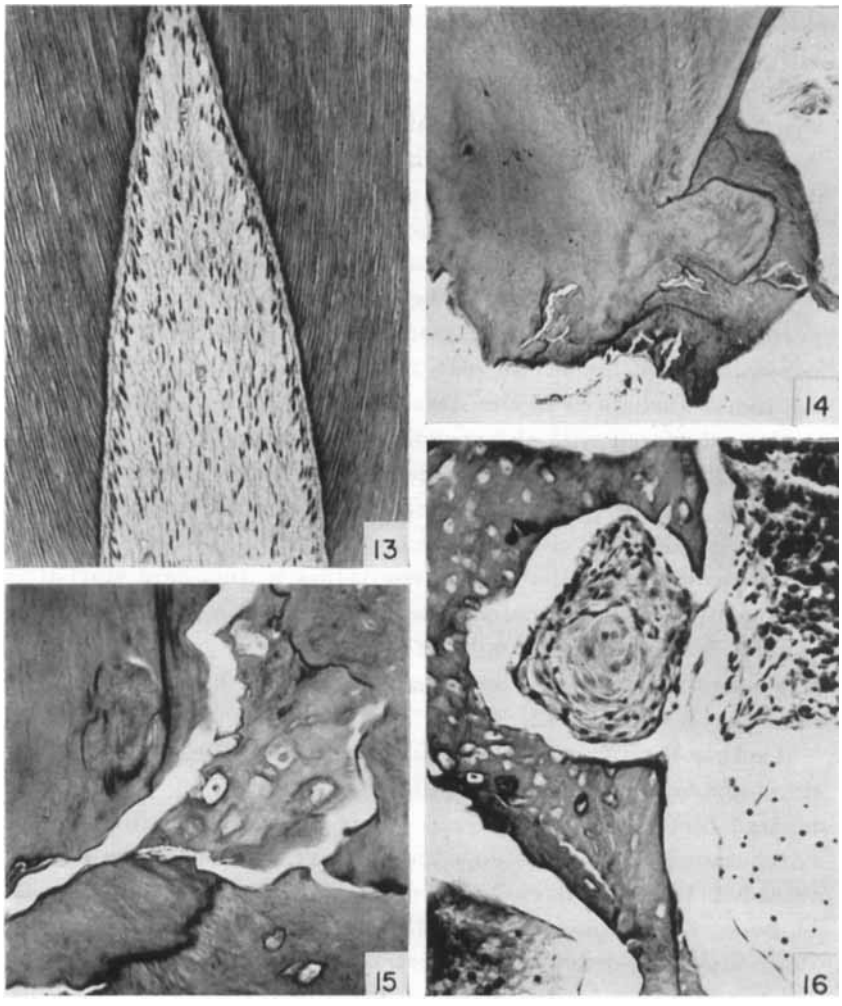
The second possibility is that local factors were the cause of the disturbances in the enamel development. Support of this is provided by —04. The disturbances in the development of the enamel due to systemic factors generally have their counterpart in the dentine in the form of disturbance in the mineralization (*cf.* rickets and fluorosis). In —04 which had badly malformed and lobate enamel, the dentine was evenly mineralized (see the microradiogram in Fig. 2 b). This tends to disprove the presence of a systemic causal factor.

Fig. 13. Decalcified section of +2. The dentine is regular, the predentine zone narrow and the odontoblastema rather regular. The pulp is normal in structure. Stain: Bock. × 325.

Fig. 14. Decalcified section of the "root tip" of —05. There is an irregular deposit of cellular cementum on the stunted root. (The section is to the side of the narrow root canal). Stain: Htx-eosin. × 70.

Fig. 15. Enlargement of a detail from the section in Fig. 14. On the left is the dentine and on the right the cellular cementum. In the centre is immature necrotic bone that has been enclosed in the root. Stain: Htx-eosin. × 450.

Fig. 16. Decalcified section of bone from the region of +2. The bone is immature, partly necrotic and surrounded by necrotic soft tissue and exudate cells. In a nitch in the bone there is stratified epithelium with evidence of keratinization. Stain: Htx-eosin. × 325.



Figs. 13—16.

In the bone fragments from the vicinity of the teeth under study evidence of non-specific chronic osteomyelitis was found. There was also an infiltration of inflammatory cells near the roots of +2 and —05; further, similar cellular infiltration was observed in the vicinity of the ameloblastema of +2. There was thus a chronic nonspecific inflammatory change in the jaw, and it seems likely that this may be *one* local factor in the disturbed development of the teeth. The degenerative calcification of some of the ameloblasts of +2 was possibly connected with the osteomyelitis. In a region where the ameloblastema is so impaired normal development of the enamel is hardly to be expected. It is possible that the chronic osteomyelitis was not only responsible for the disturbances in the development of the enamel but that it also interfered with the growth of the roots.

Osteomyelitis of the jaw is not unique for our case of osteopetrosis. *Thoma* (1950) considers its occurrence fairly common in connection with osteopetrosis.

It is conceivable, that the osteomyelitis in this case started in the vicinity of the deciduous teeth, where a marginal alveolar osteitis was evident. Gingivitis and marginal alveolar osteitis in connection with the shedding process has been described in detail by *Westin* (1942).

Another local factor that may conceivably have influenced the development of the teeth should be mentioned. A condition of normal tooth growth and eruption is active remodelling of the surrounding bone. In osteopetrosis the resorption of the bone is retarded. In the "root tip" of —05 there were enclosed fragments of bone, a pathologic feature that might well be attributed to the reduced tendency of bone resorption.

Lack of space for the growing dental germ might in these patients be another contributory factor in the disturbed development of the enamel and the roots. The tendency observed in osteopetrosis for the teeth to remain unerupted is probably connected both with the absence of bone resorption and with the osteomyelitis and the arrested development of the roots.

According to *Weinmann, Svoboda & Woods* (1945) who studied changes occurring in the teeth in some cases of amelogenesis imperfecta, the reduced enamel epithelium forms a protective layer on the enamel. Where there is premature de-

generation of this epithelium the process of eruption of the tooth is disturbed. In the present material the ameloblastema was found to be impaired. In the light of the opinion advanced by *Weinmann* and co-workers the impaction of the teeth in osteopetrosis might be ascribed in part to injury to the enamel organ.

The systemic and local factors as the causes of disturbances in the development of the teeth in osteopetrosis have so far been discussed separately. Their combined occurrence is not an inconceivable cause of the anomalous development of the teeth.

A detailed study of the decalcified enamel of +2 revealed the occurrence of spaces or notches. The spaces seen in Fig. 7 might possibly be considered as obliquely sectioned hypoplastic defects of the enamel containing illdefined keratin-like hyaline bodies, while the space in Fig. 8 with fair certainty consisted of the lumen of a blood vessel containing erythrocytes in varying stages of decomposition. Blood vessels in the enamel organ have been described in detail by *Jump* (1938). Such vessels are, however, generally not enclosed in the enamel, and the reason for one being so in this case is difficult to understand. A local abnormal tendency to proliferation of the ameloblastema is not impossible. Again, the vessel may have been enclosed in the enamel in connection with a local inflammation in which the vessel would have been a part of the granulation tissue. Both these explanations are, however, purely hypothetical.

Another feature that is difficult to explain and that is to some extent associated with the above are the spaces in the enamel with little or no mineral content near the dentine of +03 and visible in the microradiograms (Fig. 10). It is not improbable that these spaces correspond to one of the two types of space observed on the decalcified sections.

The dental changes have been discussed above against the background of their conceivable connection — direct or indirect — with the skeletal disease from which the patient was suffering. As in the earlier case one cannot exclude the presence of other factors. A disturbance of, for example, the mineral and vitamin supply shortly after birth may have contributed to the enamel defects and the poorly mineralized areas in the dentine. However, it appears most likely that the dental changes in osteo-

petrosis are connected wholly or principally with the skeletal disease which, on the other hand, probably has no primary influence on the development of the teeth. The disturbances in the dental development appears instead to be indirectly connected with the skeletal disease and to be caused by the retarded remodelling process of the bone in combination with the apparently not uncommon chronic osteomyelitis of the jaw bone.

SUMMARY

Five teeth with parts of the covering alveolar bone from a five-year-old girl suffering from osteopetrosis were studied by means of histologic and microradiographic techniques. None of the teeth was erupted, all of them having been removed surgically. One of them was almost normal in shape, while the others were badly deformed.

The development of the enamel was disturbed in all teeth studied and the crowns were so malformed in three instances that they could not be identified other than by virtue of their position in the jaws. In the enamel of one of the teeth a blood vessel was found. Near the ameloblastema there were inflammatory cells of which some were leucocytes. Such cells were met with also at the site where the pulp opened towards the surrounding soft tissue.

The dentine presented practically even mineralization and the structure was regular. In limited areas there were sites of low mineralization with interglobular spaces.

The roots were stunted and deformed and in one instance necrotic bony tissue was found to have been enclosed in the root.

The bone was immature and partly necrotic and in the marrow there was marked cellular infiltration with numerous leucocytes (osteomyelitis). Epithelial cells, partly arranged as epithelial pearls, occurred between the bone trabeculae.

The connection between the skeletal disease and the observed pathologic changes in and near the teeth are discussed. The fact that the teeth had not erupted, together with the disturbance of the development of the enamel and the roots may no doubt in some measure be attributed to local factors indirectly connected with the skeletal disease.

RÉSUMÉ

ÉTUDES SUR LES TISSUS DENTAIRES MINÉRALISÉS

VII. Modifications dentaires observées dans un cas d'ostéopétrose (suite)

Cinq dents et certaines parties de l'os alvéolaire avoisinant provenant d'une fillette de cinq ans atteinte d'ostéopétrose ont été examinés selon les techniques histologique et micro-radiographique. Les dents n'avaient pas percé et avaient été extraites chirurgicalement. L'une était de forme presque normale tandis que les autres étaient très difformes.

Sur les dents examinées la croissance de l'émail était troublée; trois des couronnes étaient si informes que seule leur position dans le maxillaire permettait de les identifier. Au voisinage de l'améloblastème se trouvaient des cellules inflammatoires, dont quelques leucocytes. Des leucocytes se rencontraient également dans la région de contact entre la pulpe et les tissus avoisinants.

La dentine présentait une minéralisation pratiquement uniforme et à structure régulière. Dans certaines régions on observait une minéralisation imparfaite présentant des espaces interglobulaires.

Les racines étaient rabougries et difformes; dans l'un des cas des particules de tissus osseux nécrosés avaient été encerclées par la racine.

La formation osseuse était inachevée et les tissus osseux atteints de nécrose. Dans la moëlle on pouvait noter une nette infiltration cellulaire avec un grand nombre de leucocytes, (ostéomyélite). Entre les trabécules se trouvaient des cellules épithéliales distribuées en perles.

La relation est étudiée entre la maladie du squelette et les modifications pathologiques observées sur les dents et les tissus avoisinants. La non-éruption des dents et la croissance troublée de l'émail et des racines sont indubitablement imputables en partie à des facteurs locaux indirectement en relation avec la maladie osseuse.

ZUSAMMENFASSUNG

STUDIEN AN MINERALISIERTEN ZAHNGEWEBEN

VII. Veränderungen an Zähnen bei Osteopetrosis.
Eine Fortsetzung früherer Untersuchungen

Fünf Zähne und die angrenzenden Teile des Kieferknochens eines fünfjährigen, an Osteopetrosis leidenden Mädchens wurden histologisch und mikroradiographisch untersucht. Keiner der Zähne war durchgebrochen, alle waren operativ entfernt worden. Ein Zahn hatte eine normale Form, die anderen zeigten mehr oder minder starke Missbildungen. Die Schmelzbildung war bei allen untersuchten Zähnen gestört. Die Kronen von drei Zähnen wiesen derart starke Missbildungen auf, dass eine Identifizierung nur auf Grund ihrer Lage im Kiefer möglich war. Bei einem Zahn wurde ein Blutgefäss im Schmelz gefunden. In der Nähe des Ameloblastems kamen Exsudatzellen vor, von denem ein Teil Leukozyten waren. Solche Zellen befanden sich auch an der Öffnung der Pulpa zum umgebenden Weichgewebe hin.

Das Dentin war, im grossen und ganzen, gleichmässig mineralisiert, und die Struktur war normal. Innerhalb begrenzter Gebiete wurden jedoch auch Zonen mit geringerer Mineralisierung (Interglobularräume) angetroffen.

Die Wurzeln waren verkürzt und zeigten Missbildungen. In einem Fall wurde nekrotisches Knochengewebe, in die Wurzel eingebaut, vorgefunden.

Das Knochengewebe war unreif und teilweise nekrotisch. In der Umgebung der Knochenbalken wurden starke Zellinfiltrationen mit kräftigem Leukozyteneinschlag gefunden (Osteomyelitis). Epithelzellen, teilweise in Form von Epithelperlen, kamen zwischen den Knochenbalken vor.

Der Zusammenhang zwischen der Skeletterkrankung und den beobachteten pathologischen Veränderungen in den Zähnen und deren Umgebung wird diskutiert.

Der unterbliebene Durchbruch der Zähne, sowie die Störungen in der Schmelz- und Wurzelentwicklung, können sicherlich zu einem grossen Teil durch lokale Faktoren, die im indirekten Zusammenhang mit der Skeletterkrankung stehen, erklärt wer-

den. Die Möglichkeit, dass der allgemeine, die Skeletterkrankung verursachende Faktor auch direkt die Entwicklung der Zähne beeinflusst, kann jedoch nicht ausgeschlossen werden. Gewisse Anzeichen sprechen jedoch gegen diese Möglichkeit.

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