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STUDIES ON MINERALIZED DENTAL TISSUES

III. Dental changes in a case of osteopetrosis

by

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INTRODUCTION

Osteopetrosis, or Albers-Schönberg disease, is an interesting hereditary disorder of the skeleton. It appears to be transmitted as a simple recessive mendelian character and the disease has not infrequently been observed in the offspring of parents who are closely related. The disturbance is often recognized at birth or soon afterwards. The symptomatology is related to the fragility of bone -- the sclerosis of the skeleton giving anemia — and to the narrowing of the foramina of the skull leading to deafness and impairment of vision. Skeletal radiographs from the skeleton in cases of osteopetrosis show very dense and characteristic bone structures. Microscopic examination of sections from the long bones reveals calcified cartilage and bone tissue throughout the whole medullary cavity and the compact bone tissue consists partly of immature coarse-fibrillar bone. In a previous paper (*Engfeldt, Engström & Zetterström, 1954*) results of a biophysical investigation of bone tissue in osteopetrosis were presented. A study of the etiology of the anemia has also been performed (*Engfeldt, Karlberg & Zetterström, 1954*). It was concluded that in osteopetrosis there is a disturbance in the mechanism leading to replacement of the immature bone tissue by adult bone. The bone tissue formed in this disease undergoes rebuilding, but once formed the calcified tissue remains unchanged in type, i.e. it does not develop into mature bone. On the basis of estimations of the endogenous carbon monoxide production it was concluded that the anemia occurring in cases of osteopetrosis is at least partly of haemolytic origin.

In the literature there are only a few descriptions of dental tissue in osteopetrosis. According to *Thoma* (1950), who has reviewed the literature, the teeth are poorly calcified and subject to caries. Delayed or incomplete eruption is not uncommon. Osteomyelitis sets in easily in the jaws in connection with caries and pulpitis. Cases have been reported in which osteomyelitis of the jaws was the cause of death. As far as we are aware no histological description of the dental tissues in cases of osteopetrosis has been published.

The aim of the present investigation is to give a report of the histological and microradiographic findings in dental tissue from a case of osteopetrosis.

CASE RECORD

A full report on the clinical findings in this case has already been published and only the main features will be mentioned here. The patient, a boy, was 6 years old when first admitted to the paediatric clinic at Karolinska sjukhuset. At the age of 10 months a skeletal disease was diagnosed and the examination disclosed features characteristic of osteopetrosis. At the age of 2 years the patient presented impairment of vision, atrophy of both optic nerves, and anemia. On examination in the paediatric clinic when the boy was 6 years he disclosed among other things severe anemia, almost blindness and impairment of hearing and also characteristic roentgenograms (Fig. 1). A biopsy was made from the rib and histological findings showed features characteristic for osteopetrosis which was also the case of the microradiograms (Fig. 2).

It was also of interest to note that only 7 deciduous teeth were present, namely the upper incisors and first molars and the lower left first molar. All of them were decayed. No permanent teeth were present.

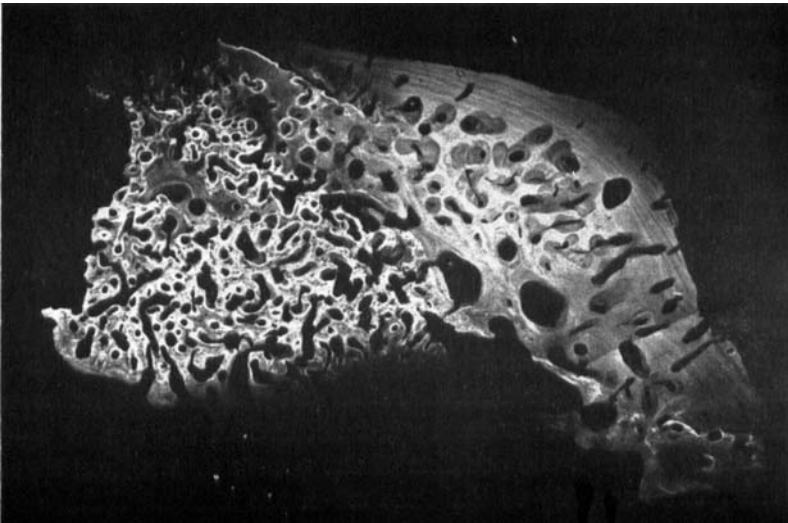
Oral radiographic examination revealed aplasia of several permanent teeth, including the lower premolars and canines, retention of most of the deciduous teeth, delayed development of the permanent tooth germs and malformation of the crowns of the upper permanent incisors. (On account of the patient's bad condition it was difficult to obtain good x-ray pictures).

Fig. 1. Roentgenogram of tibia from a case of osteopetrosis showing typical dense bone tissue with cross striation of the metaphysis.



Fig. 2. Microradiogram from an undecalcified cross section from the 7th rib. Note the normal bone tissue to the upper right with Haversian systems with different degree of mineralization (different x-ray absorption). The irregular bone tissue typical for osteopetrosis is seen to the left. It shows among other things broad and highly mineralized cementing lines and coarse fibrillar bone tissue.

12X.



Some months after this hospitalization the boy was readmitted because of increasing anemia and an acute alveolar periostitis with fever. The periostitis was cured by antibiotics.

MATERIAL AND METHODS

The material available comprises three deciduous teeth fixed in formalin (02+, +02 and +04). 02+ was decalcified, embedded and cut for histological examination while ground sections 60—100 μ in thickness were prepared from the other teeth after embedding in methyl methacrylate. The content and distribution of mineral salts in the dental hard tissues were determined by microradiography. A Philips diffraction unit with a copper target served as radiation source. The tube was operated at a P.D. of 25 kV at 12 mA. The ground sections were mounted on thin mica sheets and placed in close contact with the photographic emulsion. The distance from focus to emulsion was 25 cm. The microradiograms were registered on Eastman Kodak spectroscopic plates nr 649 which have a resolving power of about 1000 lines per mm. The microradiograms were enlarged by photomicrography on Kodak O 250 plates.

RESULTS

The three teeth had several features in common.

1. The roots presented superficial resorptions (Figs. 3, 4, 7 & 10). These resorption processes did not show the same behaviour as is normally seen in deciduous teeth during the eruption of the permanent teeth. In the latter case the resorption usually starts at the root tip or in the bifurcation and proceeds towards the crown until the root is completely resorbed. Instead the resorption in this case was of a widespread type which may occur in association with osteitis of the jaws. As this patient had had several attacks of alveolar osteitis the abnormal root resorption might have been due to them, thus having no direct connection with the skeletal disease.

2. As is seen in the microradiograms, there were in the crown dentine areas of poor mineralization (Figs. 4 & 6). These areas in the dentine are normally mineralized at the age of 6—12 months. Although the position of the poorly mineralized tissues corresponds roughly to the time when the skeletal disease was

diagnosed, it does not necessarily mean that the disturbed mineralization was associated with that disease. Similar changes are found in other disturbances of the mineral metabolism -- for instance in rickets. Cf. *Westin's* "winter dentine" (1937).

A prominent feature of the microradiograms is the even and high mineralization of that part of the dentine which was formed during the intrauterine life (Fig. 6). The same is true of the enamel (Fig. 8).

3. In the circumpulpal dentine of the roots the tubules were scattered and irregularly twisted (stratified secondary dentine. Figs. 7 & 11). It is possible that this change has something to do with the general skeletal disease; however, it must be stated that similar changes may occur as a result of local factors -- pulpitis, for instance -- or result from vitamin C deficiency (*Westin*, 1931).

In one of the teeth cementum caries was observed (Fig. 10). Beneath this lesion poorly mineralized secondary dentine was found with cell inclusions in the predentine zone (Fig. 12). Caries in the cementum is not a common feature in a six-year-old child and in this case it could perhaps be attributed to the fact that the surface of the root was exposed as a result of the alveolar osteitis.

DISCUSSION

It is of course not possible to draw any definite conclusions concerning the connection between the skeletal disease and the changes in the dental tissue found in this case. However, with the aid of the histopathological and microradiographic investigations of the teeth it is possible to date the following changes. About six months after birth there was a disturbance in the mineralization of the teeth; this was evident from widened Owen's lines, easily detected in the microradiograms (Figs. 4 & 6). The same disturbed mineralization can also be traced as hypoplastic pits in the enamel of +02 (Fig. 9). During the following years the development of the teeth was apparently normal until the roots were fully, or almost fully, developed. At an age of about 4 years the deciduous teeth are usually fully developed. If it is assumed that the development of the roots in this case was no more rapid than is normal, a new disturbance

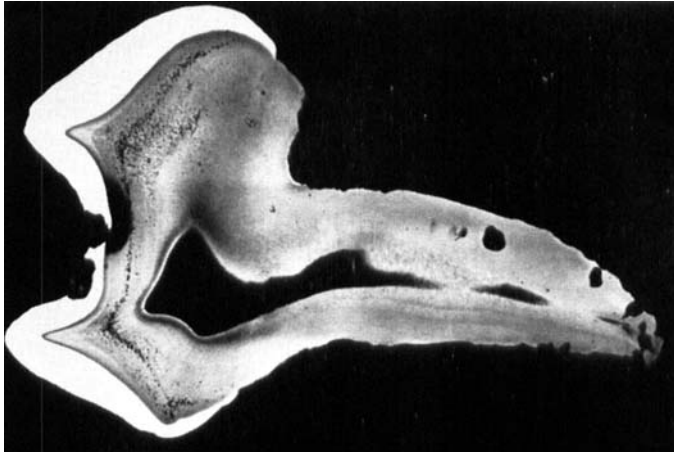


Fig. 4. Microradiogram of the section in Fig. 3. The widened Owen's line is plainly visible as a band with low x-ray absorption. $\bar{7}\times$.

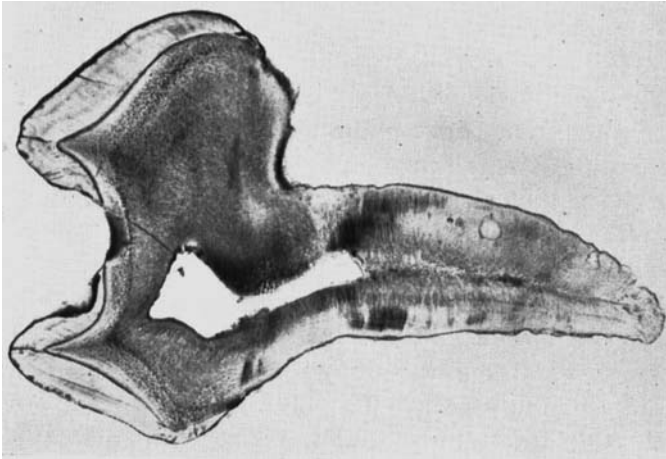


Fig. 3. 100 μ thick ground section through +04 in transmitted light. Widened Owen's line in the crown dentine. Superficial resorptions along the root surface. Fissure caries. The neonatal line is visible in the left cusp. (Part of the enamel lost near the fissure). $\bar{7}\times$.

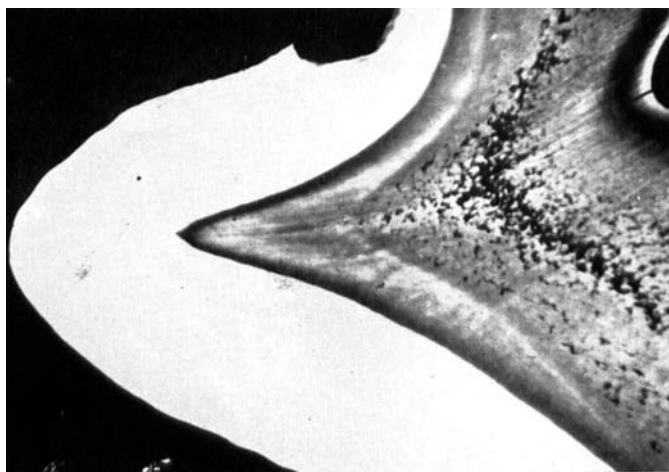


Fig. 6. Microradiogram of the same structures as in Fig. 5. The prenatally formed dentine has a higher x-ray absorption than the postnatal dentine. The latter is poorly mineralized which is indicated by the great interglobular spaces, which are in part arranged as a widened Owen's line. 30 \times .



Fig. 5. Detail of the left cusp in Fig. 3. 30 \times .

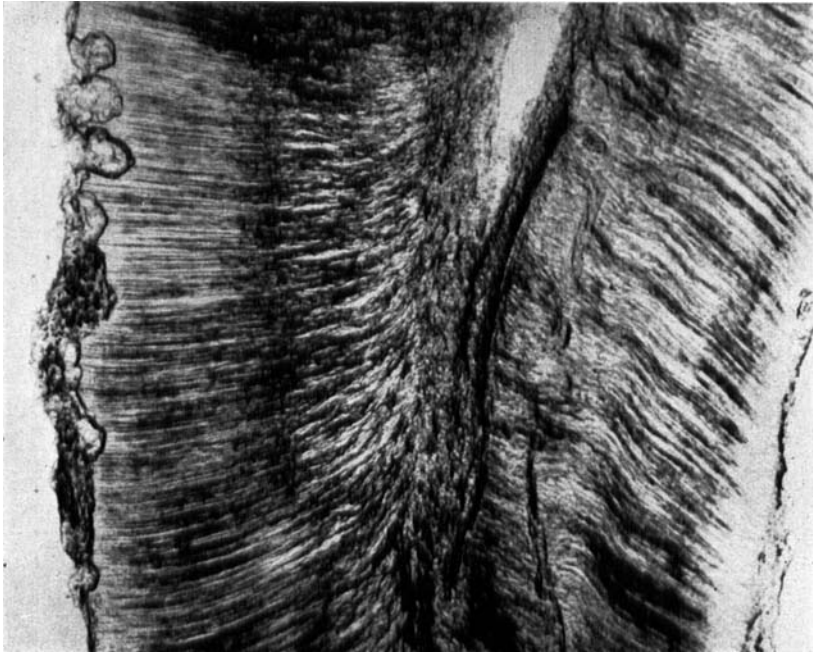


Fig. 7. Longitudinal ground section through the lingual root of +04. Resorption cavities along the root surface, in part filled with bone tissue. The circumpulpal dentine is of an irregular type.
40 \times .

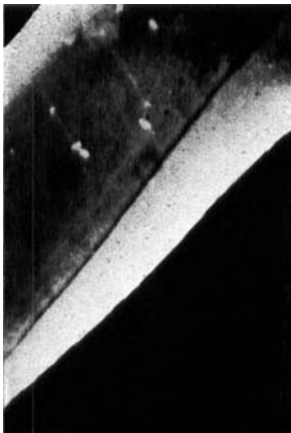


Fig. 8. Microradiogram of the cusp tip of +04. The neonatal line visible as a band with low x-ray absorption. The prenatally formed enamel has a higher x-ray absorption than the postnatal enamel. The superficial part of the enamel has a high x-ray absorption. (The dark area in the lower right corresponds to the dentine).
40 \times .



Fig. 9. 60 μ thick ground section through +02. The neonatal line visible as a dark band in the enamel. Hypoplastic pits in the enamel.
30 \times .

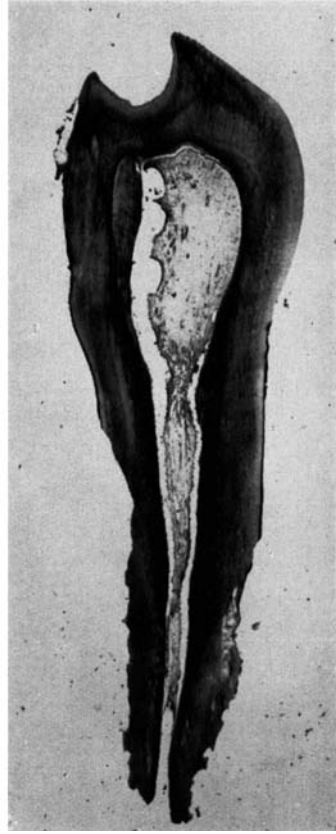


Fig. 10. Decalcified mesio-distal section through 02+. Proximal caries and caries in the cementum with secondary dentine formation. Widespread superficial resorptions of the root.
7 \times .

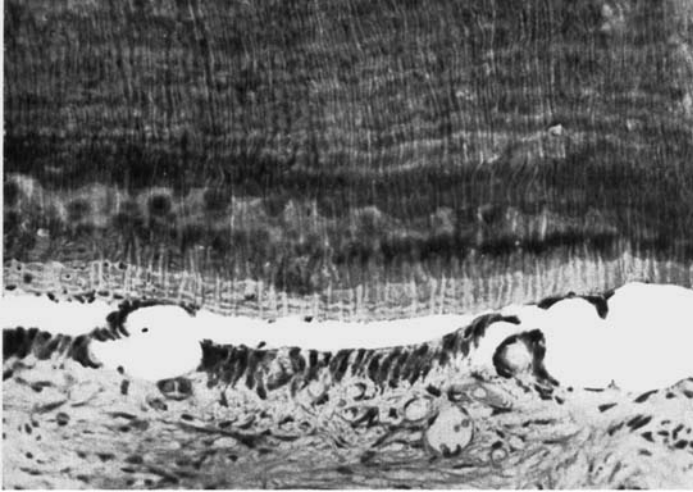


Fig. 12. Detail of the secondary dentine beneath the carious lesion in the cementum from Fig. 10. Poor mineralization of the secondary dentine. Cell inclusions in the predentine zone. Vacuolic degeneration of the odontoblastoma.

200 \times .



Fig. 11. Detail of the dentine near the root tip from Fig. 10. Laminated circum-pulpal secondary dentine. Narrow predentine zone. The odontoblastoma lost.

200 \times .

in the mineralized dental tissue occurred at the age of about four to six. This disorder manifested itself as a formation of irregular dentine on the pulpal walls of the root and as a widespread surface resorption of the roots. These changes might have been caused by local factors (pulpitis, alveolar osteitis). The formation of irregular dentine on the pulpal walls could also be explained by general disorders such as C-hypovitaminosis; however, the possibility of the skeletal disease being directly connected with these changes cannot be excluded.

It is generally maintained that patients with osteopetrosis present definite changes in bone structure in all skeletal parts that are formed in cartilage. On the other hand, the membranous bones such as the flat segments of the skull are altered little. That the dental hard tissues do not show such pronounced and characteristic features as certain parts of the bone tissue may be attributed to the same factor as the above-mentioned difference.

SUMMARY

Deciduous teeth from a six-year-old boy with the clinical and histopathological diagnosis of osteopetrosis have been investigated using histological and microradiographic procedures. In the crown dentine were found areas with a low degree of mineralization corresponding to the areas normally formed during the age of 6–12 months. The roots showed widespread surface resorptions while the apical parts of the pulpal walls presented formation of irregular dentine. The findings are discussed from the aspect of a possible relationship to the skeletal disorder.

ZUSAMMENFASSUNG

STUDIEN AN MINERALISIERTEN ZAHNGEWEBEN

III. Veränderungen an den Zähnen in einem Falle von Osteopetrosis

Die Milchzähne eines sechsjährigen Knaben, mit der klinisch und histopathologische gesicherten Diagnose einer Osteopetrosis, wurden histologisch und mikroradiographisch untersucht. Bezirke des Kronendentins, die normalerweise im Alter von 6–12 Monaten gebildet werden, zeigten einen nur geringen Mineralisa-

tionsgrad. Die Wurzeln wiesen an der Oberfläche ausgedehnte Resorptionsbezirke auf, während die apikalen Anteile der Wände der Pulpenkanäle die Bildung irregulären Dentins erkennen liessen. Mögliche Zusammenhänge zwischen diesen Beobachtungen und den Störungen am Skelett werden diskutiert.

RÉSUMÉ

ETUDES SUR LES TISSUS DENTAIRES MINÉRALISÉS

III. Changements dentaires dans un cas d'ostéopétrose

Les dents temporaires d'un garçon de six ans où le diagnostic clinique et patho-histologique avait déterminé un cas d'ostéopétrose, ont été étudiées avec des méthodes histologiques et microradiographiques. Dans la dentine coronaire se trouvaient des zones moins minéralisées qui correspondaient aux zones normalement formées lorsque l'enfant a de 6 à 12 mois. Les racines montraient des résorptions superficielles étendues tandis que les parties apicales des parois pulpales présentaient une dentine irrégulière. Ces découvertes sont discutées du point de vue d'une relation possible à un défaut du squelette.

REFERENCES

- Engfeldt, B., Engström, A., & Zetterström, R.*, 1954. Biophysical studies on bone tissue III. Albers Schönbergs disease. *Acta Paediat.* 43: 152.
- Engfeldt, B., Karlberg, P., & Zetterström, R.*, 1954. Studies on the skeletal changes and on the etiology of the anaemia in osteopetrosis. *Acta Path. Microb.* (in the press).
- Thoma, K. H.*, 1950. *Oral Pathology*. Third Edition. The C. V. Mosby Company. St. Louis.
- Westin, G.*, 1931. Ueber Zahnveränderungen in Fällen von Skorbut bei Homo. Band 1. Fahlerantz, Stockholm.
- Westin, G.*, 1937. An investigation into questions of social hygiene in the counties of Västerbotten and Norrbotten, Sweden. Part III, Chap IV. Håkan Ohlsson, Lund.

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