Dental Findings in a Child with Osteopathia Striata with Cranial Sclerosis (OS-CS): A Case Report

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The dental management of an 8-year-old girl with osteopathia striata with cranial sclerosis (OS-CS) is described. The girl presented with various oral abnormalities. The aim of this case report was to describe in detail the dental findings in a patient with OC-CS and the precautions to be taken when planning treatment. In the present case, many dental anomalies, such as delayed eruption of the permanent teeth, obliteration of the dental pulp, short roots, fused roots and taurodontism, were detected. In patients with OS-CS, routine dental care from an early stage is recommended to manage this anomaly properly.

Keywords*: osteopathia striata with cranial sclerosis (OS-CS), pulp obliteration, short root, deficient root resorption of primary teeth, delayed eruption of permanent teeth*

INTRODUCTION

Osteopathia striata with cranial sclerosis (OS-CS) is a specific bone dysplasia characterized by linear striations of the long bones and skull and longitudinal striations visible on radiographs of the bones, osteosclerosis of the cranium, and extra-skeletal anomalies owing to increased osteoblast activity.[1]

The etiology of the disease was not known for a long time. Half of the reported cases of this disease were sporadic.[2] Recently, the WTX gene, an inhibitor of WNT signaling, was identified as one of the disease-causing genes.[3,4] WNT signaling plays an anabolic role in bone formation by osteoblasts, and interference of this pathway is known to be involved in other sclerosing bone dysplasias.[4-6]

The characteristic features of the cases reported were macrocephaly, frontal bossing, hypertelorism, broad nasal bridge, conductive deafness, cleft palate and mental retardation.[7,8] OS-CS is a rare disease. To our knowledge, fewer than 100 cases have been reported, most of which demonstrated radiographic features. It has been reported that 30.7% of OS-CS patients have dental anomalies.[9] However, there are few reports of dental findings.[10,11] Therefore, in this paper, we report the dental findings and discuss the dental problems in an OS-CS patient.

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Case Report

An 8-year-old girl was referred to our department with the chief complaint of delayed eruption of the permanent teeth. She was the second child, born at 39 weeks of gestation after a normal pregnancy. Her birth weight was 3770 g (< +2SD), length 50 cm, and head circumference 37 cm (> +2SD). Since birth, she had constant severe respiratory distress, and was diagnosed with OS-CS. She was the only child in the family affected and there was no history of consanguinity. Investigations revealed that she had no WTX gene abnormalities.

The patient presented with several abnormalities including submucous cleft palate (SMCP), macrocephaly, frontal bossing, a broad depressed nasal bridge, hearing loss and atlantoaxial subluxation.

Intraoral examination at the initial visit revealed that spontaneous shedding of primary teeth had not occurred, and only the lower first molars had erupted in the permanent dentition. Her oral hygiene was poor and most of her primary teeth were carious. She had open bite, anterior cross bite, and a narrow maxillary dental arch (Figure 1).

The radiographs revealed generalized sclerosis of the mandible, with linear striations in the ramus, wavelike striations in the body of the mandible, and homogeneous sclerosis in the periapical regions of the erupted teeth and around the dental follicles of the permanent teeth (Figure 2). The primary tooth root components appeared to be more or less fused. The root canal patency of the primary molars was not visible. The tooth roots were obviously shorter than normal (Figure 3). Caries were detected on many teeth. (Figures 2 and 3).

The following treatment was provided to the patient:

1. Dental caries prevention: Oral hygiene instructions, oral prophylaxis, fluoride application and fissure sealants on permanent first molars.

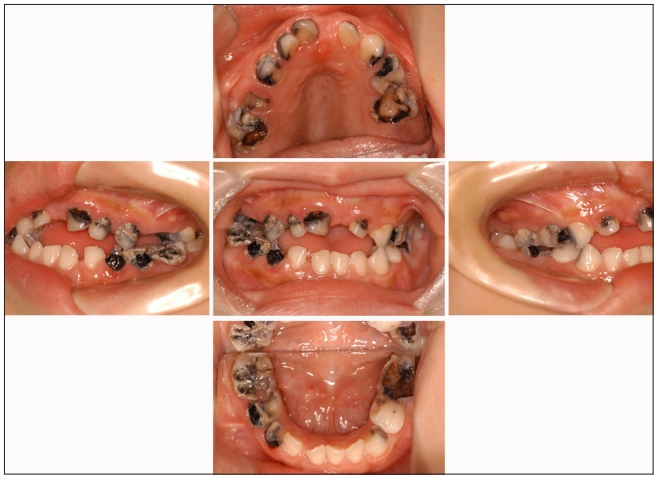

Figure 1. Intraoral photograph at the initial visit.

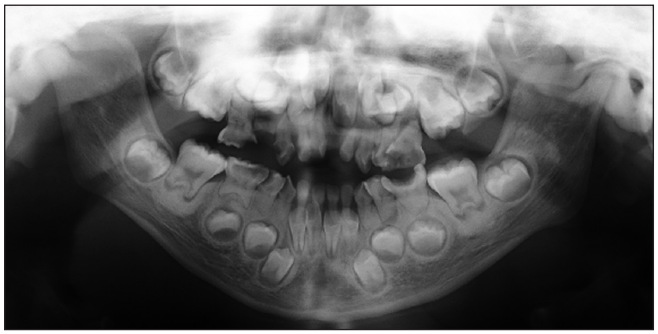

Figure 2. Panoramic radiographs taken at 8 years and 1 month of age.

2. Restoration of carious teeth (composite resin for anterior teeth and stainless steel crowns for posterior teeth).
3. Extraction of painful teeth with apical periodontitis.
4. Delivery of a space maintainer.

In spite of sclerosis of the alveolar bone, we were able to extract the teeth without difficulties and complications.

The panoramic radiographs at the age of 8 years and 1 month and at 9 years and 8 months showed that there had been little movement of the unerupted canine and premolar in the mandible over the previous 1 year and 6 months. However, after extraction of the maxillary deciduous molars, the permanent successors showed a tendency to erupt (Figures 2 and 4).

We observed the pulp chamber morphology of the extracted maxillary second primary molar in detail using micro-CT (Scan Xmate-L090, Comscan, Japan). Then, three-dimensional software (3D-BONE, Ratoc System Engineering Limited, Japan) was used for visualization and analysis of the data. The 3D image revealed obliteration of the root canal from just below the root canal orifice to the apex of the root (Figure 5).

DISCUSSION

One of the abnormalities seen in our patient was subcutaneous cleft palate (SMCP). The treatment of SMCP generally includes surgery; however, surgical intervention is not indicated for asymptomatic patients.[12] The SMPC was mild in our patient and had not affected

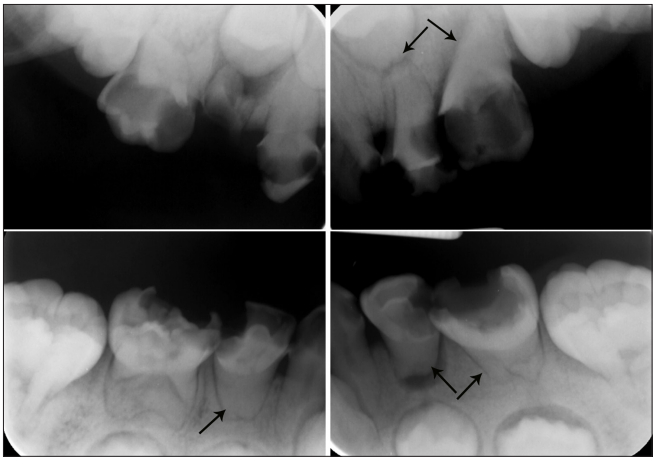

Figure 3. Periapical radiographs of the upper and lower primary molars: Some primary molars have fused roots (arrows).

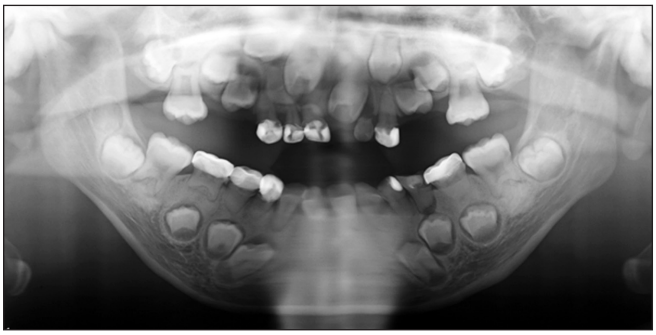

Figure 4. Panoramic radiographs taken at 9 years and 7 months of age: The succedaneous teeth in the areas where the corresponding primary teeth had been extracted tended to erupt; however, in the other areas, this change was not noted.

speech. Therefore, she was only followed up without treatment.

Our patient was mainly concerned about her malocclusion and decayed teeth. Therefore, we scheduled to refer her to an orthodontist after caries treatment. However, she had obliteration of the dental pulp cavity, short roots, fused roots, and taurodontism. Furthermore, she exhibited delayed shedding of the primary teeth and delayed eruption of permanent teeth, which complicated the dental treatment. As for obliteration of the dental pulp cavity, pulp calcification is regarded as part of the aging process when the pulp is subjected to long-standing local irritation, such as abrasion, erosion, caries lesion or dental restoration.[13] In this patient, the root canals of the permanent first molars which were immature were already narrow. Therefore, the obliteration may not be due to reparative dentin. A previous study postulated that reduced vascular flow in the pulp could result in pulpal respiratory depression, ultimately leading to pathological mineralization within the pulp.[14] Therefore, it is assumed that the blood flow to tooth pulp is impaired by sclerosis in the periapical regions of the teeth. This may lead to narrowing of the root canal.

The abnormal condition of the dental pulp and progressive osteosclerosis may result in failure of the normal exchange between the primary and permanent teeth. The lower central incisors and the first molars had erupted spontaneously in this case; it is believed that the tooth formation had been completed early and these teeth

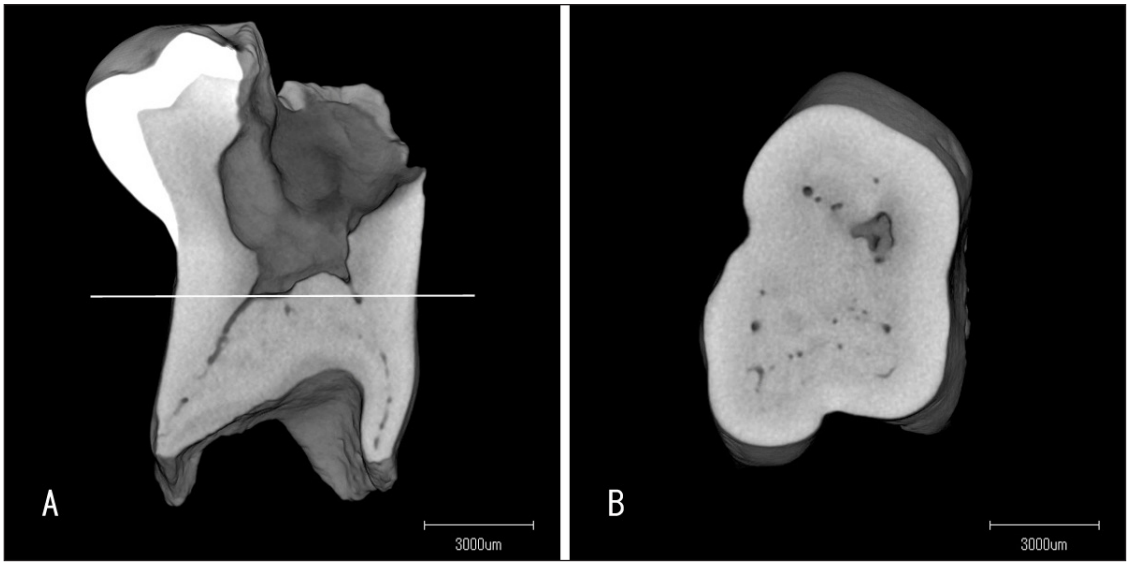

Figure 5. Three-dimensional image of the pulp chamber of maxillary right second primary molar. **A**, Longitudinal section of mesial root. **B**, Horizontal section of sub-root canal orifice (white line in A).

had moved to the outer layer of bone before osseous hardening progressed. In our patient, the extraction of the primary teeth induced a greater tendency of eruption of the successive permanent teeth. We consider that removal of primary teeth at a suitable time by the dentist may have encouraged the eruption of their successive permanent teeth.

As for short roots, there is a possibility that normal root formation was arrested by the abnormal condition of the dental pulp and progressive osteosclerosis. Short root anomaly has been associated with an increased tendency to root resorption under excessive stress such as severe orthodontic forces and trauma.[11] Additionally, a previous study has reported an increased frequency of pulp necrosis after orthodontic treatment of teeth with pulp obliteration as compared to teeth without obliteration.[14] Therefore, it cannot be ignored that obliterated teeth in subjects with OS-CS might have a higher susceptibility to pulpal disease during orthodontic tooth movement. The orthodontist should be aware of the risk associated with the abnormal tooth pulp and short roots in such patients, and the treatment plan should be adapted accordingly.

CONCLUSIONS

This OS-CS patient exhibited root canal obliteration in both the primary and permanent teeth. Therefore, more emphasis must be given to prevention of dental caries in patients with OS-CS. Timely extraction of retained primary teeth will promote and guide eruption of the succedaneous permanent teeth. In conclusion, routine dental care from an early stage is recommended to properly manage this anomaly.

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