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◆ 증 례

Oral findings and dental management of a patient with Moebius syndrome: a case report

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Abstract

Moebius syndrome is a rare, congenital neurological disease involving facial paralysis and limitation of eye movements. It results from maldevelopment of the sixth and seventh cranial nerves. Dental features of this syndrome include micrognathia, microstomia, tongue deformity, cleft palate, hypoplasia of the teeth, and congenital missing teeth.

A 7-year-old female with Moebius syndrome was referred from a local dental clinic for caries treatment. She presented with facial paralysis and microstomia. Oral findings included multiple caries with enamel hypoplasia, congenital missing teeth, and tongue deformity. Dental treatments including restorative and preventive procedures were performed.

Oral findings and management aspects of Moebius syndrome for this case are discussed. Early evaluation and multidisciplinary care are needed for children with Moebius syndrome. 〔J Korean Dis Oral Health Vol.10, No.2: 101-105, December 2014〕

Key words : Moebius syndrome, Palsy of facial nerve, Palsy of abducens nerve, Oral findings

Ⅰ. Introduction

Moebius syndrome is a congenital neurological disease characterized by partial, unilateral or bilateral palsy of the facial and abducens cranial nerves. This syndrome was defined by Paul Moebius in 1888. It is an extremely rare disorder and estimated to be present in 0.002% of births[1].

The etiology remains unclear, but genetic and environmental factors such as drugs use during pregnancy are possible etiological hypotheses[2].

The typical characteristics of Moebius syndrome include impairment of eye movement and loss of facial expression. Also, this syndrome often involves facial deformities and anomalies of the extremities and brachial musculature[3].

From a dental standpoint, the most frequent findings include microstomia, micrognathia, cleft palate, high arched palate, tongue deformity, bifid uvula, multiple congenitally missing teeth, hypoplasia of the teeth, nursing bottle caries, hypoplastic upper lip, and lack of lip seal[2,4-8]. These manifestations can cause many dental problems.

The purpose of this case report is to present the general characteristics, oral manifestations, and den-

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tal management of a child diagnosed with Moebius syndrome.

Ⅱ. Case Report

A 7-year-old female patient visited our clinic for treatment of multiple caries. The patient was born after 36 weeks' gestation with 2.7 kg weight by cesarean delivery. At birth, general hypotonia, small mandible, and cleft palate were observed. She was fed by gavage due to a sucking problem in the neonatal period. She was diagnosed Moebius syndrome with no limb malformations and normal intelligence. She had plastic surgery to treat cleft palate at ages 3 years. Because of chronic otitis media, she is followed periodically by pediatricians.

In extraoral examination, the patient exhibited loss of facial expression (mask-like face) and incapacity to move the eyes from side to side. Many features of Moebius syndrome - including micrognathia, lack of lip seal, everted and thin upper lip, broad and flat nasal bridge, thin eyebrows, protruding ears, and indistinct philtrum - were present (Fig. 1).

Oral findings included microstomia, narrow arch, limited mouth opening, restricted tongue movement with short lingual frenum, bifid tongue, and dry lips. She also had enamel hypoplasia in upper incisors and all permanent first molars, and poor oral hygiene with multiple caries and several residual roots. And anterior crossbite (functional class Ⅲ malocclusion) related with early loss of maxillary primary incisors was observed (Fig. 2).

In radiographic views, congenitally missing teeth including the first primary molars on the left maxilla and right mandible, the first and second premolars on the right maxilla and mandible, and the second premolar on the left maxilla were observed (Fig. 3A). Lateral cephalogram showed micrognathia and slight midfacial hypoplasia (Fig. 3B).

Treatment of caries, tongue-tie operation, preventive procedure such as fluoride application and fissure sealing were planned. Especially, preservation of primary teeth without permanent successors would be important for the patient. And removable appliance would be helpful for anterior crossbite.

Indirect pulp capping of all permanent first molars with calcium hydroxide and glass ionomer restoration was performed. Eighteen months later, normal root development of these teeth was observed. But secondary caries of both maxillary first molar were presented, so additional treatment will be needed (Fig 3C). Now, the patient is checked regularly with oral prophylaxis, and other remaining treatments will be performed .

Fig. 1. (A, B) Extraoral photographs of a patient aged 8 years. (Photographs obtained at 18 month after first visit). These images show micrognathia, everted and thin upper lip, lack of lip seal, thin eyebrows, broad and flat nasal bridge, protruding ears and indistinct philtrum.

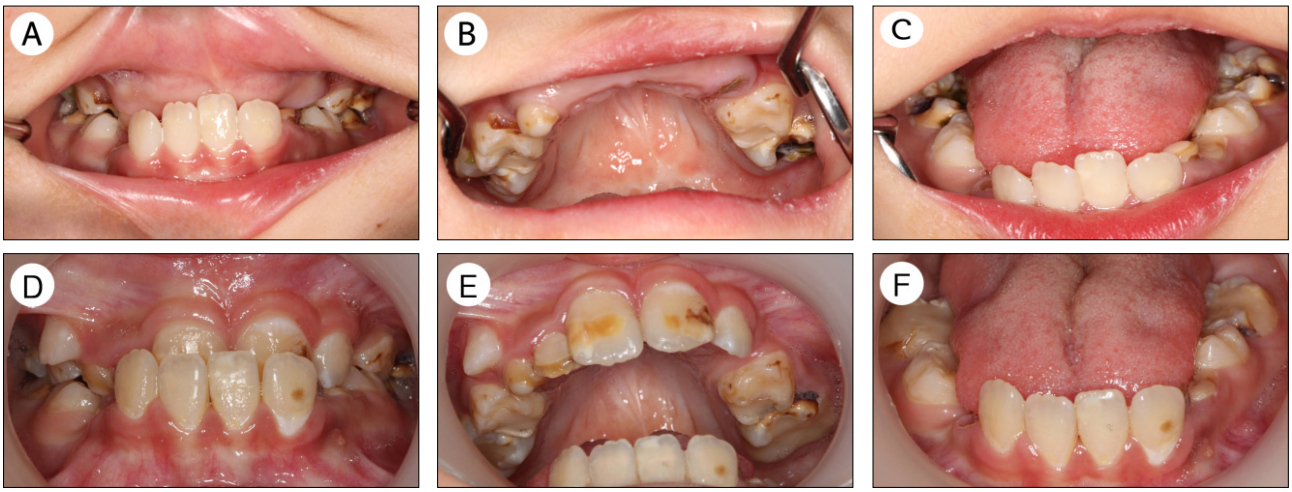

Fig. 2. (A, B, C) Initial intraoral photographs of a patient aged 7 years. These images show microstomia, narrow arch, limitation of mouth opening, dry lips, bifid tongue, restriction of tongue movement (failure of upward movement), dental caries of all first permanent molar with hypoplastic enamel and early exfoliation of upper primary incisors. (D, E, F) Intraoral photographs at 18 months after first visit. Anterior crossbite, enamel hypoplasia of upper incisors, dental caries of left mandibular lateral incisor and ectopic eruption of right upper canine are observed.

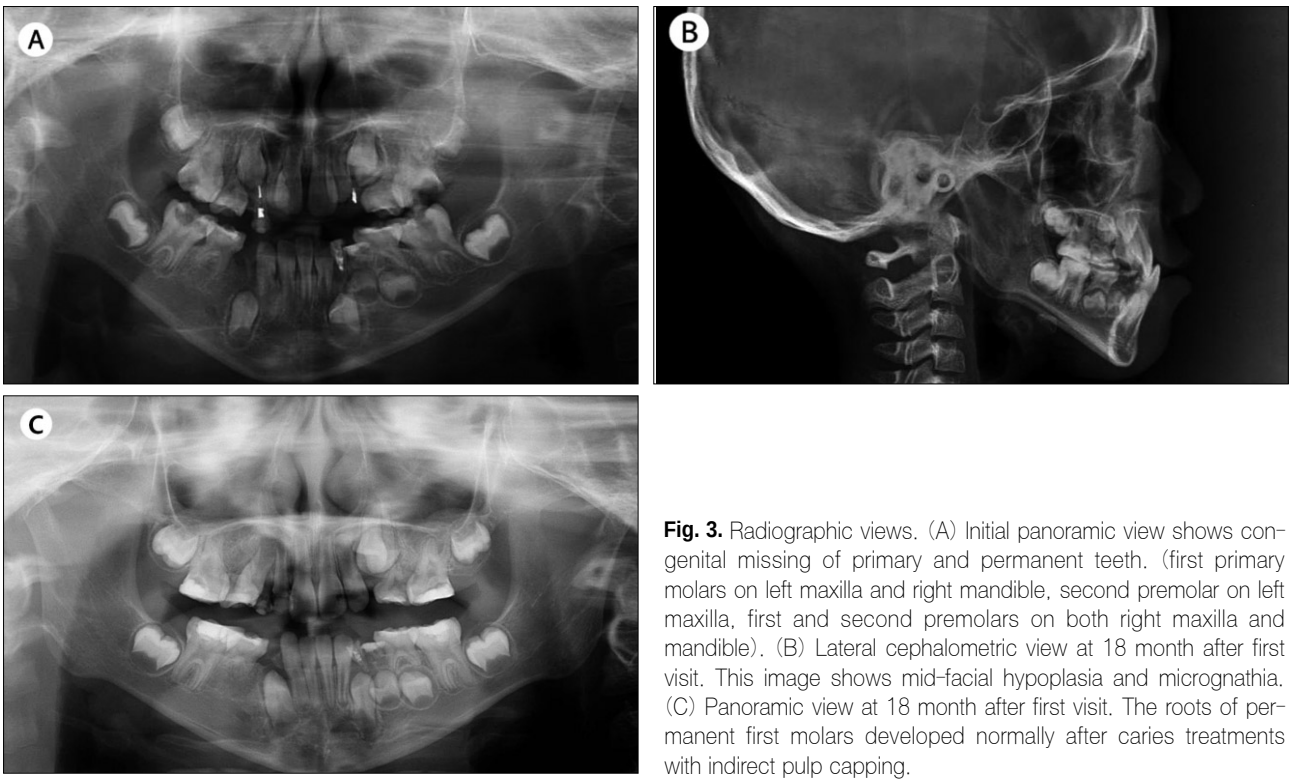

Fig. 3. Radiographic views. (A) Initial panoramic view shows congenital missing of primary and permanent teeth. (first primary molars on left maxilla and right mandible, second premolar on left maxilla, first and second premolars on both right maxilla and mandible). (B) Lateral cephalometric view at 18 month after first visit. This image shows mid-facial hypoplasia and micrognathia. (C) Panoramic view at 18 month after first visit. The roots of permanent first molars developed normally after caries treatments with indirect pulp capping.

Ⅲ. Discussion

Moebius syndrome was first described by van Graefe in 1880 and later defined by Moebius in 1888. Typical characteristics of Moebius syndrome include inability to show emotion by facial expression and to move the eyes from side to side. This syndrome may or may not involve congenital malformation of the extremities and brachial musculature including digital anomalies, clubfoot, and pectoralis muscle defect. In 1998, Abramson[9] proposed a grading system and categorization for Mobius syndrome verified with the acronym CLUFT (C, cranial nerve; L, lower extremity; U, upper extremity; F, facial structural anomaly; T, thorax). Each of the above areas is rated on a scale of zero to three, and a higher grade means a more severe anomaly[7]. Although several patients have reportedly had diminished intelligence, mental retardation is reported just 10% of cases[10].

The etiology of this syndrome remains unclear, but genetic defects and environmental factors have been hypothesized as causes. Genetically, autosomal dominant or recessive inheritance patterns have been reported[4], and translocation between chromosomes 1p34 and 13q13 is a possible etiological hypothesis[11,12].

An ischemic event in the brainstem might be caused by environmental toxicity during an early stage of gestation[13]. The most commonly associated environmental factors are (1) hyperthermia, (2) generalized hypoxia, (3) infection (rubella), and (4) the use of drugs (misoprostol, thalidomide, cocaine, benzodiazepines, and alcohol) in the first trimester of pregnancy[2]. Shepard[14] hypothesized that ischemic events follow the contractions induced by misoprostol and affect cranial nerve nuclei 6 and 7 of the fetus, resulting in facial paralysis.

A patient with Moebius syndrome might have orofacial dysfunctions, and proper treatments are needed. During the neonatal period, the patient probably will have feeding problems because there may be low tone in the masticatory system, pharyngeal dysfunction, and in some cases, cleft[3,6]. A specially made bottle or feeding tube can be helpful to provide adequate nutrition for a patient with feeding difficulties and aspiration problems[2].

Dysfunction of self-cleansing is expected due to restricted tongue movement and hypotonic masticatory muscles[8]. Hypoplastic teeth and dry mucosa caused by a lack of oral seal commonly appear. The patients tend to eat a soft-consistency diet and use a baby bottle longer than normal[6,15]. Thus, dental caries and periodontal problems are frequently observed. However, the small oral orifice and narrow arch make dental treatment difficult. The pediatric dentist should consider dietary counseling and tooth-brushing instruction for the patient's parents[16]. Prophylactic procedures such as sealant and fluoride application should be performed, as well as caries treatment, in these children[5]. In addition, frenectomy of a shortened lingual frenum is helpful for proper oral hygiene care, mastication, and speech[2].

Various orthodontic problems can occur with micrognathia, high arched palate, and narrow arch. An orthopedic appliance such as an expansion plate may be helpful to prevent malocclusion and glossoptosis[6]. In the permanent dentition, final orthodontic treatment can be performed to improve cosmetics and orofacial function. Orthognathic surgery is recommended after the end of growth.

The lack of facial expression and orofacial anomalies lead to problems with social competence, and the patient may be viewed as mentally retarded[17]. Microsurgical procedures with muscle and nerve grafts for facial animation and correction of lip paralysis would be recommended. Bianchi et al.[18] reported a case of upper lip augmentation and genioplasty as well as gracilis muscle transplantation for a patient with Moebius syndrome. This surgery can improve cosmetic appearance and function (oral competence and speech), thereby providing rehabilitation for the patient.

Ⅳ. Summary

In this case, general and oral manifestations of a patient with Moebius syndrome were observed, and detailed knowledge was obtained by clinical examination.

As is typical with Moebius syndrome, the patient showed loss of facial expression (mask-like face) and inability to move the eyes from side to side as typical clinical features.

And micrognathia, lip and tongue deformity, and missing and hypoplastic teeth were observed. Because of these characteristics, patients with Moebius syndrome have a high risk of caries, so active oral hygiene care is needed. Regular check-ups are important to maintain good oral health for a lifetime. Furthermore, orthodontic treatment for malocclusion and reconstructive orthognathic surgery can be helpful for these patients.

It is important that Moebius syndrome be diagnosed early, and for optimal care, meticulous evaluation of each child with Moebius syndrome and a multidisciplinary team approach are needed for adequate treatment.

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