Mobius syndrome: a case report

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Mobius Syndrome is characterized by showing unilateral facial nerve palsy of the sixth and seventh nerves, lack of facial expression, inability to smile and to tightly close the right eyelids. In this report, a 7-year-old-boy with Mobius syndrome is presented. He had asymmetry of facial expression, anomalies of fingers and severe tooth decay. After dental treatment, the periodic re-care visits should be done according to the eruption pattern.
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INTRODUCTION

Mobius syndrome is clinically characterized by congenital non-progressive facial diplegia and restricted lateral eye movements.[1] It is usually bilateral and can be followed by other cranial nerves palsy (third, fifth, ninth and tenth). In Mobius Syndrome the craniofacial anomalies vary greatly and include micrognathia, tongue malformations, facial and oral clefts, oligodontia and paresis of various cranial nerves.[1,2]

The etiology of this syndrome was speculated to be a congenital absence of the motor nuclei of the abducens and facial nerves, but genetic and environmental factors can also be involved. Exposure to infections, alcohol, cocaine, thalidomide or misoprostol was also related in association with Mobius syndrome. It was found in most cases that this syndrome could be the result of infarction of brain system nuclei during fetal life.[3-5] In some of the cases it was also shown vascular malformations as capillary hemangiomas in mesencephalic and pontine segments.[6]

Facial nerve palsy is usually bilateral (but not always) and incomplete, involving the lower part of the face. The resulting "mask-like facies" makes the diagnosis obvious on initial inspection. These children have difficulty relating to people in their environment because of inability to convey a reaction of joy or sorrow and are often incorrectly assumed to be mentally retarded. It seems easier to interpret these moods as they get older, but it is difficult to document any actual change in facial nerve innervation.[7]

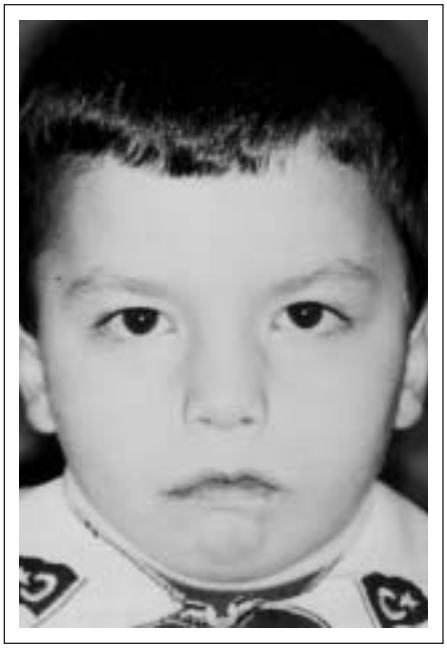

Figure 1. The facial view of the child.

The purpose of this report is to describe clinically and radiographically a boy, who suffered from Mobius syndrome.

CASE REPORT

The subject of this report is 7-year-old boy, born by a cesarean section. Intrauterine growth retardation was not reported. Birth weight 2.55kg, for a gestational age of 33 weeks. The parents were not related. Two siblings died.

The patient had bilateral paralysis, rigidity, a poor suck reflex, clubfeet, and hypoplastic left hand, lack of facial expression, and short tongue at birth. At fourteen

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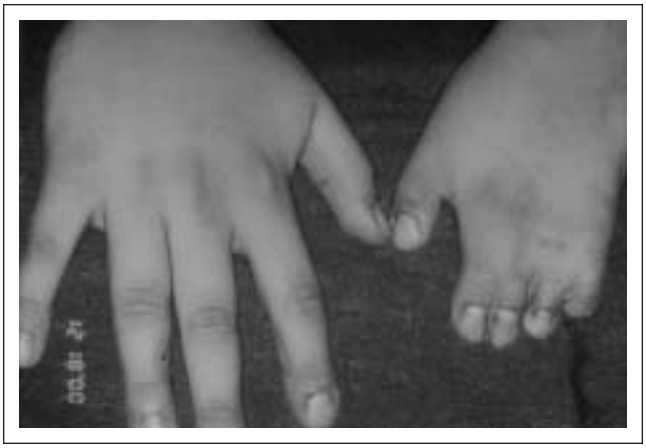

Figure 2. Hypoplastic left hand of the child.

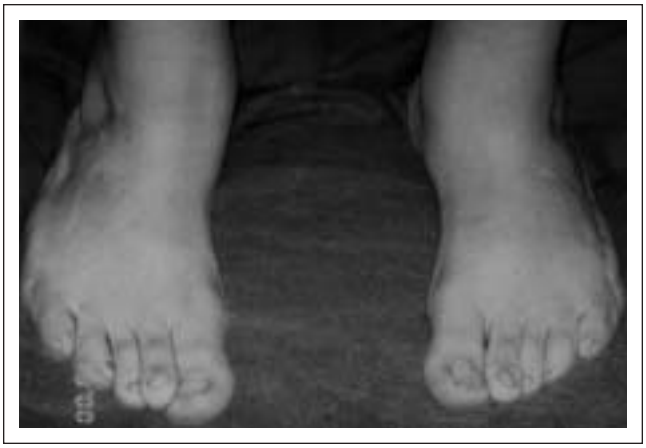

Figure 3. Typical view of the club feet.

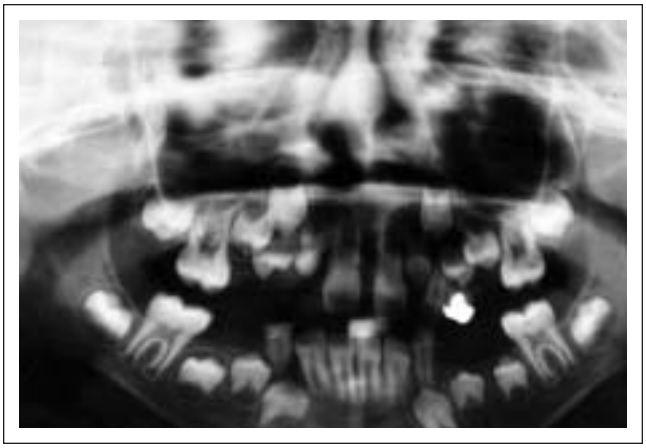

Figure 4. Panoramic radiograph showing all permanent teeth in jaws.

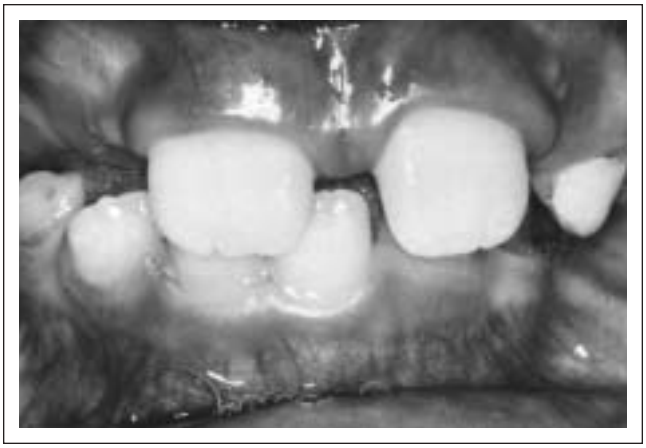

Figure 5. Frontal view of teeth.

months, it was noticed the absence of lateral eye movement, inability to smile, blinking and visual problems. He had speech difficulties, hearing problems, lack of balance. He also had PE tubes placed at this time, noting mild to moderate hearing loss. He had been diagnosed as Mobius syndrome at Istanbul University Faculty of Medicine Department of Pediatrics. At three years of age, it was noticed that he had difficulty in spelling the letters m, p and b.

The patient was referred at 7 years and two months of age to the Dental Clinic Istanbul University, Faculty of Dentistry, Department of Pediatric Dentistry for multiple caries: 53, 54, 55, 65, 74,75, 84 and 85 numbered teeth had deep caries lesions, 63, 64, 83 numbered teeth had dentinal caries. After radiographic examination, he was taken to the operating room, where dental restorations and extractions were completed (Figures 1 to 7).

DISCUSSION

Cases diagnosed as Mobius syndrome were found most frequently in the ophthalmologic literature,[7] which had more complete description of ocular findings, but often included minimal information about the presence of type of oral and systemic anomalies. The orofacial findings noted in the Mobius syndrome include: micrognathia, cleft palate, tongue anomalies, ear malformations, and bifid uvula.[7] Limb malformations may range from syndactyly, clubfeet, toe deformities and symbrachydactyly to more severe absence of digit (ectrodactyly) or amputation defects of the limbs.[7]

In the present case there were the lack of tongue movements and masticatory function. His high caries rate is probably due to this disturbed function.

It is interesting that these children do not necessarily have reading problems although their horizontal movements may be minimal and their tracking thus abnormal necessitating constant head movement. Many show normal progression in school, and in those that do not, other factors may be involved. These patients would tend to refine the concept that tracking problems cause dyslexia.

In this syndrome there is a surprising lack of serious speech defects in spite of the remnants or very small tongue. In addition to tongue malformations dysarthria

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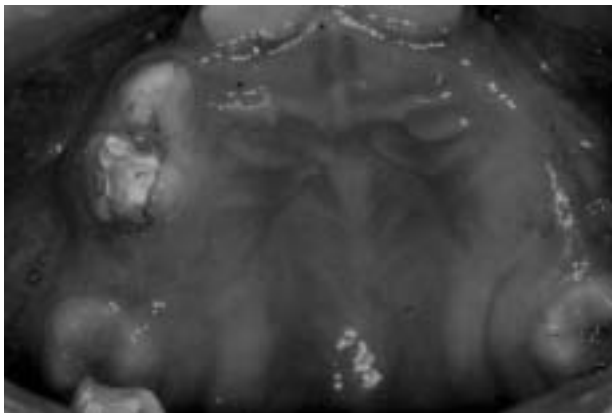

Figure 6. Upper occlusal view of teeth.

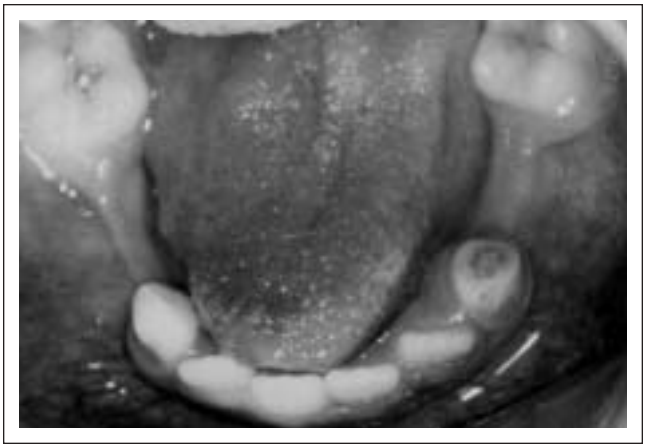

Figure 7. Lower occlusal view of teeth.

may also result from facial nerve palsies.[7] Speech therapy is frequently indicated, but almost all patients had intelligible speech. The children in this series possessed normal intelligence, as did most in cases, although the literature suggests 10% to 15% may show some mental retardation.[7] They are assumed to be mentally retarded, and this assessment, often inaccurate, may contribute to psychological problems.

Our case represents example of congenital facial diplegia that an adequate etiology is not present. The cranial nerve numbers 3, 7, 8, 11 are affected. As most of other cases he had normal intelligence and shows normal progression in his school. He had difficulty in spelling the letters m, p and b.

Another major problem for the patient with Mobius syndrome is the inability to smile. Zuker[8] has treated seven patients with this syndrome by vascularized muscle transplantation, and believed that muscle transplantation to relieve the paralysis of Mobius syndrome offers excellent potential for smile reconstruction and results in improved social habitation.

CONCLUSION

Mobius syndrome is a rare disorder. Craniofacial anomalies including severe dental caries are frequent occurrences in the syndrome. When we also consider the lack of tongue movement and masticatory problems of these children, dentists play a significant role in managing the dental anomalies of Mobius syndrome patients. Because of the systemic problems, early dental evaluation and parental counseling for preventive dental regimens have an important role in these medically compromised patients.[9]

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