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CASE REPORT

The Oral Manifestation Impact of Moebius Syndrome towards Oral Health-related Quality of Life: A Case Report

Felicia Paramita,[1] Harum Sasanti,[2] Siti Aliyah Pradono[2]
[1]Oral Medicine Postgraduate Program, Faculty of Dentistry, Universitas Indonesia, Jakarta, Indonesia
[2]Oral Medicine Department, Faculty of Dentistry, Universitas Indonesia, Jakarta, Indonesia
Correspondence e-mail to: felicia.paramita@gmail.com

ABSTRACT

Background Moebius syndrome is a rare disease characterized by unilateral or bilateral congenital facial palsy and limited ocular abduction. It may also involve other cranial nerve palsy. Moebius syndrome has many orofacial anomalies, such as cleft palate, micrognathia, gothic palate, tongue weakness, tongue atrophy, microstomia, and open bite. **Objective** This case report highlights the rare presentation of Moebius syndrome and denture stomatitis and its management considerations. **Case Report** A 22-year-old female patient came to the oral medicine clinic of Dental Hospital Universitas Indonesia, referred by a prosthodontist with a working diagnosis of denture stomatitis. On examination we found microstomia, tongue defect with movement limitations, and a gothic palate with an erythematous patch on the denture-bearing area. The patient denied any relevant medical history. However, she admitted that she was not satisfied with her oral condition. We performed a further examination based on the signs presented and found impaired ocular abduction. Furthermore, we gave her an Oral Health Impact Profile (OHIP)-14 questionnaire to assess her oral health-related quality of life. These examinations resulted in the diagnosis of Moebius syndrome with denture stomatitis and poor oral health-related quality of life. We planned a full mouth rehabilitation consisting of denture stomatitis healing, improvement of oral hygiene, root canal therapy, and renewing the denture. **Conclusion** This case underscores the importance of interdisciplinary collaboration and individualized care in managing complex conditions like Moebius syndrome, ensuring comprehensive care that addresses both functional and aesthetic concerns.

Keywords: Moebius syndrome, oral health-related quality of life, OHIP-14

INTRODUCTION

Moebius syndrome is a rare congenital disorder characterized by underdeveloped facial (VII) and abducens (VI) nerves. It was first described in the 1880s by Moebius and von Graefe.[1] This disorder is not progressive and may also be associated with other cranial nerve palsies, orofacial anomalies, and limb defects.[2] Paralysis of the facial nerve is responsible for the absence of mimicry and facial expression, as well as the suction ability.[1] Ocular involvement includes defect abduction and adduction ocular movement, crossed fixation, exotropia, or vertical eye misalignment.[1]

Oral findings in Moebius syndrome patients vary greatly, but almost all have gothic (high-arched) palates.[3] Structural anomalies of Moebius syndrome in the oral cavity can be described as hypoplastic upper lip, microstomia, mouth angle drooping, cleft palate, tongue weakness, fissured tongue, tongue atrophy, and open bite.[3] Besides structural anomalies, functional limitations are also found in Moebius syndrome patients, i.e., the inability to perform mandibular lateral movements and protrusion.[3] These oral findings result in chewing, swallowing, and speech difficulties and subsequently affect the oral health-related quality of life (OHrQoL).[4]

Despite the significant oral and functional challenges faced by patients with Moebius syndrome, the condition has not been widely studied, with most available information derived from individual case reports. To our knowledge, there has not been a comprehensive case report that explores the impact of Moebius syndrome on oral health-related quality of life (OHrQoL). This case report aims to fill this gap by discussing a patient with Moebius syndrome, assessing their OHrQoL score, and detailing the necessary treatment modifications for their oral condition.

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CASE REPORT

A 22-year-old female patient visited the Oral Medicine Clinic, Dental Hospital, Universitas Indonesia. She was referred from the Prosthodontics Clinic with a working diagnosis of chronic atrophic candidiasis on the palate under the upper removable denture. She came to the prosthodontist to have new removable upper and lower dentures made because the old dentures had been worn out.

She had already worn the removable denture, made by a dental technician, for seven years. She did not take off the dentures at night, but as suggested by the prosthodontist, in the last past week, she took them off at night and wore them the next morning. The patient did not feel any pain or burning sensation on the palate. She did not notice that there was an erythematous lesion on her palate because of the dentures.

Figure 1. Intaglio surfaces of the removable dentures

Subjective examination of medical history revealed that the patient was born prematurely in the 8th month with a birth weight of 1,200 grams. Unfortunately, detailed records of her growth and development milestones are not unavailable. The patient complained that she has swallowing and speech difficulties. Neither the patient nor her parents were aware of the specific condition affecting her.

Figure 2. Erythema on the denture-bearing area of the palate (*left*) and tongue defect (*right*) at the first visit.

Extraoral examination showed the patient's inability to ocular abduct, frown, lift the eyebrows, and pucker the lips. Intraoral examination showed a tongue defect with movement limitations, a gothic palate, and a relatively small oral cavity. There was also erythema and hyperplasia on the denture-bearing area of the palate. We also took a panoramic radiograph and found several retained dental roots

that needed to be extracted. Based on the examination, the patient was diagnosed with Moebius syndrome and denture stomatitis.

Figure 3. Panoramic radiograph showed multiple remained dental roots in the upper and lower jaw and radiolucency around the apex which all need extractions.

We also gave her the Oral Health Impact Profile-14 (OHIP-14) questionnaire to assess the OHrQoL of this patient related to the Moebius syndrome. The OHIP-14 questionnaire revealed that the patient consistently reported severe functional limitations, physical pain, psychological discomfort, and social challenges, indicating a significant impact on her OHrQoL due to Moebius syndrome.

Based on the examinations, we decided to give her an antifungal to be swished throughout the oral cavity and then swallowed four times daily. The patient was also instructed on proper denture cleaning and taking them off at night. The follow-up visit was two weeks after the first visit.

At the 1st follow-up visit, the erythema of the palate was not significantly improved. We evaluated the treatment and found that swishing the antifungal was ineffective because of the tongue movement limitation. Therefore, we decided to modify the treatment by instructing the patient to apply the antifungal to the denture and put on the denture during the day. We also scheduled her for scaling, extraction, root canal therapy for the remaining teeth, and fabrication of new removable upper and lower dentures.

Figure 4. The erythema area on the palate at the 2nd visit still notable related to the denture bearing area of upper jaw.

On the 2nd, 3rd, and 4th follow-up, the erythema was slightly improved but not fully subsided. On the 4th visit, we also checked the patient's unstimulated salivary flow rate. The unstimulated salivary flow rate was 0.2 ml/min with high viscosity and foamy. On the 5th follow-up, approximately two months after the 1st visit, the erythema of the palate disappeared.

Figure 5. Unstimulated whole saliva was collected and measured. In the measure tube it showed foaminess and only a little amount of saliva.

Meanwhile, the patient had already underwent dental treatment which were dental scaling and extracted six teeth. At the time manuscript was written, the patient was still having root canal therapy with the endodontist and had already made an appointment with the prosthodontist.

Figure 6. Gingiva and palate condition at the 5th follow-up

DISCUSSION

Moebius syndrome was first encountered by Paul Julius Moebius in 1888.[5] The key characteristics of Moebius syndrome are unilateral or bilateral palsy of Abducens (VI) and Facial (VII) Cranial Nerves.[5,6] Paralysis in the Abducens Nerve causes impairments of ocular abduction and paralysis of the facial nerve. These abnormalities cause a lack of facial mimicking, poor or absent sucking, fixed gaze, incomplete eyelid closure during sleep and ptosis.[2] In addition to the involvement of the 6th and 7th cranial nerves, many patients with Moebius syndrome also present with palsies of other cranial nerves, including the hypoglossal (XII), vagus (X), glossopharyngeal (IX), oculomotor (III), vestibulocochlear (VIII), trigeminal (V), trochlear (IV), and accessory (XI) nerves, in descending order of frequency.[1,2,4]

Moebius syndrome lacks established diagnostic criteria, but the presence of abducens and facial nerve paralysis is usually used as the marker of this syndrome. Besides those cranial nerves involvement

previously described, individuals with Moebius syndrome might present with abnormalities of the lower extremity (i.e., talipes equinovarus, syndactyly, ankylosis, absent phalanges), upper extremity (i.e., digital hypoplasia or failure of differentiation, ectrodactyly), facial structure anomalies (i.e., cleft palate, micrognathia, microtia, microphthalmia), or thorax manifestations (i.e., scoliosis, pectoral hypoplasia or breast anomaly, chest wall deformity, breast or pectoral aplasia).[2]

In our case, the patient exhibited an inability to move the eyeballs laterally, frown, pucker the lips, and lift the eyebrows. She also has difficulties with speech and swallowing. Based on these clinical signs, patient's abnormalities consistent with Moebius syndrome diagnosis. The paralysis of the abducens and facial nerves was sufficient for this diagnosis.[2,7] Other manifestations of this syndrome in this patient were glossopharyngeal nerve paralysis, which causes speech and swallowing difficulties, and tongue movement limitation. However, no abnormalities were noted in the upper or lower extremities.

The oral anomalies observed in this patient, including macroglossia, microstomia, tongue defect, and a gothic palate, were among the most common oral manifestations of Moebius.[7] The patient also presented with multiple missing teeth and poor oral hygiene, likely resulting from facial nerve impairment, which can lead to ineffective self-cleansing due to impaired chewing movements.[4,8] While some studies have reported decreased unstimulated salivary flow rates in Moebius syndrome patients, our patient's was below the normal range but not low enough to be classified as hyposalivation. The saliva had high viscosity, which may indicate changes in salivary composition.[4] This needs to be further evaluated. These oral anomalies lead to poor oral hygiene and multiple missing teeth that need to be restored with prostheses.

Our patient also presented with erythema of the denture-bearing area and was diagnosed with denture stomatitis. According to Samaranayake's oral candidiasis classification modified by Axell in 1997, denture stomatitis is one of the candida-associated lesions.[9,10] The first line treatment for local oral candidiasis, such as denture stomatitis, involves the use of nystatin oral suspension.[9,11] The drug is used four times a day to be swished throughout the oral cavity.[9,11] At the first visit, we instructed the patient to swish the nystatin oral suspension for four times a day. Still, at the follow-up visit, we found it ineffective because of the limitation of tongue movement. Therefore, nystatin was applied to the denture four times daily to optimize the contact between the antifungal drug and the palate. At the next follow-up, the treatment was successful, marked by the disappearance of erythema in the denture-bearing area.[9]

Oral health is inseparable from general health and quality of life.[12,13] Therefore, we assessed the patient's oral health-related quality of life to capture her quality of life-related to the oral limitations she experiences. The oral health related quality of life was measured by using the Oral Health Impact Profile-14 (OHIP-14) questionnaire, which has been previously validated in Indonesia.[13,14]

In response to the OHIP-14 questionnaire, the patient reported persistent difficulties with pronunciation, taste disturbances, discomfort while eating, and a range of psychological and social limitations, all of which are consistent with the challenges associated with Moebius syndrome. These findings align with previous reports, such as in Bogart and Matsumoto (2010), which highlighted lower social competence in Moebius syndrome patients compared to control groups, though no significant relationship between the syndrome and overall life satisfaction was observed.[15,16]

Based on the OHIP-14 results, we concluded that our patient was not satisfied with her oral conditions, affecting her oral health-related quality of life. Chewing, swallowing, and speech difficulties, along with the presence of multiple missing and carious teeth, adversely affected her overall well-being. Therefore, we aimed to restore the oral function and improve the oral health-related quality of life through oral hygiene, restoration and root canal therapy for the remaining teeth, and renewal of upper and lower prosthesis.[3,5,17]

CONCLUSION

Dentists, particularly oral medicine specialists, must be aware of patients' oral conditions and their association with overall health. Moebius syndrome, being a rare and challenging condition to diagnose, requires a comprehensive examination to achieve an accurate diagnosis. Treatment in patients with this condition should be individualized, with modifications made as necessary to suit each patient's

unique needs. With proper management, successful treatment outcomes can be achieved, leading to an improved quality of life for the patient.

CONFLICT OF INTERESTS

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