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Moebius Syndrome: Oromandibular-limb Hypogenesis Spectrum - A Case Report

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Abstract: ***Moebius syndrome is one of the rare disorders amongst the disorders of oromandibular limb hypogenesis with congenital non progressive bilateral or unilateral, complete or partial paralysis of the 6th and 7th cranial nerves. Occasionally, the V, X, XI, and XI1 cranial nerves are involved, resulting in difficulty of chewing, swallowing, and coughing, which often leads to respiratory complications. It is of unknown etiology with sporadic occurrence. This case report illustrates clinical features and dental abnormalities in a rare case of Moebius syndrome.***

Keywords: Moebius syndrome, Congenital facial diplegia syndrome, Congenital oculofacial paralysis, Mobius syndrome, congenital facial nerve palsy

1. Introduction

Moebius syndrome was first described by Van Graefe[1] in 1880. In 1888, German neurologist Paul Julius Moebius [2][3] defined it as a non-progressive congenital neurological disorder characterized by unilateral or bilateral facial paralysis and defective extraocular eye movements secondary to congenital paresis of the facial (VII) and abducens (VI) cranial nerves. The condition is one of the rare disorders amongst the disorders of oromandibular limb hypogenesis. The incidence is 1 in 50,000 of live births. The prevalence of Moebius syndrome in the population is yet undetermined. It affects both males and females equally [4](Stroemland et al., 2002). Despite the fact that most cases occur sporadically, familial recurrence can occur. There is an approximately 2% risk for such an occurrence [5][6][7][8] (Bencze, 1967;MacDermot et al., 1990; Rizos et al., 1998; Verzijl et al., 2003).

The most obvious symptoms of this syndrome include a loss of facial expression and compromised function of the stomatognathic complex. Moebius syndrome presents a number of typical characteristics, such as limb deformities (syndactyly, polydactyly, brachydactyly, digit agenesis, talipesequinovarus),occasional anomalies in the ears, strabismus, deafness, dysphagia, dysphonia, and sensitivity disorders in areas supplied by the trigeminal(V) nerve [9] (Lin and Wang, 1997). The ocular presentations include restricted lateral eye movements, ptosis, nystagmus, conjunctivitis and inability to close the eyelids. Orofacial anomalies are common and include microstomia, microglossia, tongue atrophy, cleft palate, cleft lip, bifid uvula, hypodontia, and dental malocclusion [10][11][12] (Martı´-Herreroet al., 1998; Fontenelle et al., 2001; Sjogreen et al.,2001). The involvement of glossopharyngeal (IX) and hypoglossal (XII) nerves is also common, the latter of which is the third most frequently affected in 25% of cases, resulting in paralysis and hypoplasia of the tongue [13] (Kumar, 1990). Mild to moderate mental retardation is known to occur in 10% of the cases. A number of studies have demonstrated a strong association between autism and Moebius syndrome: 30% to 40% of the individuals affected by the syndrome exhibit autistic behavior [12][14][15][16][17](Gillberg and Steffenburg, 1989; Miller et al., 1998; Sjogreen et al., 2001;Bandim et al., 2003; Suvarna et al., 2006). Cardiovascular abnormalities [17] associated with Moebius syndrome are uncommon, but Suvarna et al. (2006) related a case of an 8-month-old child with anomalous pulmonary venous connection.The synonyms of Moebius syndrome are

- Congenital facial diplegia syndrome
- Congenital oculofacial paralysis
- MBS
- Mobius syndrome

We report a case of a female patient presented with clinical manifestations of Moebius syndrome.

2. Case Report

A 49- year old female patient visited the department of oral and maxillofacial surgery, with a complaint of multiple carious upper and lower teeth present since one year. On general examination, she was poorly built and nourished. All vital signs were within normal limits. The patient presented with unilateral non-progressive facial paralysis with Bells phenomenon, inability to smile, difficulty in blowing/whistling and lack of forehead wrinkling since birth(Figure 1).



Figure 1: Showing typical Moebius face with lack of expression especially on left side

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The patient's past history reveled that she was born to normal parents with no history of consanguineous marriage. There was no history of birth trauma. No relevant family history was present.Extra oral examination revealed inability to close the upper eyelid of the left eye (lagophthalmos) with loss of motor function on the left side of face giving an expressionless face, epicanthal folds broad and flat nasal bridge and asymmetry of the mouth were noticed (Figure 2).



Figure 2: Shows inability to close the left eyelids and loss of wrinkles on left side of forehead

Her sensory functions were not affected. The patient revealed limb abnormalities like syndactyly in relation to fingers of the right hand (Figure 3).Intra oral examination showed fissured tongue with depapillated areas (Figure 4) . Hard tissue examination showed multiple root stumps in relation to the upper arch and deep carious teeth in relation to lower arch.



Figure 3: Showing fissured tongue

Based on the history given by her and clinical findings the case was diagnosed as Moebius syndrome. The patient received complete extraction of the non-restorable carious teeth followed by prosthodontic rehabilitation. The patient is also on a close review.



Figure 4: Showing syndactyly of right hand

3. Discussion

Moebius syndrome is a rare congenital disorder characterized by complete or partial paralysis of cranial nerves VI and VII along with other cranial nerves givingthese patients an expressionless face. Usually this condition deprives people of the capacity to protect their emotions through facial expressions. The lack of facial expressivity might lead to a decrease even in parental bonding [18].Moebius described it as a rare syndrome with congenital non progressive bilateral facial and abducent nerve palsy. It may be associated with other craniofacial dysmorphisms and congenital abnormalities of the extremities. It is usually detected in infancy. This relatively uncommon congenital anomaly has been given various names such as Congenital facial diplegia, Congenital nuclear agenesis, Congenital oculofacial paralysis, Smileless syndrome [19].

Nerves commonly affected in this syndrome are facial (in all cases), abducent (in 75% cases), hypoglossal (in 20%) cases and oculomotor (in 5% cases). Speech problems are reported in 75 to 90% of these patients [20].

A classification was proposed by Towfighi et al.[8] based on the pathologic differences observed in various case studies of the patients with this syndrome. They are as follows:

- Group I: Simple hypoplasia or atrophy of cranial nerve nuclei.
- Group II: Primary lesions in peripheral cranial nerves.
- Group III: Focal necrosis in brain stem nuclei.
- Group IV: Primary myopathy with no central nervous system (CNS) or cranial nerve lesions

Abramson et al [21] classified and graded the syndrome on the basis of the clinical findings of cranial nerve palsies and musculoskeletal anomalies by using the acronym CLUFT (cranial nerve, lower limb, upper limb, face, and thorax). This includes cranial nerve features of either partial or complete 6th or 7th nerve palsies or both; lower extremity findings of talipesequinovaris, ankylosis, longitudal, or transverse deficits; upper extremity involvement with digital hypoplasia or failure of formation; structural facial findings

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of cleft palate, micrognathia; and thoracic findings of scoliosis, pectoral hypoplasia, or other chest wall deformity
.
The criteria for diagnosis are difficult to define; however, the following guidelines should assist in making a diagnosis of Moebius syndrome.

4. Criteria for Diagnosis

1) Complete or partial facial nerve paralysis is essential for the diagnosis of Moebius syndrome.
2) Limb malformations (syndactyly, brachydactyly or absent digits, and talipes) are often present.
3) The following additional clinical features in association with complete or partial facial nerve paralysis may also be present and should be helpful in making a clinical diagnosis of Moebius syndrome: bilateral or unilateral ocular nerve palsies (commonly of the abducens and less commonly of the oculomotor and trochlear nerves); hypoplasia of the tongue owing to hypoglossal nerve paralysis; swallowing and speech difficulties owing to trigeminal, glossopharyngeal, and vagus nerve palsies; malformations of the orofacial structures (bifid uvula, micrognathia, and ear deformities); other anomalies of the musculoskeletal system, for example, Klippel-Feil anomaly, absence of the sternal head of the pectoralis major, rib defects, and brachial muscle defects are also present [22][23].

Similar disorders such as Moebius syndrome, Hanhart syndrome, hypoglossia-hypodactyly, and glossopalatinusankylosis have been described and recognized as terminal transverse defects with orofacial anomalies or oromandibular limb hypogenesis syndrome. These are characterized by craniofacial and tongue alterations, cranial nerve palsies, and limb anomalies. Micrognathia and aplasia of the pectoral muscle in association with Moebius syndrome characterize Poland-Moebius syndrome [24].

Inability to suck can be the first sign for children with Moebius syndrome. Therefore these children's require meticulous evaluation involving multidisciplinary care and support with parental co-operation. Feeding difficulties and aspiration often lead to failure to thrive during infancy. Restricted tongue movements and masticatory muscle functions often lead to difficulties in performing oral; hygiene procedures predisposing them to risk of dental caries and periodontal diseases. Eyes usually require symptomatic care and must be protected against exposure keratitis. Dietary care and topical fluoride application are some of the important preventive measures to be taken care.

However, little has been done in the development of effective treatment methods for individuals affected by Moebius syndrome. It is believed that this is related to the rareness of the condition and a lack of experienced professionals in diverse healthcare fields, including dentistry.

Recently, microvascular muscle and nerve transplant for reanimation of the face and correction of lip paralysis have been introduced. Facial reanimation using free pectoralis minor transfer is an invaluable aid to such patients which allows for greater social interaction by means of the ability to smile. Bilateral transfer of the anterior third of the masseter muscles to the comers of the mouth also has been attempted [25][26]. However, it is impossible to restore a true smile in these masks-like expressionless faces. Other associated abnormalities of the facial skeleton like micrognathia may require othognathic surgical correction. Surgical correction of the strabismus and special therapy for the deafness could be possible for these patients. Meyerson [27]recommends the recognition and reinforcement of strengths and resilience in younger patients to help maximize their professional and personal success as adults.During genetic counseling a brief discussion on the possible pathogenesis, natural history of disease and supportive measures for long term rehabilitation should be included.

5. Conclusion

An increase in clinical case studies is important to better understanding of the etiopathological aspects and clinical manifestations of this syndrome. Consequently, measures for perfecting treatment can be established with the aim of improving the quality of life of these individuals and their families.

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