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Case Report

Moebius Syndrome: A report of 2 cases with review

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Abstract: Moebius syndrome is a rare congenital disorder that is characterized by lifetime paralysis, involving a group of cranial nerves. It involves the abducent, facial, oculomotor and hypoglossal nerve. In literature, the definition and diagnostic criteria vary among different authors. Here we report 2 interesting cases of Moebius syndrome in a 12 ¼ year and a 5-year old female patients with peculiar signs and symptoms.
Keywords: Moebius syndrome, Cranial nerves, Paralysis.

INTRODUCTION

Moebius syndrome is an extremely rare congenital neurological disorder which is characterized by facial paralysis and inability to move eyes from side to side. They include cranial nerve involvement especially abducent, oculomotor, facial and hypoglossal nerve [1].

Paul Julius Moebius, a german neurologist, in 1892 was the first to describe this condition. This anomaly is also been described as congenital facial diplegia, congenital oculofacial paralysis, nuclear agenesis and congenital nuclear aplasia [2]. It has been recently suggested that the characteristic criterion for Moebius syndrome is the facial palsy with ocular impairment [3].

In about 10% of the cases, mild to moderate mental retardation is noted. Children with Moebius syndrome may have delayed speech because of paralysis of muscles that move the lips, soft palate, and tongue root. However, some children with Moebius syndrome are mistakingly labelled as mentally retarded or autistic because of their expressionless faces, strabismus, and frequent drooling [3].

The exact incidence of Moebius syndrome is unknown. Researchers estimate that the condition affects 1 in 50,000 to 1 in 500,000 newborns. In a nationwide survey reported in 2003, the prevalence of this syndrome was atleast0.002% of births for the year 1996 to 1998 [4].

The purpose of this article is to illustrate 2 cases of Moebius syndrome who reported to The Oxford Dental College and Hospital,Bangalore.

CASE REPORT 1

A 12 ¼ year old female patient had visited our dental O.P.D with a complaint of forwardly placed upper front teeth since 2 years. On eliciting her history of present illness her mother stated that she had a continuous thumb sucking habit since childhood, which is even associated with difficulty in lip closure.

The child's past medical history revealed that she had defective left eye and mentally handicapped by her physician. She was born to consanguineously wed parents with her elder siblings normal. The mother had an abdominal trauma during her pregnancy of this child. She noticed a delay in all the developmental milestones in her child.

General physical examination revealed thin built with poor nourishment and decreased height for her chronological age (135 cm in height and weighed 27.3 kg). All vital signs were within normal limits. There were no detected abnormalities in her upper and lower limbs.On extraoral examination, loss of motor functionson right side of the face giving an expressionless face, microphthalmic eye with squint; wide nasal apertures and incompetent lip were noticed. Intra oral examination showed depapillated areas over the dorsum of the tongue with a history of burning sensation on having spicy food hard tissue examination showed presence of carious 37 and 85 and mixed dentition (Fig-1).

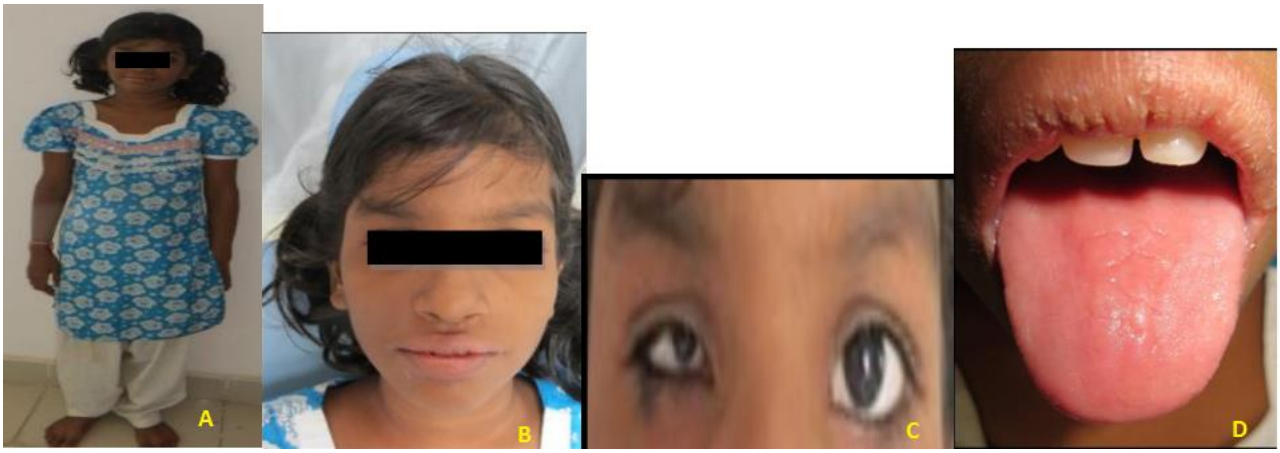

Fig-1: (A) Physical examination showing thin built and nourishment, (B) Patient having expressionless face, (C) Microphthalmic and squint left eye, wide nasal apertures and incompetent lip, (D) Presence of depapillation over the tongue.

Based on above history and clinical findings a provisional diagnosis of depapillation of tongue due to anaemia was suspected.

Radiographic examination of O.P.G and lateral cephalogram showed a mixed dentition stage and mild anterior teeth proclination respectively (Figure 2).She was referred to St. Johns medical hospital for a complete systemic evaluation.

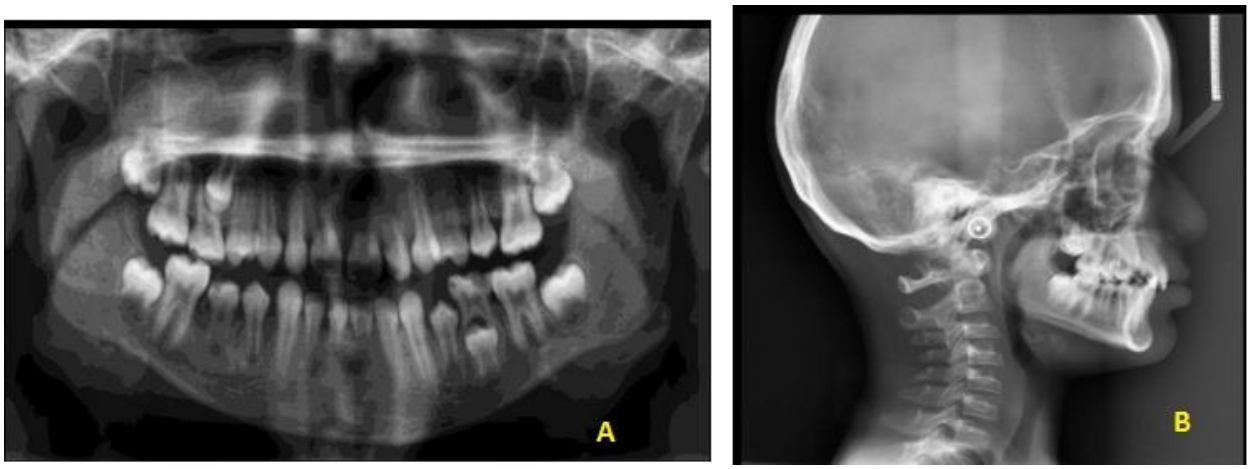

Fig-2: (A) Orthopantomogram showing mixed dentition, (B) Lateral cephalogram showing mild anterior teeth proclination.

Haematological investigation revealed 11.8gm% of haemoglobin and all other values in the complete blood picture being in normal range.

Gynaecological examination revealed absence of axial and pubic hair growth or sexual development. Ophthalmologist opined microcornea associated with coloboma, epistropia and chororetinal degeneration involving macula of her right eye. Psycological analysis by Dukes questionnaire revealed I.Q. level of a 6-year old child. Nutritionist advised her to follow a food chart as she was of thinness Grade II (As per Cole, 2012)

Based on all these findings and after correlating with literature a final diagnosis of Moebius syndrome with protein energy malnutrition was given.

The patient got eye glasses and underwent restorative treatment for her carious teeth. The patient is on a close follow up presently.

CASE REPORT 2

A 5- year old female child visited our hospital with a complaint of deeply carious upper and lower teeth since 8 months. On further eliciting history of present illness, her mother always placed a honey dipped pacifier during sleep, and also had maintained a poor oral hygiene. Her mother also gave a history of inability to close the left eye and the mouth from childhood.

She was born to normal parents but with a history of consanguineous marriage. On general examination, she was poorly built and nourished. Extra oral examination revealed that she had a

microphthalmic and strabismus left eye with loss of motor function on the left side of face. Her sensory functions were not affected.

Intraoral examination showed a sign of hypoglossal nerve involvement with a deviation of tongue to the paralysed side. Hard tissue examination revealed that she had generalized deeply carious primary teeth with more than 2/3rd of lost crown structure (Fig-3).

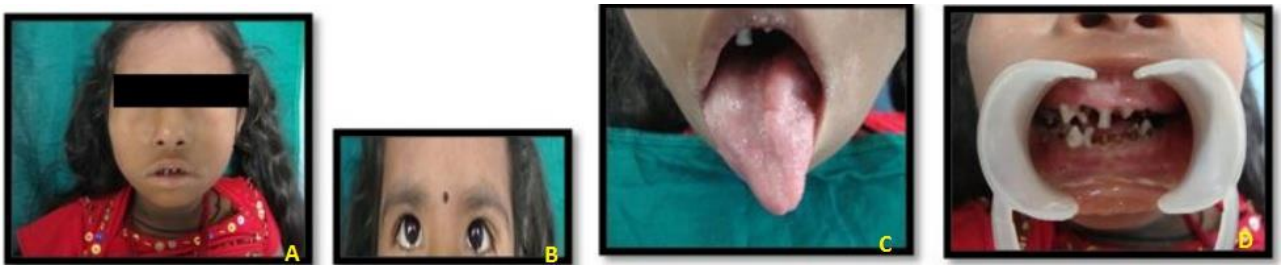

Fig-3: (a) Extra oral view showing expressionless face and protruded lower lip, (b) Impaired ocular movements and strabismus, (c) Deviation of tongue on the affected side (showing hypoglossal nerve involvement, (d) Presence of multiple deeply carious teeth.

Based on the history given by her and clinical findings the case was diagnosed as Moebius syndrome. The patient was further referred for further investigations including CT scan of the skull but her parents were unwilling for further evaluation and treatment as they had financial issues. The patient received a complete extraction of the primary teeth with prosthodontic rehabilitation. The patient is also on a close review.

DISCUSSION

We have described two cases of young Indian girls with Moebius syndrome. Although this disorder commonly is diagnosed soon after birth, but these cases were diagnosed at 12 ¼ and 5 years of age respectively.

Moebius syndrome is a rare congenital disorder which is characterized by complete or partial paralysis of cranial nerves VI and VII along with other cranial nerves giving these 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 [2].

A number of studies have demonstrated that the prevalence of autism associated with the syndrome is greater than the prevalence of autism in the general population: 30% to 40% of the individuals affected by the syndrome exhibit autistic behaviour [2].

Cardiovascular abnormalities associated with Moebius syndrome are uncommon, but Suvarna et al, related a case of an 8-month-old child with anomalous pulmonary venous connection [2]. This heterogenous condition is said to be due to vascular insufficiency, proceeding to the 6th week of gestation involving the proximal 6thintersegmental artery, consequently leading to a ‘subclavian artery supply disruption sequence [5].

The exact etiology of Moebius syndrome is unknown as there are a variety of clinical findings. The suggested etiological factors includes: genetic causes like dysplastic or degenerative developmental mishap, environmental factors like myopathies, peripheral neuropathies, vascular etiology, exposure to drugs, or any trauma during gestation period [1,2,6].

Genetic mechanism plays a minor role in the etiology of the Moebius syndrome, as most of the cases are sporadic. Genetic mechanisms could cause hypoplasia or aplasia of the cranial nerve nuclei. Cytogenetic studies have suggested 2 loci for Moebius syndrome: 1p22 and13q12.13. Nishikawa et al., [12] and Hanisson et al., [13] reported 2 cases with abnormal karyotypes and 1 case of identical twins with Moebius syndrome documented genetic factors for this disorder. Few familial cases with autosomal dominant transmission have also been documented[7].

Certain studies have suggested the that the occurrence of the disorder is often related to an interruption of blood flow in the area of subclavian artery which lead to foetal cerebral hypoxia/ischemia during the first trimester of pregnancy, mainly involving the 6th to 7th week of intrauterine life.

A few autopsy studies found evidence of brainstem pathology in Moebius syndrome. The simultaneous occurrence of anomalies of multiple organ systems in the Moebius sequence suggests a disruption of normal morphogenesis during a critical period in the development of embryonic structures. The mechanism involves haemorrhage due to early uterine contractions of various etiologies, leading to transient ischemic/ hypoxic injury to the embryo or foetus [8].

Drugs like misoprostol had been administered and used as an abortion drug. This drug is a methyl ester of prostaglandin E1, which is also used for treating peptic ulcer, as it was a better protector of antisecretion

activity than normal prostaglandin, which can even lead to uterine contractions and consequent vaginal bleeding [9].

CLASSIFICATION

A classification was proposed by Towfighi et al. 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 [7].

Clinical manifestations include an immobile facial feature with various glaze palsies, external ocular palsies, including ptosis accompanying facial palsies in 80% of the cases. Oral manifestations include micrognathia, hypoplastic upper lip and mandible, mouth-angle dropping, fissured tongue, gothic palate and open bite. In our cases we found glossitis, V,VI and VII cranial nerve involvement [2].

Kallmann syndrome is usually seen in association with Moebius syndrome which is characterized by anosmia and hypogonadism [10]. Certain Moebius syndrome patients present with anomalies like talipesequinavarus, brachydactyly, syndactyly, congenital amputations, arthrogryposis, and smallness of limbs and occasional hypoplasiaor absence of pectoralis major muscles which is termed as Poland anomaly [11].

No relevant laboratory investigations are present specific to Moebius syndrome. Only few cases have been described in the radiological literature. Common CT and MRI findings include hypoplasia of the pons or medulla, depression of the 4th ventricle, absence of the hypoglossal prominence suggestive of hypoglossal nuclei hypoplasia, calcification in the pons in the region of the abducens nuclei, and cerebellar hypoplasia [3].

The treatment for Moebius syndrome involves a multidisciplinary approach. The dental treatment of children with Moebius syndromepresent a number of difficulties due to their characteristiclimitations of the condition, such as the small and inelasticoral orifice and a dry lip mucosa[11].

Eyes usually require symptomatic care and must be protected against exposure keratitis. Dietary counselling and topical fluoride application are other important preventive measures in these children.

Recently, microvascular muscle and nerve transplant for reanimation of the face and correction of lip paralysis have been introduced. During genetic counselling a brief pathogenesis of the condition should be explained to the parents [4].

CONCLUSION

An increase in clinical case studies can give more detailed and a clear picture of this disease. These Gods favourite children are treated as abnormal and separated from society and have an extremely painful experience for social acceptance. An oral diagnostician therefore plays a crucial role in diagnosing and managing this condition along with the other medical doctors.

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