Dentoskeletal abnormalities associated with Mobius syndrome

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Abstract

Mobius syndrome is a rare congenital disorder that causes paralysis of facial nerves. In a high percentage of patients, dentoskeletal abnormalities are described. Aim of this review was to identify most common dentoskeletal abnormalities associated with Mobius syndrome. Most common dentoskeletal anomalies are micrognathia, hypoplastic upper lip, microstomia, hypoplasia of mandible, gothic palate, open bite and II class malocclusions and severe carious lesions.

Introduction

Mobius syndrome is a rare congenital disorder characterized mainly by paralysis, unilateral or bilateral, of the facial nerve associated with the involvement of other cranial nerves such as abducer or hypoglossal.1-15 Von Graeae first described congenital facial diplegia in 1880.16 Mobius lent his own name to the established syndrome in his articles published in 1888, 1892.17-18 Prevalence rate of this syndrome is approximately 1 in 100,000 neonates.19 Peculiar feature of these patients is the complete or partial absence of facial mimicry. We speak of "children without smile", associated in most cases with the absence of eye laterality movements1-20.1

In addition, significant dysfunction of cranial nerves III through XII also has been reported, especially cranial nerve VI.1-21 Multiple limb deformities (syndactyly, brachydactyly, polydactyly, adactyly, ectrodactyly) are often present.21 Bilateral or unilateral, complete or partial paralysis of cranial nerves VI and VII as well as limb deformities are found in 50% of the patients.21 Other anomalies of the muscular-skeletal system also are associated with this syndrome. For example, those affected by the syndrome often display brachial muscle defects22, rib defects, hypoplasia or absence of the pectoralis muscle, absence of the sternal head of the pectoralis major23 and Klippel-Feil anomaly. Malformations of the oral-maxillofacial structures associated with this syndrome include cleft lip and palate, bifid uvula, under development of the maxilla, micrognathia, ear lobe deformities, ear deformity, hypertelorism21. Moderate mental retardation has been reported in 10% to 50% of the patients21. Other associated anomalies have been reported such as congenital heart disease, deafness, seizure, hydrocephalus, diabetes insipidus, hypopituitary hypogonadism, 1±1 antitrypsin deficiency and premature thelarache.22

Mobius syndrome is frequently associated to oral abnormalities and orthodontic issues. Aim of this systematic review is to analyze case reports in the literature in order to identify dento-skeletal alterations associated with Mobius Syndrome.

Methods

A systematic review of the literature was performed on Pubmed medical database in order to identify case reports describing oral alterations in patients with Mobius syndrome.

Keywords used were â€œMobius syndromeâ€, â€œOrthodonticsâ€, â€œOral abnormalitiesâ€.

After a careful analysis, 9 case reports were found.

Results and discussion

The case of a 17-year-old black female was described by Rizos in 1998.24 She had, in addition to common facial abnormalities including facial asymmetry with competent and protrusive lips, microstomia, multiple missing teeth (1, 6, 7, 8, 23, 24, 25, 26, 27, and 32), severe bony defect of the alveolar process on the anterior regions of both jaws. The maxillary dental arch form was V-shaped, while the mandibular arch was U-shaped. Dentally, the patient presented with a class II molar relationship on the right side and class III on the left side. An anterior open bite of 12 mm and a bilateral posterior crossbite were present. Cephalometrically, the patient presented a class II skeletal apical base relationship with a hyperdivergent growth pattern.

Scarpelli, in 2008, described the case of a 5-year-old male child with hypoplastic upper lip, microstomia, micrognathia, gothic palate, tongue weakness, tongue
atrophy, malocclusion, open bite and extensive
carious lesions observed in the deciduous molars.\textsuperscript{25}

In 2002, De Serpa Pinto described oral findings of 12
patients with Mobius syndrome.\textsuperscript{26} Facial weakness,
hypoplastic upper lip (in all patients), microstomia (in
all patients), mouth-angle drooping (in all patients),
hypoplasia of mandible (in all patients), cleft palate (in
2 patients) gothic palate (in all patients), tongue
weakness (in all patients), fissured tongue (in 8
patients), tongue atrophy (in 6 patients) and open bite
(3 patients) were found.

In 2003, Ha described the case of an 18-year-old
Hispanic male with history of Mobius syndrome,
self-abusive behavior, severe mental retardation and
labored breathing.\textsuperscript{21}

At dental examination, multiple severely carious teeth
and a possible draining fistula that may have
developed secondary to an odontogenic abscess were
observed. The patients had severe medical
complications when he was hospitalized for oral
rehabilitation during general anesthesia.

In 2006, Magalhães described cases of 29 patients
with Mobius syndrome.\textsuperscript{27} All patients presented
micrognathia, lack of lip seal, high arched palate and
weak soft palate. The use of orthopedic appliances
was recommended to all 29 patients, but only 13
adhered to treatment and were monitored for at least
24 months. Authors observed that, after 24 months of
treatment, the palate was expanded and micrognathia
became less severe in the majority of the cases
suggesting that the early use of orthopedic appliances
is important to prevent malocclusion and glossoptosis.

In 2012, Cai described 3 cases of patients aged 17 to
24 years with Mobius syndrome and severe skeletal
open bite.\textsuperscript{28} Patients were evaluated and treated with
preoperative orthodontics, orthognathic surgery, and
postoperative orthodontic management. One patient
was treated by bilateral V osteotomies of the
mandibular body and one patient with bilateral V
osteotomies of the mandibular body plus a Le Fort I
osteotomy. The third patient had bilateral mandibular
ramus sagittal split osteotomies in combination with
maxillary osteotomies.\textsuperscript{28} Postoperative and
and severe micrognathia.\textsuperscript{29} The outcome of the patient
treatment was optimal.

Ghosh, in 2017, described 2 cases of Mobius
syndrome.\textsuperscript{19} A patient was a 20-year-old female that
had incompetent lips scar mark over the upper lip and a
high-vault palate, decays of maxillary left second
premolar, first molar and mandibular right and left first
molar. The other described patient was a 10-year-old
male that had high-vault palate, decayed maxillary
right and left first permanent molar, and maxillary left
first deciduous molar were noted.

Always in 2017, Magnifico described a case of a
23-year-old man with bilateral complete palsy of facial
nerve and dysfunction of lateral movements in both
eyes, convex profile, reduced lower anterior facial
height, open nasolabial angle, severe micrognathia,
incompetent lips with interlabial separation at rest of
13\(\pm\)4\% mm and intraoral dental class II, division 1, with
increased overjet (6\(\pm\)3\% mm) and overbite
(4\(\pm\)4\% mm), retroinclination of upper incisors, II molar
and canine class on both side, deviation of lower
midline, complete dental formula, crowding in the
lower jaw, and scissor bite of elements 2.7 and 2.8.\textsuperscript{30}
The patient was successfully treated with a combined
surgical-orthodontic strategy.

Conclusions

Among patients with Mobius syndrome, the incidence
of dentoskeletal alterations is high. Most frequent
alterations are micrognathia, hypoplastic upper lip,
microstomia, hypoplasia of mandible, gothic palate
and severe carious lesions. Open bite and II class
malocclusions are also frequent. The combination of
orthodontic treatment and orthognathic surgery is
effective in treating these abnormalities.

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