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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 Graaefe 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 movements[1-20].

Â 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 defects[22], rib defects, hypoplasia or absence of the pectoralis muscle, absence of the sternal head of the pectoralis major[23] 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, hypertelorism[21]. Moderate mental retardation has been reported in 10% to 50% of the patients[21]. Other associated anomalies have been reported such as congenital heart disease, deafness, seizure, hydrocephalus, diabetes insipidus, hypopituitary hypogonadism, Î±1 antitrypsin deficiency and premature thelarche.[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.Â 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.[25]

In 2002, De Serpa Pinto described oral findings of 12 patients with Mobius syndrome.[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.[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, Magalhaes described cases of 29 patients with Mobius syndrome.[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.[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.Â Postoperative and post-orthodontic stability was good in 2 cases, whereas a 5-mm anterior open bite developed after treatment in 1 case and additional orthodontic management was required to re-establish good occlusion.

Bianchi, in 2013 described the case of a patient treated with multiple orthognathic surgery procedures due to congenital bilateral complete palsy of cranial nerves VI and VII, palsy of the right cranial nerve XII and severe micrognathia.[29] The outcome of the patient treatment was optimal.

Ghosh, in 2017, described 2 cases of Mobius syndrome.[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 13â€‰mm and intraorally dental class II, division 1, with increased overjet (6â€‰mm) and overbite (4â€‰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.[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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