Review Article

Alterations of the myofunctional orofacial system in the Parry-Romberg syndrome: a critical literature review

Alterações do sistema miofuncional orofacial na síndrome de Parry-Romberg: revisão crítica da literatura

FLÁVIA MARQUES RIBEIRO[1]
LAURA DAVISON MANGILLI[1]
FERNANDA CHIARION SASSI[2]
CLAUDIA REGINA FURQUIM DE ANDRADE[2]*

■ **ABSTRACT**

Introduction: This qualitative literature review analyzed international scientific publications on possible orofacial myofunctional alterations in patients with Parry-Romberg syndrome by using PubMed. **Methods:** The survey was conducted in English, between 2002 and 2012, and was limited to human beings of any age. Publications without full access, duplicated by overlapping keywords, literature reviews, letters to the editor, and those not directly related to the research topic were excluded. **Results:** We identified 719 studies, of which 21 were within the established criteria. Based on the selected studies, patients with Parry-Romberg syndrome may show changes in soft and hard tissues such as atrophy of the sternocleidomastoid, masseter, and pterygoid muscles; atrophy in the cheek region and depression of the nasolabial fold; deviation of the lips and nose; unilateral tongue atrophy; atrophy of the mouth angle; progressive resorption of the maxilla and mandible bone; atrophy of the zygomatic arch and frontal bone, and facial asymmetry; atrophic root development or pathological resorption of permanent tooth numbers; and jaw reduction and delayed eruption of the upper and lower teeth. **Conclusion:** Despite the growing interest in the diagnosis and symptomatic description of individuals with Parry-Romberg syndrome, publications that address functional and interdisciplinary treatments are scarce. Therefore, specific studies aimed at improving the quality of life of these patients are needed.

Keywords: Facial hemiatrophy; Stomatognathic system; Face; Facial bones; Tissue.

■ **RESUMO**

Introdução: Esta revisão qualitativa da literatura analisou publicações científicas internacionais sobre possíveis alterações miofuncionais orofaciais em pacientes acometidos pela Síndrome de Parry-Romberg, por meio da base de dados PubMed. **Métodos:** O levantamento realizado limitou-se a seres humanos, de qualquer faixa etária, no idioma inglês, entre os

Institution: Study performed at the Departamento de Fisioterapia, Fonoaudiologia e Terapia Ocupacional and Hospital das Clínicas of the Faculdade de Medicina of the Universidade de São Paulo (USP), São Paulo, SP, Brazil.

Article received: July 12, 2013.
Article accepted: October 30, 2013.

DOI:10.5935/2177-1235.2015RBCP0126

[1] Instituto Central do Hospital das Clínicas, Divisão de Fonoaudiologia, Faculdade de Medicina, Universidade de São Paulo (USP), São Paulo, SP, Brazil.
[2] Departamento de Fisioterapia, Fonoaudiologia e Terapia Ocupacional, Faculdade de Medicina, Universidade de São Paulo (USP), São Paulo, SP, Brazil.

anos 2002 e 2012. As publicações sem acesso completo, repetidas por sobreposição das palavras-chave, revisões de literatura, cartas ao editor e as não relacionadas diretamente ao tema foram excluídas. **Resultados:** Foram identificados 719 estudos, sendo 21 dentro dos critérios estabelecidos. Com base nos estudos selecionados, pacientes acometidos pela Síndrome de Parry-Romberg podem apresentar alterações dos tecidos mole e duro, tais como atrofia dos músculos esternocleidomastoideo, masseter e pterigoideos; atrofia na região da bochecha e depressão da prega nasolabial; desvio dos lábios e nariz; atrofia unilateral da língua; atrofia do ângulo da boca; reabsorção progressiva do osso da maxila e da mandíbula; atrofia do arco zigomático, do osso frontal e assimetria facial; desenvolvimento atrófico das raízes ou reabsorção patológica dos números de dentes permanentes; redução da mandíbula e erupção atrasada dos dentes superiores e inferiores. **Conclusão:** Apesar do crescente interesse pelo diagnóstico e pela descrição sintomatológica de indivíduos com Síndrome de Parry-Romberg, a escassez de publicações que abordem tratamentos funcionais e interdisciplinares é evidente. Verifica-se a necessidade da realização de estudos mais específicos que visem à melhoria da qualidade de vida desses pacientes.

Descritores: Hemiatrofia facial; Sistema estomatognático; Face; Ossos faciais; Tecido conjuntivo.

INTRODUCTION

The Parry-Romberg syndrome, also known as progressive facial hemiatrophy[1-16], is usually unilateral[4-6,9,15-17], affecting the skin[1-3,5,7,8,10-18], muscles[1-3,5,7-13,16-18], fat[6,9,11,18], cartilages[2,6,10], and bones[1-3,5,6,8-18].

Among the possible etiological causes, the most common are local facial trauma[2,5,6,9,19]; endocrine disorders[5,6,9]; autoimmunity[2,5,9,10,19,20]; heredity, hyperactivity, or underactivity of the sympathetic nervous system[2,5,6,9,19,20]; trigeminal nerve abnormality[9,19]; and viral infections[2,5,6,9,19].

Atrophy appears in the first 20 years of life[1,2,4-6,9,10,12,13,16,17,21], progressing from 2 to 10 years[1,2,9] and stabilizing thereafter[1,2,5,9,10]. It is more common in women[4,5,9].

The main symptoms associated with the pathology described in the literature are neurological changes[2,11-13,17,18] such as migraines[2,16-18], epilepsy[2,11-13,17,18], and facial pain[2,16,18], in addition to ocular complaints[2,11,18] and hair loss[2,11]. Generally, patients take medical help regarding aesthetic complaints[5] and neurological symptoms.

The most important characteristics of the syndrome are facial asymmetry[6,21], which may extend from the chin to the forehead[21], mouth and nose deviation to the affected side[6], and unilateral teeth exposure when the lips are engaged[5,6,18]. When the forehead is affected, linear scleroderma can be diagnosed, and the sign in this facial region is called *en coup de sabre*[2,11,18,21].

Treatment is multidisciplinary, involving physicians, dentists, speech therapists, and psychologists[5]. Surgical procedures are indicated when the disease reaches a steady status[3]. In addition, patients who develop facial atrophy may need counseling[5].

No specific studies in the literature in the field of speech therapy discuss muscular and functional alterations of the Orofacial Myofunctional System in patients with the syndrome. Thus, before beginning clinical research on this condition, it would be necessary to determine the characterization/modification of soft tissue in patients with the syndrome, as described in related literature.

The aim of this study was to analyze scientific papers published in a refereed database that discourse on muscle and/or functional alterations of the orofacial myofunctional system in Parry-Romberg syndrome.

METHOD

This was a critical literature review performed to verify the possible orofacial myofunctional alterations in patients with Parry-Romberg syndrome. For the establishment of the research method, the precepts of the *Cochrane Handbook* were followed[22].

The literature review was performed by using the PubMed database, and the following descriptors were crosschecked: "facial hemiatrophy," "progressive facial hemiatrophy," "stomatognathic system," "stomatognathic," "muscle," "facial," "facial bones," "skin disease," and "connective tissue." The criteria for the inclusion of articles were as follows: research in humans, written in English, and published within the 10-year period (January 2002 to July 2012).

Articles in the database were searched individually by three researchers to minimize possible quote losses. Each quote recovered in the database was analyzed independently by the study researchers in

order to determine their relevance to the research and whether to include them in the study.

The quotes that did not allow access to the full text and duplicated citations by overlapping keywords were excluded from the study. Of the complete texts obtained, literature reviews and articles were excluded when not directly related to the subject, such as surgical methods, brain atrophy, specific complaints of vision, medical treatments, and life story of the authors of the studied disease.

The articles that were effectively related to the research proposal were analyzed. All stages of the study were conducted separately by the researchers, and when there was disagreement among the researchers, only articles in which the final position was consensual were included.

According to the crosscheck of the descriptors, 719 articles were initially found. Of these, only 326 were not duplicated citations because of overlapping keywords. Next, an individual analysis of the articles found by the researchers was conducted, listing 21 articles on the final consensus.

Analysis of the articles was performed, dividing the studies into two segments as follows: case reports and original articles. The parameters studied in case reports were participants' characteristics, sex, age, facial side involved, complaints, clinical findings, evaluations, treatments, time to intervention, results, and conclusions. For original articles, objectives, methods, results, and conclusions were selected as parameters.

Figure 1 shows the flowchart of the search and selection of articles for analysis in the study.

RESULTS

The study results are briefly described in Chart 1 (case studies) and Chart 2 (original studies).

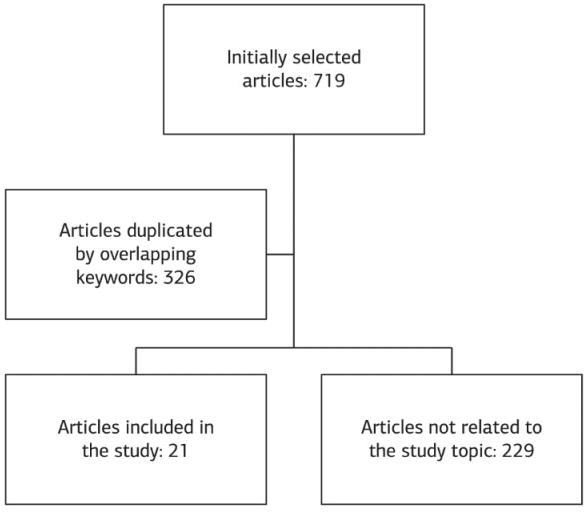

Figure 1. Flowchart of full texts selected for analysis.

DISCUSSION

On the findings of this review, we observed that only a few original articles are related to the topic. Most studies report isolated cases, addressing the clinical and objective assessment, and methods for diagnosing the syndrome. We also observed that only few studies discourse about the treatment, which when approached, describes the control of neurological symptoms.

Among the case reports, 16 showed single-case descriptions[1,2,4,5,7,9-17,19,20], one described the disease in two participants who were inbred cousins[21], and one described the disease in 23 patients[3].

In the 16 case reports of single-case descriptions[1,2,4,5,7,9-17,19,20], male patients were the most affected[6,7,9-11,13-15,20]. According to the literature, the highest incidence of the disease is among female patients[4,5,9]. The left facial side has a higher prevalence[1,2,4,6,10-12,14,15]. The literature does not show any predominance of hemifacial involvement in the disease[18].

Regarding age, we found a mean age of 20.6 years among the study patients. The youngest patient was less than 22 months old[15], and the oldest patient was 65 years[1]. As already described in the literature, the age of atrophy onset is between the first and second decades of life[1,2,4-6,9,10,12,13,16,17,21]. However, in case studies, one should take into consideration that cases that are distinct from the standard are often of interest to describe.

A study that describes the Parry-Romberg Syndrome in consanguineous cases is worth reporting[21]. Given that the etiology of the syndrome remains unknown[1-8,17,19-21], the authors pointed out the need to further investigate genetic characteristics in order to objectively define the diagnosis.

With regard to complaints, aesthetic compromise was the most frequently reported[3,6,9,15,19,20], followed by epilepsy[10,12,13,16] and pain[2,7,11,14]. The symptoms most often cited in the literature are migraine[2,16,17,18], epilepsy[2,11-13,17,18], facial pain[2,16,18], and aesthetics[5].

The assessments presented to the description of the cases were imaging tests such as radiography, magnetic resonance imaging, computed tomography, and electroencephalography[2,4-7,9-12,14-16,20]; laboratory tests[1,4,7,9-12,14,16,19,20]; physical examination[2,3,6,9,11,13-16,21]; neurological examination[4,7,9,10,12,13,15,16]; ophthalmological examination[12,14,20]; tissue biopsy[1,11]; electromyography[2,4]; orthodontic evaluation[5,6]; thermal test[2], and hearing examination[15].

With respect to methods of treatment, eight studies did not describe the use of treatment[1,4-6,9,14,15,20], seven reported drug treatment[2,7,10,11,16,19,21], two described the association between medical and surgical treatments[12,13], and one described only surgical treatment[3]. The treatments often cited in the literature for this syndrome are drug and surgical treatments[18].

Chart 1. Summary of study cases.

Reference	Sample	Complaint and Treatment	Results
Carvalho et al.[19]	A 22-year-old woman; right side affected.	Aesthetic complaint. Laboratory tests. She underwent 10 months of drug treatment and 12 months of follow-up.	Patient had areas of skin hyperpigmentation; oral changes involving the jaw and teeth. Aesthetics improved with drug treatment.
Duymaz et al.[9]	A 35-year-old man; right side affected.	Aesthetic complaint. Clinical examination, laboratory tests, imaging studies, and evaluation of cranial nerves. Treatment was not described.	Parry-Romberg syndrome associated with atrophy of the inferior contralateral member. The abnormalities found suggest autoimmune disorder as an etiological mechanism of the disease. Facial asymmetry was observed, with marked hypoplasia in the face and scarring alopecia affecting the temporal region. In the facial computed tomographic images, atrophy was observed in the sternocleidomastoid, masseter, pterygoid, and soft tissues of the right side of the face.
Longo et al.[10]	A 6-year-old boy; left side affected.	Complaint of epilepsy and seizure crises. Imaging tests, laboratory tests, and serological and neurological examinations. Drug treatment. Follow-up for 2 years.	During the first 6 months of treatment, seizure episodes have decreased from 25% to 50%. After 2 years, a diagnosis of progressive facial hemiatrophy was made. At the last follow-up, the patient had a mild learning disability and denied headaches. The results of this study indicate the association between Parry-Romberg syndrome and encephalitis/Rasmussen syndrome.
Menascu et al.[11]	A 13-year-old boy; left side affected.	Complaint of migraine. Physical examination, laboratory test, diagnostic imaging, and biopsy. The patient was followed up for 18 months with drug treatment.	Patient with *en coup de sabre*, whose examination led to the diagnosis of the syndrome. The patient did not improve with initial drug treatment. When he began treatment with dexamethasone, his migraine ceased. After 18 months, the patient had mild migraines. The symptoms were controlled by using mild analgesics.
Gomez-Diez et al.[1]	A 65-year-old woman; left side affected.	Anatomopathological studies, laboratory tests, and serological test. Treatment was not described.	The patient had progressive facial hemiatrophy at age 45 years, with bruises that evolved into a suggestive morphea plate and with bone lesions involving the maxilla and mandible. Progressive resorption of the maxilla and mandible were observed, showing significant bone tissue atrophy and pathological mandibular fracture. The left half of the lips showed a decrease in thickness. The diagnosis was based on clinical and histopathological results, and patient history.
Paprocka et al.[12]	A 10-year-old girl; left side affected.	Neurological examinations, laboratory tests, diagnostic imaging, and eye examination. Medical and surgical treatments for seizure control.	The patient had characteristics specific to encephalitis/Rasmussen Syndrome. The patient had about 40–50 seizures per day, with indication for implementation of vagus nerve stimulator as treatment.
Hulzebos et al.[20]	A 7-year-old boy; right side affected.	Aesthetic complaint. Laboratory tests and imaging studies. Treatment was not described.	The patient had progressive facial hemiatrophy on the right side of the face, with prominent unilateral chin loss and hyperpigmented but not hardened and retracted skin. A deviation from the middle jawline to the right and *overbite* were observed. A diagnosis of progressive facial hemiatrophy was made, with no intracranial lesions.
Shah et al.[13]	A 7-year-old boy; right side affected.	Physical and neurological examination. Medical and surgical treatment. Monitoring for 1 year after surgery.	The patient had Parry-Romberg Syndrome and epilepsy seizures that were difficult to control. The clinical diagnosis and evaluation led to the diagnosis of encephalitis/Rasmussen syndrome. According to the authors, this is the first report of a pathologically proven case of encephalitis/Rasmussen Syndrome associated with Parry-Romberg Syndrome.

continue...

Chart 1. ...Continuation

Reference	Sample	Complaint and Treatment	Results
Viana et al.[2]	A 37-year-old woman; left side affected.	Complaint about facial pain. Physical examination, thermal testing, diagnostic imaging, and electromyography. Drug treatment.	The patient had gradual loss of tissue, including chewing muscle volume, which led to mild facial asymmetry and difficulty in chewing. There was atrophy of the soft tissue in the left cheek region and the zygomatic arch; mild depression of the nasolabial fold, and *en coup de sabre* sign. The patient had difficulty chewing, the EMG signal in the chewing muscles on the left side remained normal compared with those on the right side.
Hu et al.[3]	6 men and 17 women with an mean age of 21.3 years; the side involved varied between subjects.	Aesthetic complaint. Classification of atrophy as mild, moderate, and severe according to physical examination results. Surgical treatment was required, and follow-up was conducted for 30.8 months.	Report of 23 patients with progressive facial hemiatrophy treated with multiple surgical techniques. With the reconstruction of skeletal and soft tissues, the occlusal plane and malocclusions were corrected, thus improving facial asymmetry.
Qureshi et al.[14]	A 10-year-old boy; left side affected.	Physical examination, eye examination, diagnostic imaging and antibody tests.	The patient had Parry-Romberg Syndrome associated with intracranial vascular malformation. Facial asymmetry was observed on the left side with hyperpigmentation spots.
Miao et al.[4]	A 22-year-old woman; left side affected.	Neurological examinations, laboratory tests and diagnostic imaging, and needle electromyography. Treatment was not described.	The patient had bilateral increased muscle tone, muscle strength, and normal sensitivity, indicating bilateral insult in the pyramidal tract. Disease progression was considered as a rare variant syndrome.
Okumura et al.[15]	A 22-month-old boy; left side affected.	Aesthetic complaint. Neurological examination, diagnostic imaging, and hearing test. Treatment was not conducted, and follow-up was conducted for only 65 months.	The patient had extensive lesions of the white matter, facial asymmetry, and skin atrophy around the region of the eyelids and maxilla. In the last follow-up, the patient had normal psychomotor development and neurological examinations.
Pinheiro et al.[5]	A 8-year-old girl; right side affected.	Complaint of dental changes. Orthodontic evaluation, physical examination, and radiography. No treatment was conducted, only follow-up.	The patient had progressive facial hemiatrophy, presenting facial asymmetry with a marked hypoplasia on the right-side face, lips and nose deviation, large linear scar, and dark *en coup de sabre* on the right side of the jaw and chin, hyperpigmentation areas of the affected skin, unilateral tongue atrophy, atrophic root development, or pathological resorption of the number of permanent teeth.
Anderson et al.[21]	A 9-year-old boy; left side affected. A 14-year-old girl; right side affected.	Complaint of pain. Clinical evaluation. Drug treatment and follow-up.	Report of two cases with Parry-Romberg Syndrome in first cousins. Case 1: developed white lesions on his left cheek, muscle weakness, upper lip anesthesia, asymmetry in his right cheek, and progression of left cheek atrophy, leading to the diagnosis of Parry-Romberg Syndrome. In the evaluation, loss of soft tissue with dentition change was observed. Case 2: developed a depression on the right forehead extending to the neck, facial asymmetry, *en coup de sabre* with skin groove and fat on the right cheek, hypoplasia of the right side of the maxilla and mandible, and malocclusion.

continue...

Chart 1. ...Continuation

Reference	Sample	Complaint and Treatment	Results
Guo et al.[7]	A 63-year-old man; right side affected.	Aesthetic and pain complaint. Physical examination, laboratory and imaging tests, Doppler ultrasonography, and nerve conduction studies. Conducted assessment and drug treatment.	Patient with progressive facial hemiatrophy with unusual features of brain atrophy detected by Doppler ultrasonography. On physical examination, right-side face atrophy, nose and lip deviations, and enophthalmos of the right eye were observed. The patient had brain hemiatrophy to the contralateral side of facial atrophy. For the authors, this is the first case of contralateral cerebral hemiatrophy. This finding extended the progressive facial hemiatrophy spectrum, indicating the possibility of a bilateral disorder.
Haldar & Mukherjee[16]	A 17-year-old woman; right side affected	Complaints of aesthetics and intractable epilepsy. Physical examination, laboratory tests, neurological examinations, and imaging tests. Assessment and drug treatment were conducted.	The patient had skin discoloration, initially diagnosed with scleroderma. Parry-Romberg Syndrome was diagnosed at the onset of facial atrophy in the temporal mandibular region. The authors concluded the report addressing the issue of the association between the two diseases.
O'Flynn & Kinirons[6]	A 7-year-old boy; left side affected.	Aesthetic complaint. Extraoral and intraoral examination, diagnostic imaging, and orthodontic evaluation. Treatment was not described. The patient was reevaluated with intervals of 6 to 12 months, with the last revaluation at 15 years old.	The patient had Parry-Romberg Syndrome and tearing of tooth root. Jaw reduction and area of skin depigmentation, angle atrophy of the mouth and tongue, delayed tooth eruption, facial atrophy, and reduction in a subcutaneous tissue, fat, and cartilage were observed. Tooth lacerations may have been caused by pressure of muscles and soft tissue in the alveolar bone.

Chart 2. Summary of original studies.

Reference	Objective	Methods	Results/Conclusion
Stone[18]	To increase knowledge on rare diseases	On an Internet site, which discusses the Parry-Romberg Syndrome and scleroderma, a research was published online for 1 year.	The participants in this study were 205 patients, of whom 85% were female. Their ages ranged from 1 to 50 years. Age at disease onset was before the age of 15 years in 71% of the cases and after the age of 25 years in 8%. No facial-side predominance was observed. As for the diagnosis, 21% of the sample reported double medical diagnosis of scleroderma with *en coup de sabre* and Parry-Romberg syndrome. With respect to etiological factors, 3% of the respondents reported that they had someone in their family with facial asymmetry. Drug treatment was reported. According to the authors, the pathophysiological mechanism of the syndrome remains unknown.
Sommer et al.[17]	To analyze the results of the evaluations of patients with progressive facial hemiatrophy and discuss possible association between hemiatrophy and scleroderma	In the period of 2000-2004, a review of medical records of patients with localized scleroderma was conducted, with 149 cases of morphea, 55 cases of linear scleroderma, 30 cases of scleroderma *en coup de sabre*, 21 cases of localized scleroderma, 11 cases of deep morphea, and 12 cases of progressive facial hemiatrophy.	This study included seven men and five women with progressive facial hemiatrophy, with a mean age of 22.8 years (range, 6-65 years). The mean age at disease onset was 8.1 years. The authors concluded that progressive facial hemiatrophy is a complex disease with various symptoms.
Blaszczyk et al.[8]	To investigate the relationship between progressive facial hemiatrophy with scleroderma and to establish involvement of the central nervous system in both diseases and possible involvement in other body sites	Participants in this study were divided into three groups. Group 1: ten patients with hemiatrophy who previously had cutaneous sclerosis. Group 2: nine patients with nonpreceded hemiatrophy of cutaneous sclerosis. Control group 3: seven patients with scleroderma. The study covered a period of 6 months.	Neurological symptoms were scarce in all of the groups. A close relationship between progressive facial hemiatrophy and scleroderma was observed.

From the original studies, the common goal was to describe the disease characteristics. The first article[18] studied the characteristics of the disease through a questionnaire posted on a specific pathological site and involved 205 patients; the second[17] retrospectively analyzed medical records of patients; and the third[8] evaluated 26 individuals to verify possible comorbidities between the syndrome and other diseases.

Finally, it is worth noting the numerous muscular and functional impairments described with the syndrome that can potentially lead to changes in the superomedial orbit (SMO).

Considering that the craniofacial complex muscles are required for the integrity and functionality of SMO, the soft tissue atrophy observed in patients with Parry-Romberg Syndrome can have a direct effect on the functions performed by this complex (chewing, swallowing, speaking, and facial expressions). In the studies presented here, the following specific changes were observed: atrophy of the sternocleidomastoid, masseter, and pterygoid[9] muscles; atrophy in the cheek region and mild depression of the nasolabial fold[2]; deviation of the lips and nose[5,7]; and tongue unilateral atrophy[5,6] and atrophy of the mouth angle[6].

Other changes described as alterations of the maxilla, jaw, and teeth can also affect the imbalance between jaw mobility and the chewing and speaking functions. Some authors also showed the involvement of hard tissue as an important atrophy of bone tissue and pathological mandibular fracture[1]; atrophy of the zygomatic arch, frontal bone, and facial asymmetry[2]; atrophic development of roots or pathological roots resorption of permanent teeth[5]; and reduction of jaw and late tooth eruption[6]. Again, this points out that the bony framework integrity of the craniofacial complex is essential for the harmonious functioning of muscles that integrates this system.

Based on this review, we consider the importance of conducting clinical studies that address issues related to speech therapy regarding the diagnosis and rehabilitation of patients with SMO changes resulting from Parry-Romberg syndrome.

CONCLUSIONS

Despite the growing interest in the diagnosis and symptomatic description of individuals with Parry-Romberg Syndrome, publications that address functional and interdisciplinary treatments are scarce. Specific studies aimed at improving the quality of life of these patients are needed.

REFERENCES

1. Gomez-Diez SG, López LG, Escobar ML, Gutiérrez LJ, Oliva NP. Progressive facial hemiatrophy with associated osseous lesions. Med Oral Patol Oral Cir Bucal. 2007;12(8):E602-4. PMid:18059248.
2. Viana M, Glastonbury CM, Sprenger T, Goadsby PJ. Trigeminal neuropathic pain in a patient with progressive facial hemiatrophy (parry-romberg syndrome). Arch Neurol. 2011;68(7):938-43. http://dx.doi.org/10.1001/archneurol.2011.126. PMid:21747035.
3. Hu J, Yin L, Tang X, Gui L, Zhang Z. Combined skeletal and soft tissue reconstruction for severe Parry-Romberg syndrome. J Craniofac Surg. 2011;22(3):937-41. http://dx.doi.org/10.1097/SCS.0b013e31820fe27d. PMid:21558914.
4. Miao J, Liu R, Lin H, Su C, Li H, Lei G, et al. Severe bilateral pyramidal tract involvement in a patient with Parry-Romberg syndrome. Am J Med Sci. 2009;337(3):212-4. http://dx.doi.org/10.1097/MAJ.0b013e31818226f9. PMid:19174689.
5. Pinheiro TPS, Silva CC, Silveira CSL, Botelho PCE, Pinheiro M, Pinheiro JJ. Progressive hemifacial atrophy--case report. Med Oral Patol Oral Cir Bucal. 2006;11(2):E112-4. PMid:16505785.
6. O'Flynn S, Kinirons M. Parry-Romberg syndrome: a report of the dental findings in a child followed up for 9 years. Int J Paediatr Dent. 2006;16(4):297-301. http://dx.doi.org/10.1111/j.1365-263X.2006.00730.x. PMid:16759329.
7. Guo ZN, Zhang HL, Zhou HW, Lan WJ, Wu J, Yang Y. Progressive facial hemiatrophy revisited: a role for sympathetic dysfunction. Arch Neurol. 2011;68(9):1195-7. http://dx.doi.org/10.1001/archneurol.2011.190. PMid:21911700.
8. Blaszczyk M, Królicki L, Krasu M, Glinska O, Jablonska S. Progressive facial hemiatrophy: central nervous system involvement and relationship with scleroderma en coup de sabre. J Rheumatol. 2003;30(9):1997-2004. PMid:12966605.
9. Duymaz A, Karabekmez FE, Keskin M, Tosun Z. Parry-Romberg syndrome: facial atrophy and its relationship with other regions of the body. Ann Plast Surg. 2009;63(4):457-61. http://dx.doi.org/10.1097/SAP.0b013e31818bed6d. PMid:19745718.
10. Longo D, Paonessa A, Specchio N, Delfino LN, Claps D, Fusco L, et al, and the Clinical and Neuroimaging Features. Parry-Romberg syndrome and Rasmussen encephalitis: possible association. Clinical and neuroimaging features. J Neuroimaging. 2011;21(2):188-93. http://dx.doi.org/10.1111/j.1552-6569.2009.00398.x. PMid:19555404.
11. Menascu S, Padeh S, Hoffman C, Ben-Zeev B. Parry-Romberg syndrome presenting as status migrainosus. Pediatr Neurol. 2009;40(4):321-3. http://dx.doi.org/10.1016/j.pediatrneurol.2008.11.007. PMid:19302950.
12. Paprocka J, Jamroz E, Adamek D, Marszal E, Mandera M. Difficulties in differentiation of Parry-Romberg syndrome, unilateral facial sclerodermia, and Rasmussen syndrome. Childs Nerv Syst. 2006;22(4):409-15. http://dx.doi.org/10.1007/s00381-005-1262-x. PMid:16247619.
13. Shah JR, Juhász C, Kupsky WJ, Asano E, Sood S, Fain D, et al. Rasmussen encephalitis associated with Parry-Romberg syndrome. Neurology. 2003;61(3):395-7. http://dx.doi.org/10.1212/WNL.61.3.395. PMid:12913207.
14. Qureshi UA, Wani NA, Altaf U. Parry-Romberg syndrome associated with unusual intracranial vascular malformations and Phthisis bulbi. J Neurol Sci. 2010;291(1-2):107-9. http://dx.doi.org/10.1016/j.jns.2010.01.003. PMid:20144465.
15. Okumura A, Ikuta T, Tsuji T, Kato T, Fukatsu H, Naganawa S, et al. Parry-Romberg syndrome with a clinically silent white matter lesion. AJNR Am J Neuroradiol. 2006;27(8):1729-31. PMid:16971623.
16. Haldar A, Mukherjee A. Parry Romberg's disease with intractable partial epilepsy. Neurol India. 2007;55(2):160-2. http://dx.doi.org/10.4103/0028-3886.32791. PMid:17558124.

17. Sommer A, Gambichler T, Bacharach-Buhles M, von Rothenburg T, Altmeyer P, Kreuter A. Clinical and serological characteristics of progressive facial hemiatrophy: a case series of 12 patients. J Am Acad Dermatol. 2006;54(2):227-33. http://dx.doi.org/10.1016/j.jaad.2005.10.020. PMid:16443052.

18. Stone J. Parry-Romberg syndrome: a global survey of 205 patients using the Internet. Neurology. 2003;61(5):674-6. http://dx.doi.org/10.1212/WNL.61.5.674. PMid:12963760.

19. Carvalho MV, Nascimento GJ, Andrade E, Andrade M, Sobral APV. Association of aesthetic and orthodontic treatment in Parry-Romberg syndrome. J Craniofac Surg. 2010;21(2):436-9. http://dx.doi.org/10.1097/SCS.0b013e3181cfe917. PMid:20216455.

20. Hulzebos CV, de Vries TW, Armbrust W, Sauer PJJ, Kerstjens-Frederikse WS. Progressive facial hemiatrophy: a complex disorder not only affecting the face. A report in a monozygotic male twin pair. Acta Paediatr. 2004;93(12):1665-9. http://dx.doi.org/10.1111/j.1651-2227.2004.tb00861.x. PMid:15918232.

21. Anderson PJ, Molony D, Haan E, David DJ. Familial Parry-Romberg disease. Int J Pediatr Otorhinolaryngol. 2005;69(5):705-8. http://dx.doi.org/10.1016/j.ijporl.2004.12.004. PMid:15850693.

22. The Cochrane Collaboration. Cochrane handbook for systematic reviews of intervention [Internet]. 2011 [acesso 2011 Maio 11]. Disponível em: www.cochrane.org/training/cochrane-handbook.

***Corresponding author:** **Claudia Regina Furquim de Andrade**
Rua Cipotânea, 51 – Cidade Universitária – São Paulo, SP, Brazil
CEP 05360-160
E-mail: clauan@usp.br