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Progressive facial hemiatrophy (Parry-Romberg syndrome), a rare cause of temporal lobe epilepsy: Case report

Felipe dos Santos Souza, Marco Orsini, Gilberto Canedo Martins Jr, Marcelo Namen, Antonio Carlos Garcia Dias Mayall, Antônio Marcos da Silva Catharino

ABSTRACT

Introduction: Parry-Romberg syndrome is also known as progressive hemifacial atrophy. It, which is characterized by a unilateral atrophy of the skin, soft tissue, muscles, and/or bones of the face, is a variant of linear morphea or an independent disorder. It has sporadic neurocutaneous involvement, occasionally it can even involve the central nervous system (CNS). In this condition, facial atrophy may be accompanied by classic linear morphea lesions on the face or elsewhere. Parry-Romberg syndrome is an uncommon disorder, more common in females, usually manifested in the first decade of life, but its etiology remains unknown.

Case Report: A woman with 49 years old, seeking neurological care due to epileptic seizures that started 21 years ago. The crises described were mostly focal, with compromised consciousness, symptomatology related to the temporal lobe with automatisms and sensory hallucinations. Physical examination: right hemifacial atrophy was observed with hemilingual atrophy and right enophthalmos.

Conclusion: The patient was diagnosed with Parry-Romberg syndrome after semiological evaluation of the neurology and investigation of the patient's clinical history. The electroencephalogram showed marked signs of right temporal cortico-subcortical dysfunction with 2–4 Hz waves, slow wave bursts. The conduct is clinical and the prognosis restricted to each patient.

Keywords: Epilepsy, Facial hemiatrophy, Neurocutaneous syndromes, Temporal lobe

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INTRODUCTION

Parry-Romberg syndrome (PRS), also known as idiopathic progressive hemifacial atrophy, is a rare neurocutaneous disease characterized by the loss of skin and subcutaneous fat from the face, muscles, and bones causing unilateral atrophy. Most patients only require soft tissue augmentation, although the syndrome has varying degrees of severity [1]. Facial hemitrophy progresses slowly and often affects one side of the face. Its pathophysiology, according to some authors, may be related to an inflammatory meningoencephalic with vasculitis, or a chronic vasomotor disorder. Ophthalmic

involvement is common, with progressive enophthalmia being a frequent finding [2].

Treatment is usually esthetic using different procedures for correcting facial atrophy of cosmetic dermatological surgery [3].

CASE REPORT

A 49-year-old woman sought neurological care due to epileptic seizures that started 21 years ago. The crises described were mostly focal, with compromised consciousness, symptomatology related to the temporal lobe with automatisms and sensory hallucinations. In addition, she also had occasional episodes of generalized seizures that occurred during sleep. Physical examination: right hemifacial hypotrophy with hemilingual atrophy and right enophthalmos were observed (Figure 1). No other significant neurological deficits were found. She reported a family history of epilepsy. On examinations: polysomnography showed increased airway resistance. Magnetic resonance imaging of the brain showed: volume reduction in the right cerebral hemisphere (Figure 2). Electroencephalogram showed marked signs of right temporal cortico-subcortical dysfunction with 2–4 Hz waves, slow wave bursts (Figure 3). Polysomnography was requested due to complaints of sleep problems, and showed mild sleep apnea-hypopnea syndrome, with an apnea-hypopnea index of 10.9/h and 124 micro-arousals and good response to continuous positive airway pressure (CPAP) with a pressure of 7 cm H_2O. She was referred for otorhinolaryngological evaluation. Performed computerized tomography (CT) of the paranasal sinuses that showed mucous thickening in the left maxillary sinus and nasal septum deviation. Despite the treatment with divalproate sodium 500 mg 12/12 h, and phenobarbital 100 mg at night, as prescribed by the neurologist who was monitoring her, lacosamide 100 mg 12/12 h was added to the therapeutic regimen with an important reduction in epileptic events that started to occur sporadically, especially when there was therapeutic failure.

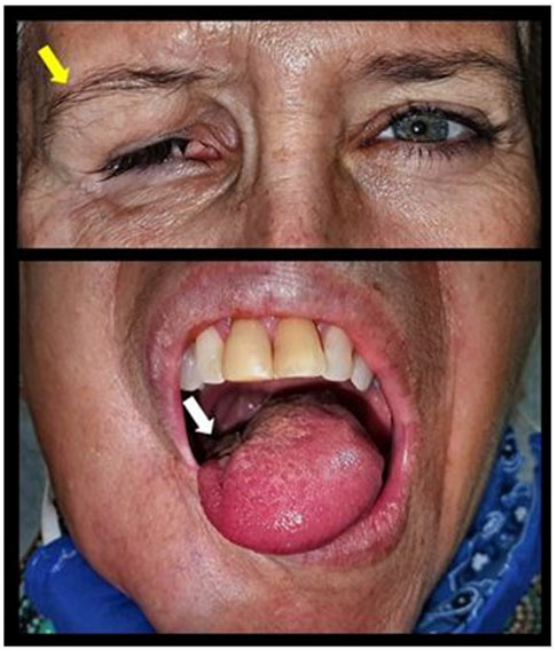

Figure 1: Facial asymmetry with right hemifacial atrophy, hemilingual atrophy (white arrow) and right enophthalmos (yellow arrow).

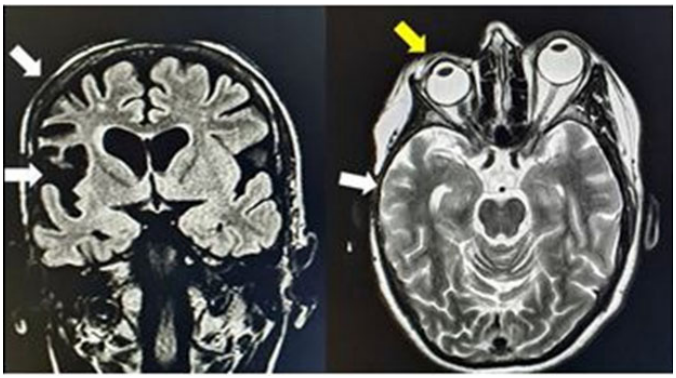

Figure 2: MRI-accentuation of cortical sulci, fissures and cerebral cisterns, especially on the right (white arrows), with ectasia of the supratentorial ventricular system, greater than expected for the age group. Slightly asymmetrical hippocampus, with smaller volume on the right associated with accentuation of the choroidal fissure in correspondence, both with preserved sign. Right enophthalmos (yellow arrow).

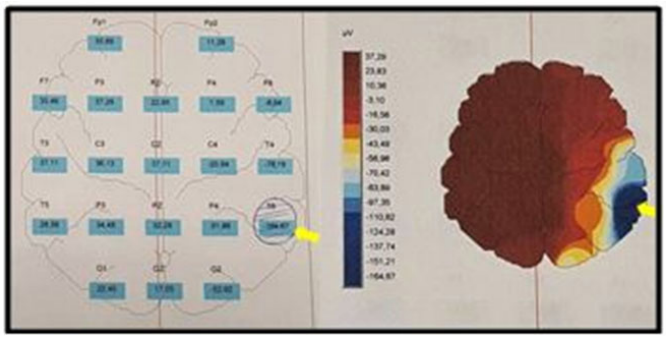

Figure 3: EEG-marked signs of structural dysfunction with an irritative component on T6, slow rhythm 1.5–4.0 Hz and spikes in the right posterior temporal region (yellow arrows) radiating to neighboring areas, alpha reduction on the right.

DISCUSSION

Parry-Romberg syndrome is described as an uncommon disorder, whose neurological symptoms include headache, trigeminal neuralgia, and focal epilepsy, more common in females, usually manifested in the first decade of life [4, 5]. Computed tomography and MRI are excellent methods for diagnosing CNS alterations, and the characteristic findings are: white matter hypersignal on T2 and FLAIR, leptomeningeal enhancement, intracranial calcifications, and cerebral atrophy [6–8].

Differential diagnosis includes diseases that present with cerebral hemiatrophy, such as Rasmussen encephalitis and Sturge-Weber syndrome, but in these we do not observe the typical hemifacial alterations like in Parry-Romberg syndrome [5]. Advanced MRI sequences are currently little explored in Parry-Romberg syndrome and may help explain its pathophysiology.

The cause of hemilateral cutaneous atrophy in the face of a patient with Parry-Romberg syndrome does not yet have a known etiopathogenesis [9–11]. It is known that immunological disorders are involved in the pathophysiology of the disease [12]. During the active phase of progressive atrophy, there are some medications that help to control and stabilize other symptoms such as headache, trigeminal neuralgia, and focal epilepsy. In the case reported here, the patient had altered brain activity in the right temporal cortico-subcortical region with 2–4 Hz waves, slow wave bursts, after performing an electroencephalogram [13, 14].

CONCLUSION

The presented case corroborates the syndromic diagnosis of Parry-Romberg syndrome. Complementary exams such as MRI and cranial CT ruled out other neurological conditions that could justify the progressive clinical presentation. The description and scientific documentation of case reports that explain how the diagnosis is made, as well as the clinical management of the disease, is relevant.

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Felipe dos Santos Souza – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Marco Orsini – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Gilberto Canedo Martins Jr – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Marcelo Namen – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Antonio Carlos Garcia Dias Mayall – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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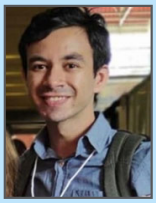

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