

CASE REPORT

PEER REVIEWED | OPEN ACCESS

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

How to cite this article

Souza FDS, Orsini M, Martins Jr GC, Namen M, Mayall ACGD, da Silva Catharino AM. Progressive facial hemiatrophy (Parry-Romberg syndrome), a rare cause of temporal lobe epilepsy: Case report. Int J Case Rep Images 2021;12:101252Z01FS2021.

Felipe dos Santos Souza¹, Marco Orsini², Gilberto Canedo Martins Jr³, Marcelo Namen⁴, Antonio Carlos Garcia Dias Mayall², Antônio Marcos da Silva Catharino⁵

Affiliations: ¹Undergraduate student of Medicine at Universidade do Estado de Mato Grosso (UNEMAT), Cáceres, Mato Grosso, Brazil; ²Programa de Mestrado em Neurologia - Universidade de Vassouras e Escola de Medicina - Universidade Iguazu - UNIG, RJ, Nova Iguaçu, Brazil; ³Adjunct Professor of the Medicine course at the Iguazu University, Iguazu University - UNIG/RJ, Nova Iguaçu, Brazil; ⁴Physician, Neurologist, Hospital Samer Rede D'or São Luiz, Resende, RJ, Brazil; ⁵Physician, Neurologist, PhD student in Neurology at the Federal University of the State of Rio de Janeiro - UNIRIO, Adjunct Professor of the Medicine course at the Iguazu University, Iguazu University - UNIG, RJ, Nova Iguaçu, Brazil.

Corresponding Author: Antônio Marcos da Silva Catharino, Rua Gavião Peixoto 70, Room 811, CEP 24.2230-100, Icaraí, Niterói-RJ, Brazil; Email: neurocurso@globo.com

Received: 06 July 2021

Accepted: 23 August 2021

Published: 12 September 2021

Article ID: 101252Z01FS2021

doi: 10.5348/101252Z01FS2021CR

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 hemiatrophy 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 H₂O. She was referred for otorhinolaryngological evaluation. Performed computerized tomography (CT) of the paranasal sinuses



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

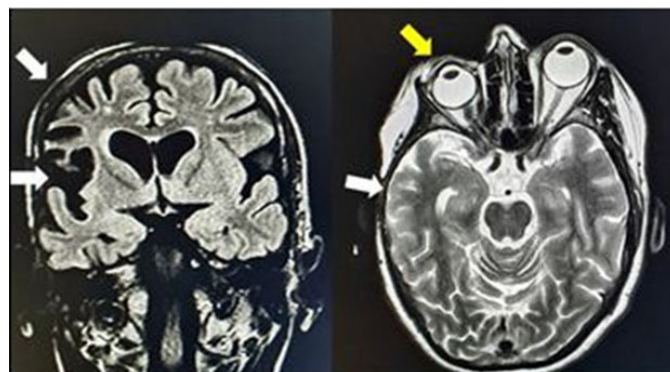


Figure 2: MRI-accentuation of cortical sulci, fissures and cerebral cisterns, especially on the right (white arrows), with ectasia of the supratentorial ventricular system, greater than expected for the age group. Slightly asymmetrical hippocampus, with smaller volume on the right associated with accentuation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

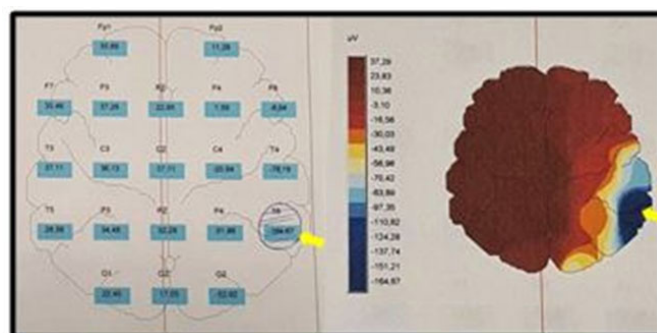


Figure 3: EEG-marked signs of structural dysfunction with an irritative component on T6, slow rhythm 1.5–4.0 Hz and spikes in the right posterior temporal region (yellow arrows) radiating to neighboring areas, alpha reduction on the right.

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.

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.

REFERENCES

1. Alfenas R, Niemeyer B, Bahia PRV, Niemeyer R, Balbi L. Síndrome de Parry-Romberg: Achados nas sequências avançadas de ressonância magnética – relato de caso. *Radiol Bras* 204;47(3):186–8.
2. Zulian F, Vallongo C, Woo P, et al. Localized scleroderma in childhood is not just a skin disease. *Arthritis Rheum* 2005;52(9):2873–81.
3. Teske NM, Jacobe H. Morphea (localized scleroderma). In: *Scleroderma: From Pathogenesis to Comprehensive Management*. Cham: Springer International Publishing; 2016. p. 91–113.
4. Provost TT, Greenberg AS, Falanga V. Localized cutaneous sclerosis. In: *Sontheimer RD, Provost TT, editors. Cutaneous Manifestations of Rheumatic Diseases*. 1ed. Baltimore: Williams & Wilkins; 1996. p. 125.
5. Sehgal VN, Srivastava G, Aggarwal AK, Behl PN, Choudhary M, Bajaj P. Localized scleroderma/morphea. *Int J Dermatol* 2002;41(8):467–75.
6. Christen-Zaech S, Hakim MD, Afsar FS, Paller AS. Pediatric morphea (localized scleroderma): Review of 136 patients. *J Am Acad Dermatol* 2008;59(3):385–96.
7. Leitenberger JJ, Cayce RL, Haley RW, Adams-Huet B, Bergstresser PR, Jacobe HT. Distinct autoimmune

- syndromes in morphea: A review of 245 adult and pediatric cases. *Arch Dermatol* 2009;145(5):545–50.
8. Peterson LS, Nelson AM, Su WP, Mason T, O'Fallon WM, Gabriel SE. The epidemiology of morphea (localized scleroderma) in Olmsted County 1960–1993. *J Rheumatol* 1997;24(1):73–80.
9. Dharamsi JW, Victor S, Aguwa N, et al. Morphea in adults and children cohort III: Nested case-control study—the clinical significance of autoantibodies in morphea. *JAMA Dermatol* 2013;149(10):1159–65.
10. Donofrio LM. Fat distribution: A morphologic study of the aging face. *Dermatol Surg* 2000;26(12):1107–12.
11. Rohrich RJ, Pessa JE, Ristow B. The youthful cheek and the deep medial fat compartment. *Plast Reconstr Surg* 2008;121(6):2107–12.
12. Shaw RB Jr, Katzel EB, Koltz PF, Kahn DM, Giroto JA, Langstein HN. Aging of the mandible and its aesthetic implications. *Plast Reconstr Surg* 2010;125(1):332–42.
13. Carruthers J, Carruthers A. A prospective, randomized, parallel group study analyzing the effect of BTX-A (Botox) and nonanimal sourced hyaluronic acid (NASHA, Restylane) in combination compared with NASHA (Restylane) alone in severe glabellar rhytides in adult female subjects: Treatment of severe glabellar rhytides with a hyaluronic acid derivative compared with the derivative and BTX-A. *Dermatol Surg* 2003;29(8):802–9.
14. Patel MP, Talmor M, Nolan WB. Botox and collagen for glabellar furrows: Advantages of combination therapy. *Ann Plast Surg* 2004;52(5):442–7.

Author Contributions

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

Antônio Marcos da Silva Catharino – 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

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

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

Copyright

© 2021 Felipe dos Santos Souza et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

ABOUT THE AUTHORS

Article citation: Souza FDS, Orsini M, Martins Jr GC, Namen M, Mayall ACGD, da Silva Catharino AM. Progressive facial hemiatrophy (Parry-Romberg syndrome), a rare cause of temporal lobe epilepsy: Case report. Int J Case Rep Images 2021;12:101252Z01FS2021.



Felipe dos Santos Souza is a medical student at the State University of Mato Grosso (UNEMAT), Cáceres, Mato Grosso, Brazil. He has published in the Brazilian Journal of Emergency Medicine (ISSN-2763-776X), Brazilian Journal of Health Review (ISSN-2595-6825), Brazilian Journal of Development Magazine (ISSN-2525-8761), Journal of Biological and Health Sciences (ISSN-1518-4595), and Annals of Clinical Case Studies (ISSN-2688-1241). His research interests include neurology, internal medicine and neurological syndromes.

Email: felipe.santos@unemat.br



Marco Orsini, Full Professor at Universidade Iguazu and Vassoras University, Rio de Janeiro, Brazil. He earned the undergraduate degree in Medicine from Rio de Janeiro Federal University/IPUB/Rio de Janeiro/Brazil and postgraduate degree from Fluminense Federal University, Department of Medicine Niterói, RJ, Brazil. He has published 420 articles/papers in national and international academic journals and authored 2 books. His research interests include Motor Neuron Diseases, Pain, and Peripheral Neuropathies. Now he is continuing his PhD in Rio de Janeiro from Federal University and Baylor.

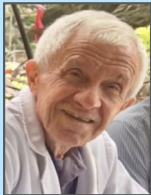


Gilberto Canedo Martins Jr, Adjunct Professor of the Medicine course at the Iguazu University, Iguazu University - UNIG/RJ, Nova Iguazu, Brazil.

Email: gilbertocmj@yahoo.com.br



Marcelo Namen is a Physician, Neurologist at Hospital Samer Rede D'or São Luiz, Resende, RJ-Brazil.
Email: marcelonamen@hotmail.com



Antonio Carlos Garcia Dias Mayall, Adjunct Professor of the Medicine course at the Iguaçú University, Iguaçú University - UNIG/RJ, Nova Iguaçú, Brazil.



Antônio Marcos da Silva Catharino is a Physician, Neurologist, PhD student in Neurology at the Federal University of the State of Rio de Janeiro - UNIRIO, Adjunct Professor of the Medicine course at the Iguaçú University, Iguaçú University - UNIG/RJ, Nova Iguaçú, Brazil. Orcid: <https://orcid.org/0000-0002-5736-1486>
Email: neurocurso@globo.com

Access full text article on
other devices



Access PDF of article on
other devices





INTERNATIONAL JOURNAL OF
CASE REPORTS AND IMAGES



VIDEO JOURNAL OF
CLINICAL RESEARCH



VIDEO JOURNAL OF
BIOMEDICAL SCIENCE



INTERNATIONAL JOURNAL OF
HEPATOBIILIARY AND
PANCREATIC DISEASES



INTERNATIONAL JOURNAL OF
BLOOD TRANSFUSION AND
IMMUNOHEMATOLOGY



EDORIUM JOURNAL OF
OPHTHALMOLOGY



Submit your manuscripts at
www.edoriumjournals.com



EDORIUM JOURNAL OF
MEDICINE



EDORIUM JOURNAL OF
CARDIOTHORACIC AND
VASCULAR SURGERY



JOURNAL OF CASE REPORTS
AND IMAGES IN ORTHOPEDICS
AND RHEUMATOLOGY



EDORIUM JOURNAL OF
PSYCHOLOGY



EDORIUM JOURNAL OF
CELL BIOLOGY



JOURNAL OF CASE REPORTS AND IMAGES IN
DENTISTRY



EDORIUM JOURNAL OF
CANCER



EDORIUM JOURNAL OF
PSYCHIATRY



JOURNAL OF CASE REPORTS AND
IMAGES IN INFECTIOUS DISEASES



EDORIUM JOURNAL OF
ANATOMY AND EMBRYOLOGY



EDORIUM JOURNAL OF
SURGERY



JOURNAL OF CASE REPORTS
AND IMAGES IN PATHOLOGY



EDORIUM JOURNAL OF
ANESTHESIA