# **ARTICLE IN PRESS**

Case Studies

# Ischemic Stroke in a Patient with Parry–Romberg Syndrome

Victoria Ebiana, MD,\*/† Sandeep Singh,§ Shaweta Khosa, MD,|| Negar Moheb, MD,\*/|| Bhavesh Trikamji, MD,¶ Neal M. Rao, MD,\*/|| and Shri K. Mishra, MD, MS, ABMS, FAAN, FNAA, FANA\*/†/||/‡

> Objective: This study aimed to discuss a case of a patient with a known diagnosis of Parry-Romberg syndrome (PRS) presenting with ischemic stroke, the second such reported case. Background: PRS is a rare genetic disorder with progressive hemifacial atrophy, which usually presents within the first 2 decades of life. Neurologic manifestations include trigeminal neuralgia with associated deafness, hemifacial pain with associated migraine headaches, seizures, movement disorders, and neuropsychiatric symptoms. Many patients have elevated antinuclear antibody (ANA) titers. However, stroke is uncommon. Case description: A 34-year-old righthanded woman, diagnosed with PRS at age 15, presented with right-sided weakness on waking up. Brain magnetic resonance imaging revealed a small infarct of the posterior limb of the left internal capsule. Vessel imaging revealed an aberrant right subclavian artery. Atrophy of the right-sided muscles of mastication is consistent with her known diagnosis of right-sided PRS. Stroke workup revealed a patent foramen ovale; however, no evidence of deep venous thrombosis was found. Hypercoagulability workup revealed an elevated ANA. The cause of stroke in this patient with PRS remains unclear, as she has no known risk factors. Conclusion: It is possible that elevated inflammatory markers associated with PRS may cause a proinflammatory state and predispose patients to small-vessel vasculopathy. It is important to note the association between PRS and ischemic stroke. Key Words: Parry-Romberg syndrome-stroke-progressive hemifacial atrophy-Romberg syndrome.

> © 2017 National Stroke Association. Published by Elsevier Inc. All rights reserved.

Received May 28, 2017; revision received August 12, 2017; accepted August 21, 2017.

Address correspondence to Shri K. Mishra, MD, MS (Administrative Medicine), ABMS, FAAN, FNAA, FANA, Department of Neurology, Keck School of Medicine of USC, 1975 Zonal Avenue, Los Angeles, CA 90089-9034. E-mail: smishra@usc.edu.

1052-3057/\$ - see front matter

## Objective

This study aimed to discuss a rare case of a patient with a known diagnosis of Parry–Romberg syndrome (PRS) presenting with ischemic stroke, the second such reported case.

#### Background

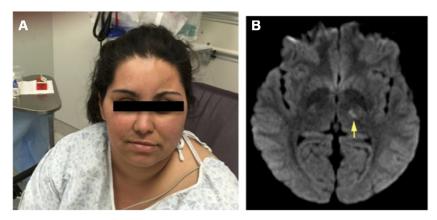
PRS is a rare genetic disorder with progressive hemifacial atrophy, which usually presents within the first 2 decades of life.<sup>1</sup> It was first described by Caleb Parry in 1825 and by Moritz Romberg in 1846.<sup>2</sup> This condition is characterized by slowly progressive unilateral facial atrophy of the skin, soft tissue, muscles, and underlying bony

From the \*Department of Neurology, David Geffen School of Medicine at UCLA, Los Angeles, California; †VA Greater Los Angeles HCS, Los Angeles, California; ‡Keck School of Medicine at USC, Los Angeles, California; §Western University of Health Sciences, Pomona, California; ∥Olive View—UCLA Medical Center, Sylmar, California; and ¶Harbor—UCLA Medical Center, Torrance, California.

<sup>© 2017</sup> National Stroke Association. Published by Elsevier Inc. All rights reserved.

https://doi.org/10.1016/j.jstrokecerebrovasdis.2017.08.032

# ARTICLE IN PRESS



**Figure 1.** (A) Right-sided hemifacial atrophy in a patient with Parry–Romberg syndrome. (B) Diffusion-weighted magnetic resonance imaging (MRI) image showing acute infarction involving the left internal capsule.

structures.<sup>3</sup> Neurologic manifestations include trigeminal neuralgia with associated deafness, hemifacial pain with associated migraine headaches, seizures, movement disorders, and neuropsychiatric symptoms.<sup>4-6</sup> Many patients have elevated antinuclear antibody (ANA) titers. However, stroke is uncommon.

# **Case Description**

A 34-year-old right-handed woman, diagnosed with PRS at the age of 15, presented with right-sided weakness on awakening. On physical examination, atrophy of the rightsided muscles of mastication consistent with her known diagnosis of right-sided PRS was noted. Brain magnetic resonance imaging revealed a small infarct of the posterior limb of the left internal capsule (Fig 1). Vessel imaging revealed an aberrant right subclavian artery. Stroke workup revealed a patent foramen ovale; however, no evidence of deep venous thrombosis was found. Workup for hypercoagulability revealed an elevated ANA titer of 1:1280; erythrocyte sedimentation rate slightly elevated at 24. Both ANA and erythrocyte sedimentation rate were stable on recheck 3 months later. The patient never developed any systemic signs of autoimmune disease. The cause of stroke in this patient with PRS remains unclear-the stroke was in a small-vessel disease territory; however, she had no evidence of traditional risk factors such as diabetes, hypertension, or hyperlipidemia. The patient showed no residual neurologic deficits, and no radiographic change was seen on subsequent follow-up.

## Conclusion

Inflammatory markers associated with PRS may indicate a proinflammatory state and predispose patients to small-vessel vasculopathy, independent of any signs of systemic autoimmune symptoms. Alternatively, as the only other reported patient with PRS and stroke was also found to have a patent foramen ovale,<sup>7</sup> it is possible that these patients may have a predisposition to small venous clots and paradoxical emboli. However, the location of the infarction in this patient is not typical of embolic stroke. This case supports the potential association between PRS and ischemic stroke in young patients without other known risk factors.

#### References

- 1. Duymaz A, Karabekmez FE, Keskin M, et al. Parry-Romberg syndrome: facial atrophy and its relationship with other regions of the body. Ann Plast Surg 2009;63:457-461. doi:10.1097/SAP.0b013e31818bed6d.
- Tolkachjov SN, Patel NG, Tollefson MM. Progressive hemifacial atrophy: a review. Orphanet J Rare Dis 2015;10:39. doi:10.1186/s13023-015-0250-9.
- 3. Jun JH, Kim HY, Jung HJ, et al. Parry-Romberg syndrome with en coup de sabre. Ann Dermatol 2011;23:342-347. doi:10.5021/ad.2011.23.3.342.
- Kister I, Inglese M, Laxer RM, et al. Neurologic manifestations of localized scleroderma: a case report and literature review. Neurology 2008;71:1538-1545.
- Doolittle DA, Lehman VT, Schwartz KM, et al. CNS imaging findings associated with Parry-Romberg syndrome and en coup de sabre: correlation to dermatologic and neurologic abnormalities. Neuroradiology 2014;57:21-34.
- Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: a retrospective review of 54 patients. J Am Acad Dermatol 2007;56:257-263.
- Tomizawa Y, Tanaka R, Sekiguchi K, et al. Cerebral infarction in a case of Parry-Romberg syndrome. J Stroke Cerebrovasc Dis 2014;23:393-394. doi:10.1016/ j.jstrokecerebrovasdis.2013.02.015.