

Teaching NeuroImages: Progressive facial hemiatrophy (Parry-Romberg syndrome) with ipsilateral cerebral hemiatrophy

Domenico Antonio Restivo, MD, PhD
Pietro Milone, MD

Address correspondence and reprint requests to Dr. Domenico Antonio Restivo, Department of Neurology, “Nuovo Garibaldi” Hospital, via Palermo 95100, Catania, Italy
darestivo@libero.it

Figure 1 Severe left facial hemiatrophy in a patient with Parry-Romberg syndrome



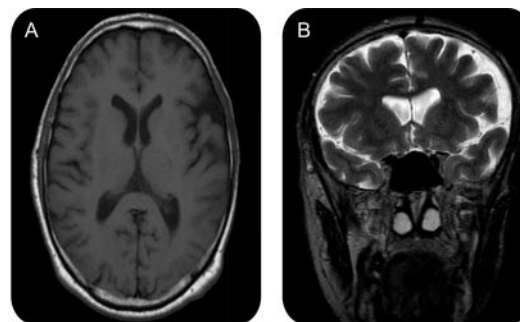
A 52-year-old man presented with left hemifacial atrophy (figure 1) beginning at age 25. There were no neurologic symptoms. Neurologic examination showed no deficits, and the limbs were symmetric. Brain MRI demonstrated left cerebral hemiatrophy (figure 2) and lack of the ipsilateral soft facial tissue.

Progressive facial hemiatrophy (PFH), or Parry-Romberg syndrome, is a sporadic disease of unknown etiology characterized by progressive shrinking and deformation of one hemiface with subcutaneous connective and fatty tissue atrophy.^{1,2} Rarely, brain MRI shows cerebral hemiatrophy, usually ipsilateral to the facial atrophy. PFH can present with neurologic symptoms such as trigeminal neuralgia and focal epilepsy.^{1,2} Histologic findings reveal a proliferative interstitial neurovasculitis.³ Chronic localized meningoencephalitis with vascular involvement may be a cause of the occasional brain involvement in PFH.³ The coexistence of brain and facial atrophy on the same side suggests that facial atrophy is not caused by brain injury. Rasmussen encephalitis, however, may also be associated with PFH, suggesting that these 2 conditions may share a common etiology.⁴

REFERENCES

1. Asher SW, Berg BO. Progressive facial hemiatrophy: report of three cases, including one observed over 43 years, and computed tomography findings. *Arch Neurol* 1982;39:44–46.
2. Lerner AJ, Bennison DP. Some observations on the aetiology of progressive hemifacial atrophy (“Parry-Romberg syndrome”). *J Neurol Neurosurg Psychiatry* 1993;56:1035–1039.
3. Pensler JM, Murphy GF, Mulliken JB. Clinical and ultrastructural studies in Romberg’s hemifacial atrophy. *Plast Reconstr Surg* 1990;85:669–674.
4. Shah JR, Juhász C, Kupsky WJ, et al. Rasmussen encephalitis associated with Parry-Romberg syndrome. *Neurology* 2003;61:395–397.

Figure 2 Brain MRI demonstrating remarkable left cerebral hemiatrophy



(A) Axial T1-weighted image shows the left cortical temporal atrophy. (B) Coronal T2-weighted image shows severe left cortical cerebral atrophy and lack of the ipsilateral soft facial tissue.

Neurology[®]

Teaching *NeuroImages*: Progressive facial hemiatrophy (Parry-Romberg syndrome) with ipsilateral cerebral hemiatrophy

Domenico Antonio Restivo and Pietro Milone

Neurology 2010;74:e11

DOI 10.1212/WNL.0b013e3181ca00af

This information is current as of January 18, 2010

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/74/3/e11.full
References	This article cites 4 articles, 2 of which you can access for free at: http://n.neurology.org/content/74/3/e11.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Clinical Neurology http://n.neurology.org/cgi/collection/all_clinical_neurology All Medical/Systemic disease http://n.neurology.org/cgi/collection/all_medical_systemic_disease Autoimmune diseases http://n.neurology.org/cgi/collection/autoimmune_diseases Developmental disorders http://n.neurology.org/cgi/collection/developmental_disorders MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

