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.
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://www.neurology.org/content/74/3/e11.full.html

References
This article cites 4 articles, 2 of which you can access for free at:
http://www.neurology.org/content/74/3/e11.full.html##ref-list-1

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
All Clinical Neurology
http://www.neurology.org/cgi/collection/all_clinical_neurology
All Medical/ Systemic Disease
http://www.neurology.org/cgi/collection/all_medical_systemic_disease

Autoimmune diseases
http://www.neurology.org/cgi/collection/autoimmune_diseases
Developmental disorders
http://www.neurology.org/cgi/collection/developmental_disorders
MRI
http://www.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/misc/about.xhtml#permissions

Reprints
Information about ordering reprints can be found online:
http://www.neurology.org/misc/addir.xhtml#reprintsus