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

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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

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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.

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