

Progressive facial hemiatrophy: Abnormality of intracranial vasculature

Article abstract—Progressive facial hemiatrophy (PFH) or Parry-Romberg syndrome is associated with ipsilateral brain lesions and neurologic symptoms. We describe a 35-year-old man with PFH and frequent hemiplegic migraine. On cerebral angiography, reversible vessel caliber changes were seen within the symptomatic hemisphere. An abnormality of the intracranial vasculature may be present in some patients with PFH and neurologic manifestations.

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Progressive facial hemiatrophy (PFH; also known as Parry-Romberg syndrome) is characterized by slowly progressive but self-limited atrophy of facial subcutaneous fat, which can be followed by wasting of associated skin, cartilage, connective or ocular tissue, muscle, and bone.^{1,2} PFH is most often unilateral and usually affects one or more of the trigeminal dermatomes, although body parts outside this distribution may be involved.^{1,2} Its relation to linear scleroderma is unclear, although some authors consider these to be overlapping conditions.¹ Symptoms usually begin in the first or second decades of life, but onset has occurred in patients older than 60 years.² Many patients are neurologically normal, although symptoms attributable to ipsilateral cerebral hemispheric dysfunction are not uncommon. Seizures are the most frequent neurologic symptom.¹ Others include cranial nerve deficits, masticatory spasms, cognitive abnormalities, and fixed focal deficits.^{1,2} Hemiplegic migraine has been the focus of only two previous reports.^{3,4}

CT and MRI findings in PFH patients with neurologic disease are usually ipsilateral and may include hemispheric calcifications, leptomeningeal enhancement, loss of cortical gyration, hyperintensities in deep white and gray matter structures, and atrophy (manifested by dilatation of the lateral ventricle).^{1,2,5} The underlying pathophysiology of these changes is unknown. Results of cerebral angiography have frequently been normal in those studied.⁵

We describe a patient with PFH and frequent hemiplegic migraine observed over 5 years. On cerebral angiography, reversible vessel caliber changes were seen within the symptomatic hemisphere.

Case report. The patient, a 35-year-old right-handed man, began having neurologic symptoms in early 1991. At that time, he developed hesitant initiation of speech when reading aloud. In late 1991, he had an episode of numbness and paresthesias in his right arm. The spell started in the fingertips and migrated, over several minutes, to the shoulder. His arm became weak and he developed an expressive dysphasia. His leg was unaffected. These symptoms lasted about 1 hour and as they resolved, the patient developed a generalized throbbing headache with nausea, vomiting, photophobia, and phonophobia. He had two similar episodes over the next 2 months, although the transient symptoms before the headache lasted 2 hours rather than 1.

At age 34 years, the patient developed an atrophic hyperpigmented patch of skin on his left forehead diagnosed as PFH. His history was otherwise unremarkable. There was no family history of neurologic illness.

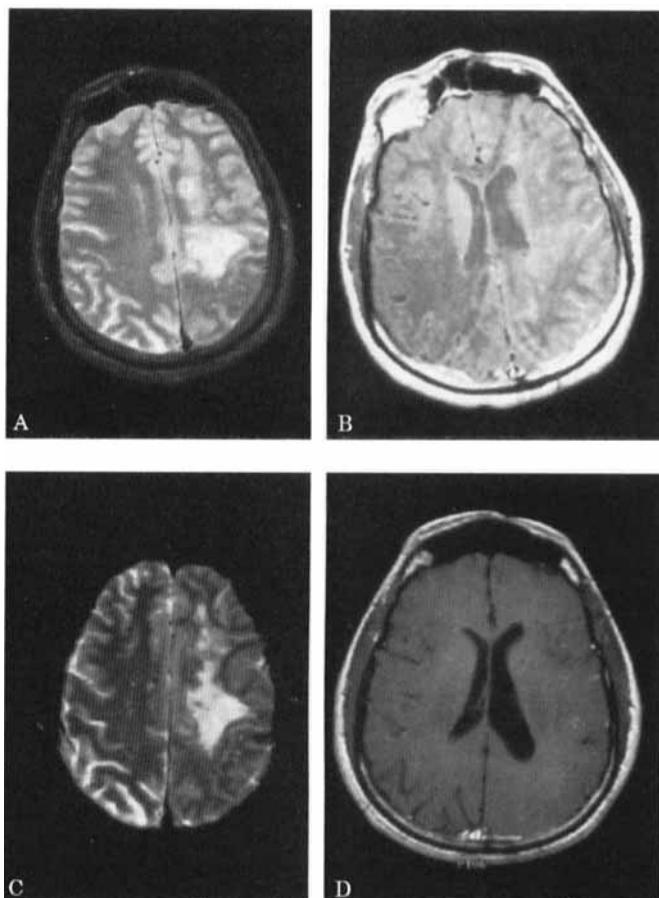


Figure 1. Image A is a T2-weighted axial MRI (time to repeat [TR] = 2400, time to echo [TE] = 80) which demonstrates abnormalities within the left hemisphere including increased signal in the subcortical and deep white matter and effacement of the normal gyral pattern. Abnormal leptomeningeal enhancement and dilation of the lateral ventricle are seen on the T1-weighted (TR = 800, TE = 20) gadolinium-enhanced scan in image B. Image C is a follow-up T2-weighted MRI (TR = 2500, TE = 100) at a similar level to image A which shows slightly more prominent white matter T2 hyperintensity with otherwise similar abnormalities. Image D is a follow-up T1-weighted (TR = 616, TE = 14) gadolinium-enhanced MRI that shows less prominent leptomeningeal enhancement.

In early 1992, he had another neurologic episode, which began in an identical fashion; however, the hemiparesis, aphasia, and headache persisted, prompting transfer to Stanford University Medical Center. On admission, general examination was remarkable only for atrophy of the subcutaneous tissue of his left forehead and nares. On neurologic examination, he was alert and combative with a global aphasia. Meningismus was absent. He had a mild left gaze preference with a right homonymous hemianopsia. Right upper motor neuron facial weakness was present with ipsilateral increased limb tone, unsustained ankle clonus, hemisensory loss, and significant hemiparesis. Reflexes were brisk on the right with a positive Babinski response.

Results of a complete blood count, hepatic and renal function, erythrocyte sedimentation rate, and CSF analysis were normal. Results of a complete hypercoagulable screen were negative. Syphilis, lyme, and HIV serology were negative. Antinuclear antibodies were present at a low titre (1:40). There was diffuse monomorphic delta activity over the left hemisphere on EEG. MRI demonstrated unilateral leptomeningeal enhancement, marked increase of T2 signal in the white matter, loss of the normal cortical gyral pattern, and enlargement of the lateral ventricle of the left hemisphere (figure 1, A and B). Several irregular intracranial vessels were seen on a left carotid angiogram (figure 2, A and C). A left frontal leptomeningeal and brain biopsy revealed minimal mononuclear inflammatory infiltrate and moderate fibrosis of the leptomeninges with mild astrocytosis in the white matter.

The patient was treated with prednisone and cyclophosphamide for possible CNS vasculitis. His neurologic deficits completely resolved over 1 week.

In the next 2 months, the patient had three more transient 1 to 2 hour episodes of right hemiparesis with aphasia followed by headache, prompting readmission to the medical center. Results of bloodwork and CSF examinations were again normal. The MRI was unchanged. Repeat left carotid angiography was normal (see figure 2, B and D). The neurologic diagnosis remained uncertain; however, he was continued on immunosuppressive therapy for 1 year.

In 1997, he was referred to the Stanford Stroke Service for evaluation of continued intermittent right hemiplegia, hemisensory loss, and aphasia followed by headache and one separate generalized seizure. The headache episodes had continued at a frequency of one every 2 to 4 weeks since 1992. He had one isolated unprovoked generalized seizure in the weeks preceding this visit. On neurologic examination, the only abnormal finding was a reduced right arm swing while walking. There was no progression of the hemifacial atrophy. A repeat MRI revealed more substantial white matter disease without other worsening (figure 1, C and D). A diagnosis of PFH complicated by hemiplegic migraine and seizures was made and the patient was started on valproic acid to use one medication for both migraine and seizure prophylaxis. There were no recurrent seizures and the migrainous episodes were substantially improved at follow-up 4 months later.

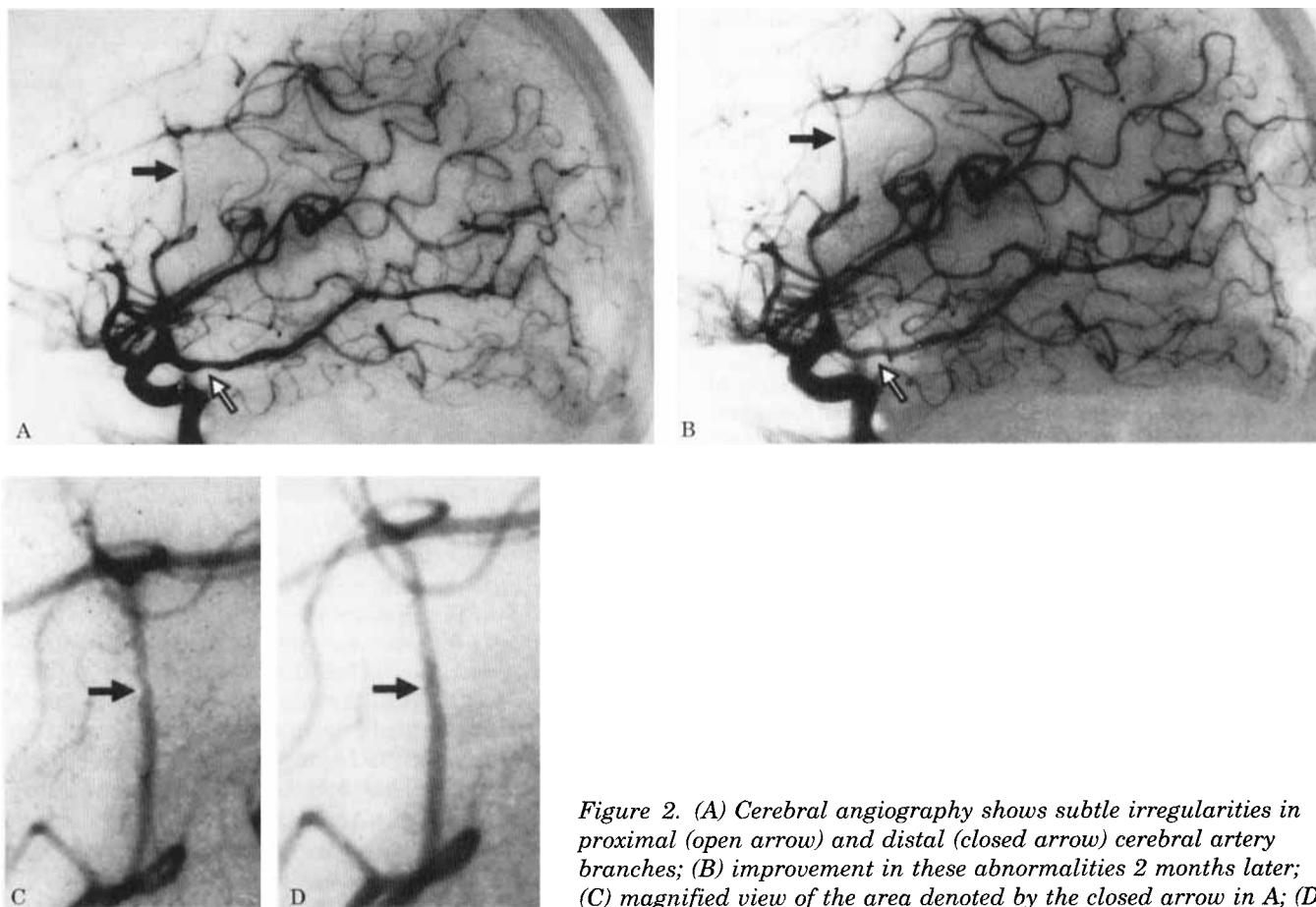


Figure 2. (A) Cerebral angiography shows subtle irregularities in proximal (open arrow) and distal (closed arrow) cerebral artery branches; (B) improvement in these abnormalities 2 months later; (C) magnified view of the area denoted by the closed arrow in A; (D) magnified view of the area denoted by the closed arrow in B.

Discussion. The pathophysiology of PFH is uncertain. Four proposed theories include cortical dysgenesis,¹ sympathetic nervous system dysfunction,⁵ chronic localized meningoencephalitis,^{2,6} and autoimmunity.^{6,7} Fry et al. have suggested that PFH is a regional disorder of the face and underlying neural structures of undetermined pathophysiology.⁸ These hypotheses attempt to explain the presence of concomitant involvement of the ipsilateral face and brain in the context of known clinical, radiologic, experimental, and limited pathologic data. The absence of definitive supportive pathology, the observation of disease onset as late as the 6th decade,² and cases (including our own) documenting new or worsening lesions on serial MRI^{3,5} argues against dysgenesis in all patients. Contradictory experimental evidence⁵ and the frequent lack of clinically apparent autonomic dysfunction in PFH cast doubt on the sympathetic theory's relevance to most patients. Pathologic data have shown evidence of perivascular inflammation within brain tissue, leptomeningeal or cortical blood vessel irregularities with meningeal fibrosis, and lymphocytic infiltration of neurovascular bundles with abnormalities of vascular endothelium in the skin.^{2,6} The development of linear scleroderma following local trauma and the presence of autoantibodies in some patients further supports an inflammatory etiology⁶ but does not account for the focal nature of the disorder. Prior pathology and our biopsy results may not be contradictory but may represent different stages of a self-limiting vascular inflammatory process. Alternatively, the pathologic data may support a heterogeneous pathophysiology (inflammation, dysgenesis, or sympathetic dysfunction) related to abnormalities of intracranial and extracranial vasculature.

Our patient satisfies clinical criteria for migraine with prolonged aura (hemiplegic migraine). Only two prior reports have specifically addressed the association of PFH and hemiplegic migraine.^{3,4} Nevertheless, this association is additionally seen within the case descriptions of numerous other series,^{2,3,5,9} suggesting that the association is more than coincidental. Although controversial, a migrainous aura is likely a primary neuronal event rather than a primary vascular event. Why some patients with PFH, whom we suspect have an abnormality of intracranial vasculature, are predisposed to hemiplegic mi-

graine is uncertain. However, stellate ganglion block has precipitated migraine with aura, which demonstrates the potential for vascular alterations to precipitate an aura.¹⁰

The presence of reversible angiographic changes in our patient lends further credence to a vascular abnormality in PFH. There are many reports of normal angiography in patients with PFH.⁵ Therefore, we conclude with caution that the reversible vessel changes observed in our patient are due to PFH. Alternatively, the findings may represent vasospasm attributable to the patient's migraine.

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