**Case Report**

A 32-year-old woman presented with intermittent headache, progressive left hemihyperesthesia (lower extremity), and right hemifacial paresthesia for 2 years. The patient recalled that the progressive facial asymmetry started 20 years previously. A physical examination revealed skull depression under the thin skin in the left frontal and right parieto-occipital region, bilateral facial hemiatrophy with alopecia, and bilateral enophthalmos, being more prominent on the left side. On a neurologic examination, the patient showed hyperactive deep tendon reflexes on both knee jerks with a left extensor plantar response, along with a peripheral facial palsy on the right side. Laboratory examinations such as blood counts, renal and hepatic function tests, hypercoagulability screening, and autoantibodies revealed normal findings.

Conventional MR images as well as DTIs were obtained by using a 3T MR imaging scanner (Signa HDx; GE Healthcare, Milwaukee, Wis). T2-weighted images and fluid-attenuated inversion recovery (FLAIR) images showed a few tiny hyperintensities in the right frontal lobe periventricular white matter. Bilateral atrophy of the right hemisphere with a few nonspecific white matter hyperintensities. Diffusion tensor imaging and fiber tractography, however, demonstrated clear fiber derangement, especially in the sensory tract of the right cerebral white matter.

**Discussion**

Up to 52% of patients with PRS have associated neuropsychiatric abnormalities such as migraine or facial pain, vision problems, depression and anxiety, and epilepsy. Among the described cases, epilepsy is the most common problem requiring patients to seek medical attention.1-3,6

In our patient, scalp and facial atrophy was bilateral but more prominent on the left frontal scalp, right parieto-occipital scalp, and left orbit. Right hemifacial paresthesia can be explained by right periorbital facial atrophy and subsequent trigeminal nerve involvement. In contrast, brain parenchymal involvement was more prominent on the right side. The right brain involvement explains clearly the left hypoesthesia of the extremities by cross-innervation of the sensory tract. In previous reports, abnormal brain lesions have been shown to be ipsilateral to the side of the face with PRS involvement.2-4,6,8

Bilaterality of PRS has been reported in approximately 2%-7.4% of cases.6 Bilateral involvement of facial atrophy and the almost unilateral involvement of the brain abnormality in our patient suggest that PRS might progress sequentially bilaterally for a long time. Bilateral involvement of facial and scalp atrophy can be visualized best by the use of 3D volume rendering images.

White matter and deep gray matter abnormalities on T2-weighted and FLAIR images were subtle in our patient. However, fiber tractography visualized clearly asymmetric involvement of the fiber tracts and larger involvement of the fiber tracts than expected from the conventional MR images. This finding implies that axonal injury in our patient was so profound that the derangement of the fiber tract could occur. Conversely, previous case reports have demonstrated relatively preserved fiber tracts despite widespread white matter abnormalities.8 Although we could not find any cortical and subcortical abnormalities on high-resolution thin-section T1- and T2-weighted images, a microscopic abnormality within the cortex might induce progressive axonal degeneration.
The pathophysiology of PRS remains uncertain. No genetic association has been found so far. A neuroinflammatory process has been suggested as a possible pathogenic mechanism, though no associated serum autoantibodies have been found. The disproportionately large involvement of the fiber tracts as compared with the white matter lesion on conventional MR images in our patient suggests a widespread underlying neurodegenerative process.

In conclusion, DTI and fiber tractography are useful in evaluating the neurologic aspects of patients with PRS with the slightest involvement of the brain.