

Progressive Facial and Cerebral Hemiatrophy: Parry-Romberg Syndrome or Scleroderma “en Coup de Sabre” ?

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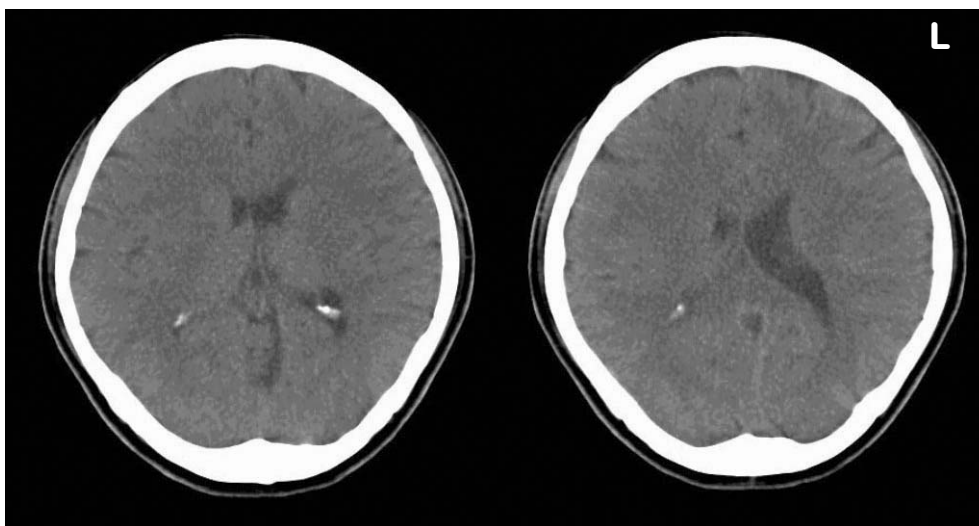


Figure. Brain CT approximately 11 years after disease onset showed moderate enlargement of the frontal horn and lateral ventricle over left hemisphere. Cortical atrophy was more apparent also over left side.

LL was a 28-year-old woman with 2 years' history of progressive skin discoloration and atrophy over left forehead. Medical and family histories were unremarkable. Examination revealed a band-like area of hyperpigmentation and atrophy over left medial forehead. Facial movement and sensation were normal. Initial impression was progressive facial hemiatrophy (Parry-Romberg syndrome, Romberg disease). Brain MRI revealed mild ventricular enlargement, especially the frontal horn and the lateral ventricle.

However, the patient was lost to follow-up for 10

years. She came back due to progression of the skin lesion which was enlarged and extended to the scalp and the nose. Laboratory tests including ANA, RA and immunoelectrophoresis were normal. Blink reflex was normal. Skin biopsy showed mild hyperkeratosis, slightly elongated rete ridges with pigmented basal cells, mild melanin incontinence, and compact sclerosis of the dermis with bound down sweat glands. Pathological diagnosis was facial hemiatrophy, but scleroderma “en coup de sabre” could not be excluded. Brain CT showed further progression of left hemisphere atrophy (Fig.).

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A clear distinction between Parry-Romberg syndrome and scleroderma “en coup de sabre” is often not possible⁽¹⁾. Both conditions are characterized by unilateral atrophy and furrow of the skin over frontoparietal area of the head and progression of the skin lesion eventually leading to facial hemiatrophy^(1,2). Although most patients are asymptomatic apart from cosmetic or functional problems, CNS involvement may occur in both disorders⁽¹⁻⁵⁾. In Parry-Romberg syndrome, the most frequent CNS symptom is focal seizure and the most frequent radiological abnormality is homolateral cerebral hemiatrophy^(2,3). Much less frequent abnormality is cortical calcification. In scleroderma “en coup de sabre”, CNS involvement may present with partial epileptic seizure or other focal neurological symptoms⁽⁴⁾. Neuroradiological abnormalities include homolateral cerebral atrophy, white matter lesions, parenchymal calcification and meningeocortical changes^(1,5).

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