Facial Myokymias In Parry-Romberg Syndrome

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Figure 1. Images showing hemifacial and hemitongue atrophy with signs of tongue bites (A) and unilateral alopecia (B). The photograph has been published with the consent of the patient.

Case Summary

A 65-year-old man came to our hospital for an outpatient neurological assessment. He had a 20-year history of hemifacial atrophy associated with hemitongue atrophy (Figure 1A), intermittent unilateral jaw spasms leading to frequent tongue bites, unilateral alopecia (Figure 1B), parotid and submandibular gland atrophy, and tooth loss. Despite the extreme discomfort experienced by the patient, he had never received a full evaluation and diagnosis. On neurological assessment, myokymias of the jaw-closing muscles (masseter and temporalis) were detected (video) and confirmed by the presence of myokymic discharges during an electromyographic study. No other neurological signs were recognized. Laboratory and neuroimaging investigations were normal. A diagnosis of Parry-Romberg syndrome was made. This is a rare neurological disorder, mostly sporadic, and characterized by hemifacial atrophy involving the skin, the fat, the connective tissue, and sometimes the bone.^{1,2} Skin hyperpigmentation or depigmentation, alopecia, enophthalmos, and deviation of the mouth and nose toward the affected side may also be observed.^{1,2} In our patient, the symptoms were partially improved by carbamazepine 800 mg. However, hemifacial atrophy is irreversible, and no treatments

able to stop the progression of the disease are available to date. The final degree of hemifacial atrophy is variable and mostly depends on the duration of the disease. Aesthetic and functional disturbances are extremely disabling for the affected patients. A careful assessment in the hospital setting is mandatory in order to properly identify the syndrome, to plan a tailored management approach for the patients, to follow the progression of the disease and its responsiveness to therapy over time. Finally, a multidisciplinary in-hospital care may contribute to improve the quality of life of patients with Parry-Romberg syndrome and their ability to cope with chronic pain.

Evidence of myokymias of the jaw-closing muscles on clinical assessment

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Authors' Note

The photograph and the video have been published with the consent of the patient.

Declaration of Conflicting Interests

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