Viral encephalitis in Parry-Romberg syndrome

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DESCRIPTION

A 28-year-old woman was admitted to our clinic for acute onset of headache, aphasia and confusion, followed by a first episode of generalised tonic-clonic seizures.

She had a history of left-sided migraine, accompanied by visual aura and right arm dysesthesia since the age of 20 years and depression. From the age of 15 years, she had experienced slow progressive atrophy of the left face's soft tissues, which was treated with fillers (figure 1). No further progression of haemifacial atrophy (HA) was noticed after the age of 25 years.

CT showed a widespread asymmetrical (left>right) white matter (WM) hypodensity. An MRI showed diffuse subcortical WM hyperintensity on T2 sequences in both cerebral hemispheres, mainly on the left side, left frontal developmental venous anomaly and left cerebellar atrophy (figure 2). Diffusion-weighted imaging excluded an acute ischaemia, and MRI spectroscopy was normal. Electroencephalography showed asymmetric amplitude of activity (higher on the right leads) and left frontotemporal Z-waves with the tendency to spread to the contralateral hemisphere. Cerebrospinal fluid (CSF) analysis detected moderate lymphocytic leukocytosis (59/mm³), a slightly increased protein level (55 mg/dL), normal glucose. Blood, CSF cultures and PCR for herpes simplex virus (HSV 1–2) were negative. Anti-HIV1-2 antibodies were absent. However, the patient was treated with acyclovir for 14 days, obtaining a complete clinical response with normalisation of CSF, thus allowing a diagnosis of viral encephalitis (VE). On discharge, the patient was almost asymptomatic, persisting only slurred speech and paraesthesia of the lips on the right side. One and 24 months MRIs were unchanged. The patient started levetiracetam due

Figure 2 Neuroimaging data. In figure parts A and B, MRI of the brain performed during viral encephalitis. Hyperintense signal on FLAIR sequences of the frontoparietal-temporal regions in the left hemisphere. Left cerebellar atrophy is shown in T1 (C) and T2 (D) sequences. FLAIR, fluid-attenuated inversion recovery.

to recurrence of tonic-clonic seizures, and she is now seizure free since 2 years.

Our patient's clinical and MRI features were consistent with Parry-Romberg syndrome (PRS) complicated by VE.

PRS is a rare neurocutaneous disorder characterised by progressive and self-limited HA.

Although PRS and en coup de sabre may coexist, in our patient, the distribution of the lesion on the trigeminal nerve and the absence of the typical linear, fibrous plaque on the frontoparietal scalp and forehead have directed the diagnosis towards PRS. Neurological manifestations occur in up to 15% of cases, including epilepsy (60.5% of cases), migraine and brain lesions ipsilateral to the HA.¹²

PRS pathophysiology is unknown, although recent papers suggest an autoimmune aetiology probably caused by viral infection.

Whether our patient's epilepsy should be considered as secondary to the VE or part of the PRS clinical spectrum still remains unclear. In PRS, epilepsy's age of onset is early, with focal and drug-resistant crises.² In our patient, the temporal relationship with VE, the presence of generalised crises and the drug sensitivity lead us to think that epilepsy is secondary to VE.

Nowadays, few cases of PRS in association with autoimmune encephalitis were described³ but, to the

Figure 1 Haemifacial atrophy in Parry-Romberg syndrome. In figure parts A and B, the patient is portrayed at the age of 19 years, after 4 years since she had experienced slow progressive atrophy of the left face's soft tissues; in figure part C 2 years later, after stabilisation of haemifacial atrophy and treatment with fillers.

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best of our knowledge, no case associated with VE was reported. This first case highlights how the onset of seizures in PRS not necessary is spectrum of disease, but a possible sign of an infectious process, which should be promptly diagnosed and treated.

Learning points

- ▶ Parry-Romberg syndrome is a rare neurocutaneous disorder characterised by progressive and self-limited haemifacial atrophy.
- ▶ Neurological manifestations occur in up to 15% of cases, including epilepsy, migraine and unilateral brain lesions, on the same side of the haemifacial atrophy.
- ▶ Seizures in Parry-Romberg syndrome are not necessary a manifestation of the disease but can be a possible sign of an infectious process, which should be promptly diagnosed and treated.

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