Viral encephalitis in Parry Romberg Syndrome
a case report

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Case report

A 30 year-old woman, affected by epilepsy by the age of 28, came to our outpatient clinic due recurrence of tonic-clonic seizures.

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

At the age of 28 the patient was admitted to infectious diseases department due to acute onset of headache, aphasia, confusional state, followed by a first episode of generalized tonic-clonic seizures. Computed tomography showed a widespread asymmetrical (left>right) white matter (WM) hypodensity. Electroencephalography showed asymmetric amplitude of activity and left fronto-temporal Z-waves with the tendency to spread to the contralateral hemisphere.

A Cerebrospinal fluid (CSF) analysis detected moderate lymphocytic leukocytosis (59 µL), a slightly increased protein level (55 mg/dL), normal glucose concentration. Blood and CSF cultures showed no bacterial growth. Anti HIV1-2 antibodies were absent. Increased of serum γ IgG were observed.

A first Magnetic Resonance Imaging (MRI) showed diffuse subcortical WM T2 hyperintensity in both cerebral hemispheres, mainly in the left side, and ipsilateral cerebellar atrophy (fig 2).

Diffusion weighted imaging excluded an acute ischemia. MRI spectroscopy was normal. A viral encephalitis (VE) was diagnosed and treated with acyclovir for 14 days. She improved by fifth day. A second lumbar puncture showed normalization of the CSF parameters. On discharge, neurological examination revealed minimally slow speech and paresthesia to the right side of the lips, spontaneously resolved within the next 10 days.

MRIs, performed at months 1 and 24 after discharge, were unchanged (fig 3).

Our patient's clinical and MRI features[1]were consistent with Parry Romberg Syndrome (PRS) who developed a VE.

Fig.1 Progressive atrophy of the left face's soft tissues at 18 and 20 years old.

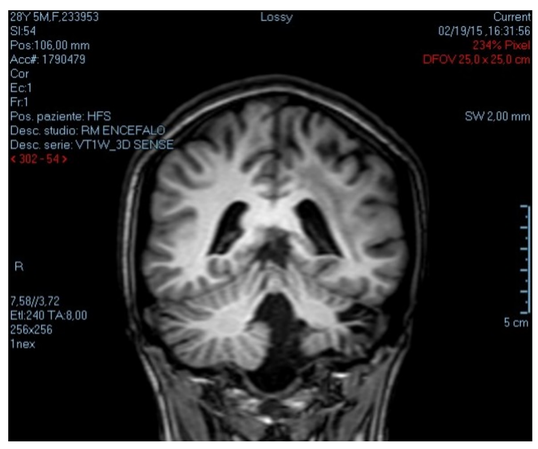

Fig.2 Ipsilateral cerebellar atrophy

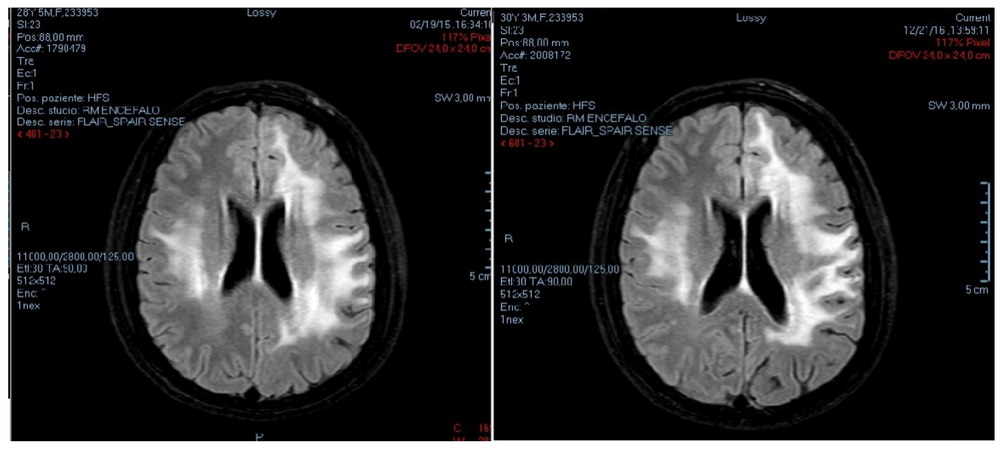

Fig.3 MRIs, performed at months 1 and 24 after discharge, were unchanged.

Discussion and conclusions

PRS is a rare neurocutaneous disorder characterized by progressive and self-limited HA. Neurological manifestations occur in up to 15% of cases, including epilepsy (60.5% of them), migraine and unilateral brain lesions, on the same side of the HA[2,1].
PRS pathophysiology is unknown although recent papers suggest an autoimmune disorder probably caused by viruses[3]. Whether our patient's epilepsy should be considered as secondary to the VE or part of the PRS clinical picture still remains unclear.
Our interpretation is that VE may have triggered the progression of neurological complications in PRS.

References

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