

A rare case of PAF comorbid with epilepsy: differential diagnosis and treatment of a patient in charge at outpatient home care clinic

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Abstract

Objective: A home care patient who was previously on outpatient clinic care with a suspicion of Parkinson's disease was taken in charge. The clinical history reported a poor response to dopaminergic therapy that had been interrupted at the initiative of the patient and his family. Therefore, the initial aim was a better diagnostic definition of a possible extrapyramidal disease and the development of a more effective treatment.

Background: during the first home visits, some signs suggesting a parkinsonism such as reduced facial expression and a certain motor slowdown with a slight axial stiffness were found, while along the way an orthostatic hypotension was added to clinical observation. A first new attempt with dopaminergic therapy had given little satisfaction, but in the meantime the presence of reported episodes of a 'lipothymic' type emerged. In order to verify the nature of these episodes, the patient performed tests that showed signs compatible with an epilepsy. **Methods:** during the observation period the patient underwent to the pressure measurement in ortho- clino- statism; the patient was subjected to electroencephalogram, electromyography, blood tests (B12 dosage) brain MR scan and DAT-Scan.

Results: from the tests performed, the patient resulted affected by epilepsy, a drastic drop in pressure when standing, signs of slight peripheral polyneuropathy, positivity of the DAT-Scan due to a reduction in the uptake of the presynaptic tracer. Therefore the patient was subjected to anti-epileptic therapy which resulted in a frequency reduction of episodes of consciousness loss, dopaminergic therapy at low doses, application of elastic stockings and hydration. To date, the patient has undergone a worsening evolution of the clinical picture relating to orthostatic hypotension.

Case Report

A male patient, aged 78, was seen for the first time in July 2020 for walking impairment and frequent falls, some of which with loss of awareness. In the past he was diagnosed with Parkinson's disease in other clinical centers and treated with dopaminergic therapy which was subsequently discontinued due to poor efficacy. At the time of our examination, the patient showed: slight hypomimia, axial stiffness, unstable walking, poor pendular movement at upper limbs, slight plastic hypertonus, no cognitive impairment. A low-dose and softly-increasing of dopaminergic therapy was tried again, according to the response of DAT-scan result, which showed pre-synaptic dopaminergic impairment of basal ganglia, but was interrupted due to poor efficacy and early presence of side effects (hallucinations). Subsequently, due to the increase in the frequency of falls, some of which associated with loss of consciousness or slight confusion, the patient was subjected during the home examination to blood pressure measurement in the sitting position and in orthostatism, which highlighted a severe fall of the values (from 140\80 mmHg to 80\n.r. mm Hg in 10 seconds with compensatory tachycardia) of blood pressure and which clinically corresponded to intense paleness and compromise to maintain the standing position. However, some of the reported episodes were characterized by elements of suspected epileptic nature consisting of tonic-clonic shaking, screaming and urine loss. This data were matched with the brain MR scan carried out which showed signs of temporal-occipital atrophy and diffuse vascular brain suffering. Consequently we suspected the coexistence of epilepsy. Therefore, an

electroencephalogram was performed which documents slow frontal-temporal dysrhythmias, by recording in the intercritical time, with clinostatic position. As a consequence the patient underwent therapy with anti-crisis drugs with an improvement in the frequency of the episodes described in the medical history by the family members, i.e. a reduction in the frequency of falls with loss of awareness preceded or not by confusional state. Moreover a control recording two months subsequent to anti-epileptic drugs medication showed an improvement of organization of the EEG.

Discussion

Pure autonomic failure (PAF) is a well known disorder of α -synuclein deposition that results in neurogenic orthostatic hypotension and other features of autonomic failure, without signs of central nervous system involvement. The interest of this clinical case consists in the difficulty of discriminating the nature of the critical episodes in a patient suffering from this rare disease, which had not previously been diagnosed despite the relevant anamnestic data of frequent and severe falls, some of which

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with significant trauma. In fact it is known in clinical practice that in Parkinson's disease, but especially in some atypical parkinsonisms, falls are a frequent and relevant symptom; however, the patient studied presented other symptoms, such as intense paleness and mental confusion when standing, which have not previously been sufficiently addressed. It is interesting to note that, although the DATscan which was performed during the diagnostic procedure had a suggestive outcome for presynaptic dopaminergic damage, the dopaminergic therapy was not effective and gave severe side effects even at low doses such as to make it impossible to use. The second element of interest consists in the difficulty of identifying the coexistence of epilepsy. In fact, even the loss of consciousness from hypotensive shock can be associated, albeit quite rarely, to an epileptic seizure due to acute cerebral hypoxia. However, in this case, the high frequency of such episodes, associated with real seizures of Grand Mal type, or focal seizures with bilateral evolution, improved with the antiepileptic treatment, which at the same time allowed to discriminate the epileptic seizures from 'simple' hypotensive seizures or lipothymias. The nature of the epilepsy in this patient is uncertain: in the absence of a familiarity, a secondary nature due to the atrophy or chronic hypoxic brain damage documented by the brain RM-scan remains possible.

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