Foix-Chavany-Marie syndrome secondary to a bilateral stroke in a patient with lupus

Síndrome de Foix-Chavany-Marie secundario a ictus bilateral en un paciente con lupu

Dear Editor:

The Foix-Chavany-Marie (FCM) syndrome, also known as opercular syndrome, was described by those authors in 1926[1]; it is characterized by a disorder of voluntary control of muscles of the face, tongue, pharynx and masticatory muscles, caused by a bilateral lesion of the opercular cortex. Patients who suffer this syndrome present severe anarthria or dysarthria, dysphagia, drooling, and facial and lingual diplegia; they are unable to open their mouth, blink an eye or protrude the tongue at will, although they retain the reflex function and can cry, smile and yawn automatically.

The most common cause in adults is an ischemic condition of both opercula. There have been two types of opercular syndrome described[2]: *a*) anterior, from damage to the front of the operculum with predominant motor disorder, anarthria and sometimes associated with aphasia, and *b*) posterior, from damage to the parietal opercular area, with a sensitive clinical picture.

Patients with systemic lupus erythematosus have an increased risk of suffering cerebrovascular events of all subtypes when compared with people of equal age and gender[3]. Patients with lupus are more likely to develop cerebrovascular events from various causes: Libman-Sacks endocarditis, vasculitis, prothrombotic state with antiphospholipid and anti-cardiolipin antibodies, although the risk factors most involved in this neurological complication appear to be hypertension, hyperlipidemia and the activity of the disease[4].

We present a case of this rare syndrome which, to our knowledge, is the first case reported in a patient with systemic lupus erythematosus.

A 33-year-old woman with a history of lupus nephritis diagnosed at age 8, with good kidney function, under treatment with prednisone at doses of 10 mg on alternate days. She had had a right carotid ischemic stroke at age 10 that left no physical sequelae; since then, she had been taking aspirin at doses of 100 mg/day. She offered no reports on this neurological complication. She had had complex partial seizures since age 5, which were treated with oxcarbazepine at doses of 450 mg/day. She showed carbohydrate intolerance (probably related to the steroid treatment), but had no arterial hypertension, hyperlipidemia or smoking habit. Her sister was also diagnosed with systemic lupus erythematosus and had had an ischemic stroke for which she was taking acenocoumarol.

She was sent to the emergency service of our hospital with symptoms of acute onset of inability to broadcast speech and weakness in the right limbs. The general examination showed no relevant changes. The neurological examination revealed anarthria, central facial diplegia, dysphagia, and paralysis of tongue and jaw movements. She could, however, yawn and laugh. She had adequate understanding of spoken and written language and could communicate by writing. No alteration of the level of consciousness or other cranial nerves was observed. Motor: strength 4+/5 at the distal level of the right limbs: symmetric and conserved myotatic reflexes; plantar cutaneous flexor reflexes; superficial and deep sensitivity without significant alterations; no dysmetria or dysdiadochokinesia.

Laboratory tests: systematic blood analysis, coagulation study, glucose, renal and liver profiles, thyroid hormones, rheumatoid factor, cholesterol, triglycerides and total protein test without relevant changes. Normal vitamin B12 and folic acid; ANA positive on titration 1/80 with a homogeneous pattern; anti-DNA, 18 U/ml (normal < 15); anti-SSA, SSB, Sm and U1 snRNP, within normal limits; complement C3, 61; C4, 11 (both low).

Hypercoagulability workup: coagulation factors, antithrombin III, protein C resistance, prothrombin gene 20210, protein C and homocysteine with no alterations. Free protein S antigen at 45%(normal > 50). Study on lupus anticoagulant study with IgG anticardiolipin antibodies positive, 52.7 (normal < 15); Exner test, 1.6 (normal, 0.8-1.2); the remaining tests showed no alterations.

Doppler of supra-aortic trunks: with no haemodynamic changes. Echocardiogram: mild mitral insufficiency; the rest was normal.

Cranial computed tomography (CT): chronic infarction in the territory of the right middle cerebral artery.

Brain MRI (Fig. 1): left Rolandic subacute infarction. Chronic ischemic injury, with underlying malacia and gliosis in the parietal-occipital junction of the right hemisphere. Cerebral angio-MRI: with no vasculitis injuries.

After five days in the hospital, the patient improved from the neurological focus with disappearance of paresis of the right limbs, as well as of dysphagia, although bilateral facio-linguo-masticatory paralysis persisted. Anticoagulant therapy was started.

Three clinical forms of bilateral paralysis have been described in the muscles innervated by cranial nerves V, VII, IX, X, XI and XII, known as bulbar, striatal and cortical[2]. Bulbar palsy is characterized by dysphonia, dysphagia, dysarthria, decrease of linguo-masticatory movement, decreased masseter reflex and atrophy with fasciculations of the affected muscles. Both voluntary and automatic movements are altered. In the striatal form, in addition to the symptoms described, the masseter reflex is usually increased and pathological reflexes such as the palmomental reflex appear, while there may be subcortical cognitive impairment and emotional lability. As occurs with extrapyramidal diseases (such as Parkinson's disease where there is facial amimia), automatic movements or those with an emotional component are reduced.

We turn our attention to paralysis by cortical affectation, known as opercular syndrome or FCM. In this case, automatic-voluntary dissociation is added to the typical symptoms of bilateral paralysis of cranial nerves. These patients are unable to open or close their mouth, protrude the tongue or close their eyes on command; however, they can still smile, cry, yawn or yell in an automatic manner.

The automatic-voluntary dissociation that characterises this syndrome is explained by the different brain location of the circuits responsible for voluntary movements and

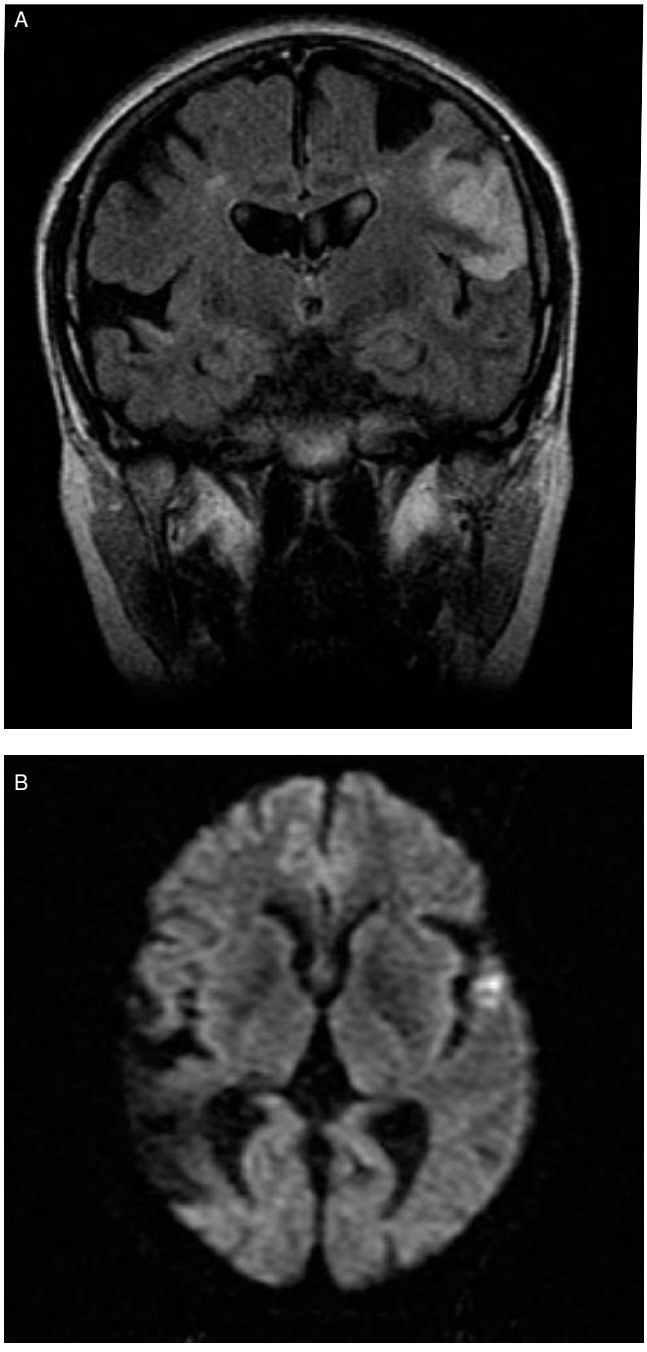

Figure 1 Brain MRI. A: coronal cut in FLAIR sequence showing signal hyperintensity at the level of the left frontal operculum. B: axial cut in diffusion sequence showing hypointensity at the right parietooccipital level that affects the right operculum and hyperintensity at the level of the left operculum indicative of right chronic ischemic lesion and left acute ischemic, respectively.

reflexes. While voluntary movements are originated in the motor cortex at the level of the primary motor and premotor area and then travel through the corticospinal pathway, automatic reflex movements follow an extrapyramidal circuit through the basal ganglia and the diencephalon. There are some reported cases of reverse opercular syndrome where the reflex movements are impaired while the voluntary movements are preserved by metastasis of both supplementary motor areas[5]. The inputs to the motor nuclei of cranial nerves V, lower VII, IX, X, XI and XII are usually bilateral. After an opercular injury, the contralateral motor cortex is capable of fulfilling the function of the damaged cortex. However, when both opercula are affected, the clinical manifestations appear. Opercular syndrome is occasionally produced by bilateral affectation of subcortical structures and even by unilateral cortical condition[6]. The reason for this is unclear, although it is believed that in the latter there would be no bilateral motor representation, being represented in a single operculum.

The typical clinical presentation of this syndrome allows its identification and the location of the lesion and its aetiology. Nevertheless, more infrequent manifestations have been described, such as the appearance of dystonia of both hands[7], which then gave rise to a hemimasticatory spasm[8]. Early recognition of this syndrome and its ischemic nature enable specific therapeutic measures, such as fibrinolysis[9], to be performed; however, our patient arrived at the hospital past this time. In most cases, cranial CT does not visualise acute ischemic lesions, as in our case; therefore, brain MRI with diffusion sequences can objectify bilateral lesions and identify which ones are acute and which are chronic[10].

The prevalence of cerebrovascular events in patients with systemic lupus erythematosus varies between series from 6.5 to 19%[3,4,11]. They normally occur within the first 5 years of diagnosis; the most common risk factor is arterial hypertension[4], anticardiolipin antibodies appear in 43%, lupus anticoagulant in 38%and autopsy findings show that 2 out of 3 patients had Libman-Sacks endocarditis[11]. Despite this, we found no data of vasculitis in our patient. In these patients, treatment with oral anticoagulants has proven effective in reducing new cerebrovascular[12] events, although we must not forget aggressive treatment of vascular risk factors[4].

In our patient, given that she had presented a new stroke while in treatment with aspirin and due to the suspicion of a hypercoagulability state later confirmed with the appearance of anticardiolipin antibodies, an anticoagulant treatment was chosen.

The presence of a sudden onset of anarthria and of paralysis of the facial and linguo-masticatory musculature should make us think about this possibility. The voluntary-automatic dissociation should not be confused with hysteria or simulation situations, enabling early diagnosis and initiation of a specific treatment for each case.

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Frontotemporal dementia and motor neuron disease

Demencia frontotemporal y enfermedad de motoneurona

Dear Editor:

Degenerative diseases are the most common cause of dementia in our environment and, within these cases, 5-10% are frontotemporal dementias (FTD)[1,2]. This percentage increases when patients are younger than 65 years old, being the second leading cause of pre-senile dementia[3]. The major neurochemical alterations are deficits in serotonin and dopamine[2]. According to Diagnostic and Statistical Manual of Mental Disorders (4th ed.) (DSM-IV) criteria, the diagnosis of FTD is mainly clinical, consisting of behavioural alterations, psychotic disorders and language disorders, among others, aided by neuroimaging and neuropsychological tests[1].

An association with other neurodegenerative diseases has been noted, including motor neuron disease (MND), a scarcely frequent[3] association, which is clinically characterised by frontal and neurological signs[4]. FTD associated with MND is classified according to whether the condition is the first, second or both motor-neurons, and amyotrophic lateral sclerosis (ALS) is the most frequent form[4].

We present the case of a patient with FTD, with an evolution of 1 year, who developed MND.

A 59-year-old woman, right handed, with no family history of neurodegenerative disease or personal history of psychiatric illness. Only a mild dyslipidemia can be highlighted. The patient had suffered from alterations in personality, talkativeness, daytime sleepiness, easy laughter, great difficult in crying and psychotic disorders for 1 year: "the paintings in her house talked to her, and an impairment of speech had appeared in the last month".

Physical examination, with cardiopulmonary auscultation, was normal. Neurological examination showed that the patient was oriented in all three areas; she presented mild dysarthria, normal cranial nerves, except for a slight left hypoglossal paresis with some fasciculation in that territory; without myoclonus, ataxia and dysmetries; the strength, tone, sensation and reflexes were normal in the limbs and there were no fasciculations. Gait was also normal. In the neuropsychological study, the results were: MMSE, 29/30; clock test, 9/10; verbal fluency, 12; Trail Making A, 48 s; FAB, 13. Additional tests performed (biochemical, haemogram, serology (including HIV), thyroid hormones, vitamin B12 and folic acid) were normal.

Cranial computed tomography (CT): temporal atrophy. Cerebral and cervical magnetic resonance imaging (MRI): temporal atrophy and small meningioma of the right frontal sulcus; the rest was normal. Conduction studies of motor and sensory nerves were normal and no blockages were observed. The electromyographic study of muscles dependent on the cervical, lumbosacral and bulbar regions revealed a diffuse and asymmetric neurogenic pattern, with greater expression in the left side, with mild to moderate acute denervation activity. Thus, the patient met the DSM IV diagnostic criteria for frontotemporal dementia, associated in this case with motor neuron disease.

The great importance of the social impact of FTD, as of MND, its pre-senile onset, higher frequency in cases of family history (up to 50% in FTD), the burden on caretakers and the possibility of carrying out genetic advice and study[3] (up to 20% of patients with an association of these two diseases may have a history of ALS and up to 40% of FTD[5]), makes us give more importance to this association since, although rare, in recent years it is being noted that it is becoming more important, perhaps because it was previously underdiagnosed.