

Slowly progressive voluntary-automatic dissociation of facial movements (Foix-Chavany-Marie syndrome)

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Abstract

We describe a case of slowly developing Foix-Chavany-Marie syndrome associated with distinctive bilateral cortical atrophy of frontal lobes in a 59-year old woman. The neurophysiological mechanism and differential diagnosis concerning the unusually slow progression are discussed.

Abbreviations

EMG - electromyography
FCM - Foix-Chavany-Marie
MRI - magnetic resonance imaging

INTRODUCTION

The rare syndrome, characterized by the loss of voluntary function of lower face muscles (anarthria, amimia, dysphonia and dysphagia) and preserved automatic movements such as yawning or coughing, was first described by Magnus in 1837 [9]. The syndrome has obtained its eponyme Foix-Chavany-Marie (FCM) according to the authors, who associated it with bilateral lesions in the operculum of the inferior frontal gyrus [6]. Most of the cases reported so far have been related to acute stroke or to encephalitis. Here we present a patient showing the signs of FCM with unusually slow progression associated to bilateral frontal lobe cortical atrophy.

CASE REPORT

The patient was a 59-year-old right-handed woman (born in 1944). She had a history of 2nd stage syphilis diagnosed in 1978, with no persisting signs of the disease after a treatment with high-dose penicillin. Further, she suffered from non-insulin dependent diabetes mellitus since 1984 and from chronic obstructive pulmonary disease with frequent acute exacerbations since 1994. Since 1995, she was treated for arterial hypertension and moderate chronic dilated cardiomyopathy associating chronic heart ischemic disease.

In 1995, she perceived first difficulties with articulation of speech. The dysarthria was progress-

ing and her voice has changed. In 1999, a neurologist described dysarthria and areflexia on lower limbs. MRI scans showed marked brain atrophy affecting predominantly the frontal lobes and intact brain stem (Figure 2a). Progressively, speech became unintelligible and swallowing difficulty appeared. A repeated MRI in 02/2001 showed white matter hyperintensities on T2-weighted images in addition to the known atrophy. Doppler ultrasonography of carotid and vertebral arteries revealed only mild atherosclerotic changes. Neuropsychological examination confirmed depression but indicated no cognitive deterioration. Anti-depressive therapy with

fluoxetine 20mg/day had been started since 02/2002. On control visit in 09/2002 the patient's speech was entirely incomprehensible and she could only communicate by writing.

In 12/2002 the patient was first admitted to our department because of sudden right-sided hemiparesis and hemiparesis. Her dysphagia and dysarthria remained unchanged. CT scan showed an ischemic lesion in the left thalamus. The right-sided hemiparesis resolved completely within 5 days but residual hemiparesis persisted. In 02/2003, complete aphonia was noted and no active movements of the vocal chords were observed on laryngoscopic examination. EMG of the laryngeal muscles did not reveal any neurogenic lesion or neuromuscular transmission disorder.

Last hospitalization at neurology occurred in 12/2004. The patient was fully cooperative, she could not speak, but she was able to communicate by writing and understood perfectly the spoken language. On neurological examination she presented anarthria, aphonia and dysphagia. When asked, she was able to close her eyes only after latency and the closure was weak (Figure 1). She was unable to frown, to show her teeth or purse her lips and she could not raise the corners of her mouth or execute any voluntary movement of lower face muscles. When she was asked to show her tongue, she only slightly opened her mouth and did not move the tongue at all. On the other hand, her blink reflex was intact, she smiled on emotional stimuli, grimaced when eating lemon, moved her tongue and chewed a bit in the mouth. The masseter reflex was normal, gag reflex was decreased, oral automatism reflexes were not present. There was no impairment of eye movements or other cranial nerves on examination. Deep tendon reflexes were decreased on the upper limbs and absent on the lower limbs. There was a symmetric distal tactile hypesthesia and pallhypesthesia in the legs. On neuropsychological testing, the patient's cognition appeared intact (excluding spoken language). However, there were more grammar mistakes in written speech; it was slower, less fluent and less well-organized than in 02/2002. The serological pattern of specific anti-TP IgM and VDRL negativity together with TPHA and specific anti-TP IgG antibodies positivity corroborated the history of syphilis without any recent disease activation. The cerebrospinal fluid was normal including the absence of VDRL, TPHA and TP antibodies. MRI showed marked cortical atrophy of the frontal lobes predominating on the right and distinct enlargement of both Sylvian fissures (Figure 2b). Compared to the examination in 1999, there was an obvious progression of the atrophy at the precentral and inferior frontal gyri (Figure 2c and 2d).

At the end of 12/2004 the patient was admitted at pneumology for the exacerbation of her pulmonary disease. In 01/2005, a heart failure occurred. She was resuscitated, but a persistent vegetative state developed as a consequence of a supposed extensive hypoxic brain damage. The patient died after 2 months on intensive care. The autopsy was not performed.



Figure 1.

- 1: no instruction, resting face
- 2: voluntary movements
 - 2a) the patient is able to close her eyes after an instruction,
 - 2b) unable to show her teeth – opens her mouth only,
 - 2c) unable to move her tongue – opens her mouth only
- 3: automatic movements
 - 3a) taste stimulation (lemon) – distinctly grimacing,
 - 3b) accidental tactile stimulation (a hair in the mouth) – obvious tongue movements

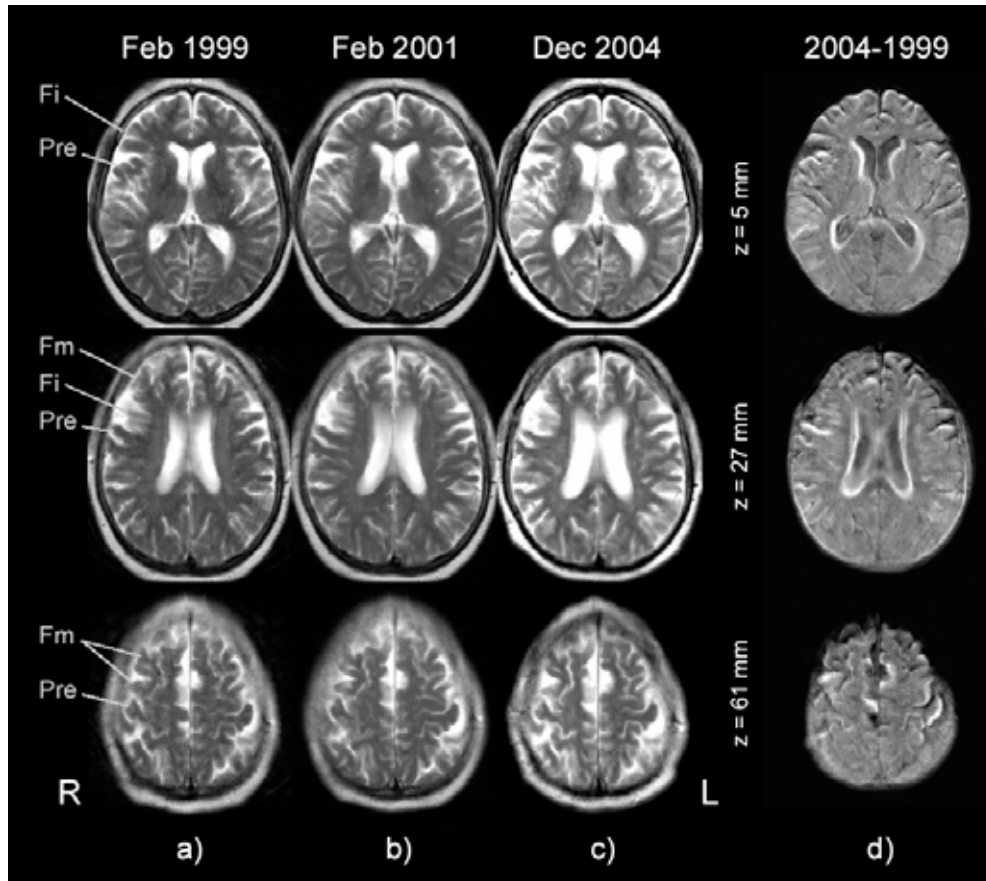


Figure 2. Columns a), b), and c) correspond to three T2-weighted MRI examinations in 1999, 2001 and 2004. The column d) shows a subtraction of MRI 2004 minus 1999. Rows represent three representative sections normalized in the standardized stereotactic space (Montreal Neurological Institute). A marked decrease of white and gray matter volume within the 5 year period is obvious. The difference is highlighted in white in the column d). The atrophy of gray matter affected almost exclusively the frontal lobes, predominantly the precentral gyrus (Pre) and inferior frontal gyrus (Fi), less middle frontal gyrus (Fm), superior and medial. White matter atrophy lead to widening of the lateral ventricles. Parietal, temporal and occipital lobes were minimally affected.

DISCUSSION

We present a patient with nearly 10 years history of gradually developing dysarthria, dysphonia and dysphagia progressing to complete loss of voluntary motility of face and mouth with preserved automatic movements. This clinical pattern of dissociation between the voluntary and automatic function of the lower cranial nerves associated with bilateral lesions of anterior frontal operculum on MRI is characteristic for FCM [2,14].

The clinical syndrome of slowly progressive dysarthria, dysphonia, dysphagia and facial palsy may cause diagnostic problems, particularly in the initial stages. The face expression can remind of parkinsonian hypomimia, however it differs by the character of dysarthria, by the presence of automatic-voluntary dissociation and by the absence of other symptoms of parkinsonism. It is more difficult to distinguish between FCM and the pseudobulbar syndrome caused by bilateral frontal lobe

white matter lesions. The pseudobulbar syndrome can similarly present by disturbances at articulation, phonation and swallowing but in contrast to FCM, there are usually signs of pyramidal tract lesions and frontal deliberation signs including emotional incontinence. Both automatic and voluntary face movement disturbances are common. Facio-linguo-masticatory apraxia is another similar syndrome that was even considered to be an equivalent of FCM [5,12]. However, this form of apraxia is based on errors in timing, sequence and space orientation of elementary movements that still can be executed separately. In contrast, FCM is characterized by complete paralysis for voluntary movements but preserved automatic functions [2]. Important features that could help to differentiate between FCM and other frontal cortical atrophies (e.g. fronto-temporal dementia, Pick's disease) are intact cognitive functions and absence of aphasia up to the late stages of the disease [3]. In our patient, intact cognition was confirmed on repeated neuropsychologi-

cal examinations. Finally, the diagnosis of myasthenia gravis was considered in the patient and diabetic cranial neuropathy or Miller-Fischer syndrome could be taken into account in the differential diagnosis as well. However, all of these are pure peripheral disorders that do not include the automatic-voluntary dissociation.

FCM is a clinical syndrome with heterogeneous etiology. Published reports showed vascular ischemic lesions in most of the cases. Usually, the syndrome developed fast and did not progress in time [6]. So far, only a few cases of slowly progressive FCM were described [3,8,11], among these were four of unclear etiology [3] and one in chronic herpetic encephalitis [11]. In our patient, there were some vascular risk factors and the history of at least one ischemic stroke. However, we documented a seven year period of slow deterioration of dysarthria, dysphonia and dysphagia between the initial symptoms and complete anarthria. Such a clinical course indicates more multiinfarct encephalopathy as the cause of biopercular lesions than bilateral strategic infarctions that are considered to be the most frequent mechanism of FCM of vascular origin [14]. It is probable that multiple small infarctions accumulated within the years being potentiated by hypoxia in chronic obstructive pulmonary disease. Indeed, repeated MRI revealed an increasing number of focal lesions cumulated under the insular cortex and in the white matter of the frontal lobes. The association with syphilis is unlikely as the patient did not show any signs of disease activity. Mononeuropathies (including an isolated facial palsy) could rarely occur in the second and more often in the third stage of syphilis [4] but again, it might not produce the automatic-voluntary dissociation.

The mechanisms of dissociation of the face muscles, tongue, pharynx and larynx function are still unclear. Motor nuclei of the 7th to 12th cranial nerves in the pons and medulla oblongata receive the supranuclear innervations from the frontal motor cortex via both the ipsilateral and contralateral corticonuclear tracts. The somatotopical areas corresponding to these nuclei are located in the periinsular cortex of the inferior frontal and precentral gyri (anterior parts of so called operculum). Thanks to the bilateral supranuclear innervation, a unilateral lesion does not induce any severe difficulties with speaking, swallowing or face movements. Conversely, bilateral lesions lead to a marked pontine and bulbar denervation producing clinical symptoms of FCM. Indeed, most works have described FCM due to bilateral cortical lesions and only single reports showed FCM after an isolated cortical or subcortical infarction [1]. Hence, bilateral supranuclear cortico-nuclear denervation causes the loss of voluntary function whereas automatic movements are preserved in the impaired region. That kind of dissociation indicates the existence of a separate, direct connection between subcortical centers and lower cranial nerves nuclei that differs from the corticonuclear tract. These direct connections would remain intact in patients with opercular lesions and therefore preserve the emotional component of face expressions and other automatic movements that

could be operated by the subcortical centers without any voluntary control.

It seems that not only simple reflexes like coughing but even more complex movement programs and stereotypes (e.g. chewing, emotional face expressions) are initiated and controlled from subcortical areas. Accordingly, these functions can be exercised independently of any voluntary intention based on adequate stimulation only. Such an independent subcortical system has never been anatomically described in humans. However, based on animal experiments and pathological literature, it can be supposed that basal ganglia and limbic system play key roles in this so called third motor system [7, 10, 13]. Thus rare cases of morphologically documented FCM represent interesting and unique view of functional brain anatomy.

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