Foix-Chavany-Marie syndrome: a case report

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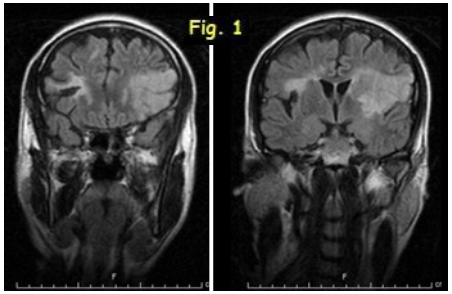
BACKGROUND

Foix-Chavany-Marie syndrome (FCMS) or **anterior opercular syndrome** was first described in detail by Foix et al. in 1926[1]. It is a rare cortico-subcortical suprabulbar palsy of the lower cranial nerves and it is characterized by anarthria, dysphagia and bilateral facial weakness with preserved reflexes and automatic or emotional movements (**automatic voluntary dissociation**). It may be congenital or acquired, persistent or intermittent. It develops after bilateral lesions of the anterior opercula or subcortical insular regions, most often due to sequential cerebro-vascular accidents. It has been also reported in patients with central nervous system infections, neurodegenerative disorders and neuronal migration disorders (dysgenesis of the opercular cortex)[2]. The spectrum of FCMS includes a reversible form in children in status epilepticus from benign rolandic epilepsy.

CASE REPORT

LG, a 45-year-old Caucasian right-handed man, was admitted to the Rehabilitation Unit of San Paolo Hospital in Milan after a left hemispheric ischemic stroke. He has a medical history of hyperhomocysteinemia, medically treated arterial hypertension and a previous right hemispheric ischemic stroke from which he had completely recovered. At admission, the patient was awake and alert, understood spoken and written language but was unable to speak. He was only able to generate a groaning sound and was drooling continually. He was unable to perform voluntary facial or tongue movements: he was neither able to perform voluntary orofacial movements nor to imitate facial motor acts (such as pursing his lips, protruding his tongue..). Emotional facial movements were preserved. His swallowing function was mildly impaired: the voluntary phase of swallowing was compromised (he was unable to initiate the action), but the reflex phase was preserved. Spontaneous swallowing of saliva was observed but with reduced frequency. A nasal-duodenal tube was positioned. He had right hypertonic hemiplegia with Babinski's sign and right sensitive hemisyndrome with somatosensory impairment for touch and position. During the first week an episode of trismus was observed with spontaneous resolution.
Brain MRI revealed a wide acute ischemic lesion in the left middle cerebral artery territory, involving the frontal lobe, insula, operculum, and extending to oval centre, and a right frontal encephalomalacia (Fig. 1). The patient underwent intensive rehabilitation treatment with recovery of language and swallowing functions. After two months he had a good verbal communication and returned to oral diet, with no restrictions or compensatory strategies.



DISCUSSION

FCMS is classically associated with sequential infarcts of the bilateral anterior opercula or subcortical insular regions. The limited data of literature describe a poor prognosis for voluntary swallowing and speech recovery.
In FCMS volitional but not reflexive movements of the face and the throat are affected. This confirms that supranuclear control of the jaw is largely bilateral.
Voluntary corticofugal system originates in the frontal opercular area, whereas spontaneous emotional and automatic movements are presumably mediated by pathways involving other structures (such as basal ganglia). The involuntary motor system, which is older phylogenetically, has less plasticity towards the learning process than the neocortical voluntary system, but it may be recruited in brain mechanisms leading to recovery after stroke.

CONCLUSIONS

Anterior opercular areas are known to be involved in orofacial movement generation[3] and are typically affected in FCMS. The exact functional mechanisms underlying "automatic voluntary dissociation" are not clear and have not been extensively studied.
In the rehabilitation process, however, the precocious diagnosis of FCMS can lead to a more appropriate assessment and therapeutic approaches to speech and nutrition disturbances. FCMS requires close cooperation within a multidisciplinary team.

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