Foix-Chavany-Marie syndrome caused by a disconnection between the right pars opercularis of the inferior frontal gyrus and the supplementary motor area

Case report

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Foix-Chavany-Marie syndrome (FCMS) is a rare type of suprabulbar palsy characterized by an automatic-voluntary dissociation of the orofacial musculature. Here, the authors report an original case of FCMS that occurred intraoperatively while resecting the pars opercularis of the inferior frontal gyrus.

This 25-year-old right-handed man with an incidentally diagnosed right frontotemporoinsular tumor underwent surgery using an asleep-awake-asleep technique with direct cortical and subcortical electrical stimulation and a transopercular approach to the insula. While resecting the anterior part of the pars opercularis the patient suffered sudden anarthria and bilateral facial weakness. He was unable to speak or show his teeth on command, but he was able to voluntarily move his upper and lower limbs. This syndrome lasted for 8 days. Postoperative diffusion tensor imaging tractography revealed that connections of the pars opercularis of the right inferior frontal gyrus with the frontal aslant tract (FAT) and arcuate fasciculus (AF) were damaged.

This case supplies evidence for localizing the structural substrates of FCMS. It was possible, for the first time in the literature, to accurately correlate the occurrence of FCMS to the resection of connections between the FAT and AF, and the right pars opercularis of the inferior frontal gyrus. The FAT has been recently described, but it may be an important connection to mediate supplementary motor area control of orofacial movement. The present case also contributes to our knowledge of complication avoidance in operculoinsular surgery. A transopercular approach to insuloopercular gliomas can generate FCMS, especially in cases of previous contralateral lesions. The prognosis is favorable, but the patient should be informed of this particular hazard, and the surgeon should anticipate the surgical strategy in case the syndrome occurs intraoperatively in an awake patient.

Key Words • arcuate fasciculus • diffusion tensor imaging tractography • Foix-Chavany-Marie syndrome • glioma • frontal aslant tract • insula • functional neurosurgery • oncology

The opercular syndrome was first described by Magnus in 183722 and was later called “Foix-Chavany-Marie syndrome” after the French authors who reported it in 1926.15 This entity is a type of suprabulbar palsy characterized by a loss of voluntary control of facial, lingual, pharyngeal, and masticatory muscles in the presence of preserved reflexive and automatic functions of the same muscles. The specific hallmark of FCMS is automatic-voluntary movement dissociation: affected individuals are unable to voluntarily open their mouth, blow their cheeks, or show their teeth on command or to speak, whereas automatic and emotional movements, such as yawning, emotional laughing, or crying, are all possible.2,15

Abbreviations used in this paper: AF = arcuate fasciculus; DTI = diffusion tensor imaging; FAT = frontal aslant tract; FCMS = Foix-Chavany-Marie syndrome; fMRI = functional MRI; SLF = superior longitudinal fasciculus; SMA = supplementary motor area.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
The most frequent causes of this syndrome are bilateral strokes affecting both opercula, but the syndrome has also been reported after infections, developmental abnormalities, status epilepticus, neurodegenerative disorders, head injury, and insular glioma surgery. In the majority of reported cases the lesion affects broad cortical and subcortical areas of both opercula and the insula, and thus does not allow one to perform an accurate structure-function correlation. Consequently, despite more than 100 documented cases in the last century, the structural substrate of the syndrome has remained unknown.

Insular glioma surgery is an extremely rare cause of this syndrome, as only 5 cases have been reported in the literature. Here, we describe an original case of FCMS that occurred during resection of a right insular WHO Grade II glioma. The patient underwent awake surgery, and so it was possible, for the first time in the literature, to accurately correlate the occurrence of the syndrome with resection of the pars opercularis of the inferior frontal gyrus. Catani et al. recently reviewed the connectivity of this area, describing a new fasciculus that connects this area with the SMA. They named this connection the “frontal aslant tract” because of its oblique shape. In the case presented here the connections of the FAT and AF with the pars opercularis of the inferior frontal gyrus were damaged during surgery. Based on these findings we discuss the pathophysiology of characteristic symptoms of FCMS, such as automatic-voluntary dissociation and dysphagia. We also analyze the implications for surgery in the insuloopercular region.

Case Report

History and Examination. A 25-year-old man with a right frontotemporoinsular tumor incidentally diagnosed in January 2011 was neurologically intact. Magnetic resonance imaging studies revealed a right frontotemporoinsular lesion compatible with a WHO Grade II glioma, that is, hypointense on T1-weighted MRI with no Gd enhancement and hyperintense on T2-weighted and FLAIR imaging. No other lesions were observed in the ipsilateral or contralateral hemisphere (Fig. 1 left). Diffusion tensor imaging tractography revealed no asymmetries in the main white matter bundles organization between the left and right hemispheres. Tractography revealed that the pars opercularis of the inferior frontal gyrus was bilaterally connected to the AF, anterior portion of the SLF, and FAT (Figs. 2A and B and 3). Functional MRI during verb generation showed that activity was predominantly located in the left hemisphere (frontal operculum, parietal lobe, and posterior temporal lobe), but significant activation was also noted in the right insuloopercular region (Fig. 4). On the preoperative Edinburgh Handedness Inventory, the patient scored +80 (right handedness).

Operation. In August 2011 the patient underwent surgery using an asleep-awake-asleep technique with direct cortical and subcortical electrical stimulation, a method extensively described by Duffau and colleagues. Surgery was performed under awake conditions for 3 reasons: 1) fMRI revealed language activation in the right hemisphere. 2) The tumor was resected through a transopercular approach, and awake surgery enabled us to determine whether it was safe to remove the posterior part of the right rolandic operculum (not infiltrated by tumor) to optimize the surgical approach to the posterior part of the insular tumor. 3) Awake surgery with a simple motor task (repetitive movements of contralateral upper and lower limbs) enabled us to accurately monitor motor function during tumor resection. Under awake conditions it is possible not only to record the occurrence of involuntary movement, but also to monitor any slight modification of the movement (slowness, arrest, or lack of accuracy).

Tumor margins in relation to the sulcal and gyral brain surface anatomy were verified with neuronavigation. Prior to tumor resection, cortical mapping was performed. A bipolar electrode with a 5-mm space between
the tips and delivering a biphasic current (square-wave pulses in 4-second trains at 60 Hz, single-pulse phase duration 1 msec, and amplitude 2–8 mA; Nimbus, Hemo-dia) was applied to the brain. Sensorimotor mapping was performed first with identification of the primary motor area of the left thumb, left face, and jaw (Fig. 5). Then the patient was asked to perform counting (regularly counting from 1 to 50, over and over) and picture-naming tasks using the DO80 picture-naming test, which consists of 80 black and white pictures selected according to variables such as frequency, familiarity, age of acquisition, and level of education.9,11–13,37 Electrocorticography was used to monitor for afterdischarges and to eliminate the possibility of language errors due to evoked or spontaneous subclinical seizure activity. Seizures were not registered during cortical mapping and tumor resection. Despite the right-handedness of the patient, cortical language sites were identified at the frontal and parietal lobes: an area of speech arrest was identified at the ventral premotor cortex, and an area of anarthria at the postcentral gyrus. Cortical areas inducing picture-naming errors were not identified.

After completion of the cortical mapping, tumor removal was begun. The insular tumor was approached through a transopercular corridor; therefore, in the first step the frontal operculum was resected from anterior to posterior. The pars oribcularis was extirpated first, then the pars triangularis, and finally the pars opercularis. While resecting the anterior part of the pars opercularis, the patient suffered sudden anarthria and bilateral facial weakness. He was unable to speak or show his teeth on command, but he was able to voluntarily move his upper and lower limbs. Given the impossibility of continuing to collaborate on the tasks, the patient was sedated and a laryngeal mask was placed. With the patient under general anesthesia, the insular component of the tumor was resected. Subcortical motor stimulation was regularly alternated with resection using the same electrical parameters as those at the cortical level.

**Postoperative Course.** The patient was extubated 24 hours after surgery. He was unable to open his mouth, move his tongue, swallow, or speak. However, he was able to yawn and incidentally able to smile; he was unable to smile when asked to do so voluntarily. Dysphagia and sialorrhea were present. Examination revealed bilateral complete palsy of the lower facial muscles, whereas upper facial and ocular movement were normal. Gag, jaw, and corneal reflexes were present. No limb paresis was noted. Unlike speech, other aspects of language were preserved, and communication was possible by writing. His writing was without any grammatical, orthographic, or...
syntactical errors, and his reading as well as his auditory comprehension was intact.

Four days after surgery the patient started to voluntarily open his mouth and utter a few sounds. Two days later, he could move his tongue and speak in short sentences. Eight days after surgery he was completely recovered, with complete normalization of facial and lingual movement, speech, and swallowing. He was discharged to home 10 days after surgery. Two months later his functional status was the same as preoperatively, and he returned to college by the beginning of October 2011.

No seizures were registered after surgery, and the antiepilepsy drugs were stopped after discharge. Histopathology revealed low-grade oligodendroglioma with a Ki 67 lower than 5%. Magnetic resonance imaging performed 3 months after surgery demonstrated a small tumor residue of 8 cm³, and thus the extent of resection was 88% (Fig. 1 right). Postoperative DTI tractography revealed that the pars opercularis of the right inferior frontal gyrus was only connected to the anterior portion of the SLF (Fig. 2C). Therefore, the connections of this area with the AF and FAT were damaged during surgery (Fig. 3 right). On the other hand, corticosubcortical primary motor structures coming from the opercular portion of the precentral gyrus were intact after surgery (Fig. 6).

Postoperative fMRI during verb generation demonstrated a pattern of activation that was similar to that preoperatively (Fig. 4).

Discussion

Identification of Specific Cortical Area Responsible for FCMS

The first task in localizing the area responsible for FCMS is to determine if the lesion affects one or both hemispheres. Most cases in the literature describe bilateral lesions. However, cases with seemingly unilateral lesions have also been reported. Starkstein et al.31 recorded a case of FCMS after a right ischemic infarction of the insula and frontotemporoparietal operculum in a right-handed patient. On macroscopic anatomopathological examination, no other lesions appeared in the left hemisphere, Cosnett et al.4 documented a nocardial abscess in the right operculum, causing FCMS in a left-handed patient; the abscess was diagnosed with CT. Note that only a few cases of FCMS with unilateral lesions have been revealed using MRI. Moragas Garrido et al.27 reported 2 cases of unilateral opercular lesions causing FCMS, one located in the left operculum and the other in the right operculum. Although even in the absence of contralateral structural lesions, some authors consider synchronous epilepsy and hypometabolism as major factors in causing FCMS.2,16,20,27,31 In the present case the possibility of contralateral structural lesions was discarded using MRI, and epilepsy did not seem to participate in the development of FCMS for the following reasons: 1) fluctuations were not observed in the symptoms that slowly regressed over the course of 8 days, 2) intraoperative electrocorticography revealed no seizures, and 3) postoperative electroencephalography revealed no seizures. Some authors have hypothesized that contralateral insuloopercular hypometabolism may prevent an efficient compensatory recruitment of these areas, leading to the occurrence of FCMS.10 However, no metabolic study was performed in our patient to permit us to confirm this hypothesis.

How a unilateral lesion can produce bilateral paralysis of the facial-lip-pharyngeal-laryngeal musculature remains poorly understood. Moragas Garrido et al.27 hypothesized that an anatomical variant with predominant connectivity in one side may exist in some individuals. A lesion within this dominant network may explain bilateral manifestations. In this sense, recent DTI tractography studies have demonstrated asymmetries in subcortical organization between the 2 hemispheres, with left lateralization of the long segment of the AF and right lateralization of the anterior segment of the AF.33 However, in the case presented here, no asymmetries were found in the main white matter bundles organization, which could have explained right lateralization of opercular function (Figs. 2A and B and 3). Moreover, fMRI during verb generation revealed that activity was predominantly located in the left hemisphere (Fig. 4), suggesting that language function was primarily located in the left side. However, crossed speech arrest was transitorily induced by intraoperative electrical stimulation of the right ventral premotor cortex in this right-handed patient. Vassal et al.34 reported the same finding in 3 right-handed patients, supporting the hypothesis of mirror organization of language networks between the right and left hemispheres in some people.

The next question to address is the role of the insular lesion in the generation of FCMS. The insular lobe is highly connected to several areas implicated in the coordination of facial-lip-pharyngeal-laryngeal musculature, such as the lateral premotor cortex, and the frontal and parietal operculum.1,14,26 Moreover, numerous studies have implicated
Identification of Subcortical Connections Responsible for FCMS

Preoperative DTI tractography revealed that the right pars opercularis was connected to the anterior segment of the SLF, the AF, and the FAT. Postoperative DTI tractography showed that this area was only connected to the anterior portion of the SLF. Consequently, surgery damaged the connections of this area with the AF and FAT (Figs. 2C and 3). The FAT has been recently described: it is an obliquely shaped bundle that connects the most posterior part of the Broca territory (that is, precentral cortex [Brodmann Area 6] and pars opercularis [Brodmann Area 44]) in the inferior frontal gyrus with the SMA and pre-SMA in the superior frontal gyrus (Brodmann Areas 8 and 9). Functional properties of this connection are unknown; however, it is possible to hypothesize about its role based on the topography within the brain and the cortical connections. The SMA and pre-SMA are implicated in the initiation of movement and speech. In fact, when this area is injured, an SMA syndrome occurs, with a reduction in spontaneous movement contralateral to the lesion and mutism. It is worth noting the similarities between the SMA syndrome and FCMS: in both entities there is a loss of control of voluntary movement, while reflexive and automatic functions of the same muscles are preserved. A plausible interpretation of these similarities is that the SMA mediates voluntary control of face, tongue, and pharynx movement through this direct connection between the SMA and frontal operculum. If the FAT is damaged, the information coming from the SMA cannot reach the oropharyngeal motor cortex, causing a selective volitional palsy. Rapid recovery of these symptoms within 2–4 weeks may be caused by recruitment of the contralateral specular network. On the other hand, automatic movement of the same muscles is preserved in both syndromes, as this type of response only requires intact extrapyramidal pathways.

Connections of the AF with the pars opercularis were also damaged in the featured case (Fig. 2C). The anatomy of the AF and other components of the SLF has been recently reviewed. In the left dominant hemisphere this tract is well known to underlie the dorsal phonological root, eliciting a conduction aphasia characterized by repetition impairment and phonemic paraphasias; however, the functional role of the right AF is poorly understood. Based on the observation of crucial language areas in the right hemisphere of right-handed patients, Vassal et al. proposed that in some right-handed patients the AF might also subserve phonological processing in the right side. A lesion in this tract on the right side could cause phonological impairment that could participate in the orofacial musculature discoordination that appears in FCMS.

Implications for Surgery at the Insuloopercular Region

Surgery in the nondominant pars opercularis occurs relatively frequently; however, FCMS has been exceptionally reported after surgery. In fact, this syndrome has been reported only after insuloopercular glioma resection. Duffau described 4 cases of FCMS in a series of 51 resected insular gliomas. Consequently, we estimate a nonnegligible 7.8% risk of the syndrome developing after this type of surgery. This risk may be explained by the fact that in this type of surgery, operculum not infiltrated by tumor is occasionally resected to gain exposure to the insular lobe (after electrostimulation confirmation that the opercula is not essential for the function). The risk of FCMS may be higher in cases with preexisting contralateral structural, functional, or metabolic damage. Consequently, in insular glioma resection, it is important to observe the following cautions: 1) be aware of situations in which contralateral brain injury is a component of the patient’s history; 2) perform a rigorous preoperative neuromaging assessment of the contralateral hemisphere to detect opercular lesions that may increase the risk of FCMS developing; 3) inform the patient and family of this risk, since it can be shocking if it is not expected, despite the favorable prognosis; 4) anticipate the surgical strategy that will be followed if the syndrome occurs during awake surgery, as the patient will be unable to collaborate; and 5) keep the opercular resection to the minimum necessary to approach the insular component of the tumor and pay special attention to resection of the pars opercularis of the inferior frontal gyrus.

The classic FCMS caused by a bilateral opercular lesion is considered a highly disabling sequelae with poor functional recovery. In fact, impairment in verbal language requires the patient to develop other means of
communication, and dysphagia increases the risk of aspiration threatening the patient’s life. However, a favorable functional outcome was reported in the present case and in other cases after surgery for an insular glioma.5,10 The following factors may be implicated in this rapid recovery: 1) slow infiltration of the insula and operculum by the tumor generates functional reshaping with the activation of regional and distant networks. 2) In insular glioma resection only one side is damaged; consequently, after surgery the contralateral hemisphere may underlie the restoration of function, as previously suggested in swallowing recovery after stroke.10 3) Intraoperative electrical stimulation allowed identification and preservation of the motor area of the face. A lesion within this area may have caused more severe or long-lasting FCMS, as this area may participate in functional recovery.5,10 4) In the current case resection was limited to the anterior part of the pars opercularis; therefore, fibers of the anterior portion of the SLF were preserved, as demonstrated in the postoperative DTI tractography study.

Disclosure

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