**FOIX-CHAVANY-MARIE SYNDROME AFTER SURGICAL RESECTION OF A RIGHT INSULO-OPERCULAR LOW-GRADE GLIOMA: CASE REPORT**

**OBJECTIVE AND IMPORTANCE:** We describe an atypical case of transient Foix-Chavany-Marie syndrome, or faciopharyngoglossomasticatory diplegia with automatic voluntary dissociation, occurring after surgical resection of a right insulo-opercular glioma.

**CLINICAL PRESENTATION:** A 26-year-old right-handed man experienced partial seizures that were poorly controlled by antiepileptic drugs during a 2-year period as a result of a right insulo-opercular low-grade glioma, leading to the proposal of surgical resection. In addition, 1 year before the operation, the patient experienced a severe brain injury that resulted in a coma. A computed tomographic scan revealed left opercular contusion. The patient recovered completely within 6 months.

**INTERVENTION:** Intraoperative corticosubcortical electrical functional mapping was performed along the resection, allowing the identification and preservation of the facial and upper limb motor structures. A subtotal removal of the glioma was achieved. The patient had postoperative anarthria, with loss of voluntary muscular functions of the face and tongue, and he had trouble chewing and swallowing. All of these symptoms resolved within 3 months.

**CONCLUSION:** These findings provide insight into the use of surgery to treat a right insulo-opercular tumor. First, surgeons must be particularly cautious in cases with a potential contralateral lesion (e.g., history of head injury), even if such a lesion is not visible on magnetic resonance imaging scans; preoperative metabolic imaging and electrophysiological investigations should be considered before an operative decision is made. Second, surgeons must perform intraoperative functional mapping to identify and to attempt to preserve the corticosubcortical facial motor structures. A procedure performed while the patient is awake should be discussed to detect the structures involved in chewing and swallowing in cases of suspected bilateral lesions. Third, the patient must be informed of this particular risk before surgery is performed.

**KEY WORDS:** Foix-Chavany-Marie syndrome, Insulo-opercular tumor, Intraoperative functional mapping, Low-grade glioma
CASE REPORT

Patient Presentation

A 26-year-old right-handed man presented with partial seizures in September 2000; examination revealed a right insulo-opercular tumor. At that time, a stereotactic biopsy was performed, which permitted a diagnosis of low-grade oligodendroglioma. No surgery was proposed to the patient. Despite the use of antiepileptic drugs (gabapentin and valproate, then gabapentin and carbamazepine), the patient still experienced approximately two partial seizures per week, during which his speech was transitorily interrupted and he drooled.

In June 2001, the patient experienced a severe head injury and coma, which necessitated his admission to our department. A computed tomographic scan revealed a diffuse subarachnoid hemorrhage associated with a mild left central concussion (symmetrical to the tumor). Artificial ventilation with tracheostomy was necessary for 15 days. One month later, the patient’s neurological status had dramatically improved, with normal consciousness and the beginning of ability to walk, but right brachiofacial paresis and cognitive disorders persisted. Neurological rehabilitation allowed a complete functional recovery within 6 months. A new magnetic resonance imaging (MRI) scan performed at that time and 6 months later revealed no residual abnormality in the left hemisphere (as assessed by T1-weighted images with and without gadolinium, T2-weighted images, and fluid-attenuated inversion recovery images). Because the seizures were not being controlled, and because the volume of the right glioma increased slightly (50 ml on the preoperative MRI scan), with invasion of the whole insula and the frontal operculum (Fig. 1), it was decided to surgically resect the tumor in August 2002, 2 years after the tumor was diagnosed. On the preoperative Edinburgh inventory (40), the patient scored +100 (right-handedness).

Intervention

The resection was conducted with the patient under general anesthesia. We used a method of intraoperative electrical motor mapping that we reported previously (12, 14) (Fig. 2). After identification of the tumor boundaries by intraoperative ultrasonography, direct cortical stimulations were performed, allowing the detection of the primary motor sites of the superior limb and face (5-mm spaced-tip bipolar electrode, biphasic current with pulse duration of 1 ms, frequency of 60 Hz, intensity of 8 mA [Ojemann cortical stimulator; Radionics, Burlington, MA]). Because the patient was intubated, it was impossible to note whether involuntary swallowing was elicited by stimulation.

The frontal operculum was resected. The sylvian fissure was opened laterally by using functional limits posteriorly provided by subcortical stimulation, which identified the pyramidal pathways of the face and the superior limb within the corona radiata. Once the insular surface was exposed, cortical mapping was performed at its level. No motor response was elicited. The portion of the glioma involving the insula was then removed, again using deep subcortical stimulations, as previously reported (13, 14).

FIGURE 1. Preoperative coronal (A and C) and axial (B and D) T1-weighted, gadolinium-enhanced MRI scans revealing a right insulo-opercular low-grade glioma. The neurological examination was normal.

FIGURE 2. a, intraoperative photograph obtained before tumor removal. The tumor boundaries identified via ultrasonography are marked by letter tags (A–G). The primary motor cortical sites of the superior limb (1, 2) and of the face (3) were detected via electrical mapping. b, intraoperative photograph obtained after tumor removal. The primary motor cortical sites and their corresponding descending corticospinal pathways (49, 50), both identified along the resection by use of repeated corticospinal electrical stimulations, represent the posterior functional boundaries of the resection. Anterior is to the left.
Postoperative Course

The patient was extubated immediately after surgery. There was no limb deficit; however, the patient experienced mutism and an inability to swallow. Examination revealed a bilateral complete palsy of the lower facial muscles. The mouth remained half-open, and the patient drooled. The tongue remained immobile inside the mouth; voluntary deglutition and chewing were impossible, so the patient was fed through a nasogastric tube. Nevertheless, some possibility of automatic swallowing seemed likely. Laryngoscopy revealed that the vocal cords were not paralyzed. The patient could not utter a sound or articulate simple words. He could not produce suction movements or throw a kiss, either on command or by imitation. No upper facial disturbance or ophthalmoplegia was observed; the upper facial motility was bilaterally intact. The corneal reflex was preserved.

Over the course of 3 days, the patient had complete anarthria and could communicate only by writing. He could write fluently, with no feature of aphasis agraphia: the phrases were semantically and syntactically correct. There was no apraxia of the right hand. Comprehension was normal. The patient was examined daily by a speech therapist. Four days after surgery, the patient began to utter a few sounds. One day later, he was able to articulate some words, and after the seventh day, he could speak in short sentences. Lower facial and lingual palsy progressively recovered in 2 weeks. The nasogastric tube was removed 3 weeks after the operation, allowing the patient to return home with rehabilitation. Two months later, the functional status was the same as it had been preoperatively; in particular, normalization of swallowing and speech had occurred.

The patient experienced no seizures after the operation; he continued therapy with the same antiepileptic drugs that had been administered before he underwent surgery. Control MRI scans performed 3 months after the operation revealed a subtotal resection according to the classification of Berger et al. (4), with less than 10 ml of residue remaining (approximately 4 ml in the posterior insula) (Fig. 3). The histological examination confirmed the diagnosis of low-grade oligodendroglioma, and no radiotherapy or chemotherapy was performed.

DISCUSSION

We describe here a unique case of FCMS. To our knowledge, this is the first case of FCMS reported after brain tumor surgery. Moreover, this syndrome occurred after a unilateral right operculoinsular lesion, and the symptoms were transient.

Postoperative FCMS

The seminal publications of Magnus (35) in 1837 and Foix-Chavany-Marie (19) in 1926 described the association of anarthria and bilateral central faciolinguovelopharyngeomotoric paralysis with automatic-voluntary dissociation. Since then, the etiologies detailed by Weller (53) in 1993 in an extensive review of the literature indicated that the causes in approximately 63 patients were as follows: vascular (5, 18, 33, 36, 37, 44, 45, 48, 51), infectious (9, 22, 39, 46), developmental (3, 7), epileptic (8, 21, 26), and degenerative (6, 30). On the basis of these findings, Weller (53) proposed the differentiation of five clinical types of FCMS. More recently, Laurent-Vannier et al. (31) reported a case of FCMS caused by head trauma in a child.

In our case, the FCMS was not directly induced by the glioma; there was no preoperative deficit despite the progressive tumor growth. This could be explained by brain plasticity mechanisms, widely described in low-grade gliomas (11, 16, 47, 49, 54). Nevertheless, the patient had a transient speech arrest and drooled during partial seizures—perhaps an equivalent of an FCMS-like disorder despite the impossibility of better detailing the patient’s neurological status during these phases because the patient completely recovered in 1 minute, which did not permit time for examination. FCMS occurred immediately after the tumor resection; thus, it was likely the direct result of the surgical act itself. Glioma surgery is known to potentially induce specific syndromes postoperatively, such as the supplementary motor area syndrome after resection of frontomesial structures (20, 28) or athymhormic syndrome after removal of paralimbic gliomas involving the right striatum (15). However, to our knowledge, no FCMS has been reported after resection of an operculoinsular tumor.

FCMS Caused by Unilateral Right Operculoinsular Lesion

Classically, FCMS is caused by bilateral cortical or subcortical damage involving the frontal operculum with or without involvement of the insula (2, 19, 53). There are only five reports of FCMS in patients with seemingly unilateral brain
lesions (9, 21, 26, 35, 48), among them two putative (structural or functional) lesions in three cases (9, 21, 26).

In our patient, FCMS occurred after the resection of a glioma involving the whole right insula and the right frontal operculum, without any cortical or subcortical contralateral damage, as revealed by multiple preoperative MRI scans (in particular, sensitive fluid-attenuated inversion recovery images). This is a surprising result because we had never before observed such a postoperative deficit in our experience of more than 40 procedures involving insular-paralimbic-opercular gliomas (11, 13, 14), nor had such a deficit ever been described in the surgical literature of this kind of tumor (29, 52, 55, 57).

On the other hand, direct electrical stimulation revealed that both the frontal operculum and the right (anterior) insula were involved in swallowing (41, 42). Transcranial magnetic stimulations (24) and functional neuroimaging methods (25, 38, 56) provide data that converge with reports of dysphagia after unilateral anterior insular lesions (10) or opercular lesions (43). Moreover, these two structures are also known to be implicated in orofacial movements and chewing (1, 41, 42, 50), as emphasized by Starkstein et al. (48), who reported a case of bilateral opercular syndrome and crossed aphasia that was the result of a right insular lesion (i.e., aphemia induced by a lesion of the right non-language-dominant hemisphere in a right-handed patient).

Therefore, taking into account these data, we might rather ask the question: why is there usually no FCMS after resection of an operculoinsular tumor? It was previously suggested that the contralateral hemisphere could compensate for the facial motor cortex (32) and the insula (11); thus, one explanation might be that not only the slow-growing, low-grade glioma but also the surgical act itself generates functional reshaping (16)—namely, the recruitment of the contralateral homologous area (23), as already indicated after supplementary motor area resection via pre- and postoperative functional MRI scans (27).

In the present case, the only difference in comparison to the other patients who underwent surgery for an insulo-opercular tumor is the fact that the patient previously sustained a brain injury with left opercular damage. Thus, even if the MRI scan revealed nothing abnormal 1 year after the head trauma, we can hypothesize that the possible persistence of a left (insula-)opercular hypometabolism (as demonstrated in the literature that describes findings revealed via single-photon emission computed tomography [51]) prevented an efficient compensatory recruitment of these areas, leading to the occurrence of an FCMS. However, no metabolic study was performed in our patient to permit us to confirm this hypothesis. The hypothesis of a prolonged postoperative epilepsy seems less likely for two reasons: 1) the symptoms slowly regressed over the course of several weeks (without fluctuation and then sudden arrest [7], and/or with shorter duration [26]); and 2) the patient’s electroencephalogram revealed nothing abnormal.

**Transient FCMS**

One classic feature of FCMS is a poor recovery, except in the particular case of status epilepticus in children (53). One explanation of the favorable functional outcome in our case could be the delayed recruitment of the contralateral hemisphere, previously suggested in swallowing recovery after stroke (23), by the lifting of inhibition via the transcallosal pathways (34), even if the functional compensation was not possible immediately after surgery for the reasons considered above (as in the supplementary motor area syndrome, lasting usually 10–20 d [17]). Moreover, the use of intraoperative corticosubcortical stimulations allowed the identification and subsequent preservation of the essential structures for the movements of the face (but not for the movements of the tongue or for deglutition—i.e., no detection of the whole corticonuclear pathways). Therefore, it seems likely that these facial motor areas participate, at least partially, in the functional recovery after a transient postoperative stage of “sideration,” which is classically observed after a corticosubcortical glioma resection limited by functional boundaries (12). According to this hypothesis, a combined resection of the insular lobe, frontal operculum, and primary motor area of the face should be carefully discussed individually on the basis of a rigorous functional preoperative assessment to try to find the areas in the ipsilateral and contralateral hemispheres that are still potentially dysfunctional (as a result of tumor or a previous injury) and areas able potentially to compensate for the function, so that postoperative defects can be predicted in cases of bilaterally symmetrical lesions.

**CONCLUSION**

This study indicates the possibility of generating an FCMS after the resection of a right insulo-opercular glioma in a right-handed patient. These findings suggest the following: 1) surgeons should be particularly cautious in the case of a potential contralateral lesion (e.g., history of head injury), even if such a lesion is not visible on anatomic images. Preoperative metabolic imaging and electrophysiological investigations should be considered to make the operative decision; 2) intraoperative functional mapping should be performed to identify and eventually preserve in selected patients the corticosubcortical primary motor structures of the face (when the insula is also invaded), even in the nondominant hemisphere—in other words, a combined resection of the insular lobe, frontal operculum, and primary motor area of the face might be avoided if contralateral compensation is not possible. Moreover, a procedure performed while the patient is awake should be discussed to detect the structures involved in chewing and swallowing in cases of suspected (anatomic, electrophysiological, or metabolic) bilateral lesions; and 3) the patient should be informed of this particular risk before the surgical procedure is performed.

**REFERENCES**


This article contributes to our knowledge of complication avoidance in operculoinsular surgery. The techniques of intraoperative functional mapping that use subcortical stimulation, as the authors describe, are particularly useful.

Robert G. Grossman
Houston, Texas

Duffau et al. report an unusual neurological course after subtotal resection of a low-grade oligodendroglioma involving the right frontal operculum and insula. In their recognition from classical neurology of the bilateral pathological substrate underlying Foix-Chavany-Marie syndrome, the authors almost certainly are correct in invoking the earlier left-sided traumatic injury. It is surprising that as sensitive a sequence as fluid-attenuated inversion recovery failed to reveal any residual injury, but the severity of the injury and the associated brachial-facial deficit at the time of that injury lend credence to the presumed contralateral disturbance. What remains most provocative is the complete and fairly rapid recovery from the surgically induced syndrome, when attribution of its occurrence to a contralateral abnormality ought to preclude the contribution of that disturbed dominant hemisphere to the recovery. Recovery of preserved but transiently dysfunctional primary motor cortex must underlie this restoration of function. Functional magnetic resonance imaging might have assessed this better than intraoperative mapping.

David W. Roberts
Lebanon, New Hampshire

Erratum
In a comment by Laligam N. Sekhar and Dinko Stimac regarding an article by Mir Jafer Ali et al., titled “Arterial Reconstruction by Direct Surgical Clipping of a Basilar Artery Dissecting Aneurysm after Failed Vertebral Artery Occlusion: Technical Case Report and Literature Review” (Neurosurgery 52:1475–1481, 2003), the phrase “posterior communicating artery” in the second paragraph should have read “posterior cerebral artery.” The publisher regrets the error.

Mitchel S. Berger
San Francisco, California

This report serves as a reminder of the potential for Foix-Chavany-Marie syndrome in tumor surgery. The majority of reports concerning this syndrome indicate that bilateral lesions must be present. Accordingly, the previous head injury in this patient must have provided the substrate for the emergence of the neurological problems after nondominant opercular surgery. Regardless, the authors illustrate the usefulness of intraoperative mapping well; this likely facilitated recovery. Foix-Chavany-Marie syndrome is a rare entity, and this report is, to my knowledge, the first to report it after tumor surgery.

Joseph M. Piepmeier
New Haven, Connecticut

Postoperative Foix-Chavany-Marie Syndrome

Duffau et al. present an interesting case of an extremely unusual neurological syndrome that normally occurs in a setting other than that associated with a tumor resection. The patient had bilateral dysfunction of the face, pharynx, tongue, and muscles of mastication, and he was unable to speak. He had a history of an injury to the dominant left operculum after trauma. The resection performed by the authors included the nondominant operculum, resulting in this transient symptom complex, even though no permanent injury involving the dominant hemisphere was observed on the control magnetic resonance imaging scan.

Surgeons should thus be aware of situations in which a contralateral brain injury is a component of the patient’s history and is symmetrical to the site of the proposed resection. This will allow the surgeon to consult with the family and predict the possibility of a transient neurological dysfunction that otherwise would not be expected to be present postoperatively. In settings like this, where there is previous traumatic brain injury, functional magnetic resonance imaging assessing subcortical white matter pathways (i.e., diffusion tensor imaging) or a metabolic imaging study such as positron emission tomography may indicate disrupted pathways or regions of hypometabolism that may predict dysfunction in the setting of a contralateral resection. This might alert the surgeon to a transient postoperative syndrome that could be discussed with the family before the surgery. Thus, this is a useful case report for all surgeons who do cortical- and subcortical-based resections.