Foix-Chavany-Marie syndrome caused by bilateral opercular lesions: right side tumor and left side ischemic stroke

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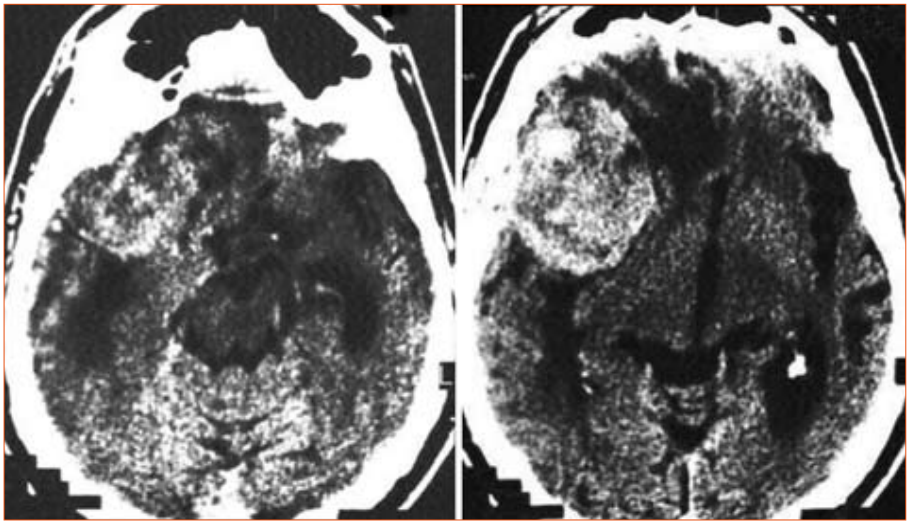

Figure. Cerebral CT scan. Right fronto-temporal infiltrative tumor, involving right operculum. Residual porencephalia after old left operculum ischemic stroke.

Foix-Chavany-Marie syndrome (FCMS) or bilateral anterior opercular syndrome is characterized by bilateral facio-labio-pharyngo-glosso-masticatory paralysis with automatic-voluntary dissociation [1]. The first case was reported in 1837 by Magnus [2], but the syndrome was described by Foix, Chavany and Marie in 1926 [1].

It is a rare disease, with a prevalence < 1/1,000,000 cases [3]. Up to date there had been reported approximately 150 de cases [3].

FCMS is characterized by bilateral cortico-subcortical suprabulbar cranial nerve paralysis (V, VII, IX, X and XII), speech dysfunctions, chewing disturbances, dysphagia, sialorrhea, and bilateral loss of facial muscles tonus. Severity of dysarthria goes from difficulties in sounds articulation (dysarthria) to complete mutism (anarthria). Patients present automatic-voluntary dissociation and lack of active emotional interacting, like in pseudobulbar syndrome. These patients have normal level of consciousness, correctly obey verbal commands and can adequately communicate using gestures or in writing. Sialorrhea occurs as a result of autonomic movements' preservation. Patients are incapable to perform any voluntary movements dependent on bulbar muscles, the tongue is flaccid without atrophy or fasciculations, the jaw jerk reflex is hyperactive, facial muscles are hypotonic and cannot be contracted voluntarily, but they can smile, cry or yawn involuntary [4,5]. Less frequently FCMS may also present with dystonia [6].

In most cases FCMS is caused by bilateral ischemic strokes [3,6-8], followed by cerebral infections [9] and reversible form of child's epilepsy [10,11]. Less common it may be secondary to tumors [12] or traumatic brain injury [13,14].

The aim of this paper is to report one case with FCMS secondary to two distinctive bilateral opercular lesions.

We report a case of a right handed, 74 years old man, admitted into the Neurosurgical Department with dizziness, speech dysfunctions (severe dysarthria), sialorrhea, dysphagia, masticatory disturbances, with progressive onset 3 weeks prior admission. Clinical exam showed left pyramidal syndrome, and bilateral V, VII, IX, X and XII cranial nerves paralysis, causing bilateral paralysis of facial, lingual, pharyngeal and masticatory muscles. All volitional movements of these muscles were absent. The face appeared atonic and completely inexpressive, the tongue showed no voluntary movements, fibrillation or atrophy. The mouth was partially opened and the patient was unable to close or open it further. Chewing and swallowing were disturbed. The automatic functions were preserved, the patient was able to smile, cry and yawn. The jaw jerk reflex was highly active. The patient obeyed commands.

Medical history was inconclusive, with possible left ischemic stroke 3-4 years ago, with no positive diagnosis, or imaging from that event.

Native cerebral CT scan showed an inhomogeneous, containing multiple necrotic areas, hyperdense (30-63 uH), poor-defined, infiltrative right fronto-temporal tumor, with massive surrounding edema and midline shift. The tumor measured 7.2 × 6.4 × 5.4 cm. The upper pole of the lesion occupied right middle gyrus, crossed corona radiata, invaded external capsule, claustrum, extreme capsule, insula, operculum and splayed M1-M2 segment of the right middle cerebral artery and the lower pole anterior reached the sphenoid horn of the right lateral ventricle. In the left temporal operculum there was a homogenous, well-defined, hypodense aria (12-16 uH), sizing 17 × 9 mm, with localized ectasia of the left middle cerebral artery in M2-M3 segment, upstream the arterial insular knee. The ventricular system was asymmetric, with 10-12 mm midline shift to the left, basal cisterns were present (Figure).

Unfortunately, the patient declined further imaging (administration of contrast agent and cerebral MRI) and surgery, against medical advisory. The outcome was poor, and the patient died after 4 months.

FCMS is a cortico-subcortical pseudobulbar palsy of the lower cranial nerves (V, VII, IX, X and XII), causing bilateral facio-labio-pharyngo-glosso-masticatory voluntary paralysis, with preserved involuntary activity of paretic muscles.

For FCMS occurrence bilateral opercular lesions are mandatory. Cerebral operculum is composed by brain covering the insular lobe. According to Penfield and Rasmussen [15] it is composed of frontal operculum (also known as rolandic operculum, consists of opercular part of F3, areas 44 and 45), temporal operculum (belonging to superior temporal gyrus, area 22) and parietal operculum (contains distal part of pre- and postcentral gyrus, supramarginal gyrus and angular gyrus. The inferior third of precentral gyrus is connected through the corticobulbar tract with nuclei of cranial nerves V, VII, IX, X, XI and XII bilaterally, except for the following nerves: VII for the lower face, X-XI for levator veli palatini and XII for genioglossus muscle, which are innervated unilaterally [16]. FCMS occurs in bilateral anterior opercular lesions, meaning bilateral damages in frontal operculum and precentral part of parietal operculum [17]. Damage of only one operculum had been described in the literature, but the exact pathopsychological mechanism was not established [18].

In 1993 Weller [17] classified FCMS in five clinical types: classic form, secondary to cerebrovascular diseases; subacute form, secondary to infections; developmental form, caused by neuronal development disorders; reversible form, associated with child epilepsy and rare form, caused by neurodegenerative disorders.

Our patient presented two different opercular lesions, namely an infiltrative tumor with right operculum invasion and old ischemic stroke in M2-M3 segment of middle cerebral artery in the left operculum. The chronological sequence of injuries was ischemic stroke, probably 3-4 years ago, with minimal clinical expression and good outcome (suspected during medical history and retrospectively confirmed by CT scan), followed by right opercular tumor occurrence, also seen on CT scan.

The majority of cases reported in the literature were caused by bilateral ischemic strokes [6-8], but other etiology may also be incriminated, and other authors reported distinctive lesions causing FMCS.

Santos et al reported two concomitant distinctive bilateral opercular lesions ischemic stroke on one side and controlateral hemorrhagic stroke [19].

Concerning the disease secondary to tumors, Duffau reported a case with mild FCMS occurring one month after resection of right insulo-opercular low grade glioma, without any detectable imaging on the left operculum [12].

Opercular atrophy and hipodense lacunar lesions can be seen on non-contrast CT, but more useful is cerebral MRI, diffusion and perfusion weighted MRI [20], SPECT [5] and electrophysiological investigations.

Differential diagnosis is made with bulbar and pseudobulbar paralysis syndromes, stroke, amyotrophic lateral sclerosis, myasthenia gravis, Brown-Vialetto-van Laere syndrome, brainstem tumors, neighboring tumors from the posterior cranial fossa secondarily invading the brainstem, brainstem hematoma, vascular malformations of the brainstem, skull base tumors involving the cranial nerves and carcinomatous meningitis.

Therapy must be individualized according to each case. Optimal treatment must target the cause of FCMS. Theoretically, in our case, first line treatment is surgical resection of the right fronto-temporal tumor, followed by adjuvant therapy, according to the histopathological diagnosis.

The tumor poses some challenges due to location in and next to eloquent areas, infiltrative nature, large size, surrounding edema and mass effect. Surgery in a patient with fronto-temporal tumor, including the right operculum and preexistent controlateral operculum lesion, requires special considerations. Intraoperative functional mapping helps to identify and preserve corticosubcortical facial and upper limb motor structures and Broca area [21]. If these structures are preserved the patient has chances to recover after a period of time following surgery. If these structures cannot be spared, in patients with giant, infiltrative tumors, FCMS may never recover. Patient and family must be informed about the possibility that symptoms may persist after surgery.

Special care must be given, within multidisciplinary approach, to speech recovery and feeding. Due to impairment in chewing and swallowing oral feeding is severely hindered. To improve feeding nasogastric tube, percutaneous endoscopic gastrostomy [4], posture [3] and/or neuromuscular electrical stimulation [22] can be used.

Unfortunately, the patients and his family refused surgery and died after 4 months. Short survival was due to tumor progression, which was large, infiltrative, with important mass effect.

In conclusion, FCMS is a rare neurological syndrome caused by bilateral opercular lesions, characterized by bilateral facio-labio-pharyngo-glosso-masticatory paralysis with automatic-voluntary dissociation. It can be caused, as in our case, by two acquired distinctive lesions, ischemic stroke on one side and tumor on the other side. After ischemic stroke involving left operculum the patient had good recovery, but once the right fronto-temporal tumor involving the right operculum progressed, the patient became symptomatic. Clinical onset of FCMS requires damage of both opercula. Careful neurological examination, medical history and imaging are the main keys in diagnosing FCMS.

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