

Recovery of Foix-Chavany-Marie Syndrome with Multi-Modal Rehabilitation Therapy: A Case Report and Review of Literature

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Abstract

Foix-Chavany-Marie syndrome (FCMS) is a rare type of pseudobulbar palsy characterized by loss of voluntary control of the facial, glossal, pharyngeal, laryngeal, and masticatory muscles with preserved automatic, involuntary movements, also called “automatic-voluntary dissociation”. These dysfunctions are usually difficult to recover. We would like to report a 50-year-old female presented as FCMS with prominent oral dysphagia and dysarthria. Therapies included speech therapy, transcranial magnetic stimulation and acupuncture. After 3 weeks’ treatment, the patient had improvements in oral facial muscle control and chewing. However, the dysarthria remained prominent. Multi-modal rehabilitation therapy may be helpful in the recovery of FCMS.

Keywords: Opercular syndrome; Foix-Chavany-Marie syndrome; Rehabilitation; Speech therapy; Transcutaneous magnetic stimulation; Acupuncture

Introduction

Foix-Chavany-Marie Syndrome (FCMS), also known as Bilateral Opercular Syndrome, is a neuropathological disorder characterized by paralysis of the facial, tongue, pharynx, and masticatory muscles of the mouth that aid in chewing. The disorder is primarily caused by thrombotic and embolic strokes, which cause a deficiency of oxygen in the brain. As a result, bilateral lesions may form in the junctions between the frontal lobe and temporal lobe, the parietal lobe and cortical lobe, or the subcortical region of the brain. FCMS may also arise from defects existing at birth that may be inherited or nonhereditary.

It most commonly occurs secondary to bilateral opercular stroke, but other cases reported in the literature include following insular glioma resection and unilateral opercular contusions following traumatic brain injury.

Case Report

A 50-year-old female with history of hypertension and prior right facial paralysis was admitted to our unit due to difficulties in speaking, swallowing, moving the tongue, and chewing for 2 months. She developed dysarthria and dysphagia as well as left-sided weakness after embolization of an aneurysm in right middle cerebral artery 2 months ago. Brain magnetic resonance imaging (MRI) and computerized tomography (CT) showed acute infarction in the right opercular region and a remote lesion in the left basal ganglia (Figure 1). After medicinal treatment and rehabilitation, the patient was able to use her left upper extremity for basic daily activities and walk independently. Her Barthel Index score was 100. However, the ability of speaking and swallowing remained impaired remarkably.

Regarding previous right facial paralysis, it was reported “right peripheral facial paralysis” happened six months ago. Brain MRI at that time accidentally found subacute hemorrhage in the left basal ganglia. However, no medical record can be retrieved for detailed review. She had slight residual dysfunctions on the right-side including weakness of showing the teeth, raising the eyebrow. Speaking, swallowing, moving the tongue and chewing were intact. Otherwise, her blood pressure has been under control. She has no history of heart disease, diabetes, smoking or drinking. She has no similar family history.

On physical examination, her mouth was half open with a 2 cm distance between both lips when relaxed. She could not voluntarily open or close her mouth and show her teeth. She was drooling constantly. Chewing was significantly impaired. The tongue was immobile, and she was not able to convey food bolus to pharynx nor initiate swallowing voluntarily. However, she was able to swallow safely once food was placed at the posterior oral cavity. No tongue atrophy, fibrillation or deviation were observed. The nasolabial sulcus disappeared on the left side and diminished on the right side. Frowning and eye closing were impaired on both sides. Taste was diminished over the right anterior 2/3 tongue but not on the left. Palmomental reflex was increased on the right. The pharynx reflex was not able to perform because of the limitation in opening mouth. She was only able to pronounce “uh”. Writing and reading was unable to assess since she was illiterate. On the other hand, she was able to close her eyes during sleep, yawn spontaneously, laugh when she felt amused and cry under appropriate conditions. Muscles strengths were 5/5 throughout, slightly weaker on the left side. There were no concerning labs results. The modified Frenchay Dysarthria Assessment (Chinese version) was 10 [1]. The standardized swallowing assessment (Chinese version) score was 33 [2].

Regarding to rehabilitative strategies, focus was on active and passive training of her orofacial and laryngopharyngeal muscles. Speech therapies included oral muscle exercises, ice stimulation, sensory stimulation, breath-holding and phonation technique, cough exercise, articulation exercise, respiratory exercise with low frequency electrical stimulation on bilateral mastoid and suprahyoid muscles. The patient received 20 minutes’ 1Hz repetitive transcranial magnetic stimulation (rTMS) daily over the contralateral motor cortex at an intensity of 100% for 10 days. She received 30 minutes’ acupuncture daily in a

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Reference	Age	Sex	Symptoms	Diagnosis	Involvement based on imaging	Recovery	Rehabilitation	Follow up
Thapa et al. (2010) Patient 1	35	F	Difficulty in swallowing and inability to speak	Cerebral infarction	Left opercular and subopercular	Marked improvement in dysarthria Partial improvement in dysphagia	N/A	7 days
Thapa et al. (2010) Patient 2	24	F	Severe global headache and inability to speak	Cerebral infarction	Large bilateral opercular	Partial improvement in dysarthria No improvement in dysphagia	N/A	3 weeks
Thapa et al. (2010) Patient 3	75	F	Inability to speak and swallow	Cerebral infarction	Large left opercular Right parietal region	No improvement in dysarthria and dysphagia	N/A	4 weeks
Thapa et al. (2010) Patient 4	50	F	Marked difficulty in speaking	Cerebral infarction	Left cerebellar and perisylvian Right insular	No improvement in dysarthria and dysphagia	N/A	N/A
Thapa et al. (2010) Patient 5	60	F	Difficulty in speaking and swallowing	Cerebral infarction	Large left opercular Small right sylvian	No improvement in dysarthria and dysphagia	N/A	2 weeks
Karaca et al. (2013) Patient 1	85	F	Inability to speak and swallow	Cerebral infarction	Bilateral perirolandic areas Left anterior operculum	Marked improvement in dysarthria and dysphagia	N/A	5 years
Karaca et al. (2013) Patient 2	77	M	Difficulty in speaking and swallowing	Cerebral infarction	Right opercular of frontal lobe	Markedly improvement in dysarthria and dysphagia	N/A	2 months
Milanlioglu et al. (2013)	81	M	Inability to speak and swallow	Cerebral infarction	Bilateral inferior frontal gyrus	No improvement in dysarthria and dysphagia	N/A	3 weeks
Theys et al. (2013)	48	M	Inability to speak	Cerebral hemorrhage	Right thalamocapsular	Markedly improvement in dysarthria	Speech therapy	6 months
Wu et al. (2013)	30	M	Inability to speak and swallow	Traumatic brain injury	Bilateral rolandic opercular	N/A	N/A	2 years
Sá et al. (2014)	76	M	Difficulty in speaking	Cerebral infarction	Left opercular	No improvement in dysarthria	Intensive rehabilitation	1 month
Ohtomo et al. (2014)	76	M	Inability to speak and swallow	Cerebral infarction	Left anterior operculum and cerebellar hemisphere Right pons	Little improvement in dysarthria Improvement in dysphagia	Rehabilitation	5 months

N/A: Not Applicable; F: Female; M: Male

Table 1: Recovery of 12 cases with different clinical and imaging features.

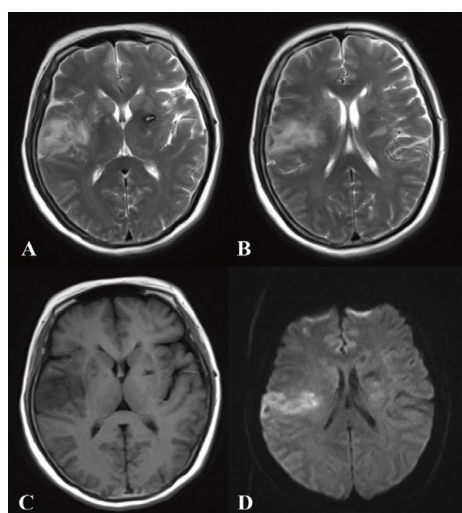


Figure 1: (A and B) Two layers of brain MRI T2-weighted images showed hyper-intensity in the right opercular region. (C) T1-weighted image showed hypo-intensity in the right opercular region. There was a remote small localized infarct in the left basal ganglia with evidence of hypo-intensity in both T1- and T2-weighted images. (D) Diffusion-weighted imaging showed associated high signal intensity in the right opercular region.

total of 15 days. According to the patient's clinical manifestations, bilateral Shàngyíngxiāng (EX-HN8), Dīcāng (ST4), Jiáchē (ST6),

Tàiyáng (EX-HN4), Jīngmíng (BL1), Sīzhúkōng (TE23), Sībái (ST2) for facial paralysis, Shǔigōu (GV26) and bilateral Tīnggōng (SI19) for difficulty in opening mouth, Liánquán (CV23) and Chéngjiāng (CV24) for immobility of the tongue, drooling and difficulties in speech and swallowing were applied.

After 3 weeks of treatments, the distance of the lips at rest decreased to 1 cm. She was able to open her mouth up to 5 cm voluntarily. She was also able to move her mandible back and forth as well as to the left and right. She was able to chew soft foods. She was able to partially protrude her tongue. She was able to pronounce “a”, “i” and “u”. However, she was still not able to speak words or sentences. The modified Frenchay Dysarthria Assessment (Chinese version) was improved by 22 points and standardized swallowing assessment (Chinese version) was improved by 1 point which was yielded from the improvement in the intensity of pronunciation. After discharge, she continued to practice by herself with help from her family. Reportedly, her functions sustained at six-month follow-up by phone.

Discussion

The operculum consists of the cortical mantle covering the insula, inferior frontal gyrus, precentral gyrus, supramarginal gyrus, angular gyrus, and superior temporal gyrus. Opercular syndrome, also known as Foix-Chavany-Marie syndrome, is characterized by voluntary dysfunction of the cranial nerves V, VII, IX, X and XII [3,4]. The voluntary controls of the muscles innervated by these nerves

are provided by primary motor cortex and pyramidal tract, whereas the involuntary and emotional-related movements are provided by thalamus, hypothalamus, and extrapyramidal tract. The isolated voluntary dysfunction in FCMS is known as “autonomic-voluntary dissociation” [4].

Cerebrovascular disease is the most common etiology of FCMS, involving branches of middle cerebral artery supplying the opercular area [3]. Encephalitis, trauma, tumors, developmental perisylvian dysplasia, vasculitis, and degenerative disease are fewer common causes. Damage to the adjacent region of operculum can cause FCMS too, most of them are bilateral. Previous fMRI study in healthy subjects found that the activated areas of tapping teeth and moving tongue tip mainly located at bilateral rolandic opercula and adjacent precentral gyri, which means lesions in those regions may cause FCMS [5]. Several other reports have delineated a unilateral lesion caused FCMS [6,7]. However, some of the studies reported that bilateral disturbance of regional cerebral blood flow (rCBF) on single photon emission computed tomography (SPECT) existed in FCMS with unilateral lesion on MRI or CT [6]. This type of FCMS remains controversial and the etiology is unclear [3]. Unilateral cortical lesions coexisting with contralateral subcortical lesions in the primary motor cortex or its descending pathways can also cause FCMS [7,8]. Ohtomo reported a unilateral opercular infarction with previous infarctions in the contralateral ventral paramedian pons presenting as FCMS, but no significant change in the contralateral hemisphere was seen on SPECT [8]. However, detailed descriptions of neurological features in this literature was limited, making it difficult to interpret. In our case, the lesions mainly located in right operculum. It's unclear if the remote lesion in the left basal ganglia contributed to the presentation. Unfortunately, further imaging examinations such as SPECT and fMRI were not performed.

Oral dysphagia and severe dysarthria are the cardinal symptoms in patients with FCMS. Weakness in the movement of the lips, tongue and jaw makes food bolus preparation and coordination difficult. Therefore, patients with FCMS are also at high risk of aspiration pneumonia, which could lead to fatal consequences. There were three case reports revealing relatively good recovery from FCMS [8-10]. In one of the case reports, the impaired jaw movement and muscle weakness resolved within 2 months. Dysphagia also improved after 5 months of rehabilitation therapy, however dysarthria persisted [8]. Another case had almost complete recovery of dysphonia after 6 months' speech therapy, however, of which CT and MRI showed a right-sided thalamo-capsular hemorrhage rather than cortical opercular areas [9]. Karaca reported two FCMS cases with relatively good prognosis. Dysphagia in one of the patients resolved completely two months later. Facial diplegia was improved, leaving only slight right central facial paralysis. In another case, speech was markedly improved two months later. It is important to note that lesions on the operculum of these two cases were

relatively small [10]. A summary of recovery of 13 cases with different clinical and imaging features was presented (Table 1). Fortunately, our patient improved significantly without complications following 3 weeks of intensive multi-modal rehabilitation therapies, compared to those with major lesions.

Conclusion

In conclusion, prognosis of FCMS may be related to the size and location of the lesions according to the literatures reviewed. Patients with bilateral, large lesions located in the typical cortical opercular areas may result in poor recovery. Those with unilateral, small lesion located in subcortical areas adjacent to opercular areas may have relatively good recovery. Intensive multi-modal rehabilitation therapy may provide hope for recovery, especially in the early stage of the disease onset.

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Statement

Written informed consent was obtained from the patient

Conflict of Interests

The author does not declare any conflict of interest.

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