Single Case – General Neurology

Foix-Chavany-Marie Syndrome due to Unilateral Anterior Opercular Damage with Contralateral Infarction of Corona Radiata

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Keywords

Foix-Chavany-Marie syndrome · Anterior operculum · Pseudobulbar palsy · Automatic-voluntary dissociation · Dysarthria · Dysphagia

Abstract

Foix-Chavany-Marie syndrome (FCMS) is a rare type of pseudobulbar palsy characterized by automatic-voluntary dissociation of movements of the face, tongue, pharynx, and masticatory muscles. Most cases are due to bilateral ischemic lesions of the anterior operculum, but the syndrome has also been described after unilateral opercular damage, either isolated or associated with contralateral cortico-nuclear tract involvement. We report a patient with FCMS due to right anterior opercular lesion with contralateral infarction of the corona radiata. The patient presented with paralysis of the face and tongue with automatic and voluntary dissociation. To our knowledge, FCMS with this peculiar lesion topography has rarely been reported. We discuss the underlying mechanism with reference to MRI and diffusion tensor imaging.

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Introduction

Foix-Chavany-Marie syndrome (FCMS) causes severe speech and swallowing disabilities due to bilateral paralysis of the face, tongue, pharynx, and masticatory muscles. The paralysis is characterized by automatic-voluntary dissociation, that is, the face and tongue muscles cannot be moved voluntarily, but move involuntarily and automatically.

The first report of this syndrome was made by Magnus [1] in 1837. A 25-year-old female patient who suffered from recurrent stroke presented with paralysis of the face and tongue with automatic and voluntary dissociation. Foix et al. [2] later reported a similar autopsy case of a 60-year-old woman who presented with almost symmetrical softening foci at the bilateral anterior opercula. After that, this syndrome was established as one type of pseudobulbar paralysis.

Pseudobulbar paralysis is divided into three types: cortical, basal ganglia/capsular, and brain stem/cerebellum. Of these, FCMS is consistent with cortical type and is caused by lesions at the bilateral anterior opercula, including the lower part of the central anterior sulcus. Unlike the other two types of pseudobulbar palsy, FCMS rarely showed urinary disturbance or forced crying or laughing, and affected muscle tone is reduced.

Here, we report a case of FCMS who showed cortical-type pseudobulbar palsy that was not due to bilateral anterior opercular lesions.

Case Report

A 55-year-old right-handed woman presented with sudden onset of dysarthria, swallowing disturbance, and right hemiparalysis. She had a surgical history of cerebral hemorrhage due to arteriovenous malformation 30 years ago, but no neurological deficits were noticed afterwards, and she reported that her everyday life has subsequently been almost normal. She has hyperlipidemia and has been taking a lipid-lowering drug.

On neurological examination, she could hardly speak or swallow. She showed mild paralysis of the right side of the face and was able to open her mouth but could not put her tongue out. Gag reflex was negative, and no soft palate upward movement was detected. She showed severe oropharyngeal dysphagia. Speech evaluation showed severe anarthria and hypophonia. The right upper limb was completely paralyzed, but paralysis of the right lower limb was mild. She could stand without assistance. Mild sensory impairment was recognized in her right side. Despite strong dysarthria and dysphagia, sneezing and yawning (reflexive/automatic exercise) and natural smiles (emotional exercise) were possible. Directed swallowing was not possible, but reflexive swallowing was maintained.

Physical examination showed no abnormalities in blood pressure, pulse rhythm, cervical bruit, etc. Routine blood tests, including a complete blood count, electrolytes, glucose evaluation, renal and liver functions, vitamin B₁₂, and thyroid function, were all normal. Chest X-ray and electrocardiogram were noncontributory.

The patient underwent a 1.5-T MRI scan of the brain, which showed a trace of old bleeding in the right operculum and a new infarct in the left corona radiata (Fig. 1). Diffusion tensor tractography showed a decreased volume of the bilateral cortico-mesencephalic tract with a marked decrease of fiber volume on the left side (Fig. 2). Fractional anisotropy of diffusion tensor imaging showed an almost intact symmetrical white matter tract from the amygdala and lateral hypothalamus (Fig. 3).

Discussion

Most FCMS cases are due to bilateral ischemic lesions of the anterior opercula, but the syndrome has also been described after unilateral opercular damage, either isolated or associated with contralateral cortico-nuclear tract involvement. Our patient had an old cerebral hemorrhage in one side of the anterior opercula and a new infarct at the opposite side in the corona radiata. This indicates that FCMS is not specific for bilateral anterior operculum lesions, and lesions from the anterior operculum to the corona radiata should be considered in diagnosing FCMS.

The primary neurons of the motor cortex show somatic localization, and the anterior operculum is adjacent to neurons that project voluntary supranuclear fibers of the face, tongue, mastication, and pharyngeal muscles. On the other hand, it is thought that emotional facial movement reaches the brain stem from the amygdala and lateral hypothalamus via the inner forebrain bundle and outer longitudinal bundle; these projections are different from those of the anterior operculum. This is the anatomical basis for the automatic-voluntary dissociation seen in FCMS.

Brain MRI in our case showed a trace of old bleeding at the right anterior operculum, with a new ischemic lesion in the left corona radiata. In diffusion tensor tractography, there was a decreased volume of bilateral cortico-nuclear tract fibers, predominantly on the left side. The decrease in right cortico-nuclear fibers is rather small, and this may be the reason why her neurological symptoms were minor until a new left infarction occurred. However, when the left cortico-nuclear damage was added, resulting in bilateral disability, the symptoms of pseudobulbar palsy became clear. Nonetheless, the damage to these cortico-nuclear fibers did not include the amygdala, outer hypothalamus, inner forebrain bundle, or outer longitudinal bundle, and this may account for the preservation of automatic, involuntary movements, despite the loss of voluntary control of the facio-pharyngo-glosso-masticatory muscles.

FCMS has been previously described after unilateral opercular damage [3–9]. Kutluay's case [4] showed a left opercular lesion on MRI and bilateral opercular disturbance of regional cerebral blood flow on SPECT. The patient's symptoms were markedly improved at 6 weeks after the onset, when SPECT showed normal perfusion in the right hemisphere. Therefore, this case was not caused by "true" unilateral lesion. Pseudobulbar palsy (i.e., bilateral involvement of the corticobulbar tract) is essential for the diagnosis of FCMS, and the existence of FCMS with unilateral lesion remains controversial. Ohtomo et al. [10] reported an FCMS case caused by a new unilateral opercular lesion with an old infarction in the contralateral pons, which involved the corticobulbar tract. We suggest that contralateral small brainstem lesions should be taken into account in cases reported as unilateral opercular lesion.

FCMS secondary to bilateral corona radiata infarcts has also been reported, but such cases are very rare [11, 12]. Our case had combined lesions in one side of the operculum and in the contralateral corona radiata. Kobayashi et al. [13] reported a similar case. They described a 75-year-old woman with FCMS who had had cerebral infarction in the left middle cerebral artery territory 23 years before. At that time, she had transient right hemiparesis but no aphasia. This time, she suddenly became mute. Neurological examination revealed severe weakness in her bilateral lower face, pharynx, and tongue. Her emotional facial movement was maintained, despite disturbed volitional facial movement. CT disclosed a fresh infarction at the right corona radiata. In this patient, lesions at the left operculum and right corona radiata with the preserved right operculum gave rise to FCMS. Marchiori et al. [14] also reported a similar case with capsular hemorrhage on the left and an old ischemic lesion of the opercular cortex on the right. Based on these reports, including ours, the volitional tract of facial move-

ment from the opercular cortex and the emotional tracts from the amygdala and lateral hypothalamus appear to run separately at the corona radiata.

Interestingly, our patient showed some swallowing improvement within 3 months of symptom onset, returning to oral feeding with only minor restrictions, and a complete resolution of her dysphagia occurred within 4 months. Conversely, marked recovery of anarthria was not observed by then. This implies a difference in recovery between speech and swallowing in FCMS [15].

In conclusion, we report the clinical and neuroradiological characteristics of a patient with FCMS due to unilateral opercular and contralateral corona radiata damage. This case extends our understanding of the possible sites of lesion associated with this rare syndrome.

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Statement of Ethics

All authors hereby declare that all work described here was conducted in accordance with the Declaration of Helsinki (1964), and the submission of the manuscript for publication has been approved by Saiseikai Shonan Hiratsuka Hospital. The patient was informed of the purpose of the case presentation and written informed consent to publish her case and images was obtained from the patient.

Disclosure Statement

All authors declare that there is no conflict of interest regarding the publication of this article.

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Fig. 1. T2-weighted MRI (TR 4,000 ms, TE 90 ms) showed a trace of old bleeding in the right operculum and a new infarct in the left corona radiata. **a** Axial. **b** Coronal.

Fig. 2. Diffusion tensor tractography showed a decreased volume of the bilateral cortico-mesencephalic tract, predominantly on the left side.

Fig. 3. Fractional anisotropy of diffusion tensor imaging showed almost intact symmetrical white matter tracts from the amygdala and lateral hypothalamus (arrows).