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Foix-Chavany-Marie syndrome due to a unilateral opercular infarction

Unilateral operküler enfarkt sonucu gelişen Foix-Chavany Marie sendromu

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Abstract

Bilateral paralysis of facial, pharyngeal, lingual, and masticatory muscles with automatic-voluntary dissociation characterizes Foix-Chavany-Marie syndrome (FCMS). The syndrome is a rare neurological disorder which usually develops secondary to cerebrovascular accidents affecting bilateral opercular regions, typically as a result of subsequent strokes. A 75-year-old woman presented with acute onset of inability to speak and swallow. Neurological examination demonstrated bilateral palsy of lower motor cranial nerves with preservation of reflex and automatic movements. Magnetic resonance images of the brain showed an acute right frontal opercular infarction and old subcortical lacunar infarctions in the bilateral hemispheres. Although FCMS occurs due to bilateral opercular lesions, rarely unilateral opercular lesion accompanied by contralateral subcortical lesions may cause the typical clinical features of the syndrome.

Keywords: Opercular syndrome; Foix-Chavany-Marie syndrome, Pseudobulbar palsy

Özet

Fasiyal, farengeal, lingual ve mastikatör kasların otomatik-volanter disosiasyon ile birlikte bilateral paralizisi Foix-Chavany-Marie sendromunu (FCMS) karakterize eder. Sendrom, tipik olarak bilateral operküler bölgeleri etkileyen ardışık serebrovasküler olaylara ikincil gelişir. 75 yaşında kadın hasta akut başlangıçlı konuşma ve yutma güçlüğü nedeniyle başvurdu. Nörolojik muayenede refleks ve otomatik hareketlerin korunması ile birlikte alt kraniyal motor sinirlerin bilateral paralizisi saptandı. Beyin manyetik rezonans görüntüleme, sağ frontal operküler bölgede akut enfarkt alanı ile birlikte bilateral hemisferlerde eski subkortikal laküner enfarktlar olduğuna işaret etti. FCMS'nun bilateral operküler lezyonlara bağlı oluşmasına karşın, nadiren kontrlateral subkortikal laküner enfarktlarla birlikte unilateral operküler bir lezyon da sendromun tipik klinik özelliklerine neden olabilir.

Anahtar sözcükkler: Operküler sendrom; Foix-Chavany-Marie sendromu, Psödobulber paralizi

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Introduction

Foix-Chavany-Marie syndrome (FCMS) or the anterior opercular syndrome is characterized by anarthria or severe dysarthria and loss of voluntary control of facial, pharyngeal, lingual, and masticatory muscles with preserved reflexive and automatic-emotional functions [1, 2]. Although Magnus was the first to describe the opercular syndrome in 1837, in 1926 Foix, Chavany and Marie reintroduced the syndrome that was later named after them [3, 4]. The syndrome is a cortico-subcortical form of pseudobulbar palsy and it is caused by bilateral frontal opercular lesions [1, 2, 5, 6] However, cases of FCMS with unilateral opercular lesion have been rarely reported [2, 7, 8]. In this report, we describe the clinical and radiological findings of a case of FCMS secondary to a right opercular infarction.

Case Report

A 75-year-old female was admitted to our hospital because of sudden onset of inability to speak and swallow. Her past medical history was notable for type II diabetes mellitus and ischemic stroke that was experienced 6 years before, leaving her with mild right hemiparesis.

The patient had no fever; her pulse was 72 beats per minute and regular, blood pressure was 120/80 mmHg, and respiratory rate was 16 breaths per minute. Neurological examination revealed that the patient was alert and fully conscious; she followed commands to move her limbs indicating preserved language comprehension and limb praxis. She appeared emotionally appropriate and aware of her situation, and involuntary laughter or crying was not observed. However, she was completely mute and aphonic. Evaluation of writing, reading and calculation abilities was not possible due to the patient's baseline literacy skills. Her pupillary light reflexes, extraocular movements and ability to follow objects voluntarily with her eyes were normal. No voluntary movements of the face, jaw, lips or tongue were present. She was unable to close her eyes, open her mouth, chew, protrude her tongue, swallow or make any facial gestures on command. Her mouth was kept half-open and drooled. Her tongue was immobile in the mouth. However, she was able to spontaneously smile, yawn, and blink her eyes. The corneal reflex was preserved and the gag reflex was decreased. Voluntary chewing and swallowing could not be made, therefore the patient was fed through a nasogastric tube. On motor examination, there was a mild right hemiparesis residual from her first stroke. The plantar reflexes were flexor bilaterally. Deep tendon reflexes were hyperactive on the right side. Sensory examination to pinprick, touch and vibration and coordination tests were normal. Gait was hemiparetic, with circumduction of the right leg.

There were no abnormal findings in routine laboratory data including complete blood count, erythrocyte sedimentation rate, C-reactive protein, urine analysis, coagulation tests, and serum levels of electrolytes, liver enzymes, urea, creatinine, protein, calcium and magnesium. Fasting serum glucose was 210 mg/dL. Radiograph of chest was normal. Electrocardiogram revealed atrial fibrillation.

Brain magnetic resonance images obtained one week after symptom onset showed a lesion on the right frontal operculum that was hypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR images, there were also areas of increased signal intensity in the lesion on T1-weighted images, indicating an acute, hemorrhagic infarction. In addition, several subcortical lacunar infarctions with variable age in the

bilateral hemispheres and the left cerebral peduncle were noted (Fig. 1, 2 and 3). Hexamethylpropylenamine oxime (HMPAO) Single Photon Emission Computered Tomography (SPECT) of the brain obtained three weeks after symptom onset. During hospitalization, treatment with intravenous heparin, warfarin and insulin was prescribed. In addition, swallowing and speech therapy was performed. After one month of hospitalization, the patient was able to close her eyes and mouth voluntarily and on command and incomplete normalization of swallowing and speech was observed.

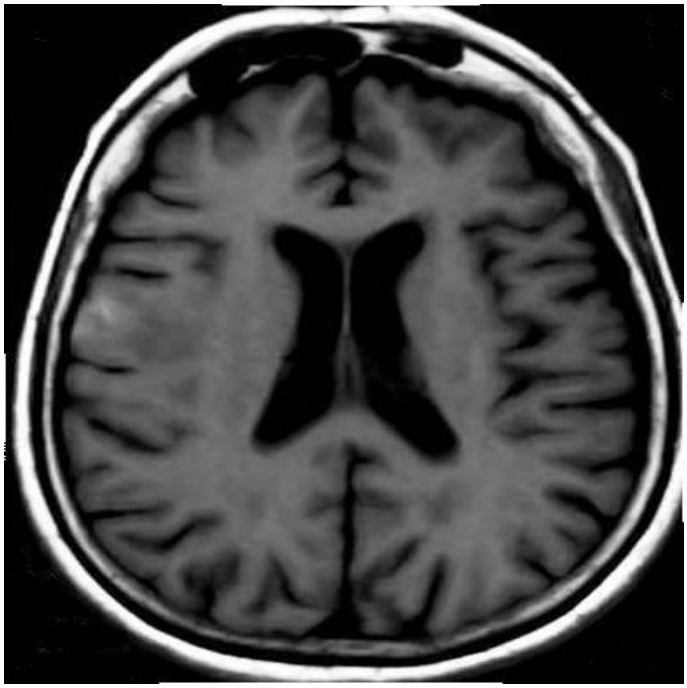

Figure 1. T1-weighted magnetic resonance image of the brain shows cerebral atrophy and decreased signal intensity and edema on the right frontal operculum. There are also areas of hyperintensity in the lesion, indicating a hemorrhagic infarction.

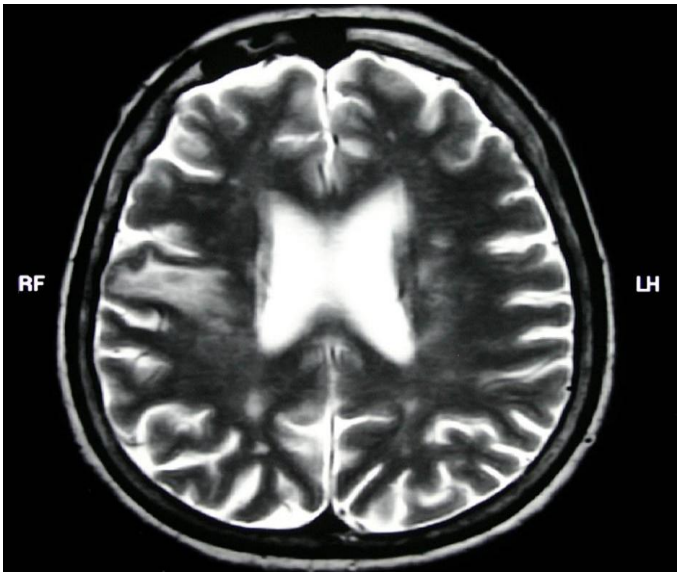

Figure 2. T2-weighted magnetic resonance image of the brain shows increased signal intensity on the right frontal operculum.

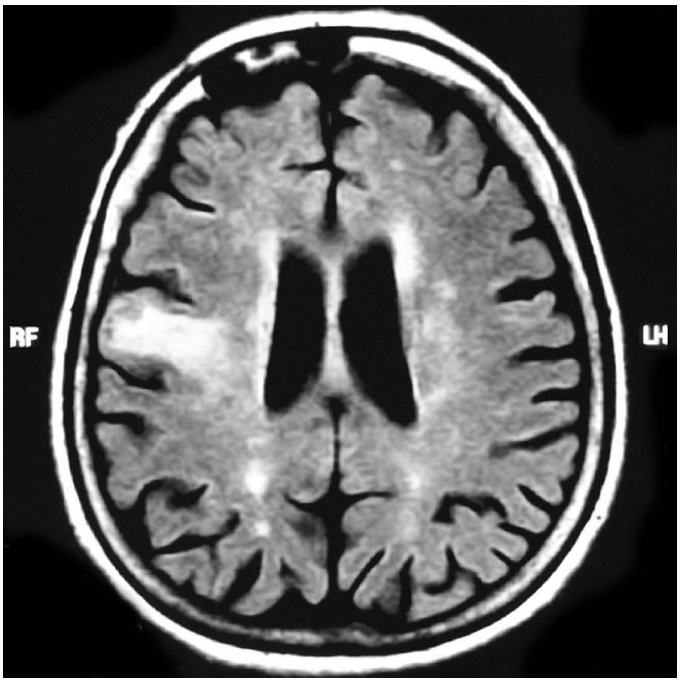

Figure 3. Fluid-attenuated inversion recovery magnetic resonance image of the brain shows increased signal intensity on the right frontal operculum and periventricular white matter and subcortical area.

Discussion

The clinical features of FCMS include anarthria or severe dysarthria, masticatory problems, facial weakness, drooling, dysphagia, a tendency for the mouth to be held half open, weakness of the tongue, absent movement of the palate and decreased or absent gag reflexes [1, 2, 5]. Patients with FCMS are unable to close the eyes, open the mouth, speak, protrude the tongue, or swallow voluntarily or on command; however, they may blink, laugh, or yawn spontaneously [2, 5] Congenital or developmental FCMS has been reported in children with bilateral perisylvian cortical dysplasia [9]. In adults, the most common cause of acquired FCMS is multiple, simultaneous or more commonly subsequent strokes, either embolic or thrombotic in nature, involving anterior operculum bilaterally [2]. Ischemic lesions of operculum or subopercular regions are caused by embolic occlusion or spasms of the slyvian branches of the middle cerebral artery [1]. Other causes of acquired FCMS include meningitis, encephalitis, toxoplasmosis, acute disseminated encephalomyelitis, multiple sclerosis, moyamoya disease, head injury, perinatal difficulties, tumors, vasculitis, neurodegenerative diseases, epileptic disorders and status epilepticus [6, 7, 9-16].

The "operculum of the insula of Reil," or operculum is formed by cortical convolutions of frontal, parietal, and temporal lobes. In FCMS, frontal part of the operculum that is composed by the posterior part of the inferior frontal convolution and the lower part of the precentral gyrus is preferentially affected [2]. There are bilateral projections originating from the precentral gyrus to the nuclei of the 5th, 7th (pars intermedia), 9th, 10th, and 12th cranial nerves. Bilateral interruption of these corticobulbar projections results in the symptoms of FCMS. The automatic-voluntary dissociation in FCMS is explained by diverse control mechanisms of voluntary and emotional movements of face, tongue and pharynx. Voluntary control of these muscles originates in the primary motor cortex, whereas emotional control of the muscles may go through pathways other than the corticonuclear tracts [17].

Cases of ‘unilateral opercular syndrome’ with unilateral cranial nerve findings due to a

unilateral opercular lesion have been rarely reported [1, 18]. More rarely, cases with 'bilateral anterior opercular syndrome' characterized by bilateral cranial nerve abnormalities and automatic-voluntary dissociation secondary to a unilateral opercular lesion have been reported [2, 7, 8]. In cases with a unilateral opercular lesion, brain SPECT may reveal disturbances on the contralateral side due to diaschisis [19]. Because brain SPECT did not demonstrate findings of contralateral disturbance of cerebral blood flow, we speculate that bilateral opercular syndrome findings were secondary to an acute right frontal opercular infarction in addition to previous contralateral subcortical infarctions in our case. Accordingly, in his review of 62 cases with anterior opercular syndrome, Weller reported two cases of FCMS due to a unilateral opercular lesion accompanied by contralateral subcortical lesions [2]. However, as brain SPECT of our patient could be obtained three weeks after symptom onset, when cerebral blood flow to the right frontal operculum had also disappeared, bilateral disturbance of cerebral blood flow may still be the cause.

FCMS must be differentiated from lower motor neuron-type bulbar palsy and subcortical pseudobulbar palsy. Bulbar palsy is characterized by the absence of jaw and gag reflexes, atrophy and fasciculations of the lower motor neuron-innervated muscles and lack of automato-voluntary dissociation. The opercular syndrome represents the cortical type of pseudobulbar palsy. There are differences between cortical and subcortical types of pseudobulbar palsy. While opercular syndrome presents with muteness, inability to move the facial, buccal, lingual and pharyngeal muscles and abolished gag reflex, pseudobulbar palsy is characterized by dysarthria, dysphagia and exaggerated gag reflex. In addition, involuntary laughter and crying, urine and bowel incontinence and dementia, which are often a part of pseudobulbar palsy, are not seen in cases of FCMS.

The management consists of speech apraxia and dysphagia treatment [20]. The prognosis of FCMS for recovery of voluntary swallowing and speech is poor except those associated with epilepsy in children. Baijens et al. reported a case of FCMS treated with neuromuscular electrical stimulation; however, although the patient returned to oral diet, control of swallow initiation did not improve [21].

In conclusion, although FCMS typically develops secondary to bilateral opercular lesions, a unilateral opercular lesion accompanied by contralateral subcortical lesions disrupting corticoulbar projections may cause the syndrome.

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