Foix-Chavany-Marie Syndrome (Anterior Opercular Syndrome) Case Report

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Abstract

Foix-Chavany-Marie syndrome is a rare cortico-subcortical suprabulbar or pseudobulbar palsy of the lower cranial nerves 5, 7, 9, 10 and 12. This syndrome consists of anarthria, bilateral volitional paresis of the facial, lingual, pharyngeal, and masticatory muscles bilaterally with preservation of the reflexive, emotional and autonomic innervations of the same muscles. We present the first case report in the Middle East of Foix-Chavany-Marie syndrome.

Keywords: Foix-Chavany-Marie syndrome, Anterior opercular syndrome, Operculum, Brain MRI.

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Introduction

Foix-Chavany-Marie syndrome (FCMS) or anterior opercular syndrome was first reported by Magnus in 1837. It is also known as facio-labio-pharyngo-glosso-masticatory paralysis with automatic-voluntary dissociation. FCMS is a rare cortico-subcortical suprabulbar or pseudobulbar palsy of the lower cranial nerves 5, 7, 9, 10 and 12. In 1926 Foix, Chavany and Marie reintroduced this specific syndrome which was later named after them. This syndrome consists of anarthria, bilateral volitional paresis of the facial, lingual, pharyngeal, and masticatory muscles bilaterally with preservation of the reflexive, emotional and autonomic innervations of the same muscles.(1)

The most classical neuro-anatomic localization of the FCMS is related to bilateral anterior opercular lesions. More recently a few cases of unilateral and bilateral cortical and subcortical (subopercular) lesions of the corticobulbar tracts have been reported.(1)

Case Report

A left-handed 72 year-old lady presented with a sudden onset slurred speech that rapidly progressed to inability to speak and swallow 18 days prior to admission to our hospital. In the first 5 days of the onset of her symptoms she was managed at a peripheral hospital as a case of stroke, then she was referred to our institution outpatient department where she was examined and FCMS was suspected, and

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consequently admitted for further evaluation. She only had history of occasional palpitations with chest tightness. On presentation she had a blood pressure of 110/70 mmHg, pulse rate was 72, regular and her temperature was 36,8°C .She was alert but couldn't speak and was able to write down her history using her left hand. There was no deficiency in comprehension, naming, reading or writing skills. Extra-ocular muscles movements, the ability to follow objects voluntarily with her eyes and, her pupillary light reflexes were normal. She was unable to whistle, puff, blow a kiss, clench her teeth, move her jaw, move or protrude her tongue, but no tongue wasting or fasciulations were seen. Bilateral facial palsy in which the lower face was affected more than the upper and the left more than the right. No movement of soft palate was observed with a very weak cough and diminished gag reflex. Corneal and jaw reflexes were normal. During physical examination we noticed that she was able to laugh in appropriate conditions and the facial diplegia improved during laughing, also spontaneous swallowing of saliva was observed but with reduced frequency, although she was unable to swallow on command. She was not emotionally labile. Motor examination revealed no weakness, and reflexes were brisk throughout with plantar flexion bilaterally. Sensory, cerebellar and gait examinations were normal. Routine laboratory tests were normal, Electrocardiogram (ECG) on admission showed sinus rhythm, but during hospital stay she developed palpitations with chest tightness and ECG showed atrial fibrillation with rapid ventricular response. Cardiology evaluation showed that she had paroxysmal atrial fibrillation and was managed accordingly, while ENT and maxillo-facial consultations were unremarkable.

Radiology investigations included brain magnetic resonance imaging (MRI) without and with contrast and diffusion weighted images (Figures 1 and 2) showed a subacute ischemic insult in the right periventricular/ opercular region and another chronic one in the left periventricular/ opercular area. Gadolinium intravenous contrast injection did not show evidence of abnormal enhancement or brain space occupying lesion.

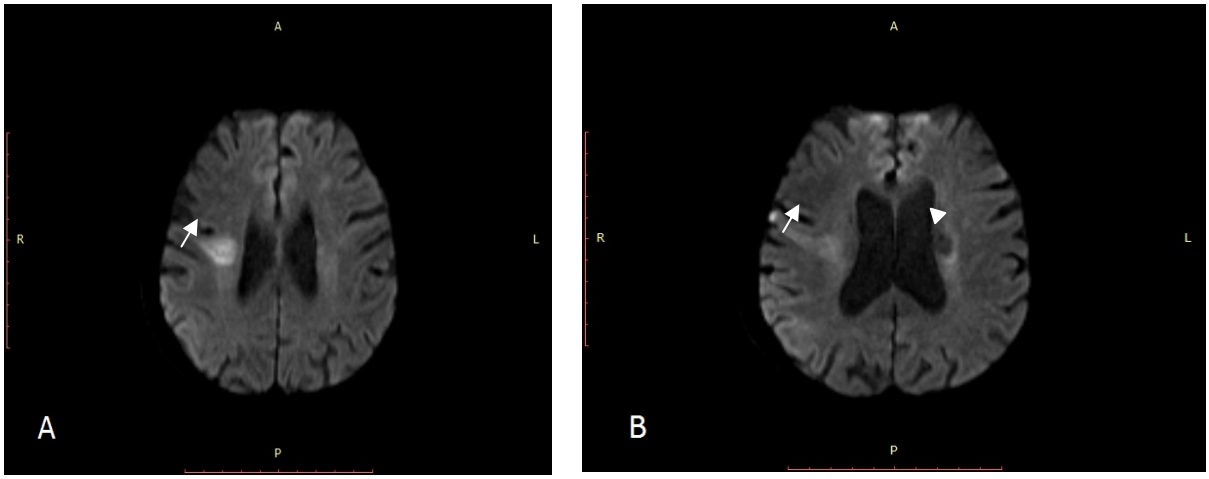

Figure 1: Axial DWI showing a subacute ischemic insult in the right periventricular (arrow) white matter (A) and a chronic infarct (arrow head) on the left side (B)

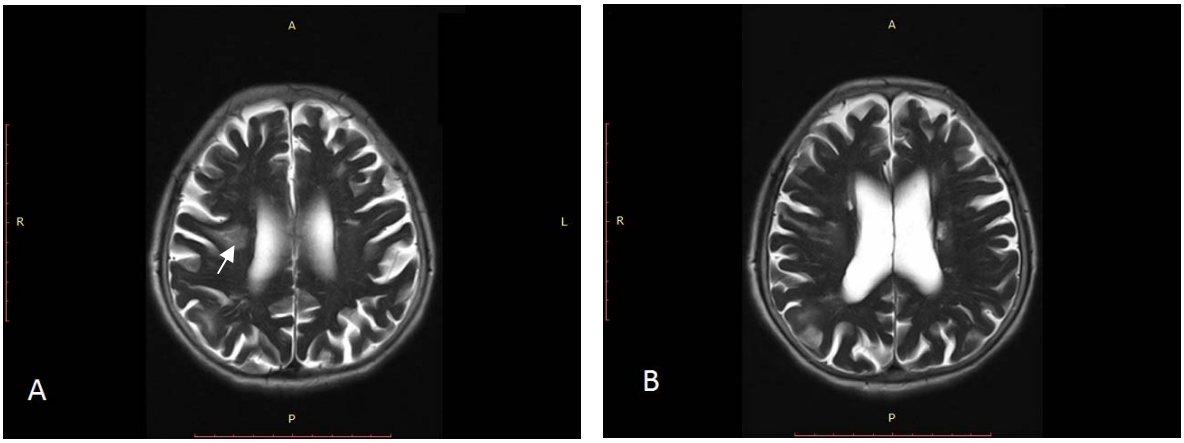

Figure 2: Axial T2 WI showing corresponding abnormal signal in the right periventricular (arrow)white matter (A) and chronic ischemic insult(arrow head) on the left side(B)

Neck magnetic resonance imaging (MRA) with i contrast showed only 30% stenosis at the origin of the right internal carotid artery. Brain MRA was normal. She was treated conservatively with antiplatelets, statin, enalpril, and speech physiotherapy. She was fed through nasogastric tube. She refused per cutaneous gastrestomy tube. Oral anticoagulants were not considered for long term therapy for social reasons. She was seen 1 month later at outpatient clinic where she showed some improvement regarding her swallowing but unfortunately she was lost for further follow up.

Discussion:

Five clinical types of FCMS were described in literature by Weller[(2)].

Volitional and emotional controls of facial and oral muscles have been described. Volitional control requires an intact primary motor cortex and its descending pyramidal pathways[(3)]. Emotional control involves deep structures, perhaps the extrapyramidal system, thalamus, and even hypothalamus.[(4)] Bilateral anterior perisylvian lesions involving the primary motor cortex within the frontal and parietal opercula can produce FCMS, which has striking neurologic manifestations. Patients with this syndrome lose voluntary control of facial, pharyngeal, lingual, masticatory, and sometimes ocular muscles. Reflexive and automatic functions of these muscles, however, are preserved. These patients may blink or yawn spontaneously, but cannot close their eyes or open their mouths on command[(3)]. Writing and comprehension are preserved in these patients as well.[(2)]

FCMS is caused by bilateral lesions involving the cortex and white matter of the anterior opercula with some rare reports of FCMS due to unilateral lesions.[(5)] Usually, unilateral acute infarcts follow a previous contralateral stroke as in our patient, while bilateral acute infarcts are seen in 1-6% of stroke patients and acute bilateral simultaneous cerebral infarcts are rarely reported in these patients.[(6,7)]

FCMS rather than "bilateral anterior opercular syndrome" is preferred, since the latter might lead us to the erroneous

assumption that the lesions as related to the clinical findings in FCMS are bilateral. First, some patients, though few, had the clinical findings of FCMS with unilateral lesions. Second, clinical and radiological correlative studies show that lesion sites other than both opercular regions can cause FCMS[8,9]. Duffau et al. reported FCMS in a patient following resection of a right insulo-opercular low grade glioma.[10]

Table 1. Clinical types of FCMS were described in literature by Weller[2]

Type I	Classical and most common form, associated with cerebrovascular disease either embolic or thrombotic with different etiologies.
Type II	A subacute form caused by central nervous system infections.
Type III	A developmental form most often related to neuronal migration disorders specially the dysgenesis of the opercular cortex or bi-opercular maldevelopment.
Type IV	A reversible form in children with epilepsy.
Type V	Associated with neurodegenerative disorders.

Table 2. Impairments distinguishing types of bulbar dys-control[3]

	Language deficits	**Bulbar voluntary movements**	**Automatic movements**	**Emotional lability**	**Gag reflex**
FCMS	_	+	_	_	Absent
Broca's aphasia	+	_	-/+*	_	Normal
Oral-buccal apraxia	_	_	-/+*	_	Normal
Pseudobulbar palsy	_	-/+	-/+	+	Heightened
Bulbar palsy syndrome	_	+	+	_	Absent

***** Usually only to complex tasks
+ Impairment present, _ impairment absent
-/+ Impairment may or may not be present

The "operculum of the insula of Reil," or operculum is formed by cortical mantle and subjacent white matter of frontal, parietal, and temporal lobes.[11] The 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]

Bilateral projections from the precentral gyrus to the nuclei of the 5th, 7th (pars intermedia), 9th, 10th, and 12th cranial nerves are present.[11] The automatic-voluntary dissociation in FCMS is explained by diverse control mechanisms of voluntary and emotional movements of face, tongue and pharynx.[11] 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.[12]

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.[2,13,14] Our patient, like all of the cases reported elsewhere, had an absent gag reflex.

The important differential diagnoses of FCMS include oral-buccal apraxia, Broca's aphasia, apraxia of speech, pseudobulbar palsy, bulbar palsy, and disorders of the peripheral nerves or of the neuromuscular junction, e.g., Guillain-Barre syndrome, botulism, and myasthenia gravis. Oral-buccal apraxia is frequently present in patients with Broca's aphasia.[15] Such patients have more difficulty in complex praxic performances[16]. (i.e., whistling, blowing, kissing). In contrast, the patients with FCMS have difficulty even in the simplest, imitative praxic tasks (open your mouth, close your eyes, protrude your tongue).[3]

Because of its abrupt onset, FCMS is often misdiagnosed as an acute brainstem stroke. However, brainstem stroke, Guillain-Barre, botulism, and myasthenia gravis can be differentiated from FCMS by the presence of abnormal eye movements, lower motor neuron signs, and absence of automato-voluntary dissociation.[3]

Prognosis for recovery of speech is poor and FCMS patients may require augmentative communication devices[3]. Although some authors reported reversal of FCMS in a patient with hydrocephalus who was treated with ventriculoperitoneal shunt.[17] Some suggest that FCMS does not usually have a poor prognosis and clinical deficits may improve in some patients.[18] In conclusion, we presented here what we believe to be the first case report in the Middle East of FCMS to draw the attention of practicing physicians to such a rare but important clinical entity due to its unique clinical anatomical correlation and its general poor prognosis especially for swallowing problems that may need long term management including insertion of PEG tube.

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متلازمة فويي – شيفاني – ماري (متلازمة الوصاد الأمامي): تقرير حالة

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الملخص

متلازمة فويي – شيفاني – ماري هي نادرة الحدوث وهي عبارة عن شلل فوق بصلي أو بصلي كاذب عادة يصيب القشرة الدماغية وما تحتها الخاصة بالأعصاب الدماغية السفلى الخامس والسابع والتاسع والعاشر والثاني عشر.

وتتكون من تعذر النطق وضعف في عضلات الوجه وعضلات اللسان وعضلات البلعوم وعضلات المضغ مع المحافظة على التغذية العصبية الانعكاسية والعاطفية وغير الإرادية لهذه العضلات. وفي هذا البحث نقدم الحالة الأولى من نوعها لهذه المتلازمة في الشرق الأوسط.

الكلمات الدالة: متلازمة فويي – شيفاي – ماري، متلازمة الوصاد الأمامي، الوصاد، تصوير الرنين المغناطيسي للدماغ.