The Classical Type of Foix-Chavany-Marie Syndrome: Assessment and Treatment of Dysphagia

I Chernev, R Petrea, M Reynolds, F Wang

Citation

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Abstract

Foix-Chavany-Marie syndrome (FCMS) or anterior opercular syndrome is a rare neurological syndrome most commonly caused by cerebrovascular accident. The anatomical substrate of the classical (clinical type A) FCMS is constituted by any combination of bilateral cortical (opercular) or subcortical (subopercular) lesion, typically as a result of subsequent strokes. FCMS is characterized by facio-labio-pharyngo-glosso-masticatory paralysis with automatic-voluntary dissociation where involuntary movements of the affected muscles are preserved. A 79 year old woman with FCMS is presented to discuss the assessment, treatment and prognosis for patient's eventual return to oral feeding.

INTRODUCTION

Foix-Chavany-Marie syndrome or anterior opercular syndrome was first reported by Magnus in 1837.[1]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. The syndrome consists of anarthria, bilateral volitional paresis of the facial, lingual, pharyngeal, and masticatory muscles bilaterally with preservation of the reflexive, emotional and automatic innervations of the same muscles.

The most classical neuro-anatomic localization of the FCMS is a bilateral anterior opercular lesion. More recently a few case reports of unilateral and bilateral cortical and subcortical (subopercular) lesions of the corticobulabar tracts have been reported. These lesion localizations suggest that any combination of cortical or subcortical lesions of the operculum or its connections on both sides of the brain can produce a syndrome indistinguishable from the classical anterior opercular syndrome.[2][3] Weller M (1993) reviewed 62 cases. He classified FCMS into five clinical types: A. the classical and most common form associated with cerebrovascular disease, B. a subacute form caused by central nervous system infections, C. a developmental form probably most often related to neuronal migration disorders, D. a reversible form in children with epilepsy, and E. a rare type associated with neurodegenerative disorders.[3] Some authors consider FCMS in children a different entity.

In the last 20 years there is increasing knowledge regarding etiology and diagnosis of FCMS however there is very little, concerning assessment, prognosis and treatment of dysphagia in this population group.

The aim of this article is to report a clinical case of classical type FCMS and discuss the assessment, treatment and prognosis for patient's eventual return to oral feeding in this specific subgroup of patients. This individual case of FCMS falls into the rare category of unilateral opercular lesion and contralateral subcortical lesion after two subsequent unilateral strokes.

CASE PRESENTATION

This 79 year-old right-handed Haitian woman with past medical history of diabetes mellitus, hypertension, microcytosis and one previous stroke was admitted to our stroke center for new onset of loss of speech, right facial droop and drooling from the right side of her mouth. Her initial neurological exam was notable for old left facial droop, new right facial droop and minimal left upper extremity weakness more in the proximal areas. Sensory exam showed bilateral lower extremity stocking and glove distribution decreased sensation, specifically for temperature and vibration, more pronounced on the left side. With regards to speech, patient was nonverbal with only occasional automatic vocalizations produced. Prior to

admission she was able to speak and ambulate without assistive devices. Initial head computed tomography (CT) showed no definite evidence of acute intracranial abnormalities, but did show old right internal capsule infarct and moderate small vessel changes. (Fig. 1) Subsequent magnetic resonance imaging (MRI) demonstrated acute left hemisphere stroke in the perisylvian area and old right subcortical infarct. (Fig. 2 and Fig. 3) Magnetic resonance arteriography (MRA) showed left middle cerebral artery (MCA) distal M1segment diffuse moderate to severe irregularities with poor visualization of the posterior division and its branches (Fig. 4).

Figure 1

Figure 1: Axial CT without contrast demonstrating regions of hypodensity seen within the right internal capsule and right basal ganglia consistent with old infarct

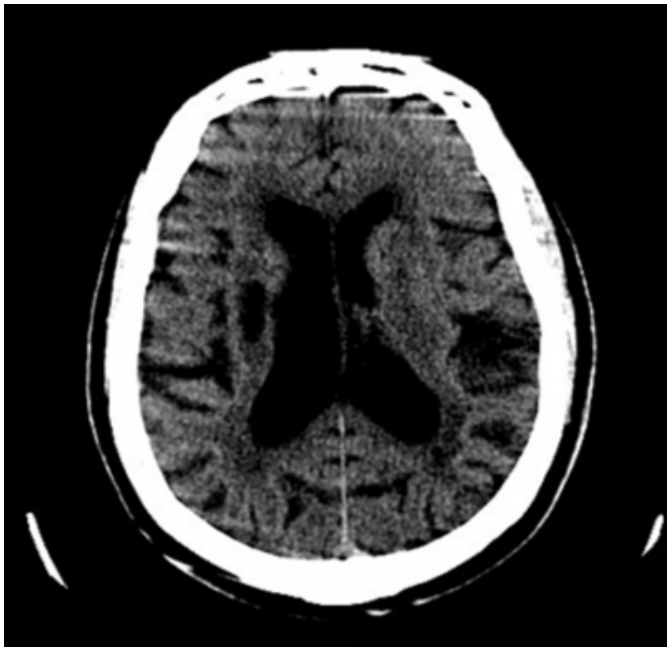


Figure 2

Figure 2: Axial DW MRI showing focus of restricted diffusion in the left posterior frontal lobe

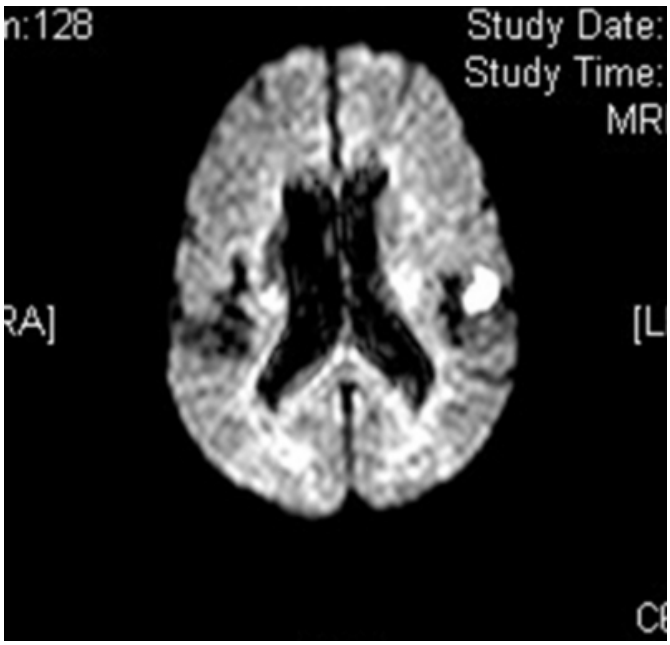


Figure 3

Figure 3: Axial FLAIR MRI showing a region of encephalomalacia in the subcortical area related to old infarct

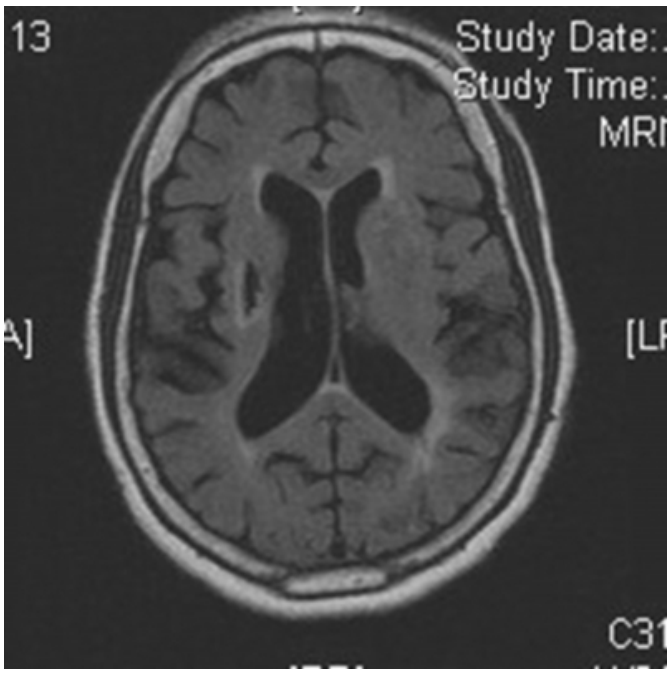
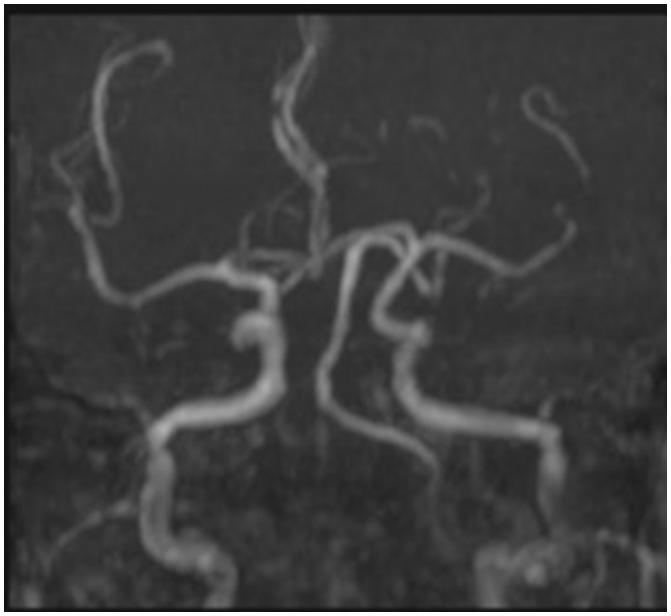


Figure 4

Figure 4: MRA at the level of circle of Willis showing irregularities in the distal M1 and lack of visualization of the posterior division of the left MCA



On hospital day (HD) 3 patient was noticed to have worsened gait with no changes in the motor strength of any of the extremities and she became completely aphonic. Repeat head CT demonstrated extension of the stroke in the same left MCA territory.

Evaluation of swallow function revealed severe oropharyngeal dysphagia and a nasogastric feeding tube was inserted for adequate nutrition.

On HD 5 patient was transferred to acute rehabilitation unit for further therapy. On admission to the rehabilitation unit the patient was non-verbal and not able to ambulate independently. Muscle strength in all four extremities was preserved with minimal deficit in the left upper extremity. Sensory exam was unchanged from admission. Deep tendon reflexes (DTR) were hyperactive but symmetrical in the lower extremities and normoactive in the upper extremities. Plantar flexor response was downgoing bilaterally.

More detailed exam in the next days showed that the patient was not able to voluntary smile, wrinkle forehead, protrude tongue, close eyes, initiate swallow. At the same time she was able to spontaneously smile, yawn, and blink her eyes. She was able to voluntary follow objects with her eyes. She was also able to voluntary follow simple one- and two-step commands with her upper and lower extremities but not able to perform multiple-step, complex tasks especially with her right hand.

Initial evaluation of speech and language revealed non-verbal patient with few audible responses characterized as spontaneous vocalizations during laughter and attempts to communicate with Haitian interpreter. Expressive language skills could not be formally assessed given patient's inability to speak, however, she demonstrated intact non-verbal communication through spontaneous use of gestures and pointing to items in the environment to communicate responses. A reliable verbal or non verbal Yes/No response could not be established secondary to non-verbal status and presence of limb and trunk apraxia. Written expression was not possible due to the upper limb apraxia as well as patient's baseline literacy skills.

Initial evaluation of the swallow function revealed mid-open mouth with anterior pooling and spillage of oral secretions at rest, secondary to inconsistent reflexive swallow and inability to voluntary contract perioral musculature. During trial of pureed solids, patient was unable to initiate labial movements to strip material from spoon or lingual movement to propel material posteriorly, when placed on anterior lingual surface. Spontaneous swallowing of saliva was observed, but with reduced frequency. Given the severity of oral dysphagia and suspected length of time required before patient would be able take sufficient nutrition, hydration and medication by mouth, a percutanoues endoscopic gastrostomy (PEG) tube was placed for nutritional maintenance.

Speech therapy interventions targeting dysphagia management focused on strengthening oral and pharyngeal musculature through oral motor exercise and therapeutic feedings. Patient was unable to participate in traditional oral motor exercises secondary to inability to volitionally initiate movement in oral structures. Visual feedback using a mirror was attempted to elicit oral movements; however, this was not effective in this patient due to paresis of oral structures during volitional movements. Therapeutic feedings were initially completed with trials of ice chips to assess integrity of pharyngeal swallow reflex. The patient's mouth remained in a mid open posture when speech-language pathologist attempted to administer ice chips, resulting in insufficient oral containment. When patient was allowed to self-administer ice chips using fingers, she was able to place ice on lingual surface and extend her head to allow ice chips to move posterioraly where pharyngeal swallow reflex was promptly triggered. Head extension was necessary as the patient was unable to facilitate anterior-posterior transit of ice chip secondary to lack of lingual movement. Once it became apparent that a pharyngeal swallow reflex was

elicited consistently during ice chip trials, a Fiberoptic Endoscopic Evaluation of Swallowing (FEES) was completed to further assess pharyngeal swallow function. FEES revealed adequate movement of epiglottis, larynx and vocal folds during reflexive swallowing of saliva. Thin liquid trials were administered via a syringe, as patient had insufficient ability to manipulate oral musculature for oral containment, bolus formation and anterior-posterior propulsion. During trials of thin liquid patient demonstrated mildly delayed onset of pharyngeal swallow, as material reached the valleculae or pyriforms prior to onset of swallow reflex. Despite delay in reflex initiation, airway protection was complete with all swallows and penetration or aspiration was not observed. Mild oral residue was observed, however, swallow reflex was triggered once residue spilled into pharynx with no penetration or aspiration observed. Based on the results of FEES, recommendations were made for continued oral trials via a syringe with the goal of gradually increasing patient's nutritional intake by mouth.

The patient was discharged from the acute rehabilitation floor on the day following the FEES, with recommendations for outpatient speech and swallow treatment.

DISCUSSION

FCMS or anterior opercular syndrome is a cortical-subcortical suprabulbar palsy of the lower cranial nerves with voluntary reflexive dissociation. In this case the FCMS was caused by new left cortical opercular lesion and old right subcortical lesion in the internal capsule after two subsequent infarcts. In his review of 62 cases with anterior opercular syndrome Weller M (1993) reported only four cases due to unilateral cortical lesion with contralateral subcortical lesion. These cases were confirmed by MRI, CT or necropsy. In our case the cortical lesion was missed by the conventional CT scan. Subsequent MRI and MRA confirmed a lesion in the region of the MCA. As stated in other previous reports, MRI is a very important image modality for diagnosis of a lesion in the opercular area.[567] The limited data in the literature indicates that the prognosis for voluntary swallow and speech recovery in the classical form of FCMS is poor.[38]Baijens LW at al. (2008) reported a single case of classical type of FCMS treated with neuromuscular electrical stimulation (NMES) three years post developing FCMS. The patient was treated with NMES simultaneous with traditional dysphagia therapy for six months. Results indicated that patient returned to oral diet with minor restrictions including altered consistency or use of compensatory strategies; however, the voluntary control of swallow initiation did not improve.[9] Improvement observed within this case can not be contributed to NMES alone, as it was completed simultaneously with traditional dysphagia therapy. Further research is necessary to support the use of NMES with this population; however, completion of a well controlled, randomized trial will be difficult given the small number of patients who are identified with this condition.

In order to maximize the patient's opportunity to return to oral feeding, timely recognition of the syndrome, early PEG placement and use of modified feeding techniques and postures may be of benefit for clinical type A FCMS patients. As the likelihood of rapid return to oral feeding is low, early gastric tube placement can avoid the adverse effects of naso-gastric tube feeding, such as nasal and esophageal irritation, erosion and infection, as well as patient agitation which may interfere with dysphagia treatment. Early initiation of liquid injection techniques and postural changes to propel the liquids further in the pharynx, avoiding the deficient musculature, is an approach which can be used to supplement nutrition to the patient and further strengthen pharyngeal musculature. FEES or videofluoroscopy prior to initiation of swallowing techniques is mandatory to confirm the intact swallowing reflex.

Conclusion: FCMS is a rare neurological syndrome not very well known by physicians. Identification of patients with neurological characteristics of this syndrome and careful physical evaluation are crucial in diagnosing the classical type of FCMS. Lesions in the opercular and subopercular area found on MRI support the diagnosis further. The awareness of this existing syndrome and timely diagnosis can lead the physician and speech and language pathologist to more appropriate assessment and therapeutic approaches, therefore improving the prognosis and the patient's chance to return to oral feeding.

References

r-0. Magnus A. Fall von Aufhebung des Willenseinflusses auf einigeHirnnerven. Arch AnatPhysiolWiss Med 1837;258-66.
r-1. Bakar M, Kirshner H.S,NiazF.Theopercular –subopercular syndrome: four cases with revew of the literature. Behavioral Neurology 11(1998) 97-103
r-2. Weller M. Anterior opercular cortex lesions cause dissociated lower cranial nerve palsies and anarthria but no aphasia: Foix-Chavany-Marie syndrome and "automatic voluntary dissociation" revisited. Journal of Neurology. 240(4):199-208, 1993.
r-3. Christen HJ,Hanefeld F, Kruse E, Imhäuser S, Ernst JP, Finkenstaedt M. Foix-Chavany-Marie (anterior operculum) syndrome in childhood: a reappraisal of Worster-Drought syndrome. Dev Med Child Neurol. 2000 Feb;42(2):122-32.

r-4. Szabo K, Gass A, RoßmanithC,Hirsch JG. HennericiMG.Diffusion- and perfusion-weighted MRI demonstrates synergistic lesions in acute ischemic Foix-Chavany-Marie Syndrome. J Neurol (2002) 249:1735-1737
r-5. Konieczny PL, Eidelman BH, Freeman WD. Teaching Video Neuroimage: Foix–Chavany–Marie syndrome. Neurology. 2008 May 20;70(21):e88.
r-6. Tohgi H, Sano M, Takahashi S, Chiba K, Matsuoka S. Bilateral anterior opercular syndrome: localizing value of SPECT and MRI. Neuroradiology. 1988;30(6):579-81.
r-7. Maw-Yuan A, On-Kee L, Yen-Lin W. Foix-Chavany-Mariesyndrome.Chinase Medical Journal (Taipei) 2001;64:540-544
r-8. Baijens LW, Speyer R, Roodenburg N, Manni JJ. The effects of neuromuscular electrical stimulation for dysphagia in opercular syndrome: a case study. Eur Arch Otorhinolaryngol. 2008 Jul;265(7):825-30. Epub 2008 Jan 8

Author Information

Ivan Chernev, MD
Department of Rehabilitation Medicine, Boston University Medical Center

Rodica E. Petrea, MD
Department of Neurology, Boston University Medical Center

Melanie S. Reynolds, MS CCC-SLP
Department of Rehabilitation Therapies, Boston University Medical Center

Feng Wang, MD
Assistant Professor, Department of Rehabilitation Medicine, Boston University Medical Center