Unmasking Bilateral Opercular Syndrome: A Rare Case of Foix–Chavany–Marie Syndrome

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Abstract

Bilateral opercular syndrome, also known as Foix–Chavany–Marie syndrome (FCMS), is a rare cortical pseudobulbar palsy caused by bilateral lesions of the opercular regions. This results in paralysis of voluntary movements of the facial, lingual, pharyngeal, and masticatory muscles, while sparing automatic and emotional functions such as laughing and crying.

We report a case of a middle-aged man in his early 60s with a prior right middle cerebral artery (MCA) stroke from the 1990s and multiple cardiovascular risk factors. He presented acutely with aphonia, inability to swallow, and complete loss of voluntary movements affecting his lips, tongue, and facial muscles bilaterally.

On neurological examination, the patient demonstrated right-sided hemiplegia accompanied by absent voluntary orofacial and tongue movements. Facial paralysis was complete, involving the entire face with no forehead sparing, yet gag and cough reflexes remained intact. Importantly, automatic and emotional movements—including smiling, laughing, and crying—were preserved. Brain MRI revealed bilateral anterior opercular infarcts, confirming the diagnosis of FCMS.

This case exemplifies the classical presentation of FCMS, characterized by cortical pseudobulbar paralysis affecting voluntary bulbar muscles (facio-labiopharyngo-glosso-masticatory paralysis) alongside preserved emotional-facial motor function, demonstrating the hallmark automatic–voluntary dissociation.

Introduction

Foix-Chavany-Marie first described this syndrome in 1926 based on his observations of patients who exhibited profound difficulty in performing voluntary movements such as speaking or chewing, yet paradoxically maintained the ability to perform involuntary or emotional movements like smiling in response to humour or yawning(Nowak et al., 2010). This "automatic-voluntary dissociation" is a hallmark feature of FCMS and reflects the underlying neuroanatomical and functional segregation between voluntary motor control (mediated by the opercular cortex and corticobulbar tracts) and involuntary emotional expressions (mediated through subcortical and limbic pathways).

The etiological landscape of FCMS is diverse. It most commonly results from bilateral ischemic strokes involving the opercular regions, often linked to vascular risk factors such as hypertension and diabetes. However, other causes include infections like encephalitis, inflammatory processes, traumatic injuries, tumours, and neurodegenerative diseases that produce bilateral damage to the opercular cortices or the corticobulbar pathways. Because these lesions are bilateral and symmetrical, they produce the characteristic pattern of bilateral facial diplegia, distinguishing FCMS from other neurological syndromes.

Clinically, patients present with anarthria or severe dysarthria, profound difficulty in chewing and swallowing, and inability to voluntarily move the facial muscles. Automatic-voluntary dissociation occurs due to the voluntary control of facial and tongue muscles is supplied by the primary motor cortex and the pyramidal tract, whereas the extrapyramidal tract, the thalamus, and the hypothalamus supply emotional and spontaneous control so these emotional and automatic functions are not lost in opercular lesions(Nowak et al., 2010).

Diagnosis is primarily clinical but is supported by neuroimaging techniques such as Magnetic Resonance Imaging (MRI), which reveal bilateral lesions in the opercular cortices. Functional imaging and electrophysiological studies may further show the extent of cortical and subcortical involvement. Given the rarity of the syndrome, awareness among clinicians is essential to avoid misdiagnosis and to guide appropriate multidisciplinary rehabilitation focusing on speech, swallowing, and motor function recovery. Recovery is usually more challenging owing to involvement of both cerebral hemispheres and neurological deficits are markedly more when compared with the unilateral hemispheric strokes.

Case presentation:

We present the case of a middle-aged right-handed man in his 60s with sudden onset of profound speech and swallowing difficulties. He also has new weakness on the right side of his body. His past medical history was significant for a previous ischemic stroke affecting the right cerebral hemisphere, which resulted in baseline weakness of the left side of his body. In addition to this, he had multiple vascular risk factors including poorly controlled hypertension, lupus anticoagulant positivity suggestive of a hypercoagulable state, a prior myocardial infarction, history of deep vein thrombosis, and a long-standing smoking habit.

The patient acutely developed aphonia, characterized by complete loss of voice, accompanied by severe dysphagia that rendered him unable to swallow even his own saliva. He was found to have complete paralysis of voluntary movements involving the orofacial muscles and tongue bilaterally. He had new weakness of the right side of his body this time. His National Institutes of Health Stroke Scale (NIHSS) score on admission was 10, indicating a moderate degree of neurological impairment.

Detailed assessment of cranial nerve function revealed complete facial paralysis affecting both upper and lower facial muscles, without sparing the forehead. Despite the severe impairment in voluntary motor control, automatic and emotional facial movements—such as spontaneous smiling, laughing in response to jokes, and tearful crying—were preserved. Reflexive functions, including gag and cough reflexes, were intact, highlighting a distinctive dissociation between voluntary and involuntary motor pathways.

Computed Tomography (CT) scan at the time depicted acute infarct of the left frontal operculum region along with old infarct and gliosis in the right cerebral hemisphere. Patient was thrombolysed in the emergency department and admitted in hyperacute stroke unit afterwards for monitory. MRI of the brain was done about 6 weeks after the presentation and it demonstrated ischemic infarcts bilaterally involving the anterior opercular regions and adjacent insular cortices. These findings confirmed the diagnosis of Foix-Chavany-Marie Syndrome (FCMS), a rare neurological condition characterized by bilateral opercular damage leading to selective loss of voluntary control over muscles innervated by the cranial nerves.

The patient received timely intravenous thrombolysis in an attempt to salvage ischemic brain tissue. However, despite aggressive medical management and intensive multidisciplinary rehabilitation focusing on speech and swallowing therapy, his dysphagia persisted, resulting in the inability to safely maintain oral nutrition. Consequently, a percutaneous endoscopic gastrostomy (PEG) tube was placed to provide long-term enteral feeding support.

Imaging:

Figure 1 shows the CT scan of the head done at presentation of the patient.

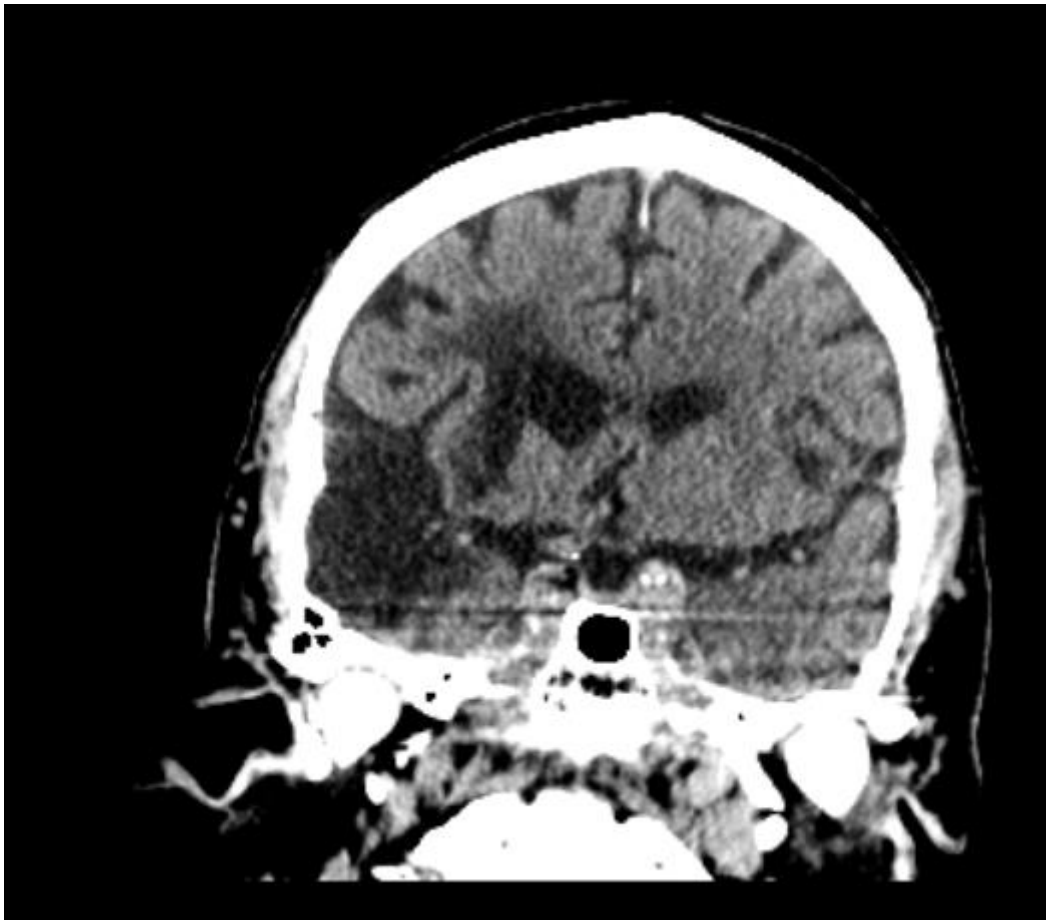

Figure 1: CT head: A small area of cortical low density shown in the left inferolateral frontal lobe just above the level of the frontal operculum. This is suggestive of an evolving cortical infarct.

MRI Head: Figure 2-4

This MRI was done about 6 weeks after the presentation of the patient.

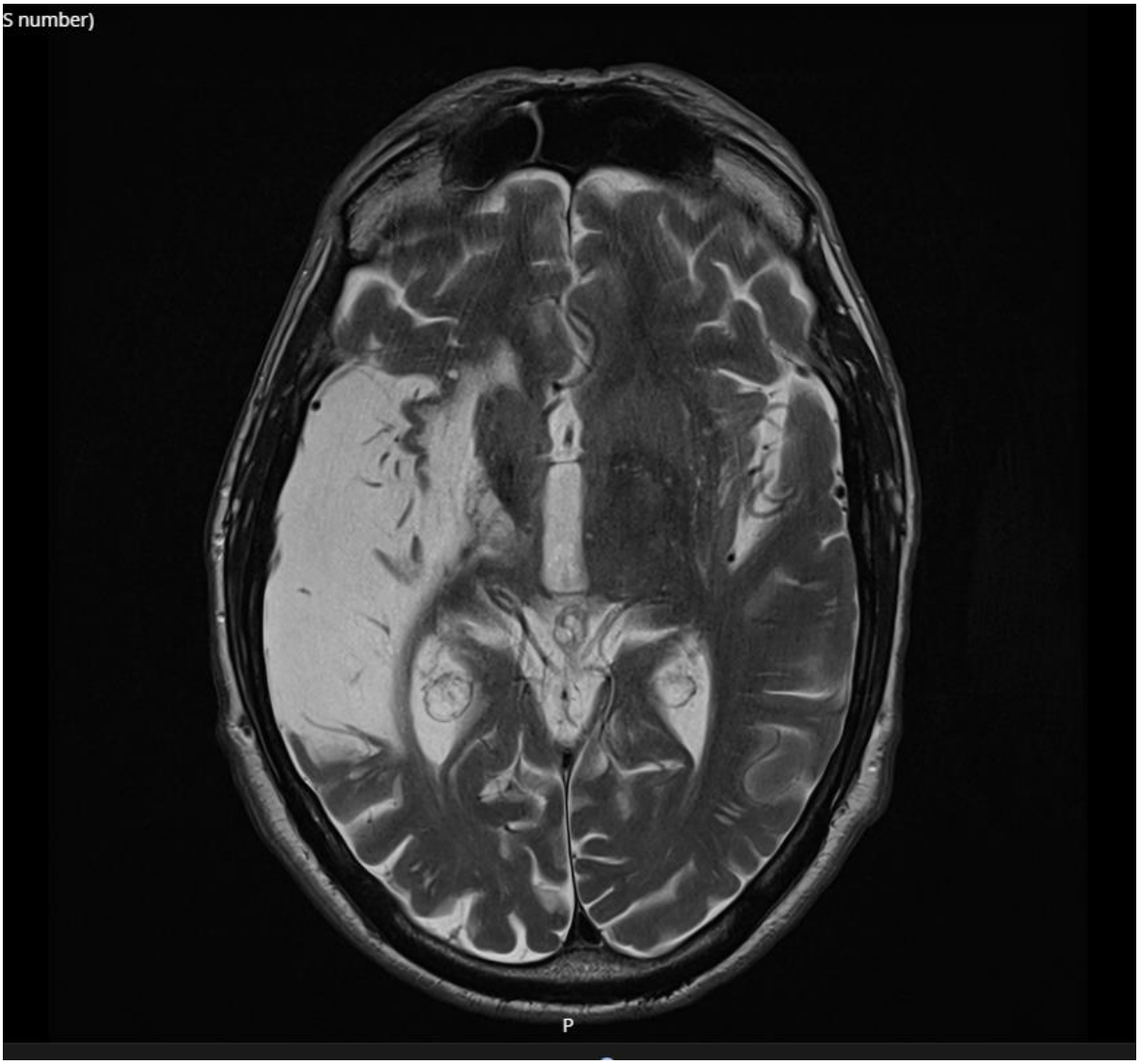

Figure 2: T2 view in the transverse plane.

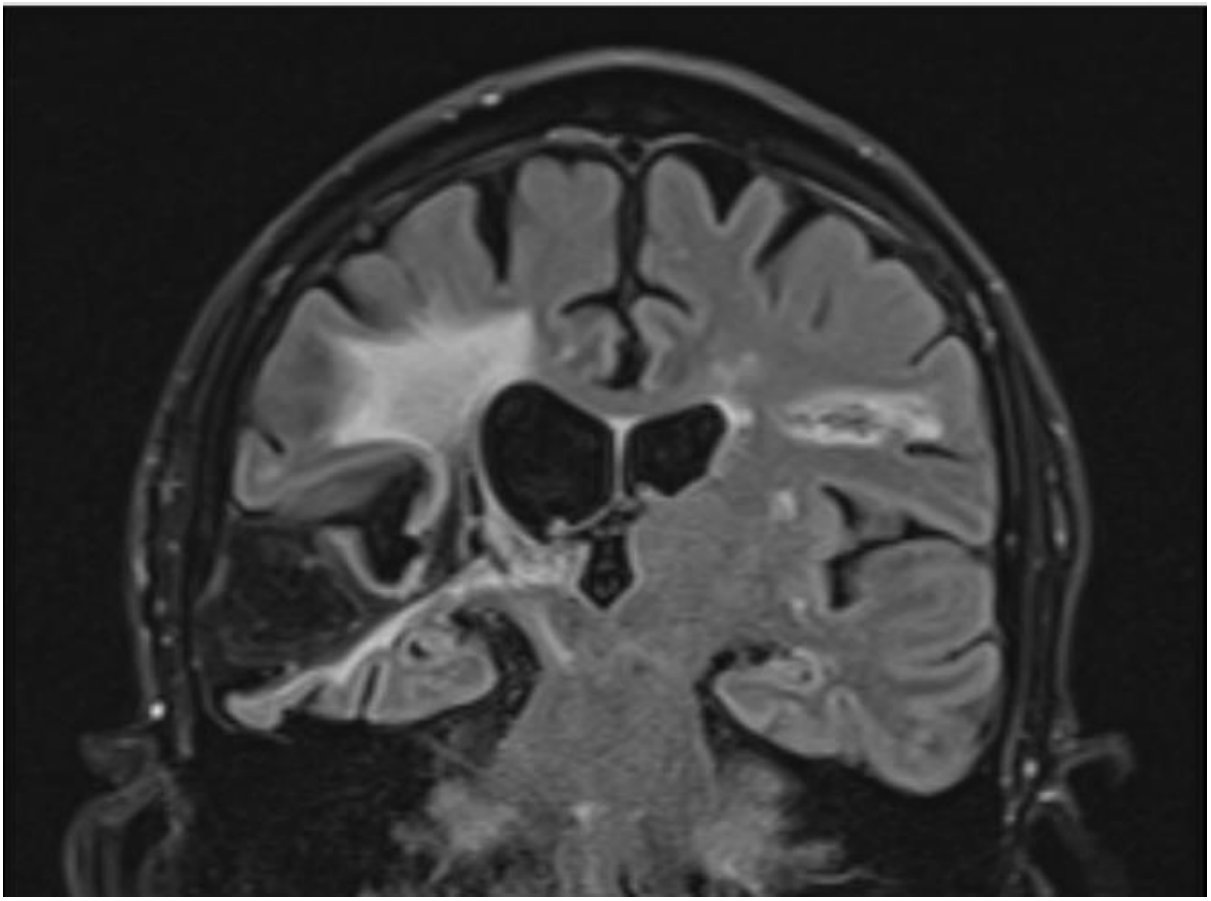

Figure 3: shows same MRI head in T2 coronal plane.

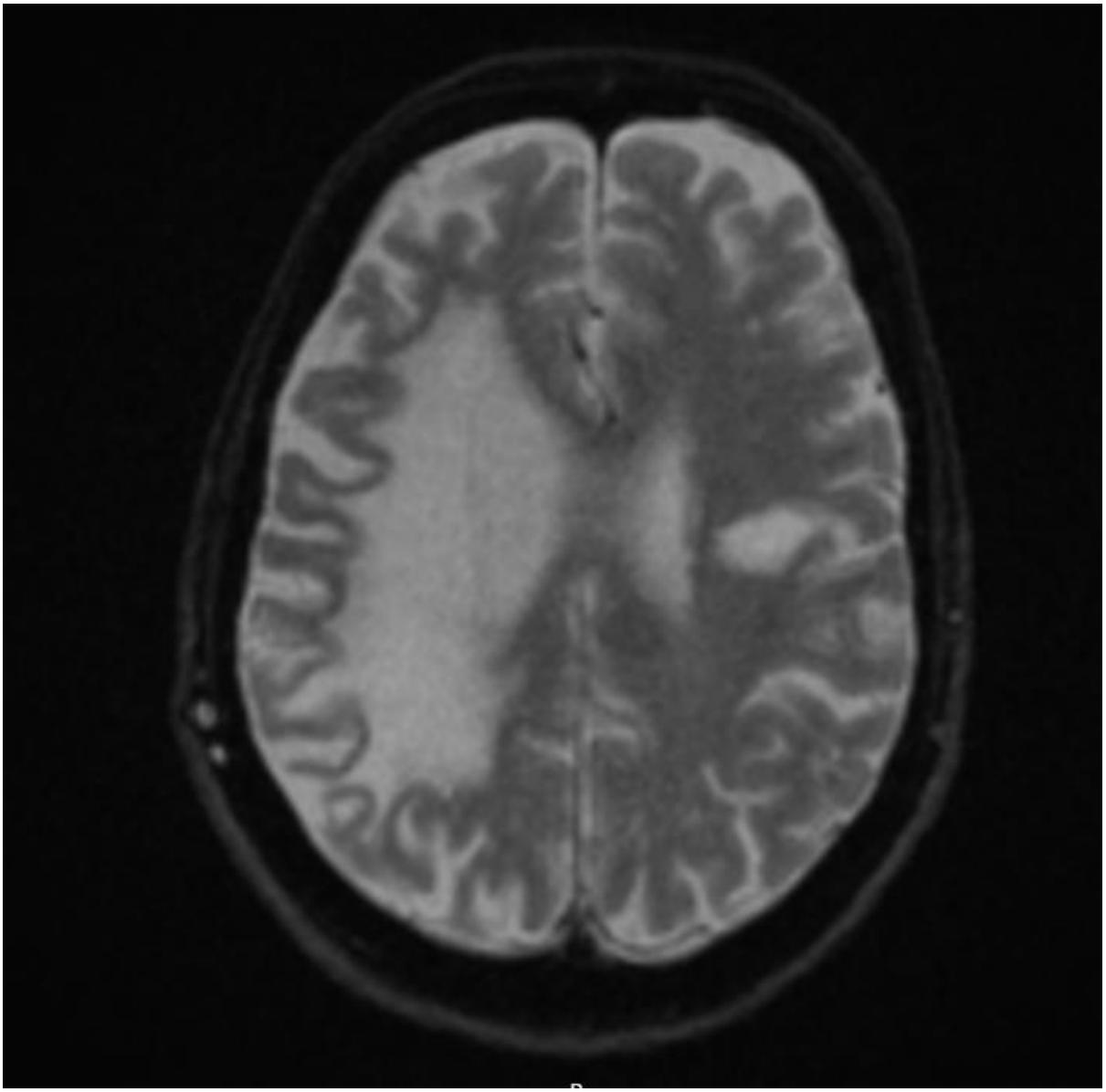

Figure 4: DWI sequence of the same MRI Head.

MRI Head report:

Well established infarct with encephalomalacia is noted in the right frontal, parietal and temporal region. Further, another old infarct is noted in the left frontal lobe. Volume loss and high signal intensity in the right cerebral peduncle likely due to previous infarct.

Scattered T2/FLAIR white matter hyperintensities most likely due to chronic small vessel disease.

MRI showed bilateral acute infarcts in anterior operculum/insula regions of both hemispheres, consistent with classic FCMS lesions.

Discussion:

Foix–Chavany–Marie Syndrome (FCMS), or bilateral operculum syndrome, remains a striking example of how bilateral cortical injury can lead to profound selective functional loss. Although the diagnosis in our patient was clinically and radiologically straightforward, the trajectory of recovery highlighted several complex challenges that distinguish FCMS from more common unilateral hemispheric strokes, particularly in terms of rehabilitation and prognosis.

One of the most striking features in this case was the near-total loss of voluntary control over the orofacial, lingual, and pharyngeal muscles, despite preservation of automatic and emotional movements(Cho et al., 2016). While this classic automatic–voluntary dissociation is well-described, what is often underappreciated is how disabling this dissociation becomes when both hemispheres are affected. In contrast to unilateral strokes, where intact contralateral pathways and cortical plasticity

often support meaningful recovery, bilateral opercular involvement eliminates this compensatory potential(Sivaprasad et al., 2021). As a result, patients like ours frequently exhibit prolonged and often irreversible deficits in swallowing, speech, and facial motor function.

In this patient, communication in the initial days following the stroke was limited to written output, which he used effectively despite the loss of speech. However, this ability was of limited utility given the complexity of care needs and the emotional frustration associated with being unable to speak or swallow. Despite engagement with specialized speech and language therapists, there was no measurable improvement in his verbal output or oropharyngeal motor control over the course of hospitalization. This resistance to conventional therapy further emphasizes the distinct cortical disconnection pattern of FCMS and highlights the limited efficacy of standard rehabilitation approaches in such cases.

From a practical standpoint, management of dysphagia in FCMS is particularly challenging. Our patient initially required a nasogastric (NG) tube to provide temporary nutritional support, but persistent and severe swallowing impairment led to the need for percutaneous endoscopic gastrostomy (PEG) tube placement. This progression is common in FCMS due to the extensive bilateral cortical damage disrupting volitional control over the swallowing mechanism, which typically does not respond well to therapy unless there is partial opercular sparing. (Richards et al., 2017)Despite initiation of comprehensive swallowing therapy, our patient showed no meaningful improvement. The Speech and Language Therapy (SALT) team contributed to this case and provided the following insights regarding the patient. According to the SALT team:

"Our patient exhibited profound oral phase impairment due to an absence of volitional oral movement to control or propel the bolus. When food or liquid was introduced into the mouth, it was not actively managed but simply flowed posteriorly by gravity. In such cases, traditional exercises or modifications to food texture (e.g., thickened fluids) are of little therapeutic value. Passive range-of-motion activities for the jaw, tongue, and facial muscles were recommended to preserve muscular flexibility. Encouragingly, the patient's reflexive pharyngeal swallow appeared intact, suggesting some ability to clear small volumes of fluid without aspiration once they reached the pharynx. Consequently, we advised cautious introduction of small amounts of water via spoon as an initial intervention. Even if aspirated, water poses a lower risk for pneumonia compared to other substances."

Beyond the swallowing limitations, emotional and psychological distress can be profound in these patients, as they retain awareness and emotional responsiveness but lack the physical means to express themselves verbally or resume basic functions such as eating. The cognitive-emotional disconnect, with intact comprehension and emotion but complete expressive limitation, makes FCMS particularly isolating.

This case also underscores the importance of early multidisciplinary involvement—including neurologists, rehabilitation specialists, nutritionists, and psychological support—to establish realistic goals and provide comprehensive care. Given the rarity of FCMS, many clinicians may not anticipate the poor prognosis for functional recovery, particularly when bilateral lesions are complete and symmetric. As seen in our patient, even with aggressive therapy, the outcomes may be limited to supportive care and adaptation to chronic disability.

In conclusion, FCMS presents not only a diagnostic challenge but a therapeutic one, as well. Bilateral opercular involvement sharply reduces the brain's capacity for compensation, distinguishing these patients from those with unilateral lesions who may show meaningful recovery(Sivaprasad et al., 2021). Long-term outcomes remain guarded, and the focus often shifts from recovery to adaptation and supportive care.

Neuromuscular stimulation as shown improved swallowing in classical FCMS. Historically, swallowing may recover before speech, but anarthria often persists long-term, especially in bilateral cortical cases. There have been a few case reports where patients swallowing function has recovered without the need of any invasive feeding tubes(Richards et al., 2017). Early intensive rehabilitation, including neurostimulation and action observation, may improve outcomes—though high-quality evidence is limited(Baijens et al., 2008).

Conclusion

This case highlights the classical yet rarely encountered presentation of Foix–Chavany–Marie Syndrome, a striking example of cortical pseudobulbar palsy resulting from bilateral opercular infarctions. Our patient developed near-total paralysis of face, tongue, lips and pharyngeal musculature, while emotional and reflexive movements remained intact—a hallmark of the syndrome. Despite early diagnosis, prompt thrombolysis, and intensive multidisciplinary rehabilitation, the functional prognosis remained poor, particularly regarding speech and swallowing recovery. Unlike patients with unilateral cerebral lesions, those with bilateral opercular involvement face a significant loss of cortical redundancy, making neuroplastic recovery far more limited(Sivaprasad et al., 2021).

The patient's inability to respond to standard language and swallowing therapy underscores the challenge of managing FCMS and the need for realistic, patient-centred goals(Chernev et al., 2009). His temporary reliance on written communication and progression from nasogastric feeding to PEG feeding further reflects the disabling nature of the condition(Baijens et al., 2008). Emotional burden, social isolation, and functional dependence are profound in FCMS, making early psychological and rehabilitative support as vital as medical stabilization.

Ultimately, this case reaffirms the importance of recognizing FCMS early, understanding its poor rehabilitative trajectory, and tailoring care to support long-term quality of life through adaptive strategies.

Learning Points

1. **Foix–Chavany–Marie Syndrome (FCMS)** results from bilateral opercular infarctions and presents with a dissociation between impaired voluntary orofacial and oropharyngeal movements and preserved reflexive/emotional responses such as smiling, crying, and the gag reflex.
2. **Diagnosis and Early Recognition** are critical. Key early features include anarthria, profound dysphagia, and bilateral lower facial weakness (without forehead sparing), with preserved cognition and written communication often serving as a temporary communication bridge.
3. **Rehabilitation is particularly limited** in FCMS compared to unilateral strokes, due to the loss of bilateral corticobulbar input. Recovery is typically slow and incomplete, especially when there is no opercular sparing.
4. **Swallowing and language therapy often show limited benefit**, and conventional compensatory strategies may not be feasible. Newer interventions such as neuromuscular electrical stimulation and task-based neurorehabilitation require further exploration.
5. **Long-term management requires a multidisciplinary approach**, often including early therapy input for nutrition, and collaboration between neurology, rehabilitation, SALT, dietetics, and mental health professionals. Clinician awareness ensures timely diagnosis, appropriate prognosis, and supportive planning.

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