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Case Report

Foix-Chavany Marie syndrome-A rare entity

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ABSTRACT
Foix-Chavany Marie syndrome (FCMS), is a rare cortical type of pseudobulbar palsy that results in paralysis of orofaciopharyngeal muscles, wwhilethe autonomic, involuntary, and reflexive functions of the above muscles are preserved. Here we present a case of 65-year-old right-handed male patient presented with sudden inability to speak and swallow, or move the tongue, along with difficulty in chewing. While his verbal and reading comprehension were intact, and he communicated through writing and gestures. Neurological examination showed preserved pupillary and corneal reflexes, normal extraocular movements, and loss of voluntary facial and tongue motor control. Early recognition and comprehensive management, including supportive therapy and addressing underlying conditions, are paramount for optimising patient outcomes.

INTRODUCTION

Foix-Chavany-Marie syndrome (FCMS), or opercular syndrome, is a rare neurological disorder characterized by bilateral paralysis of the facial, lingual, pharyngeal, and masticatory muscles with preserved involuntary and emotional movements. It is caused by bilateral lesions in the anterior opercular cortex affecting corticobulbar pathways. [1,2]

In India, FCMS remains extremely rare, with fewer than 50 cases reported in literature. Limited awareness and access to advanced neuroimaging may contribute to underdiagnosis.[3]

History

A 65-year-old right-handed male with hypertension and type 2 diabetes mellitus presented with acute-onset inability to speak, swallow, chew, or move his tongue. He remained alert and attempted to communicate using gestures and writing. There was no limb weakness, altered consciousness, or seizures.

On clinical examination

The patient was alert, oriented, and cognitively intact. Verbal and reading comprehension were preserved. He followed commands and responded through writing, indicating intact language processing despite mutism.

All cranial nerves examination is normal except CN V: Voluntary jaw movement and chewing were weak; jaw jerk reflex was brisk.CN VII: Bilateral lower facial weakness on voluntary movement (e.g., smiling on command). Involuntary expressions like yawning and laughing were preserved.CN IX & X: Palatal movement was reduced during voluntary phonation; gag reflex was preserved. Swallowing was impaired voluntarily.CN XII: Tongue was immobile on command; no atrophy or fasciculations observed.Muscle tone, bulk, and strength were normal in all limbs. Reflexes were brisk and symmetric; plantar responses were bilaterally flexor. Sensory modalities (pain, touch, proprioception) were intact.

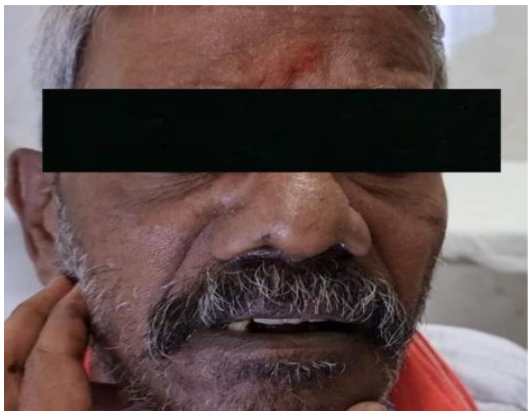
No signs of incoordination or ataxia.

The primary differential included psychiatric mutism (e.g., catatonia or conversion disorder). However, preserved comprehension, absence of affective or psychotic symptoms, and consistent neurological deficits argued against a psychiatric etiology. Moreover, the presence of involuntary emotional movements (e.g., yawning, spontaneous smiling) with loss of voluntary control is a key feature distinguishing FCMS from functional disorders.

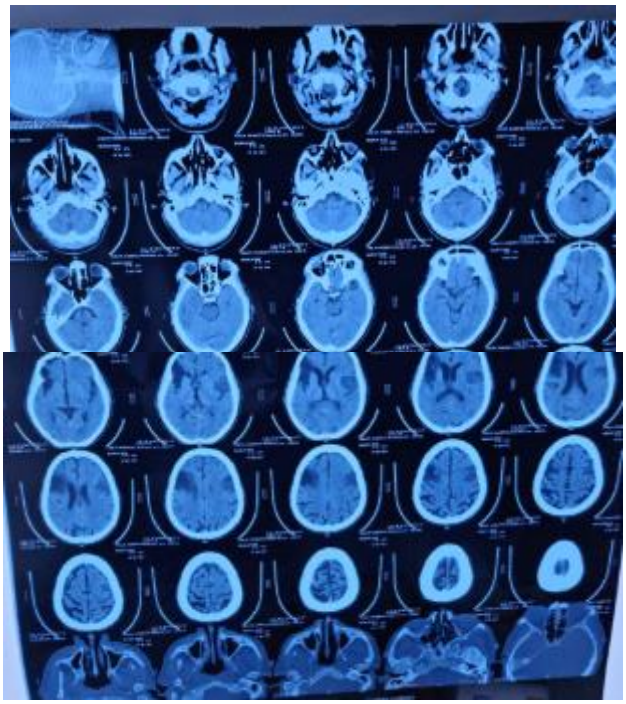
Investigations: On routine checkup, Complete blood count (CBC), Renal function tests (RFT), liver function tests (LFT) were done. Fasting blood sugars were 121 gm/dl, Post prandial blood sugars were 207 gm/dl, Triglycerides was 133 gm/dl, Low density lipoprotein was 97 gm/dl, High density lipoprotein was 32 gm/dl, Blood Urea was 24 mg/dl, Serum creatinine was 0.9 mmol/L, Serum electrolytes 136/4/98,Whole blood clotting time-normal(clotted within 20 minutes),Pseudocholinesterase was 8654

An NCCT was done to identify any organic cause"which revealed acute bilateral opercular infarcts. A diagnosis of FCMS secondary to bilateral ischemic stroke was made.

Management included antiplatelet therapy, statins, risk factor control, and supportive care. A nasogastric tube was placed for nutrition. The patient was enrolled in comprehensive speech and swallowing rehabilitation.



On-NCCT-



Impression- Bilateral symmetrical involvement of the anterior opercula extending into the insular cortices, suggestive of opercular syndrome. No evidence of acute hemorrhage, mass effect, or midline shift. Ventricles and cisterns appear normal. Findings are consistent with Foix-Chavany-Marie syndrome; clinical correlation advised for characteristic cranial muscle paralysis with automatic-voluntary dissociation.

DISCUSSION

The current mapping of the divisions of the operculum has demonstrated that stimulation causes motor and language deficits as well as somatosensory and oropharyngeal symptoms.[4] The anterior operculum contains the voluntary motor fibers for the the cranial nerve nuclei via the corticobulbar tract.[5,6] Injury to this area can cause FCMS bilateral voluntary paralysis of facial, masticatory, pharyngeal, laryngeal, and buccal muscles.[7,8] Autonomic-voluntary dissociation in FCMS is explained by the presence of alternative pathways for facial emotional expression and automatic movements, hypothesized to be mediated through the inner forebrain and outer longitudinal bundle that connect the amygdala and hypothalamus to the brainstem. [5,6]

A unilateral lesion can cause FCMS either independently or if a patient has experienced contralateral cortical-subcortical lesions prior to the recent injury.[4,9] Magnus et al reported the first case in 1837,[10] and in 1988 Starkstein et al[11] reported a patient with a lesion in the right insula and frontotemporoparietal operculum. However, these cases were reported before the advent of MRI or nuclear scanning. We found three fairly recent cases of an isolated unilateral lesion resulting in FCMS; each report described a patient who developed FCMS after unilateral damage to the pars opercularis, two due to a stroke and one due to iatro genic surgical error.[12–14] Previous case reports have described how a new unilateral lesion in combination with older lesions can result in FCMS.[15,16] Our patient's brain MRI revealed some pre vious brain damage contralateral to the acute infarction, which appears to have contributed to FCMS.

Some hypotheses as to why a contralateral lesion causes FCMS include (1) contralateral subcortical lesions interrupt the corticobulbar projections from the anterior opercular cortex to the brainstem nuclei, and (2) a unilateral representation of the motor centers dominantly and bilaterally innervates the affected muscles.[5] Martino et al used tractography to correlate the occurrence of FCMS to the resection of connections between the frontal aslant tract and arcuate fasciculus and the right pars opercularis for the first time in the litera ture.[13] A case of unilateral FCMS with right infarction of the corona radiata revealed decreased volume of bilateral cortico nuclear tract fibers in tractography; when left cortico-nuclear damage was added, it resulted in bilateral disability.[6]

Bilateral ischemic stroke is the most probable cause. Neuroimaging (MRI/CT) is essential to confirm this. Management includes stroke treatment (e.g., thrombolysis), speech and swallowing therapy, and nutritional support.

While early intervention may improve outcomes, persistent speech and swallowing deficits are common. Prompt diagnosis and tailored rehabilitation are crucial to optimize recovery and enhance the patient's quality of life.

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