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CASE REPORT

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Foix–Chavany–Marie syndrome due to unilateral opercular infarction—A case report

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Key Clinical Message

Unilateral opercular lesions can result in Foix–Chavany–Marie syndrome, which is marked by acute anarthria, automatic-voluntary movement dissociation—most notably the absence of voluntary facial and tongue movements—and a generally better prognosis. Better patient outcomes are mostly dependent on early detection, management, and rehabilitation.

Abstract

Opercular syndrome is a rare neurological disorder caused by bilateral or unilateral lesions of the operculum that result in symptoms related to speech and swallowing difficulties with dissociation of automatic-voluntary movements in affected muscles. 78-year-old female presented with acute onset dysarthria, left sided facial deviation and difficulties in chewing, speaking, and swallowing. CT head revealed ischemic changes in left frontal operculum and was diagnosed with the unilateral opercular syndrome. The case was managed according to ischemic stroke protocol. The patient was discharged after 7 days of hospital stay, with MRS 2, NIHSS 9 and secondary stroke preventive measures. At 4 months follow-up, her MRS was 1, with mild dysarthria, that could be understood, and her swallowing improved to some amount of drooling while feeding. Early recognition, treatment, and rehabilitation play important role in prompt improvement of symptoms.

KEYWORDS

aphasia, dysphasia, hypertension, opercular syndrome

1 | INTRODUCTION

The "Opercular Syndrome," also known as Foix–Chavany–Marie syndrome(FCMS), was first reported by Magnus in 1837 and was first described by Foix in 1926.[1] It is a rare cortical type of pseudobulbar palsy that results in paralysis of facial, lingual, pharyngeal, and masticatory voluntary muscles, while autonomic, involuntary, and reflexive functions of the above muscles are preserved.[2] This syndrome arises from lesions in the cortical or subcortical

regions that affect the anterior opercular area surrounding the insula, which is formed by the gyri of the frontal, temporal, and parietal lobes.[3] It is distinguished from bulbar palsy by preserved jaw jerk and pharyngeal reflex, the absence of fasciculation, atrophy, and denervation phenomenon, and the absence of pathological laughter and emotional disturbances seen in pseudobulbar palsy.

2 | CASE DESCRIPTION

A 78-year-old elderly right-handed female, an active smoker, with occasional alcohol consumer, not being treated previously for any comorbidities, past history of metal plate fixation in right leg, presented with acute onset loss of speech, inability to swallow, inability to move her tongue, difficulty in chewing foods, and deviation of the angle of the mouth to the left side with drooling of saliva within 8 h of onset, with no history and features suggestive of raised intracranial pressure, or arrhythmia.

On presentation, her blood pressure was 200/120 mmHg, normal pulse rate and respiratory rate. Her neurological examination revealed that she was aphasic, but verbal perception and comprehension was normal. Hearing and understanding were normal. She was unable to protrude her tongue beyond the lower incisor, chew, or swallow. Pupillary, corneal reflex, and extraocular movements were normal. The loss of the nasolabial fold on the right side was observed. She was unable to close her right eye on command, but she was able to blink, close her eyes completely while sleeping, and move the jaw and facial muscles with spontaneous emotional responses like laughing or crying. The upper and lower limb muscle strengths were 5/5. Pronator drift was present in the right arm. The deep tendon reflex was normoactive, and the Babinski response was absent. Extrapyramidal, coordination, and sensory examinations were normal. All other systems, including the cardiopulmonary system, were normal. Her NIHSS was calculated to be 10, and MRS 2.

The laboratory investigations including complete cell count, liver and kidney function, urinalysis, and erythrocyte sedimentation rate were normal. Her ECG showed normal sinus rhythm, and carotid doppler reported normal findings. A transthoracic echocardiogram showed concentric LVH, diastolic dysfunction, and a left ventricular ejection fraction of 60% without vegetation, thrombus, valve, or wall motion abnormalities. As the patient had metal implant, CT head was opted for imaging modality. CT head showed hypodensity in both gray and white matter of the left frontal operculum, left temporal lobe, and insular cortex (Figures 1 and 2). However, CT angiography was not performed. Her BP was initially managed

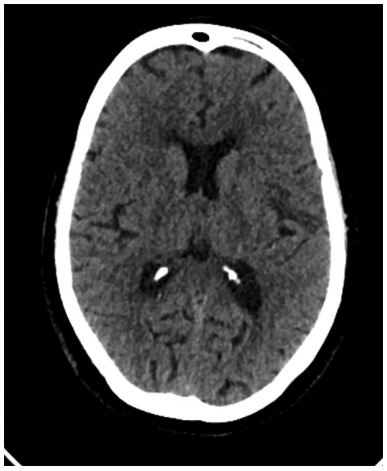

FIGURE 1 Hypodensity in both gray and white matter on the left frontal operculum.

with intravenous labetalol. No thrombolytic therapy was given, and she was managed medically with 300 mg acetylsalicylic acid stat and 75 mg daily, statin 20 mg daily, and was placed on a nasogastric tube for feeding due to lack of swallowing function. Physiotherapy and speech therapy was done. At the end of the first week, there was some improvement in swallowing and speech but no improvement in chewing.

Four months later, she had improvement in her swallowing ability and was able to tolerate oral feeds with occasional drooling. Her MRS was 1, with mild dysarthria, that could be understood.

3 | DISCUSSION

Usually, most opercular syndrome presents with bilateral corticobulbar involvement and rarely unilateral involvement. In the case of opercular syndrome (OPS), there is a distinct phenomenon of "autonomic-voluntary dissociation" characterized by selective paralysis of voluntary muscle weakness in both unilateral and bilateral involvement.[1] In our case, we observed the typical manifestation of autonomic-voluntary dissociation, with features of unilateral involvement.

A case series of opercular syndrome was reported in Nepal presenting classical features of cortical pseudobulbar palsy, with bilateral involvement in 4 and unilateral in 1 case. Interestingly, in cases of bilateral involvement, there was no improvement in swallowing function. However, in patients with unilateral involvement, there was a partial improvement in swallowing.[4] Similar improvement was seen in our patient.

A case of 55 years female with unilateral opercular damage was reported, with an additional old cerebral

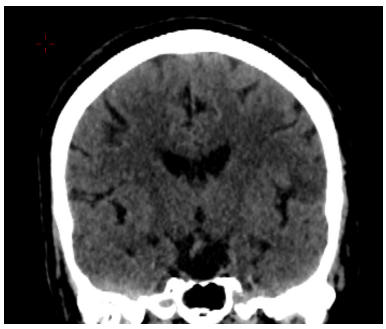

FIGURE 2 Coronal section showing hypodensity in the left opercular area.

hemorrhage on 1 side and a new infarct on another side; depicting damage of bilateral corticonuclear tract fibers, explaining the neurological disabilities.[5] Another case of 62 years male reported FCMS secondary to acute ischemic stroke, with excellent functional outcome with time.[1] Rivas et al. reported a case of unilateral FCMS, where the patient had previous brain damage leading to the development of FCMS, with gradual improvement in voluntary function.[6]

From the above case reports, we can deduce that the prognosis of opercular syndrome (OPS) varies depending on the severity of the clinical condition, underlying etiology, and timing of diagnosis. The prognosis for unilateral FCMS tends to be better than bilateral OPS.[6] In our case, there was an improvement in swallowing and speech by the end of the first week, and chewing functions by 4 months.

In the abovementioned cases, the new unilateral lesion was described in combination with previous lesions. It can be caused by various factors such as head trauma, developmental perisylvian dysplasia, multiple sclerosis, acute disseminated encephalomyelitis, vasculitis, and neurodegenerative diseases, among which ischemic stroke-related OPS is the most common classic type.[7] In our case, no predisposing brain damage or neurological disability was noted. However, based on risk factors of elderly age, active smoking status and LVH changes suggestive of long standing hypertension, the etiology is likely ischemic stroke-related OPS. But MRI could not be performed, thus, whether the FCMS was due to an independent unilateral lesion or, with the presence of a contralateral white matter lesion could not be commented on. Based on the improving status and relatively good prognosis of the case, the lesion is more likely to be unilateral in nature.

Although cases of FCMS due to unilateral lesions have been reported in the past, actual unilateral FCMS existence still remains controversial and requires further imaging and investigations.

4 | CONCLUSION

Unilateral opercular lesions even though rare can cause FCMS which is characterized by acute anarthria, and automatic-voluntary movement dissociation in the form of the absence of voluntary facial and tongue movements, which have relatively better prognosis. Early recognition, intervention, and rehabilitation are important key factors in better outcomes for the patient.

AUTHOR CONTRIBUTIONS

Pramodman Singh Yadav: Conceptualization; writing – original draft. **Leeza Shah:** Conceptualization; data curation; resources; writing – original draft; writing – review and editing. **Anusha Rayamajhi:** Writing – original draft; writing – review and editing. **Binod Mehta:** Writing – review and editing. **Min Raj Bhurtel:** Writing – original draft. **Pratik Adhikari:** Writing – original draft; writing – review and editing. **Manisha Shrestha:** Writing – original draft; writing – review and editing. **Sashank Bhattarai:** Writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

The datasets supporting the conclusions of this article are included within the article.

ETHICS STATEMENT

Not applicable.

GUARANTOR

All the authors are the guarantor of the study.

CONSENT

Written consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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