A case of Opercular-Subopercular Syndrome with Favorable Prognosis

Sir,

Bilateral voluntary facial, pharyngeal, lingual, masticatory paralysis with automatic-voluntary dissociation is the hallmark of "Foix Chavany Marie Syndrome" (FCMS), also known as Opercular syndrome.[1] Though classical FCMS results from the involvement of bilateral anterior opercular cortex, it has also been reported in a few cases with unilateral involvement, when previous insults to contralateral operculum coexist.[2] Reports of involvement of contralateral subcortical structures are limited.[3] We present a rare case of FCMS following an acute unilateral opercular stroke in a patient with a chronic asymptomatic contralateral white matter lesion suggestive of "Opercular-Subopercular syndrome."[4]

A 70-year-old right-handed male, hypertensive, chronic smoker presented with acute onset inability to speak, open mouth, chew and swallow, when he woke up from sleep one early morning. Examination revealed anarthria, decreased lingual, palatal movements with preserved gag reflex, no pooling or drooling of saliva [Video 1]. His comprehension was preserved and communicated through gestures. At the same time, he was able to open mouth effortlessly while yawning, laughing to a joke, and also had preserved reflex swallowing. He had emotional lability with preserved sensory functions, limb power, exaggerated deep tendon reflexes, and bilateral equivocal plantar. Thus, our patient had preserved automatic/reflex movements in the absence of voluntary pharyngeal, lingual, masticatory actions pointing toward opercular syndrome. His routine hemogram, glycemic status, and renal and hepatic functions were normal. Early computed tomography of brain was unremarkable. Magnetic resonance imaging (MRI) of brain showed acute infarct in the left anterior operculum and right frontal subcortical subacute infarct [Figure 1]. Cardiac evaluation including echocardiogram and 24 h Holter monitoring was normal. He improved with supportive care, nasogastric tube feeding, aspirin, statin, antihypertensives, and rehabilitative measures, but swallowing difficulty persisted. On follow-up after 4 weeks, the patient's symptoms had partially improved. He was able to open his mouth and could speak better, but mild difficulty in swallowing and occasional choking persisted; hence, nasogastric tube feeding was continued. On follow-up after 8 weeks, patient was able to feed orally, only mild dysarthria persisted.

Opercular region comprises the cortices which surround the insula, from which arise corticobulbar fibers to 5, 7, 9, 10, 12 cranial nerve nuclei. Our patient had features of suprabulbar palsy with "automatic-voluntary dissociation" characterized by preservation of automatic, reflex movements (yawning, gag reflex, reflex swallowing) with paralysis of voluntary action of cranial nerve musculature, the hallmark feature of FCMS.

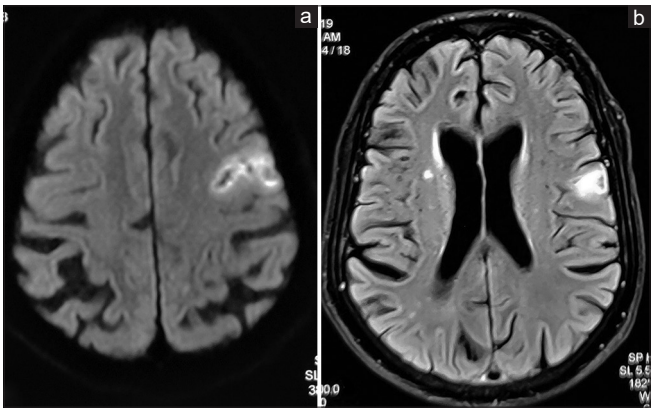

Figure 1: MRI brain (a) diffusion-weighted sequence showing diffusion restriction in the left anterior operculum. (b) FLAIR sequence showing additional right frontal subcortical subacute infarct

The voluntary-automatic dissociation in FCMS is explained by the existence of alternative pathways connecting the amygdala and hypothalamus to the brainstem. The classical FCMS has bilateral involvement of the anterior operculum but our patient had only unilateral opercular involvement with coexistent contralateral subacute infarct in right frontal subcortical region. A unilateral anterior opercular lesion with contralateral white matter lesion could present as FCMS, as it could interrupt the projections from the anterior opercula.[3]

The prognosis associated with bilateral opercular lesions tends to be poor, with the majority of patients having persistent anarthria and dysphagia. In our case of FCMS due to predominant unilateral lesion, patient had relatively better functional recovery. A study by Theys *et al*. on the neural correlates of functional recovery from FCMS highlighted the role of contralateral anterior opercular activation in recovery from anarthria.[5] Promptly detecting the automatic-voluntary dissociation clinches the diagnosis. Clinical recognition of the favorable prognosis of opercular-subopercular syndrome and early initiation of active rehabilitative measures can bring better functional outcomes.

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Conflicts of interest

There are no conflicts of interest.

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