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 action of cranial nerve musculature, the hallmark feature of FCMS.

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.

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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Heterozygous Variant of the PLP1 Gene

The proband was a 47-year-old unmarried female. She has non-consanguineous parents. The proband first visited the department of neurology at the age of 42, presenting gait disturbance and voiding difficulty. These symptoms started manifesting in her early adulthood. As a child, the proband could not run fast. In her late 30s, she complained of weakness in her legs but remained ambulatory. She underwent transverse carpal tunnel release surgery at the age of 36. She has one younger sister and two brothers: None of them have complaints associated with the mutation of the X-linked gene, manifesting with broad-spectrum symptoms, including early-onset nystagmus, hypotonia, cognitive impairment, and shortened life span. Making a final diagnosis of PMD or SPG cannot be based solely on the causative gene.

Spastic paraplegia (SPG) is a genetically heterogeneous disease entity that comprises more than 70 subtypes based on the causative gene. SPG2 mainly manifests as paraparesis with or without central nervous system (CNS) involvement. Using whole-exome sequencing (WES), we identified a novel heterozygous duplication mutation in PLP1, causing the condition.

Genetic testing revealed homozygosity for the previously identified c.520dupG variant in the PLP1 gene, confirming [Figure 1c]. DNA samples extracted from white blood cells were captured through an Illumina NovaSeq 6000 (San Diego, California, USA) and the captured libraries were sequenced by an Agilent SureSelect Human All Exon V6 (Santa Clara, California, USA). Using the exome pairs of 46, we performed commercially available WES. Briefly, the potential tests showed a central conduction defect in the left of the bilateral median nerve (>6.0 ms). Somatosensory evoked potential tests showed high intensities on the internal capsule posterior limb (Figure 1d). Brain magnetic resonance imaging performed at the age of 41 showed high intensities on the internal capsule posterior limb. As a result of the bilateral median nerve stimulation, the median nerve showed conduction velocities, with markedly prolonged terminal latency (ulnar nerves) and motor (36.6–49.5 m/s in the upper extremities) slow sensory (28.1–38.8 m/s at the distal part of median and tibial nerves) and motor (36.6–49.5 m/s in the upper extremities) slow sensory (28.1–38.8 m/s at the distal part of median and tibial nerves) and motor (36.6–49.5 m/s in the upper extremities) slow sensory (28.1–38.8 m/s at the distal part of median and tibial nerves). Nerve conduction studies (NCS) demonstrated image [Figure 1b]. Nerve conduction studies (NCS) demonstrated.

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