DOI: 10.7860/JCDR/2014/8105.4450

Case Report

Internal Medicine Section

Bi-opercular Syndrome: A Case Report and Minireview

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ABSTRACT

Opercular syndrome is a well known but neglected entity and is quite common, but it is difficult for non-neurologists to diagnose this entity because of lack of awareness. Inability to speak and swallow with dissociation of automatic voluntary movements in the affected muscles are the essential features of this syndrome. The aetiology in most of the reported cases is vascular (thrombosis or embolism) involving branches of middle cerebral artery supplying the opercular area. We are reporting a case of "bilateral opercular syndrome" caused by stroke in a young patient. He had sudden onset of bilateral facial and tongue palsy, inability to speak and swallow, but with preserved automatic functions. He was mute but verbal comprehension was normal. CT head revealed bilateral perisylvian infarcts.

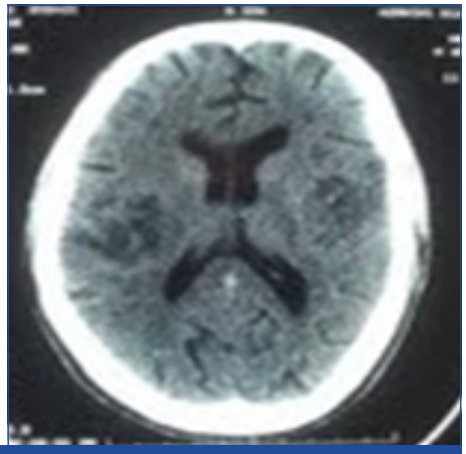
Keywords: Foix-marie-chavany, Stroke-in-young, Syndrome, Voluntary-automatic dissociation

CASE REPORT

A 40-year-old right handed healthy man had sudden onset of loss of speech and inability to swallow. He did not have any prior history of similar complaints or seizures. There was no history of headache, fever, vomiting or visual symptoms. He was neither a diabetic nor a hypertensive. He was a chronic smoker and alcoholic. On examination, his pulse was 72 per minute, regular and all peripheral pulses were well felt and BP was 130/80 mm Hg. Neurologically, he was conscious and responding to commands, but, he could not vocalize and he could not protrude his tongue or swallow on comm and [Table/Fig-1]. However, opening of mouth was noted during yawning and laughing [Table/Fig-2]. He had bilateral central facial paresis with preserved involuntary facial movements. Jaw jerk, palatal and pharyngeal reflexes were normal. Pronator drift of left upper limb and bilateral brisk deep tendon reflexes with bilateral extensor plantars were noted. Fundi and other examinations were normal. Cardiovascular, respiratory and per-abdomen examinations revealed no abnormality. CT head revealed bilateral perisylvian infarcts [Table/Fig-3]. MRI brain and angiography could not be done due to financial constraints. Complete hemogram, renal function tests, serum electrolytes and serum transaminases were normal. ECG and echocardiogram including transesophageal echo were normal. Bilateral carotid and vertebral doppler did not reveal any obstruction or stenosis. Transcranial doppler was normal. Stroke in young workup including serum homocysteine level, collagen vascular profile, APLA antibodies and thrombophilia workup were normal. Serum VDRL and HIV I and II were negative. Fasting lipid profile revealed total cholesterol of 296 mg/dl and LDL cholesterol of 138 mg/dl. He was put on Ryle's tube for feeding. He was started on antiplatelets and statins. A psychiatrist consultation was sought for advice regarding de-addiction. Speech and swallowing assessment was done by the therapist. He has shown no improvement in speech and swallowing at discharge and over 4 weeks follow up. He has been advised regular follow ups.

DISCUSSION

The patient presented here had classical features of bilateral opercular syndrome characterised by cortical pseudo-bulbar paralysis (facio-labio-pharyngo-glosso-masticatory paralysis). A salient aspect of this syndrome is voluntary – automatic dissociation, so that the patient cannot voluntarily smile or cry, not even swallow food but has preserved automatic or emotional movements of those muscles (corneal reflex, threaten reflex, laughing, crying, yawning) [1-6].



[Table/Fig-1]: Patient not able to protrude tongue on command (voluntary action) **[Table/Fig-2]:** Patient smiling on hearing a joke (automatic action) **[Table/Fig-3]:** Non-contrast CT Brain shows bilateral perisylvian hypodense lesions (infarcts)

The differential diagnosis for such a clinical presentation include non-cortical suprabulbar palsy (pseudobulbar palsy), catatonia, akinetic mutism, oral buccal apraxia, Broca's aphasia, bulbar palsy secondary to myasthenia gravis, GB syndrome and brainstem strokes. The prompt ability of the patient to follow commands involving limb muscles differentiates from catatonia or akinetic mutism. Broca's aphasia, even when associated with muteness does not involve the total loss of voluntary movements in the cranial musculature. Oral buccal apraxia also has automatic-voluntary dissociation but, patients are not completely mute. It differs from bulbar palsy by preservation of jaw jerk, pharyngeal reflex and by the absence of fasciculation, atrophy and phenomenon of denervation. In myasthenia gravis and GB syndrome, automatic-voluntary dissociation is lacking in eye and facial movements. Unlike in pseudobulbar palsy, the pathological laughter and emotional ability, partial bipyramidal syndromes with dysarthria, urinary incontinence, frontal lobe release signs are lacking in FCMS [2,6,7].

Anatomically, 'operculum' refers to the cortices which surround the insula, including the inferior frontal, pre and postcentral, supramarginal and angular (inferior parietal) gyri and superior temporal convolution with variable involvement of subjacent white matter [1,5]. The opercular syndrome was first described by Magnus in 1837 and is also known as Foix-Chavany-Marie syndrome (FCMS), facio-labio-glosso-pharyngolaryngo-brachial paralysis or cortical type of pseudobulbar paralysis [1-5]. The clinical picture appears in its most characteristic form when bilateral lesions are present as in the patient presented in this case report.

The aetiology in most of the reported cases is vascular (thrombosis or embolism) involving branches of middle cerebral artery supplying the opercular area [1], especially in adults including our patient.

In children, the most common cause is bilateral perisylvian cortical dysplasia. Less common causes include trauma, CNS infections(tubercular meningitis, Herpes simplex encephalitis, brain abscess), multiple sclerosis, neuronal migration disorders, neurodegenerative disorders, partial status epilepticus in benign epilepsy of childhood with rolandic spikes ("reversible opercular syndrome") [2-5,8].

The precise neuroanatomic basis of automatic-voluntary dissociation has remained obscure. It proves divergent corticobulbar pathways for voluntary and automatic control of craniofacial muscles [1,2]. Possible hypothesis put forward to support this are [1,7]:

A) Posed emotional expression are processed by the voluntary corticofugal system whereas spontaneous emotional expression is presumably mediated by pathways involving the basal ganglia, B)Differential functions of traditional pyramidal and extrapyramidal pathways, C) the voluntary and emotional motor impulses originate from different cortical or subcortical areas or take different courses through brain, D) Physiological tonic inhibition of the emotional pathway in the basal ganglia by the voluntary motor system of the pyramidal tracts, E) Involuntary movements are initiated in the limbic system or the prefrontal cortex but then use the same corticofugal efferent pathway as the voluntary motor system; the separation of both pathways is just caudal prior to brainstem motor nuclei, F) Involuntary motor system is phylogenetically older and may be less plastic and less amenable to learning processes than the neocortical voluntary system, G) The preservation of emotional expression in this syndrome is explained by activation of the cranial muscles by pathways arising from outside the primary motor cortex.

CONCLUSION

The typical clinical presentation of inability to speak and swallow with dissociation of automatic voluntary movements in the affected muscles are the essential features of this syndrome. Neuroradiology is conclusive. Vascular aetiology predicts a poor recovery of deficits.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Nov 22, 2013**
Date of Peer Review: **Mar 26, 2014**
Date of Acceptance: **Apr 14, 2014**
Date of Publishing: **Jun 20, 2014**