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Case Report

A rare young case of Foix Chavany Marie syndrome

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ABSTRACT

Foix Chavany Marie syndrome, also called as bilateral opercular syndrome (OPS) was first described in 1837 by Magnus and further defined by Foix, Chavany and Marie in 1926 after whom it was named so. Here, we present a case of 37-year-old female, with known recurrent CVA who presented with sudden onset aphasia, dysphagia and difficulty in opening and closing her mouth and drooling of saliva. On imaging, patient was found to have ischemia of bilateral operculum.

Keywords: Foix Chavany Marie syndrome, OPS, Automatic voluntary dissociation, Pseudobulbar palsy

INTRODUCTION

Opercular syndrome (OPS) also called as Foix-Chavany-Marie syndrome is a rarely seen cortical form of supranuclear (pseudobulbar) palsy caused by bilateral anterior opercular lesions. It is an "automatic-voluntary dissociation" syndrome characterized by paralysis of facial, lingual, pharyngeal, and masticatory voluntary muscles with preservation of autonomic, involuntary, and reflexive functions.[1] In 1926, Foix a French physicians, first described OPS, facio-labio-glosso-pharyngo-laryngo-brachial paralysis.[2] Gag reflex is usually decreased or absent.[3] The prevalence of OPS is not known till now. In classical cases, bilateral abnormality of the opercular cortex is noticed. It can present at any age and most commonly caused by multiple strokes and lesions. We report a young case of 37-year-old female with OPS presented with acute anarthria, and absence of voluntary facial and tongue movements with automatic–voluntary movement dissociation.

CASE REPORT

A 37-year-old female, home maker with history of recurrent cerebrovascular accident involving the right side presented with sudden onset aphasia, difficulty in swallowing, difficulty in closing and opening mouth, difficulty looking upwards and on left lateral side. No history of weakness of limbs, up rolling of eyes, loss of consciousness, tongue bite. No history of deviation of angle of mouth. No history of double vision, drooping of eyelids. Patient was newly diagnosed with type 2 diabetes mellitus.

On physical examination, patient was conscious, oriented with inadequate mouth closure and drooling of saliva. She was afebrile. Vitals were found to be normal.

CNS examination revealed normal higher mental functions. NIHSS score at presentation was 9 points. Examination showed bilaterally absent upward gaze with abduction lost on the left side, XIth and Xth cranial nerve palsy were present. Gag reflex could not be elicited as mouth opening was restricted. Chewing was impaired and tongue was immobile and she was not able to pass the food bolus adequately to her pharynx. Angle of mouth was slightly deviated to left side. She was able to pronounce 'uh'. Motor system examination revealed normal power and tone of left upper and lower limb, while on the right-side power of 4/5 in the upper limb and lower limb with

preserved reflexes. There was no sensory system involvement. Her Barthel index score was 90 points (totally independent). On the other hand, she was able to laugh when amused, yawn spontaneously, cry under appropriate situations. MRI brain showed old left peri-sylvian lesion with new right peri-sylvian infarct. Patient was treated with antiplatelets, statins and other neuroprotective drugs. Rehabilitative therapy was initiated by providing speech therapy, phonation therapy, exercise to oro-pharyngo-laryngeal muscles and articulation exercises. After 4 weeks, patients mouth opening was upto 5cm, she was able to protrude tongue partially and able to chew soft food to some partial extent an was able to produce words that were comprehensible to an extent. Patients progress was static at 6 month follow up. Patient improved symptomatically during the course in the hospital and got discharged.

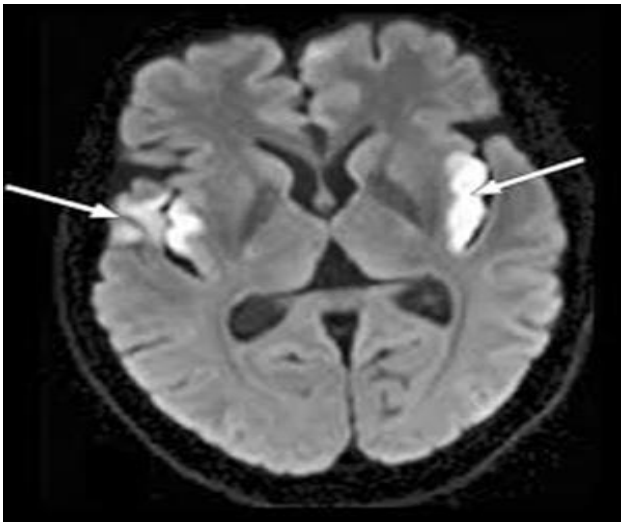

Figure 1: MRI brain T1W showing hyperintensities suggestive of old and new perisylvian infarcts.

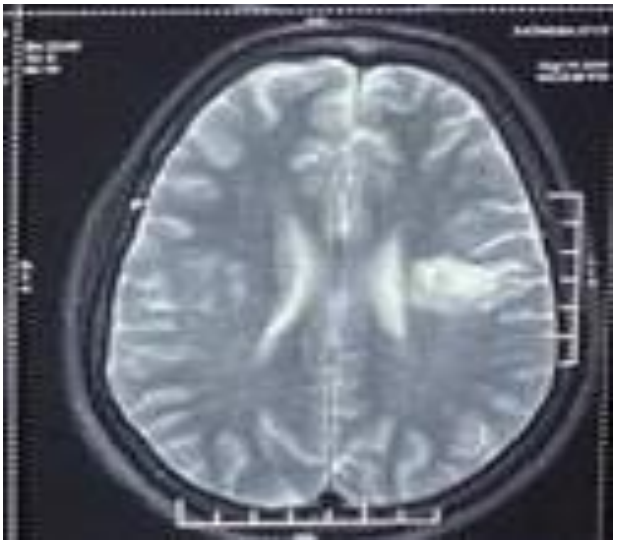

Figure 2: MRI brain T2W of old and new peri-sylvian infarcts.

DISCUSSION

Anarthria and paralysis of the facial, pharyngeal, masticatory, tongue, laryngeal, and brachial voluntary muscles are the classical features of OPS. Oral dysphagia and severe dysarthria are cardinal symptoms of FCMS. Weakness of jaw, mouth and tongue are common predisposing the patient to develop aspiration pneumonia. Hence appropriate measures to prevent it must be taken.

Cerebrovascular insult is the most common cause of FCMS. Other common causes are trauma, tumour, encephalitis, vasculitis, degenerative diseases. In children, hypoxic insults can also be the cause. 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.[4] Ischemic stroke related OPS is the most common type especially in adults.[5] Our patient had history of recurrent cerebrovascular accident involving the right side and presented with classical symptoms.

Lesions in posterior part of the inferior frontal gyrus and inferior part of precentral gyrus leads to OPS. There exist numerous connections between bilateral precentral gyrus and cranial nerves 5, 6, 9, 10, and 12. Damage of these corticobulbar tracts bilaterally leads to OPS. The voluntary muscle that controls face, tongue and pharynx are provided by primary motor cortex and pyramidal tract support, Likewise the spontaneous and emotional controls are regulated by thalamus, hypothalamus, and extrapyramidal tract.[6] This selective paralysis of voluntary muscle weakness in OPS is named as "autonomic-voluntary dissociation. The dissociation between the presence of reflexive and automatic muscle action and voluntary palsy in FCMS is explained by the existence of alternative pathways connecting the amygdala and hypothalamus to the brainstem.

Three tests to diagnose are automatic-voluntary dissociation assessment, psycholinguistic testing and neuropsychological testing. In our case, the patient presented with loss of voluntary actions with preserved involuntary skills. Treatment should be based on the onset, etiology which involves a multidisciplinary approach. We treated the patient with anti-platelets, statins and other neuroprotective drugs and speech physiotherapy. She improved symptomatically during the course in the hospital and got discharged.

CONCLUSION

The presence of acute anarthria and absence of voluntary facial and tongue movements with automatic–voluntary movement dissociation are the typical features of OPS. So FCMS should always be considered in such a setting. In our case, the probable etiology is vascular insult. Early identification of these uncommon signs and symptoms is

critical in making the diagnosis and treatment. Unilateral anterior operculum lesions can cause FCMS and can have good recovery if the lesion is small. Diagnosis should be confirmed by neuroradiology. Intensive Rehabilitation with multi modal approach is vital in such cases.

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