

Case Report

FOIX-CHAVANY-MARIE SYNDROME-A RARE PRESENTATION

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ABSTRACT

“Foix-Chavany-Marie syndrome” or “bilateral opercular syndrome” is a severe form of pseudobulbar palsy with dissociation of automatic-voluntary movements in the affected muscles. It is due to bilateral lesions of perisylvian cortex or subcortical connections and the major etiology is vascular. It is a rare neurological syndrome with only a few reported cases in the literature. Here we describe a case of Foix-Chavany-Marie syndrome due to two separate vascular events- an old infarct in the right opercular area and acute bleed in the left opercular area.

Keywords: Foix-Chavany-Marie Syndrome, Opercular

INTRODUCTION

Foix-Chavany-Marie syndrome is a rare cortical form of supranuclear palsy caused by bilateral anterior opercular lesions; in this syndrome there is an “automatic-voluntary dissociation” of motor function of lower cranial nerves. Manifestations include volitional paralysis of masticatory, facial, pharyngeal, and lingual muscles innervated by cranial nerves V, VII, IX, X, and XII, with preserved autonomic and emotional innervation of these muscles. Most of the reported cases are due to thrombosis or embolism involving branches of middle cerebral artery supplying the opercular area. Herein, we report a 64-year-old male presenting with acute onset of aphonia, and facio-labio-glosso-pharyngo-laryngeal paralysis with preservation of involuntary and emotional activities.

CASES

Our patient was a 64 years old male, right handed, manual labourer, with no formal education. He presented to us with complaints of acute onset inability to speak and difficulty in swallowing. The past medical history was remarkable for cerebrovascular accident-left hemiparesis 1 year back and then the weakness resolved completely within one month and thereafter he was not on any medications. There was no history of any other significant medical illness in the past. The family history was unremarkable. On examination, the patient was conscious and alert. He was afebrile, with pulse 68/minute and blood pressure 150/90 mms of Hg.

On neurological examination, he was aphonic; but his comprehension of spoken language was normal. The pupillary, corneal reflexes and extraocular movements were intact. He was not able to close the eyes, protrude the tongue, show the teeth, whistle, chew, and swallow; his mouth was mid-open and was unable to move the lips. The tongue was on the midline, spastic and immobile. No tongue atrophy, fibrillation, and deviation were observed. The taste sensation was intact. Palatal, laryngeal, blink, and gag reflexes, spontaneous smiling, and yawning were protected.

There was pronator drift in the right arm indicative of minimal weakness, normal power in all other limbs, normoactive deep tendon reflexes, bilateral flexor plantar responses, no sensory deficits, no meningeal or cerebellar signs. Fundus and other examinations were normal. Hematological and biochemical parameters, ECG and echocardiography were normal.

MRI scan of brain revealed acute bleed in the left posterior frontal-opercular area (5×3.2×2.3 cm), multiple old infarcts involving the right mid-frontal opercula, right inferior parieto-occipital, right anterior basal ganglia and bilateral inferior cerebellum (Figure 1 & 2). Doppler ultrasonography of carotid and vertebral arteries was unremarkable.

This patient had all the features of bilateral anterior opercular syndrome characterized by cortical pseudo-bulbar paralysis (facio-labio-pharyngo-glosso-masticatory paralysis) with automatic-voluntary dissociation

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as evident by preserved movements of facial muscles; due to two vascular events -an old ischemic lesion in one operculum and acute haemorrhage in the other.

He was given anti-cerebral edema and other supportive measures. After 2 weeks of inpatient care, he had incomprehensible sounds, but there was no improvement in chewing, and swallowing functions.

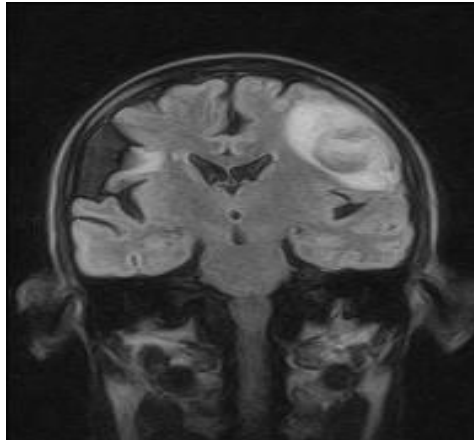


Figure 1: MRI-T1-coronal section showing acute bleed in the left operculum and old infarct in the right opercular area

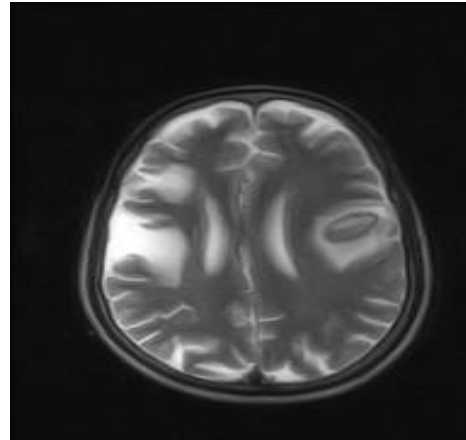


Figure 2: MRI-T2 FLAIR-axial section showing acute bleed in the left operculum and old infarct in the right opercular area

DISCUSSION

The opercular syndrome is a rare disorder due to bilateral lesions of opercular cortex surrounding the insula, which is separated by the ascending and the posterior rami of the lateral sulcus into (a) Frontal operculum formed by posterior part of the inferior frontal gyrus (i.e. pars-triangularis, pars-opercularis and even by the caudal portion of the pars-orbitalis); (b) Fronto-parietal opercula formed by the lowermost part of the precentral and postcentral gyrus and the anterior and lowermost part the inferior parietal lobule; and (c) Temporal opercula formed by the superior temporal gyrus (Bruyn *et al.*, 1969).

The syndrome was first described by Magnus (1837) and later more extensively investigated by French neurologists Charles (1882-1927) and Jean (1892-1959) with French pediatrician Julien (1899-1987) in 1926 and so the eponym Foix-Chavany-Marie syndrome (FCMS) (Foix *et al.*, 1926), facio-labio-glossopharyngo-laryngo-brachial paralysis or cortical type of pseudobulbar paralysis (Posteraro *et al.*, 1991). Further, it is classified based on the site of lesion as bilateral anterior opercular syndrome (lesion in both anterior or frontal operculum)(Mao CC et al, 1989), opercular-subopercular syndrome (lesion in opercular cortex one side and the subopercular lesion on the other side) (Bakar *et al.*, 1998), subopercular syndrome (lesions in subcortical corticobulbar projections only) (Starkstein *et al.*, 1988), unilateral anterior syndrome involving frontal operculum, and posterior syndrome involving frontoparietal operculum. Its manifestation includes central type of facial paresis, difficulty in opening of mouth, chewing the food, protrusion of tongue and inability to speak and swallow. Additional features include trismus, ageusia and pseudo-ophthalmoplegia (i.e. ptosis with weakness of conjugate gaze to opposite side and deviation of head and eyes to the side of lesion). In addition unilateral anterior syndrome is reported involving frontal operculum and presenting with contralateral and upper limb paresis and inability to speak (Piere Marie's anarthria). It differs from bulbar palsy by preservation of jaw jerk, pharyngeal reflex and by the absence of fasciculation, atrophy and phenomenon of denervation and unlike pseudobulbar palsy the pathological laughter and emotional disturbances are lacking.

The etiology in most of the reported cases is vascular (thrombosis or embolism) involving branches of middle cerebral artery supplying the opercular area. Other lesions producing the syndrome include astrocytoma, developmental bilateral perisylvian cortical dysplasia, herpes simplex encephalitis, progressive supranuclear motor system degeneration, bilateral toxoplasmosis in AIDS, and in multiple

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sclerosis. Weller *et al.*, (1993) reviewed 62 cases and classified opercular syndrome based on etiology; i.e. (a) Classical form most often related to vascular etiology; (b) Subacute form due to central nervous system infections; (c) Developmental form most often related to neuronal migration disorders; (d) Reversible form in children with epilepsy; and (e) Rare type related with neurodegenerative disorders.

Our patient had an old thromboembolic stroke involving the right opercular area and acute haemorrhagic stroke involving the left operculum. This is a very rare pattern with hardly any cases reported in the available literature.

The treatment and prognosis of opercular syndrome is related to underlying etiological factors. However, clinical improvement is usually poor. Chewing, swallowing, and speech functions do not usually recover completely. Patients with this syndrome have a significant risk for aspiration pneumonia. Therefore, during acute treatment and rehabilitation process, speech dysfunction and feeding are two most important issues. In our case, there was minimal improvement of speech, but no improvement in swallowing, and chewing functions was observed. Preserving patients from opercular syndrome can be possible via preventive treatment of repetitive stroke.

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