Bilateral Anterior Opercular Syndrome

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Abstract

Opercular syndrome, also known as Foix-Chavany-Marie syndrome, is characterised by paralysis of the facial, masticatory, pharyngeal, laryngeal, brachial and tongue muscles. It is a cortical form of pseudobulbar palsy which is commonly caused by a vascular aetiology. The clinical presentation is anarthria, weakness of voluntary muscles involving face, tongue, pharynx, larynx, and masticatory muscles. However, autonomic reflexes and emotional functions of these structures are preserved. We report a case of a 61 year old man who had difficulty in chewing, swallowing and vocalising since one and a half month, which on imaging was found to be opercular syndrome.

Introduction

Opercular syndrome (OPS) is a rare cortical type of pseudobulbar palsy that is characterized by paralysis of volitional facial, lingual, pharyngeal and masticatory muscles, with preservation of autonomic, involuntary and reflexive functions. OPS also known as facio-labio-glosso-pharyngo-laryngo-brachial paralysis, was first described by French physicians, Foix, in 1926. However, the first case was reported by Magnus (from Germany) in 1837. The lesions are usually located at the anterior part of the operculum, so it is also called anterior OPS.

Patient Presentation

An 61-year-old right-handed male presented with complaints of inability to swallow and difficulty in chewing, drooling of saliva from angle of mouth, regurgitation of food on lying down and alteration of voice since one and half months. His speech was almost incomprehensible. There was no history of vomiting, diplopia, seizures. He had no past medical history of diabetes mellitus, hypertension, coronary artery disease, benign prostate hypertrophy, tuberculosis. History of bidi smoking was present. The family history was unremarkable.

On examination, pulse was 88/min and BP was 124/84 mm Hg. On neurological assessment, he was conscious, speech was incomprehensible, but his comprehension was normal. He was able to answer questions with sign-language. Pupillary, corneal reflexes and extraocular movements were intact. The left nasolabial fold was flattened. There was right side deviation of tongue; no atrophy or fibrillations were observed. He was not able to open the mouth and protrude the tongue, nor was he able to bare the teeth, whistle, chew, or swallow on command. However, opening of mouth was seen during laughing and yawning. Gag reflex was absent and uvula was central. Taste sensation was preserved. Babinski response was absent. Extrapyramidal, coordination and sensory examinations were normal.

Laboratory investigation including liver and kidney function, complete cell count, urinalysis and erythrocyte sedimentation rate was normal. An echocardiogram showed global left ventricular hypokinesia with left ventricular ejection fraction 15-20 % and a thickened sclerosed aortic valve. Doppler ultrasonography of carotid and vertebral arteries was unremarkable. Magnetic resonance imaging (MRI) of brain indicated chronic ischemic changes in bilateral inferior frontal gyrus and insular cortex infarct with chronic ischemic changes in periventricular areas and centrum semiovale (Figure 1). He was treated with acetylsalicylic acid and hypolipidemic drugs daily and was placed on nasogastric tube feeding due to lack of swallowing function. At the end of the 3rd week, his speech comprehension had improved and he was able to open the mouth. However, there was no improvement in speech, chewing, and swallowing functions at the time of discharge.

Discussion

The most characteristic feature in this patient is that he could not voluntarily smile or swallow food but had preserved automatic activity like spontaneously laughing on hearing a joke or crying. This phenomenon often referred to as facio-labio-pharyngeal-linguo-masticatory paralysis with automatic-voluntary dissociation is seen in Opercular syndrome, also known as the Foix-Chavany-Marie syndrome (FCMS). The patient presented here had classical features of bilateral opercular syndrome characterised by cortical pseudo-bulbar paralysis.²,³

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.² This was seen specifically in the MRI in this patient.

The aetiology in most of the reported cases is vascular (thrombosis or embolism) involving branches of middle cerebral artery supplying the opercular area³ which was also the case in our patient.

The differential diagnosis for such a clinical presentation include pseudobulbar palsy, catatonia, akinetic mutism, oro-buccal apraxia, Broca’s aphasia, bulbar palsy secondary to myasthenia gravis, GB syndrome and brainstem strokes. The patient was however able to move his limb muscles on command ruling out catatonia and akinetic mutism. Broca’s aphasia, even when associated with mutism does not involve the total loss of voluntary

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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, pathological laughter and emotional lability, partial bipyramidal syndromes with dysarthria, urinary incontinence, and frontal lobe release signs are lacking in FCMS.²

The prognosis is variable, dependent on the severity of clinical disease, the type of the aetiology and the time of diagnosis.¹ However, clinical improvement is usually poor. Chewing, swallowing, and speech functions do not usually recover completely. These patients have a significant risk for aspiration pneumonia. Therefore, during acute treatment and rehabilitation process, speech dysfunction and feeding are two most important issues.¹

**Conclusion**

There is not much that can be done for the patients of opercular syndrome because as it was seen in our patient chewing, swallowing and speech functions do not improve significantly. Since the most common cause is a vascular insult, preserving patients from OPS can be possible via preventive treatment against repetitive strokes.

**References**