



## AN INTERESTING CASE OF DYSPHAGIA

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### ABSTRACT

Hughling Jackson<sup>1</sup> was the first to describe buccofacial apraxia (oral apraxia) as a case of non-protrusion of the tongue. These patients have difficulty with skilled movements of the face, lips, cheeks, tongue, pharynx and larynx. This refers to the condition in which the patient cannot perform learned skilled movements of the lips, mouth, tongue, throat in the absence of motor paralysis of concerned muscles. It is usually associated with Brocas aphasia. Our patient is an elderly female with bucco-facial apraxia with dysarthria.

**Key words: apraxia, aphasia, dysarthria.**



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## CASE DISCUSSION

A 75 yrs old female came to the medical office with complaints of difficulty in swallowing, inability to speak for three months. She was apparently normal 6 months prior, then had insidious development of difficulty in speech; difficulty in pronouncing the words initially which gradually progressed to unintelligible speech over the next three months and the patient was aphasic for 3 months. No fatiguability of speech. She was able to produce sound 'ahh' with difficulty, but unable to pronounce words; she was able to communicate with writing and sign language. She had no difficulty in understanding written and spoken language. She had difficulty in swallowing for three months for both solids and liquids, gradual in onset and progressive in nature with no diurnal variation. Patient had dribbling of liquids from her mouth on attempted swallowing and had been unable to sip using a straw. She was able to swallow freely when water was poured directly to the back of the throat. No nasal

regurgitation or using pressure maneuvers for swallowing and no pain during swallowing or difficulty in chewing. No difficulty in using upper and lower limbs. No involuntary movements. No sensory disturbances in the face, trunk and limbs. No bowel or bladder disturbances. No difficulty in co-ordination of the limbs. History of difficulty in walking due to pain and deformity of the knees and ankles was present. No difficulty in vision, hearing, ocular movements. Patient was unable to voluntarily puff her cheeks/ whistle/ blow air. But she had no difficulty in spontaneous smiling/ frowning. She also had no difficulty in closing her eyes/ wrinkling her forehead voluntarily. She had no problems with picking up taste and flavor of food. No difficulty in appreciating sensation over the tongue, or giddiness, vomiting; difficulty in voluntary movement of the tongue. No past history of systemic hypertension, diabetes mellitus, tuberculosis and epilepsy, TIA.

### ***On examination***

#### ***Nervous system examination***

1. Higher mental functions: Patient was right handed, memory was normal. No hallucinations or delusions, oriented to time, place and person, speech- aphasia.
2. Cranial nerves: V-sensory normal, muscles of mastication normal, jaw jerk normal. VII - Voluntary movements of the upper half of the face were intact. The patient was unable to voluntarily perform or mimic whistling, pouting, blowing air or purse lips. But patient was able to hold a glass of water between lips and facial expressions were intact. No emotional incontinence. IX & X & XI CN - gag reflex normal, uvula midline, effective cough was present XII CN - tongue movements were restricted in all directions, on palpation -tongue is normal in consistency, no wasting or fasciculation. All other cranial nerves were intact.
3. Motor system: normal; release reflexes were absent
4. Sensory system: normal
5. Cerebellum: normal
6. Examination of skull and spine: normal.
7. Carotids: no bruit heard

#### ***Differential diagnosis for dysarthria + dysphagia***

- 1) PSEUDOBULBAR PALSY
- 2) BULBAR PALSY
- 3) BULBAR MYASTHENIA GRAVIS
- 4) AMYOTROPHIC LATERAL SCLEROSIS

### **Investigations**

1. CT scan of the BRAIN revealed hypodensity in left gangliocapsular region and bilateral frontal white matter suggestive of lacunar infarct with microvascular ischemic changes.(MRI couldn't be done because the patient was claustrophobic.)
  2. Anti musk and anti acetyl choline receptor antibodies were negative
- Patient was being treated with anti platelet agents and gravitor prescribed by neurologist outside, but due to absence of diurnal variation and nasal regurgitation precipitated by exertion the possibility of buccofaciolingual apraxia was considered and also it was pointed out all the facial muscles were active during smiling, crying etc.

## **DISCUSSION**

### **Apraxia**

Apraxia<sup>2</sup> is defect in the ability to carry out known acts in the absence of motor weakness, sensory loss or ataxia.

Types :

- 1) ideomotor apraxia
- 2) ideational apraxia
- 3) buccofacial apraxia
- 4) limb kinetic apraxia
- 5) constructional and dressing apraxia

### **Buccofacial apraxia with aphasia**

Hagling Jackson was the first to describe buccofacial apraxia (oral apraxia) as a case of non-protrusion of the tongue. These patients have difficulty with skilled movements of the face, lips, cheeks, tongue, pharynx and larynx. Most do not improve with imitation, but often improve when using an actual object. 90% of patients with Broca's aphasia have oral apraxia. But both can also be completely dissociated, suggesting that the anatomic system that mediates facial praxis is not the same as that which mediates movements of speech but are closely associated. Oral apraxia is also often found in patients with conduction aphasia . It

can be considered that verbal and oral apraxias are points along a continuum with a common underlying mechanism. Speech requires more finer coordination than does simple facial movements. Therefore the phonologically inaccurate speech of nonfluent aphasia can be caused by an apraxic disturbance affecting speech more than oral movements.

### **Buccofacial apraxia**

This refers to the condition in which the patient cannot perform learned skilled movements of the lips, mouth, tongue, throat in the absence of motor paralysis of concerned muscles. It is usually associated with brocas aphasia.

### **Etiology**

- 1) Vascular-infarction/haemorrhage
- 2) Trauma
- 3) Metabolic – hypoglycemia (transient apraxia/ aphasia)
- 4) Tumours involving left frontal lobe
- 5) Degenerative – alzheimers disease, parkinsonism, corticobasal degeneration

Mutism following brain trauma is quite common, is usually transient, and recovery of speech is essentially the rule. Lasting total absence of speech without aphasia is highly unusual. Three such patients, two of traumatic and one

due to vascular origin showing buccofacial apraxia (BFA) and computerized tomography (CT) evidence of bilateral frontal lesions are reported. It is suggested that complete lasting mutism associated with BFA is a result of

bihemispheric lesions affecting mainly the opercular part of the inferior frontal gyrus and immediate adjacent regions.

### **Crossed buccofacial apraxia**

Buccofacial apraxia in right-handers is thought to be mediated by the left hemisphere. Ranjit B. et al<sup>3</sup> presented the case of a right-handed man without family history of sinistrality who exhibited a buccofacial apraxia following a unilateral right parietal infarction. Other findings such as intact language, visuospatial impairment, and hemi-inattention are more characteristic of right posterior lesions. Facial apraxia, as well as aphasia, has been associated with lesions in the cerebral hemisphere contralateral to the dominant hand. Nitta N<sup>4</sup>, Shiino A, Watanabe described a patient with severe facial apraxia caused by contusion in the right frontal operculum, premotor area and primary motor cortex ipsilateral to the dominant hand. The patient had no aphasia or limb apraxia. Magnetic resonance images of the brain reveal no abnormality of the hemisphere contralateral to the dominant hand. Thus, in some individuals, facial praxis is controlled by the hemisphere non-dominant for both handedness and language.

### **Foix-chavany-marie syndrome<sup>5</sup>**

Cortical- subcortical suprabulbar/pseudobulbar palsy of lowercranial nerves, characterised by severe dysarthria and dysphagia asso With B/L central facio-glosso-masticatory paralysis with prominent automatic-voluntary dissociation(AVD) in which involuntary movements of affected muscles are preserved . Patients present with acute onset bilateral paresis of facial,lingual,pharyngeal and masticatory muscles(V,VII,IX,X & XII cranial nerves). Reflexive,emotional and automatic innervations of these muscles are preserved and smiling,crying ,yawning under natural circumstances possible. Facial appearance atonic .mouth half open, speech disturbance severe – mute. Chewing and swallowing (oral stage) is impaired severely, but swallowing reflex is adequate if bolus is delivered directly

into pharynx; the tongue is almost immobile and jaw jerk exaggerated.

### **Orofacial apraxia in motor neuron disease**

Amyotrophic lateral sclerosis (ALS) is typically characterized by progressive degeneration of the motor neurons. Recent studies, including neuropsychological testing, imaging and neuropathology examination has demonstrated that frontal lobe involvement is common, although with variable clinical expression. Diffusion-tensor MRI shows widespread involvement, including the temporal, parietal and occipital lobes. Apraxic syndromes have been very rarely described in ALS. A 78-year-old right-handed woman<sup>6,7,8</sup> was referred with a history of slowly progressive slurred speech followed by mild difficulty in swallowing, which had started 3 years before evaluation. patient presented with dysarthria but she progressed to prominent OFA that started asymmetrically and evolved to affect both sides. Later on, she showed signs of dementia, establishing the probable diagnosis of MND-FTD. Although this association has not been mentioned in recent years, in 1894 Strumpell described the association of pseudobulbar syndrome and apraxia (the patient did not sustain eye closure or move the eyes on command, but had normal spontaneous movements). Dereux reported Parinaud's<sup>7</sup> syndrome with normal involuntary movements in a patient with ALS. Lapresle et al. reviewed the previous reports and described 2 cases of facial automatic-voluntary motor dissociation in ALS patients. Abe et al. reported 3 patients with ALS who had difficulty voluntarily opening or closing their eyelids, associated with disturbance in the executive function. Pinto et al. reported an ALS patient with respiratory apraxia, without clinical dementia or other apraxic syndromes. Duffy et al. reported progressive apraxia of speech as a sign of ALS, which consisted of impaired capacity to plan commands that direct speech movements.

## **CONCLUSION**

We present this case report because this condition even though prevalent is either under-diagnosed or misdiagnosed and treated for a long time without any improvement. Our patient improved with physiotherapy and speech therapy.

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