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Dysarthria and Dysphagia in Foix-Chavany-Marie Syndrome

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ABSTRACT Foix-Chavany-Marie Syndrome (FCMS) is a severe form of pseudobulbar palsy, characterised by facio-labio-pharyngo-glosso-masticatory paralysis with automatic voluntary dissociation where involuntary movements of the affected muscles are preserved. It is due to bilateral lesions of perisylvian cortex or subcortical connection and the major etiology is vascular. We report a case of 37 years male with Cerebrovascular accident leading to Foix-Chavany-Marie Syndrome . He was reported to our department with difficulty in speaking and swallowing. Detailed evaluation had been carried out in our department. The result showed severe oral weakness with drooling of saliva and dysphagia. His comprehension was adequate. He expressed his needs through pointing and gestures. Foix-Chavany-Marie Syndrome is a well known entity among Neurologist, but it is difficult for Speech Language Pathologist to diagnose the entity because of the lack of awareness. This study is done to create awareness among Speech Language Pathologist.

KEYWORDS : Foix-Chavany-Marie Syndrome, Cerebrovascular accident, Dysarthria, Dysphagia

Introduction:

Foix-Chavany-Marie Syndrome (FCMS) is also known as Opercular Syndrome. It was first reported by Magnus in 1837. In 1926 Foix, Chavany and Marie reintroduced this specific syndrome which was later named after them. Operculum represents the cortex surrounding the insula and can be divided into three parts; the frontal operculum, the frontoparietal operculum and the temporal operculum, with variable involvement of the subcortical white matter. Bilateral lesions in these areas clinically mimic a pseudobulbar palsy in the distribution of the V, VII, IX, X and XII cranial nerve. Volitional control of facial, oral and pharyngeal musculature demands intact motor cortices and pyramidal pathways, pathology in this areas leads to a selective palsy of voluntary use of these muscle groups. Emotional or spontaneous use of these muscle groups, however require intact extrapyramidal pathways as well as parts of the hypothalamus and thalamus, thus explaining the dissociation between automatic and voluntary movements.

The etiologies are vascular (thrombosis or embolism involving branches of middle cerebral artery supplying the opercular area), astrocytoma, bilateral lesions of perisylvian cortex or subcortical connections, herpes simplex encephalitis, progressive supranuclear motor system degeneration and bilateral taxoplasmosis in Acquired Immunodeficiency Syndrome (AIDS). It can occur at any age and may be congenital or acquired, persistent or intermittent. It is a cortico-subcortical suprabulbar or pseudobulbar palsy of the lower cranial nerves, characterized by severe dysarthria and dysphagia associated with bilateral central facio-pharyngo-glosso-masticatory paralysis. It is the dissociation of automatic-voluntary movements in the affected muscle. However, the reflexive, emotional and automatic innervations of these muscles are preserved and smiling, crying or yawning under natural circumstances is possible. 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 of the lesion).

In 1993, Weller M classified Foix-Chavany-Marie Syndrome into five clinical types, 1) classical and most common form associated with cerebrovascular accident 2) A subacute form caused by central nervous system infections. 3) A developmental form probably most often related to neuronal migration disorders. 4) A reversible form in children with epilepsy 5) A rare type associated with neurode generative disorders.

Method and Materials:

We report a case of 37 years male, who had a cerebrovascular accident and was admitted at Neurology Department in Rajiv Gandhi Government General Hospital, Chennai on 19th September 2016. He had a history of hypertension for the past 5 years and was not under regular medication. He was a chronic alcoholic, consumes alcohol daily. Neurological assessment revealed normal muscle tone with IX (Glossopharyngeal) and X (Vagus) Cranial Nerve palsy. Cardiac evaluation had been carried out by cardiologist showed stable cardiac status. MRI (Magnetic Resonance Imaging) Scan of Brain taken on 3th October 2016 showed,

- Acute infarct in left tectum and right thalamus and right basal ganglia.
- Multiple chronic lacunar infarct in right corona radiate and right ganglio-capsular region and right cerebellum.

He was reported to our department on 4th October 2016 with the complaint of difficulty in speaking and swallowing. He was fed through nasogastric tube.

Detailed Speech and Swallowing Assessment had been carried out in our department. Frenchay Dysarthria Assessment was carried out for differential diagnosis of Dysarthria. Manipal Manual of Swallowing Assessment was administered for comprehensive analysis of swallowing disorders. Hearing sensitivity was assessed by using Pure Tone Audiometer.

Results:

Informal Assessments:

Oral Peripheral Mechanism Examination (OPME):

Table 1:

Structure	Appearance	Movement
Lips	Normal	Asymmetrical
Teeth	Normal	
Tongue	Normal	Protrusion, Elevation, Lateral movements-absent
Hard palate	Normal	
Soft palate	Normal	Immobile
Uvula	Normal	
Nose	Normal	
Jaw	Loosely hanged	

Formal Assessment:

Frenchay Dysarthria Assessment:

Section 1: Reflex

Cough: Occasional difficulty with choking or food sometimes going down the wrong way

Swallow: Patient was unable to swallow – on a nasogastric tube

Dribble/Drool: Very obvious dribbling/drooling when at rest, but not continual

Section 2: Respiration

At rest: Patient was asked to take a deep breath through the mouth and let out audibly and slowly as possible, he was unable to perform the task.

In Speech: Patient was asked to count from 1 to 20 as quicky as possible on one breath, he was unable to perform.

Section 3: Lips
At rest: Lips slightly drooping apart or asymmetrical only noticeable to a skilled observer

Spread: Patient attempts to start the task but both spread and elevation are minimal

Seal: Patient unable to maintain any pressure.

Alternate: Patient was asked to repeat "oo ee" 10 times. Patient was unable to make any movement recognizable as representing either shape.

In speech: No observable production of bilabials or movement of lips in speech attempt.

Section 4: Jaw

At rest: Jaw loosely open majority of time

In speech: No apparent movement of jaw in attempt to speak.

Section 5: Soft Palate

Fluids: Patient has continual difficulty with fluid or food

Maintenance: Palate shows no spreading or elevation

Section 6: Laryngeal

Time: Patient unable to maintain a clear phonation on /AH/ for 3 seconds

Pitch: The patient was aphonic.

Volume: Patient had no vocalisation

In speech: Patient had no vocalisation

Section 7: Tongue

At rest: Tongue shows minimal deviation

Protrusion: Patient able to protrude tongue to lip only

Elevation: Patient unable to elevate the tongue

Lateral: Patient unable to move the tongue in lateral direction

Alternate: Patient was asked to say "ka la", there was no change in the tongue position

In speech: There was no apparent movement of the tongue during speech.

Swallowing evaluation results:
The patient was conscious and appropriately positioned to perform Dry Swallow. The patient was not able to do dry swallow even after three to four repeated trials. Different consistency of food was administered to the patient for swallowing. The patient had more difficulty in transferring the food from the spoon to the oral cavity due to the reduced labial movements. Spillage of food takes place as the mouth was half-open. The retraction of the tongue was absent so the patient had difficulty in propelling the thin and thick consistency of food due to immobile tongue. Aspiration was present. Pharyngeal constriction was minimal. There was no laryngeal elevation due to X cranial nerve palsy. This revealed that patient had oro-pharyngeal dysphagia.

Hearing assessment results:
The hearing assessment was done with pure tone audiometry. The patient had bilateral mild sloping sensory neural hearing loss He was differentially diagnosed from Pesudobulbar Palsy, Bulbar Palsy and Oral Apraxia

Table 2:

	Oral Apraxia	Bulbar Palsy	Pseudobulbar Palsy	Opercular syndrome
Site of lesion	Left hemisphere (frontal and parietal lobes)	Lower motor neuron.	Brainstem (corticobulbar tract).	Opercular area or its sub cortical projections
Reflexes	Present	Absent	Present	Present
Weakness	Absent	Present	Present	Present
Dysphagia	Absent	Present	Present	Present
Fasciculation	Absent	Present	Absent	Absent
Facial weakness	Absent	Present	Present	Present
Emotional disturbance	Absent	Absent	Present	Absent

Discussion:
The patient reported here had classical features of Foix-Chavany-Marie Syndrome characterised by cortical pesudobulbar paralysis ie) facio-labio-pharyngo-glosso-maticatory paralysis. The salient feature of this syndrome is voluntary – automatic dissociation. Hence the patient cannot voluntarily smile or cry, not even swallow food but had preserved automatic or emotional movements. [7]. The interesting fact is that, this patient developed Foix-Chavany-Marie Syndrome due to Cerebrovascular Accident as Weller et al classified cerebrovascular accident as the Classical and most common cause associated with the Opercular Syndrome.[8]. Bilateral perisylvian lesions, involving the primary motor cortex within the frontal and parietal operculum can result in opercular syndrome. From the literature showed that Foix-Chavany-Marie Syndrome can also result due to lacunar infarcts[8], lesion in thalamus and brain stem [1]. Patient reported here had lacunar infarcts in corona radiata and thalamus. Thus, the presentation of case study supports the literature. This feature seems to highlight the previous clinical findings of Thapa Lekjung et al (2010). Foix-Chavany-Marie Syndrome is treated based on its etiology. Speech and swallowing strategies can be framed according to the following steps which are as follows.

Management strategies:
Swallowing intervention plan:
Swallowing therapy includes direct and indirect swallowing treatment.

Table 3: Indirect technique:

SL. NO	GOALS	ACTIVITY
1	OROMOTOR EXERCISE: To increase the range of tongue movements	Elevation of the anterior part of the tongue to alveolar ridge is practised. Then the patient is made to hold the tongue at the level of alveolar for 1 second.

2	To increase the labial tension	Rounding the lips tightly and spreading the lips broadly is practised.
3	Diet modification	Swallowing therapy is started with spoon-thick consistency

Table 4: Direct swallowing treatment:

SL.NO	GOALS	ACTIVITY
1	SWALLOWING MANEUVERS To prevent the food from aspiration	The patient is instructed to take a deep breath and by holding it he is asked to swallow. Immediately after the swallow the patient is asked to cough.
2	POSTURAL TECHNIQUES	The patient is asked to touch the chin to the neck and made to swallow

Speech intervention plan:

In Speech Management, only expression has to be worked upon as the comprehension of the patient is adequate. Initially, oral sensori-motor facilitation technique can be given to strengthen the lips, cheeks, jaw and tongue.

Conclusion:

Here we conclude that Foix-Chavany-Marie Syndrome affects all the structures that are responsible for speech production and swallowing. Thus the patient with Foix-Chavany-Marie Syndrome has difficulty in speaking and swallowing. The management strategies include the Swallowing therapy and Speech therapy. This study will provide better understanding about differential diagnosis of Dysarthria and Dysphagia in Opercular Syndrome. It can be differentially diagnosed from pseudobulbar palsy by the presence of voluntary movements, absence of gag reflex and absence of pathological laughter and emotional disturbance. It can be differentially diagnosed from bulbar palsy by preservation of jaw jerks, pharyngeal reflex and by the absence of fasciculation and atrophy of muscles.

References:

[1] Bogousslavsky J, Dizerens K, Regli. F, Despland PA, Opercular Cheiro-Oral Syndrome, US National Library of Medicine, National Institute of Health, PubMed.gov, https://www.ncbi.nlm.nih.gov/pubmed/2039391

[2] I Chernev, R Petrea, M Reynolds, F Wang, Assessment and Treatment of Dysphagia, The Internet Journal of Neurology. 2008 Volume 11 number 1, https:// ispub. com/ IJN/11/1/9244

[3] M. P. Pender and S. M. Ferguson, Dysarthria and Dysphagia due to Opercular Syndrome in multiple sclerosis, multiple sclerosis 2007,13(6): 817-819, https:// www.ncbi.nlm.nih. gov/ pubmed/17613613

[4] P. Gillardin, R. Van Eetvelde, T Kostermans, R. De Potter, D. Dewilde, M. Lemmerling, Foix-Chavany-Marie Syndrome or Opercular Syndrome, JBR-BTR, 2015, 98:56, https://www.ncbi.nlm.nih.gov/pubmed/26223075

[5] Sarin S. M., Sarosh Kumar K.K., Kadeeja Beevi, Vinu J. And Balakrishnan Valliyot, Foix-Chavany-Marie Syndrome, Indian Journal of Medical Case reports, 2014, Vol. 3(4), www.cibtech.org

[6] Sibel Karaca, Basak Karakurum Goksel, Meliha Tan, Ozlem Alkan, Foix chavanny Marie Syndrome due to cerebral infarctions, Neurol Sci (2013) http:// journals.ju.edu.jo> JMJ>download

[7] S. Praveen Kumar, K. Pramod, Bi-Opercular Syndrome, Journal of Clinical and Diagnostic Research, 2014 Jun, Vol-8(6), http:// www.ncbi.nlm.nih.gov/ pmc/articles/ PMC4129278

[8] Thapa Lekhjung, Paudel Raju, Rana PVS, Department of Neurology, College of medical science-Teaching Hospital, Neurology Asia 2010, www.neurology-asia.org>articles