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Foix-Chavany-Marie Syndrome Secondary to Unilateral Anterior Opercular Infarct without Pre-Existing Lesion

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Abstract: ***Foix-Chavany-Mariesyndrome (FCMS) is a cortical-subcortical pseudobulbar palsycharacterized by voluntary lingual,facial,pharyngeal,and masticatory muscle paralysis with preserved autonomic and involuntary function.We here report a case of FCMS caused by an acute unilateral anterior opercular lesion without a pre-existing lesion in the contralateral opercula.Our case is a rare presentation of FCMS secondary to acute unilateral lesion in the opercula***

Keywords: Foix-Chavany-Marie syndrome, pseudobular palsy, opercula

1. Introduction

Foix-Chavany-Marie syndromeis a rare type of pseudobulbar palsy with paralysis of voluntary facial, pharyngeal, and masticatory muscles with preservation of involuntary and automatic movements.The usual presentation is facial weakness,difficulty in speaking, and difficulty in swallowing, FCMS is usually a result of vascular insults on bilateral opercular and adjoining subcortical areas. FCMS secondary to acuteunilateral lesions with preexisting lesions is rare and even rarer is the one secondary to acute onset unilateral anterior opercular lesion. Our patient is a case of FCMS following acute onset unilateral infarct in the opercular, parietal lobe.

2. Case Report

A 50-year-old male with a known medical history of diabetes, hypertension, and congestive cardiac failure presented to our emergency department with a 4-day history of right-sided weakness, and difficulty in swallowing and speaking. The patient did not have any previous neurological deficits. Patient's vitals were Pulse-110/min B.P-100/60mmhg and Spo2-96% on O2 .Initial neurological examination revealed the patient hada right-sided weakness, severe dysphagia, and dysarthria. Power was 4/5 in the right upper and lower limb, with right extensor plantar reflex (positive Babinski sign).The gag reflex was absent.In view of severe dysphagia Ryles tube feeding was initiated. Involuntary movements like sneezing andyawning were intact.

His Haemogram, liver, and kidney function tests were unremarkable, except for 2D-ECHO findings which were suggestive of Dilated Cardiomyopathy with all four dilated chambers with EF-25% without evidence of clot or vegetations

MRI Brain withcerebral and carotid Angiography was suggestive of a large acute non-hemorrhagic infarct involving the left insular cortex anterior parietal cortex and subcortical white matter.Another similar infarct was noted in the left parietooccipital cortex and subcortical white matter.Cerebral and Carotid Angiography was normal.

Patient was started on Tab aspirin 75mg od ,tab atorvastatin 10mg hs, tab Apixaban 5mg bd, tab Dapagliflozin 10mg od, tab torsemide 5mg + tab spironolactone 50mg, tab digoxin 0.25mg

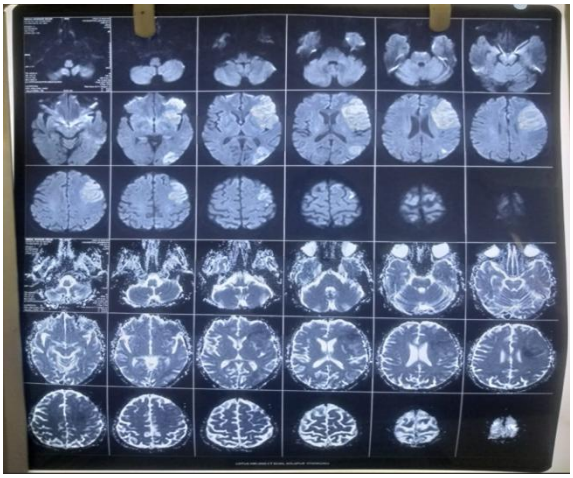

Figure 1: MRI Brain (DWI)

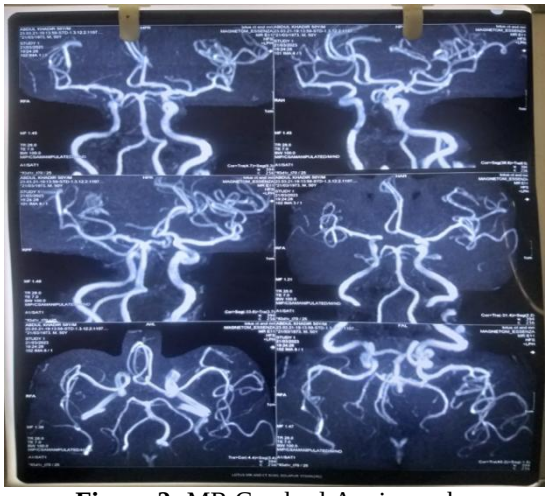

Figure 2: MR Cerebral Angiography

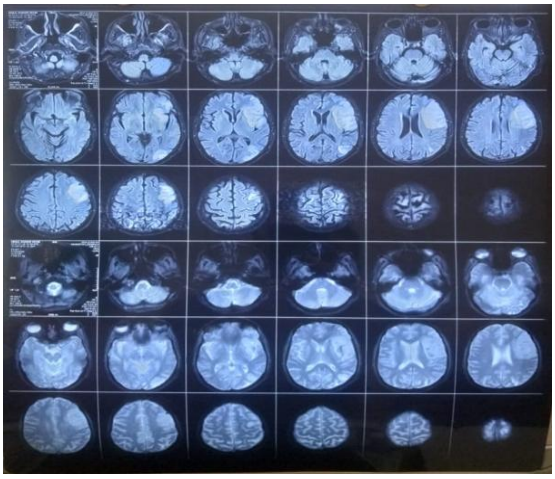

Figure 3: MRI Brain (T2)

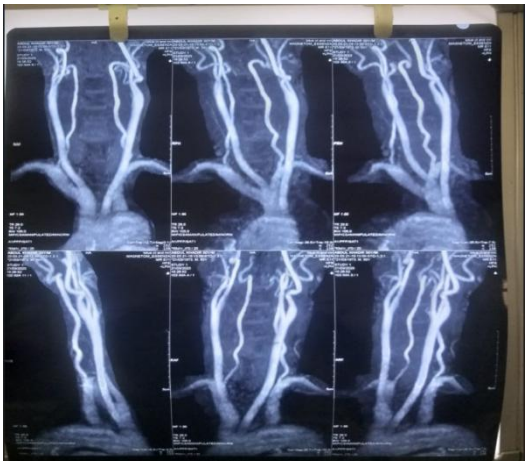

Figure 4: MR Carotid Angiography

3. Discussion

The operculumis the cortex adjacent to the insula encompassing three lobes with a role ina variety of neurologic and psychiatric conditions including sensory, motor, and cognitive processing. Anterior opercula in particular contains voluntary motor fibres for the muscles associated with deglutition and facial expressions (facial, glossopharyngeal, vagus, and hypoglossal).These fibres travel to respective cranial nerve nuclei via the corticobulbar tract. Any lesions involving this area bilaterally can cause FCMS with complete voluntary paralysis of the pharyngeal, laryngeal, and facial muscles.

Autonomic and involuntary movements in these patients are through the inner part of the forebrain and outer longitudinal bundle connecting areas like the hypothalamus and amygdala to the brain stem.

Our patient's MRI Brain and Angiography revealed an acute infarct involving the left insular cortex with subcortical white matter (areas of opercula) apart from infarct in the left parietooccipital cortex and subcortical white matter. There was no evidence of previous lesions involving the contralateral cortico-subcortical matter.

Although rare cases of FCMS have been reported secondary to acute unilateral lesions on preexisting chronic white matter lesions on the contralateral side.Our case is extremely rare with acute infarct in the left insular cortex with subcortical involvement manifesting as FCMS.Our proposed theory is that as the patient was right-handed with left dominant cortex acute infarct in the left cortical-subcortical lobe(dominant) may precipitate FCMS in some individuals.

A 6 month follow up of this patient was done and had improvement in dysphagia.

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