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Clinical Reasoning: A 33-year-old with left sided hemiparesis and anarthria

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Section 1

A 33-year-old man with a history of cardiac thrombus (noncompliant with warfarin) and a prior stroke presented with left-sided weakness and inability to speak. Head CT showed a hyperdense right middle cerebral artery. He received IV-tPA followed by thrombectomy.

On day 2 he had worsening left sided weakness. Repeat CT scan showed hemorrhagic conversion within the right basal ganglia with 6mm of midline shift. On exam, he was alert, oriented, and followed complex commands. Phonetically, he could only grunt, requiring responses to be obtained through writing and gestures. He had a left facial droop and was unable to open his mouth, puff out his cheeks, protrude his tongue, and manage his saliva. On motor testing, he had a left hemiparesis with hyperreflexia. He had no fasciculations.

He ultimately required PEG placement for dysphagia and was discharged to an inpatient rehabilitation facility.

Of note, his prior stroke occurred 4 years earlier. Per chart review, he presented with a right facial droop, dysarthria and expressive aphasia, and was found to have infarcts in the bilateral frontal lobes. He made a successful recovery with no residual deficits.

Questions for Consideration:

- 1) *Describe his speech/language disorder. Is this apraxia, aphasia, or anarthria?*

Section 2

Distinguishing between various speech motor and language disorders is critical for both localization and prognosis after stroke. Aphasia is defined as a disturbance in language and is classically divided into receptive and expressive aphasia. In receptive aphasia, patients have difficulty understanding written and spoken language and thus will be unable to follow commands or respond appropriately (1). Typically, these patients are unaware of their deficits, and will speak in long-winded sentences that make little sense. Lesions responsible for receptive aphasia occur in the posterior temporal area of the dominant hemisphere, known as Wernicke's area. Patients with expressive aphasia, on the other hand, have no trouble understanding and following spoken or written commands. When speaking or writing however, these patients often have improper grammar and difficulty with word finding. The difficulty with word finding can manifest as neologisms, word substitutions, and difficulty with pronunciation. Expressive aphasia classically localizes to Broca's area or the inferior frontal gyrus.

Orofacial apraxia is an inability to perform skilled movements of face, tongue and oropharynx, despite normal understanding and no motor weakness (2). When asked to show how to blow a kiss or lick their lips, patients are unable to perform these tasks. However, automatic movements such as licking their lips when dry is preserved. Speech is also affected in a similar manner, with retained ability to carry on automatic conversations such as greetings, but distortion of lower frequency words as patients struggle to properly coordinate the muscles of speech production. Ability to read and write is maintained. Orofacial apraxia is almost always caused by lesions in the anterior insula and adjacent frontocentral operculum of the left hemisphere.

Anarthria is a complete loss of speech motor ability.

Given that our patient could not eat, chew, swallow, or produce any vocalizations, yet had preserved written language production and language comprehension, we concluded that our patient was anarthric and not aphasic. Further examination showed a positive jaw-jerk reflex, a negative gag reflex and no fasciculations of the tongue, oropharynx and laryngeal muscles. Cerebellar testing was without tremor or dysmetria on finger to nose testing, and he had normal rapid alternating movements. Notably, his face moved well when yawning or spontaneously laughing.

Questions for Consideration:

- 1) *Based on the patient's clinical characteristics, where would you localize his lesion?*

Section 3

Anarthria can localize to the bilateral cortex, bilateral basal ganglia, cerebellum, brainstem, or bilateral lower motor neurons (3). Given our patients' hyperreflexia, lack of fasciculations, and retained ability to yawn and laugh, we concluded that a lower motor lesion affecting the cranial nerve nuclei or peripheral facial nerves was unlikely. Normal cerebellar exam also made a cerebellar lesion unlikely.

To differentiate between basal ganglia and cortical lesions, MRI was reviewed (figure 1B), showing subacute infarcts in the right temporal operculum, subcortical frontal operculum, insula and putamen. There was encephalomalacia from previous infarcts, involving the left frontal and temporal operculum, confirming bilateral cortical localization of his syndrome.

Imaging from the previous hospitalization was obtained and reviewed (figure 1A), demonstrating acute infarcts in the left insula, left temporal operculum and right frontal operculum.

Figure 1

Questions for Consideration:

- 1) *Our patient's constellation of symptoms constitutes what syndrome?*
- 2) *What is the expected recovery for this patient?*

Section 4

The clinical and neuroimaging findings of orofacial paralysis, anarthria, retained yawning and laughing, as well as bilateral inferior frontal lobe strokes are characteristic of bilateral opercular syndrome, also known as Foix-Chavany-Marie syndrome (4).

Bilateral opercular syndrome is a rare form of pseudobulbar palsy. It is differentiated from bilateral peripheral cranial nerve or brainstem lesions by the preserved ability to smile, laugh and yawn. This is because spontaneous facial expressions are produced through contributions of the thalamus, hypothalamus and extrapyramidal tract. The selective paralysis of voluntary muscle weakness is termed, “autonomic-voluntary dissociation”.

Currently, there is little information regarding long-term recovery in these patients. Most case reports have described “poor recovery” with respect to dysarthria, anarthria, and dysphagia after 4 weeks of initial insult and intensive therapy. One case report described slow, incomplete improvements in speech and dysarthria after extended periods of rehabilitation (5).

We followed the patient over a 6-month period following stroke in order to characterize his course of recovery. Over this time period, he regained the ability to lift his arm in flexor synergy and move his fingers individually. Speech improved from complete anarthria to unintelligible dysarthria at 4 weeks, and finally to moderate dysarthria with only some words requiring repetition at 6 months. Cheek puff improved from complete inability at onset, to a weak cheek puff with air escaping on compression. At 6 months, dysphagia improved to the point that he could tolerate a regular diet with thin liquids. Saliva management remained a problem and he frequently had to wipe his mouth with a napkin.

Video

Discussion

Despite documentation of “expressive aphasia”, our patient likely suffered from bilateral opercular syndrome after his first stroke, in which MRI showed acute strokes with lesions in the bilateral perisylvian, opercular regions. He made a complete recovery within 6

months and had no residual deficits until 4 years later when he suffered a new right-sided stroke. The second, purely right-sided stroke led to a recurrent bilateral opercular syndrome by affecting the portion of brain that had undergone neuroplastic reorganization to recover bulbar motor functions after the first stroke.

Bilateral opercular syndrome often involves bilateral lesions to the frontal operculum that includes the inferior portion of the precentral gyrus, the pars triangularis, and the opercularis of the inferior frontal gyrus. The precentral component involves the primary motor cortex, the portion of the motor homunculus controlling the face, lips, tongue, and pharynx (6). Thus, bilateral opercular syndrome is largely a disorder of facial muscle paresis and oropharyngeal motor control, not one of language. It is therefore plausible to conclude that a stroke causing bilateral opercular syndrome should follow a recovery pattern similar to other strokes causing central facial paresis.

Two prior studies have characterized orofacial recovery after stroke. One study followed patients for two years after stroke (median enrollment 40 days after stroke), and found negligible improvements in maximum lip force, maximum bite force, chewing efficiency, and hand grip strength in patients receiving no rehabilitation therapy (7). Another study enrolled patients 2 days to 10 years after stroke with dysphagia. After 5 weeks of oral training, all patients, regardless of time since stroke, improved significantly in both lip force and swallowing capacity (8).

The improvement in dysphagia seen in this study, is consistent with recent studies demonstrating recovery of task specific activities of the upper extremities in the setting of proper rehabilitation (9). One promising strategy involves a rehabilitation technique called constraint induced movement therapy, in which patients are forced to perform activities using their affected arm and prevented from using their unaffected limb. Reasons for improvement with intensive therapy are likely multifactorial, including but not limited to: preventing learned non-use, overcoming neuromuscular adaptation, and providing adequate therapy intensity and duration to promote motor learning of complex movements (10).

The patient presented in this report improved with early, persistent rehabilitation, along with home exercises and practice. The improvement in function over a 6 month period in this individual with serial damage to bilateral motor networks supports the potential of neuroplasticity and active rehabilitation efforts to promote recovery.

Appendix 1: Authors

Name	Location	Contribution
Sean Schowalter, MD	Beth Israel Deaconess Medical Center, Boston	Designed, conceptualized and drafted manuscript
Douglas I. Katz, MD	Boston University School of Medicine, Boston	Advised and reviewed manuscript for intellectual content
David J. Lin, MD	Massachusetts General Hospital, Boston	Advised and reviewed manuscript for intellectual content

Video - <http://links.lww.com/WNL/B241>

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Figure 1: Axial MRI Images of Prior and Recent Stroke.

Prior stroke (figure 1A), produced bilateral perisylvian, opercular ischemic lesions.

The red arrow indicates the right sylvian fissure; *, stroke in right frontal

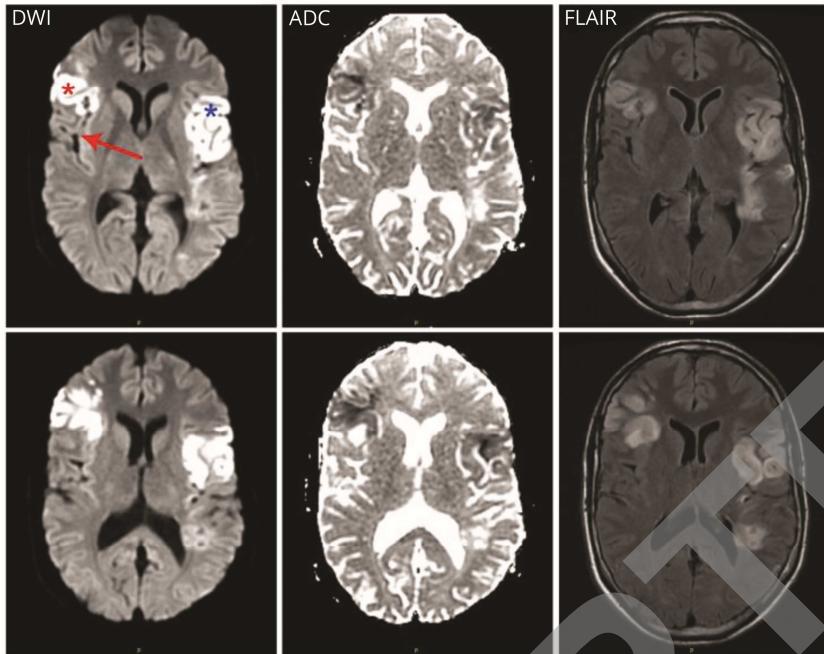
operculum; *, stroke in left frontal/temporal opercular and insular regions. The

recent stroke (figure 1B) displays an acute ischemic lesion in the right

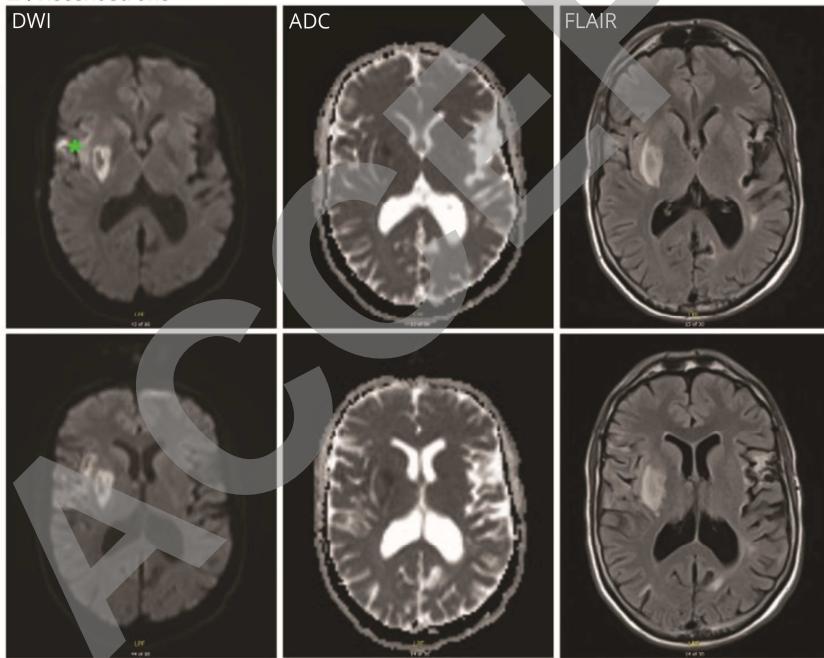
frontotemporal operculum (*) and in the right basal ganglia (putamen).

Encephalomalacia from the prior stroke in the left frontotemporal operculum is still visible.

A. Prior stroke



B. Recent stroke



Video: Physical Exam at 4 Weeks, 3 Months and 6 Months, Showing Gradual Improvement in Orofacial Function and Dysarthria.

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