Neurosciences Quiz

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A lady with facio-linguo-pharyngeal-masticatory diplegia

Clinical Presentation

A 70-year-old woman, with a history of hypertension and poor compliance with medications, presented with 2 episodes of neurological dysfunction. During the first episode that occurred 2 years previously, she developed the sudden onset of non-fluent speech with associated anomia, impaired repetition of sentences, and preserved verbal comprehension and transient right hemiparesis. Her speech problems improved significantly over the subsequent days. In the recent/second episode, her troublesome symptoms included mutism, dysphagia (with brisk gag reflex bilaterally and preserved cough reflex), drooling of saliva, and difficulty in chewing and bilateral lingual paresis without tongue wasting. The jaw jerk was not brisk. When angry, she was able to produce words and short phrases. However, she did not produce any verbal output volitionally or in response to questions. There was no pseudobulbar affect or emotional incontinence during the period of her hospital stay. She was fully ambulant without any significant arm/leg weakness. Electrocardiogram and echocardiography were unremarkable. A CT angiogram of neck and intracranial arteries revealed only minimal atherosclerotic changes. Her recent cranial CT is shown in **Figure 1**.

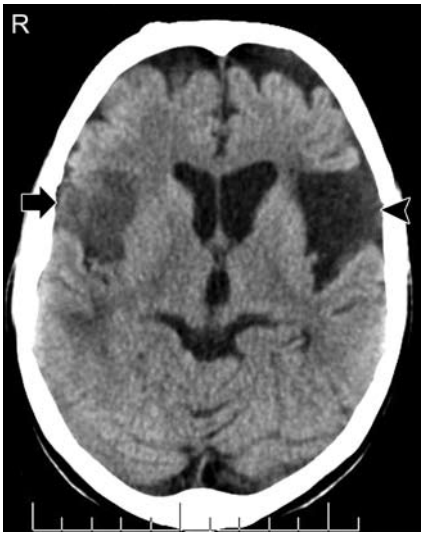

Figure 1 - Axial non-contrast brain CT.

Questions:

1. What does the CT brain (**Figure 1**) show?

2. What is the reason for her recurrent neurological symptoms?

3. What is the syndromic diagnosis for her recent troublesome symptoms?

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Answers

1. Non-contrast CT brain shows a recent hypodensity (short arrow) in the right fronto-temporal operculum and insular region (suggestive of acute infarction in the territorial distribution of anterior division of right middle cerebral artery), and an old hypodensity (arrowhead) in the corresponding region on the left side, along with minimal left sided ventricular dilatation (suggestive of old infarction in the distribution of anterior division of left middle cerebral artery).
2. During the first clinical episode, she had developed expressive aphasia (and transient right hemiparesis) due to infarction in the anterior division of the left middle cerebral artery and had made significant clinical improvement. In the second episode, she had troublesome facio-linguo-pharyngeal-masticatory diplegia due to subsequent new infarction in the anterior division of right middle cerebral artery. Thus, she had recurrent neurological dysfunction due to sequential ischemic strokes involving the anterior opercular and insular regions on both sides. Her vascular risk factors included old age and hypertension. The presence of infarctions in different vascular distributions (left and right side of cerebral circulation) warranted cardiac evaluation to look for the source of the recurrent thrombo-embolism. However, the evaluation did not reveal persistent atrial fibrillation, thrombus, valvular disease, vegetations or myxoma. Nor did the cerebral vascular imaging disclose any significant arterial stenosis or dissection.
3. The presence of suprabulbar palsy (facio-linguo-pharyngeal-masticatory diplegia with brisk gag reflex and preserved reflex cough response) with automatic-voluntary dissociation (while she remained mute or anarthric for responding to questions and for volitional speech, she produced few words/sentences when emotionally disturbed) in the setting of bilateral fronto-temporal opercular infarctions suggests anterior opercular syndrome, eponymously known as Foix-Chavany-Marie syndrome.

Discussion

Our patient presented with pseudobulbar or suprabulbar diplegia (resulting in anarthria-dysarthria, dysphagia, sialorrhea, lingual paresis, and masticatory weakness) in the setting of bilateral anterior opercular infarctions. This symptom-complex is characteristic of opercular syndrome, also known as Foix-Chavany-Marie syndrome.[1-5] The salient feature is the presence of automatic-voluntary dissociation. The volitional control of the facio-linguovelo-pharyngeal-masticatory muscles is affected, while the reflex and emotional movements are preserved. This clinical feature suggests the presence of divergent cortico-bulbar pathways for the control of volitional and automatic/emotional cranio-facial movements.[1-5] There are only a few cases of this syndrome published in the literature and the most common etiology is either simultaneous or sequential infarctions in the bilateral opercular regions.[1,2] Other reported causes include epilepsy, herpes simplex encephalitis,[3] trauma,[4] a variant of fronto-temporal dementia,[5] multiple sclerosis, cerebral toxoplasmosis in the setting of acquire immunodeficiency syndrome, glioma, and development disorders. In the present patient, the opercular syndrome was due to sequential infarctions in the bilateral middle cerebral artery distribution. Apart from the conventional vascular risk factors, the reason for her recurrent stroke could not be ascertained. She was treated with antiplatelet drugs, antihypertensive medications (with an emphasis on drug compliance), statins, and naso-gastric feeding. She demonstrated only mild improvement of her persistent suprabulbar symptoms, when reviewed 3 months after her second stroke.

References

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