Section Editors: Daniel Strbian, MD, PhD, and Sophia Sundararajan, MD, PhD

Supplementary Motor Area Stroke Mimicking Functional Disorder

Paul Mathew, MD; Prem Pradeep Batchala, MD;
Thomas Jose Eluvathingal Muttikkal, MD

Case Description

A 52-year-old right-handed women was referred from an outside hospital with a diagnosis of stroke. On arrival at our hospital, her National Institutes of Health Stroke Scale was 9 (Table I in the online-only Data Supplement). She had a strength of 0/5 in right upper and lower extremity muscle groups. There was decreased sensation to finger touch in the right lower extremity. She was oriented to person, place, and time. She did not have spatial neglect, motor neglect, apraxia, or abulia. Speech was fluent with intact comprehension, naming, and repetition. Cranial nerve examination was normal.

Interestingly, the patient was able to bear weight and demonstrated a positive Hoover sign that raised suspicion of functional paresis. She was outside the intravenous thrombolysis window but within the thrombectomy window. She had history of nonischemic cardiomyopathy, polymorphic ventricular tachycardia, and had an implantable cardioverter defibrillator.

She underwent noncontrast computed tomographic (CT) examination of the brain along with a multiphasic CT angiography of the intracranial circulation and CT angiography of the neck. The noncontrast examination was negative for acute stroke (Figure 1). However, CT angiography demonstrated an abrupt cutoff in distal portion of the callosomarginal artery branch of the left anterior cerebral artery (ACA; Figure 2). Bilateral common and internal carotid arteries did not show stenosis or atherosclerotic disease. She was admitted to the stroke service for further management. Magnetic resonance imaging of the brain was not done because the compatibility of the implantable cardioverter defibrillator could not be determined. A follow-up noncontrast CT brain was performed after 12 hours which revealed a small nonhemorrhagic acute infarct in the posterior third of left medial frontal gyrus corresponding to the supplementary motor area (SMA; Figure 1). She was empirically started on anticoagulation with apixaban. The implantable cardioverter defibrillator was interrogated and showed an episode of tachycardia ≈2 months before this admission. Being a single lead implantable cardioverter defibrillator, atrial fibrillation could not be ruled out and a strong suspicion for cardioembolic source remained. EKG on admission and during the stay showed normal sinus rhythm. Subsequent telemetry showed 3 episodes of paroxysmal supraventricular tachycardia lasting 5, 7, and 18 beats with rates of 140 to 160 bpm, in addition to supraventricular premature beats in couplets and triplets. Transthoracic echocardiography showed normal cavity size and wall thickness of left ventricle and an ejection fraction of 55%. At the time of discharge 2 days after admission, she regained variable amount of strength (2/5 to 5/5) in the muscle groups of her right upper and lower extremities. At follow-up appointment 2 months later, she had regained near normal strength on her right side. There was no language deficit during the course of events. The final diagnosis was SMA syndrome from occlusion of the left ACA callosomarginal branch.

Discussion

SMA is located in the medial aspect of Brodmann area 6, which is an area in the posterior third of the medial aspect of superior frontal gyrus. It is bounded inferiorly by cingulate sulcus and gyrus and posteriorly by the precentral motor gyrus controlling the lower limb (Figure 3). The functional importance of this area remained ambiguous for long since its discovery in 1951 by Penfield and Welch.[1] Subsequent studies linked this area to self-initiated motor movements, planning and sequencing the motor action, response inhibition, and bimanual movements. The recent concept is that SMA is a functionally heterogeneous area, and it may be involved in wider cognitive functions, like timing, spatial processing, numeric cognition, working memory, and language.[2–4] It is also thought that there are 2 subareas within SMA, namely pre-SMA and SMA proper.[2] Pre-SMA area is rostral, has connections to prefrontal cortex, and is mostly involved in planning process. SMA proper is caudal, has connections to primary motor cortex, premotor cortex, and spinal cord, and is involved in execution of movements. Both areas also have connections to basal ganglia. There is also anterior to

Received August 16, 2017; final revision received November 27, 2017; accepted November 30, 2017.

From the Department of Neuroradiology, University of Virginia Health System, Charlottesville.

The online-only Data Supplement is available with this article at http://stroke.ahajournals.org/lookup/suppl/doi:10.1161/STROKEAHA.117.019106/-/DC1.

Correspondence to Thomas Jose Eluvathingal Muttikkal, MD, Department of Neuroradiology, University of Virginia Medical Center, Charlottesville, 1215 Lee St, VA 22908. E-mail tje6d@virginia.edu

(*Stroke*. 2018;49:00-00. DOI: 10.1161/STROKEAHA.117.019106.)

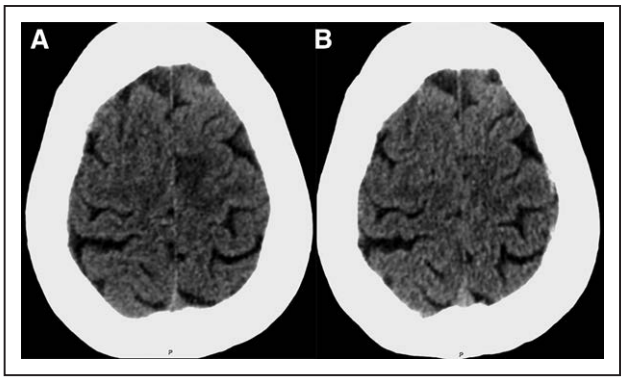

Figure 1. A, Noncontrast computed tomography (CT) head done 12 hours after the initial CT showing an acute infarct involving the supplementary motor area on the left. **B**, Unremarkable initial noncontrast CT head at the same axial level as (**A**).

posterior somatotopy in the SMA proper area with language being anterior followed by face, upper limb, and inferior limb.

SMA's arterial supply is often from callosomarginal branches of ACA. Isolated ACA arterial infarcts are uncommon with literature quotes of only 1.1% to 1.3% of all intracranial ischemic strokes.[5,6] Furthermore, infarcts restricted only to SMA are even rarer with only few case reports in literature. These cases can shed more light on the functional aspect of this area.[7] Atheroembolism is the most common pathogenesis of these infarcts. Typically, SMA syndrome is seen in the setting of acute ACA infarcts or neoplasms in that location or after surgical resection involving the region. The symptomatology of SMA syndrome is variable but generally consists of impairment of volitional movements, hemineglect, and dyspraxia of contralateral limbs. The other characteristics of the syndrome are preserved muscle tone, near complete to complete recovery of mutism, and contralateral weakness in a relatively short period of time (weeks to months).[7]

Stroke mimicking functional disorder is not a well-known entity in contrast to functional disorder mimicking stroke. There is limited literature on the incidence of stroke initially misdiagnosed as functional disorder. Booij et al[8] described 2 young female patients with posterior circulation stroke, who were initially misdiagnosed as having functional disorder which resulted in delay in initiating appropriate treatment. In the series by Crimlisk et al,[9] of 64 patients misdiagnosed as functional disorder, 1 was because of stroke. Stone et al[10] did an extensive systematic review of studies from 1965 to 2003 to estimate how often patients with an initial diagnosis of functional symptoms are subsequently given a disease diagnosis that explained their original symptoms. They found that the reported rate of misdiagnosis of functional symptoms has on average been 4% since 1970. The most common misdiagnosis were related to gait and movement disorders. However, this study did not provide any data on incidence of stroke in these patients. To the best of our knowledge, there are no prior reported cases of SMA stroke mimicking functional disorder.

Functional disorder is diagnosed based on clinical criteria provided by the Diagnostic and Statistical Manual (fifth edition). The diagnosis can be effectively ruled out when there is an adequate explanation of the neurological symptom. Our patient presented with 0/5 power in the right upper and lower limbs but was able to bear weight and showed positive Hoover sign raising the suspicion of functional disorder or malingering. Neuroimaging provided an adequate explanation to the apparent disparity between the patient symptoms and neurological signs by demonstrating an acute SMA infarct.

TAKE HOME POINTS

- Supplementary motor area is located in the posterior third of superior frontal gyrus anterior to primary motor area for the lower limb.
- Supplementary motor area is often supplied by callosomarginal branch from anterior cerebral artery, and isolated infarct in this region is rare. Atheroembolic disease is the most frequent cause for infarct in this region. Computed tomography angiographic examination can be helpful when a magnetic resonance imaging examination cannot be performed in these patients.
- Functional disorder is a diagnosis of exclusion. Stroke is rarely known to mimic functional disorder. Neuroimaging can play a vital role when clinical examination cannot differentiate functional pathogenesis from stroke.

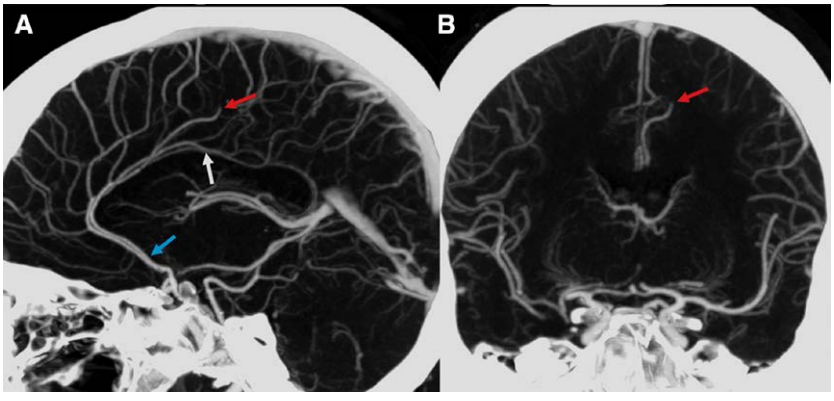

Figure 2. A, Computed tomographic (CT) cerebral angiography thick maximum intensity projection sagittal image showing abrupt cutoff of a callosomarginal artery branch (red arrow) supplying the left supplementary motor area. Also seen are the patent A2 segments of the anterior cerebral arteries (blue arrow) and the patent left pericallosal artery (white arrow) arching around the corpus callosum. **B**, CT cerebral angiography thick maximum intensity projection coronal image showing abrupt cutoff of a callosomarginal artery branch (red arrow) supplying the left supplementary motor area.

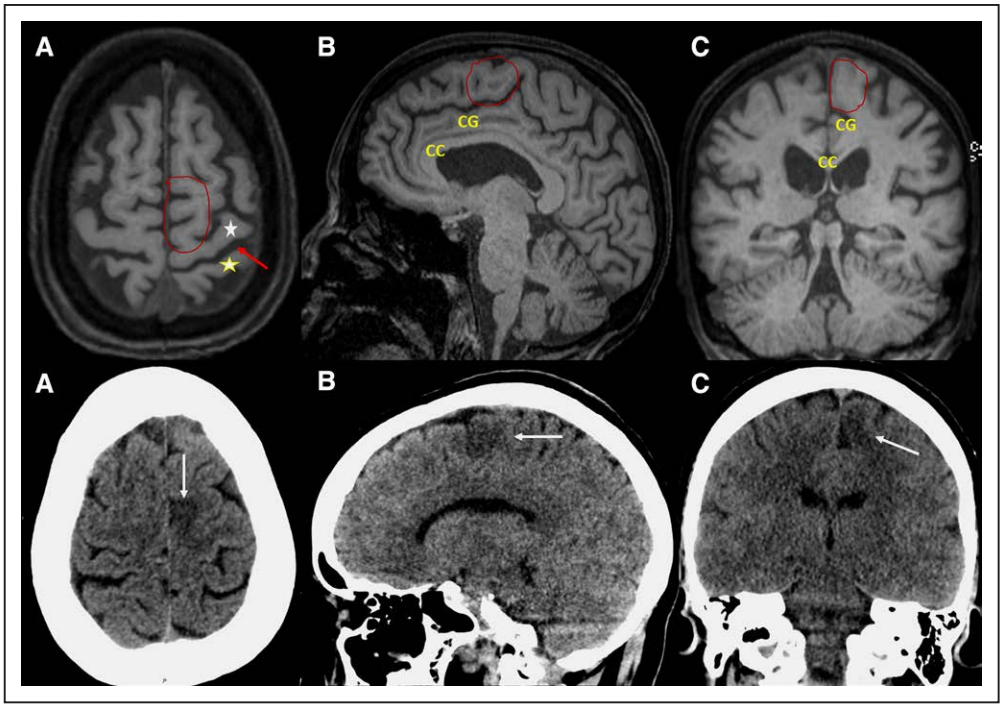

Figure 3. Supplementary motor area. Top row, Axial (**A**), sagittal (**B**), and coronal (**C**) magnetic resonance brain image of an example case depicting the approximate location of the supplementary area location in the posterior part of superior frontal gyrus within the schematic red line. The other important landmarks in the axial image are precentral gyrus (white star), postcentral gyrus (yellow star), and central sulcus (red arrow). Bottom row, Noncontrast head computed tomography of our index patient, axial (**A**), sagittal (**B**), and coronal (**C**) corresponding to the top row, demonstrating the acute infarct localized to supplementary motor area (white arrow). CC indicates corpus callosum; and CG, cingulate gyrus.

Disclosures

None.

References

1. Penfield W, Welch K. The supplementary motor area of the cerebral cortex; a clinical and experimental study. *AMA Arch Neurol Psychiatry*. 1951;66:289–317.
2. Cona G, Semenza C. Supplementary motor area as key structure for domain-general sequence processing: a unified account. *Neurosci Biobehav Rev*. 2017;72:28–42. doi: 10.1016/j.neubiorev.2016.10.033.
3. Chung GH, Han YM, Jeong SH, Jack CR Jr. Functional heterogeneity of the supplementary motor area. *AJNR Am J Neuroradiol*. 2005;26:1819–1823.
4. Hertrich I, Dietrich S, Ackermann H. The role of the supplementary motor area for speech and language processing. *Neurosci Biobehav Rev*. 2016;68:602–610. doi: 10.1016/j.neubiorev.2016.06.030.
5. Kang SY, Kim JS. Anterior cerebral artery infarction: stroke mechanism and clinical-imaging study in 100 patients. *Neurology*. 2008;70(24 pt 2):2386–2393. doi: 10.1212/01.wnl.0000314686.94007.d0.
6. Kumral E, Bayulkem G, Evyapan D, Yunten N. Spectrum of anterior cerebral artery territory infarction: clinical and MRI findings. *Eur J Neurol*. 2002;9:615–624.
7. Alonso A, Gass A, Rossmanith C, Kern R, Griebe M, Binder J, et al. Clinical and MRI patterns of pericallosal artery infarctions: the significance of supplementary motor area lesions. *J Neurol*. 2012;259:944–951. doi: 10.1007/s00415-011-6289-1.
8. Booij HA, Hamburger HL, Jöbsis GJ, Beuerle EY, Kruyt ND. Stroke mimicking conversion disorder: two young women who put our feet back on the ground. *Pract Neurol*. 2012;12:179–181. doi: 10.1136/practneurol-2011-000152.
9. Crimlisk HL, Bhatia K, Cope H, David A, Marsden CD, Ron MA. Slater revisited: 6 year follow up study of patients with medically unexplained motor symptoms. *BMJ*. 1998;316:582–586.
10. Stone J, Smyth R, Carson A, Lewis S, Prescott R, Warlow C, et al. Systematic review of misdiagnosis of conversion symptoms and "hysteria". *BMJ*. 2005;331:989. doi: 10.1136/bmj.38628.466898.55.

KEY WORDS: anterior cerebral artery ■ cerebral angiography ■ defibrillators, implantable ■ malingering ■ stroke

Stroke
Journal of the American Heart Association

Supplementary Motor Area Stroke Mimicking Functional Disorder

Paul Mathew, Prem Pradeep Batchala and Thomas Jose Eluvathingal Muttikkal

Stroke. published online January 8, 2018;
Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 0039-2499. Online ISSN: 1524-4628

SUPPLEMENTAL MATERIAL

Table I. NIH Stroke Scale

Level of Consciousness Responsiveness	0
LOC Questions	0
LOC Command	0
Best Gaze	0
Visual	0
Facial Palsy	0
Motor Arm RIGHT	4
Motor Arm LEFT	0
Motor Leg RIGHT	4
Motor Leg LEFT	0
Limb Ataxia	0
Sensory	1
Best Language	0
Dysarthria	0
Extinction/Inattention	0
NIHSS Initial	9