Moyamoya Presenting after Whole Body Cryotherapy

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Abstract

Background

Purpose: Moyamoya syndrome is the progressive stenosis of intracranial carotids with secondary collateralization. Whole body cryotherapy (WBC) involves external cooling and is used in holistic and sports medicine, its neurologic effects are unknown.

Case report: We report a first case of symptoms of moyamoya syndrome presenting following WBC and diagnosed with classic MRI ("Brush Sign", "Ivy sign") and digital subtracted angiography.

Conclusion: WBC may provoke symptoms of moyamoya syndrome possibly through hyperventilation or vasoconstriction. Practitioners should be aware of possible consequences of WBC in patients with poor cerebrovascular reserve.

Keywords: Stroke, cerebrovascular disease, moya moya , rt-PA, cryotherapy

Acta Neurol Taiwan 2020;29:64-66

INTRODUCTION

Moyamoya syndrome involves progressive stenosis of the intracranial internal carotid arteries, which can result in decreased cerebral blood flow and symptoms of cerebral ischemia. We describe a manifestation of moyamoya syndrome presenting with acute aphemia following whole body cryotherapy.

CASE REPORT

A 32-year-old Caucasian woman with no past medical history presents with acute word finding difficulty and right sided weakness. After four minutes into a whole-body cryotherapy session at "presumed -127 Fahrenheit", she felt a generalized unwellness and stopped early. This was her first and only whole-body cryotherapy session. Upon leaving the chamber she had sudden right arm weakness and was unable to produce words. This was captured on self video at the time and verified to be the last known well. She did not develop a headache and had no seizure, migraine, paroxysmal neurologic episodes or neck trauma history.

Upon arriving in the emergency room as a stroke alert, her exam showed an expressive aphasia, intact writing and comprehension, and right arm and leg weakness. She was

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Received March 6, 2020. Revised March 23, 2020.
Accepted March 27, 2020.

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able to type words on her cellphone but not able to produce verbal speech. A non-contrast CT head demonstrated subacute to chronic left occipital and frontal white matter infarcts. CT angiogram of head and neck showed moderate narrowing of the left paraclinoid internal carotid artery (ICA) with near occlusion of the ICA terminius and proximal M1 middle cerebral artery (MCA). There was also severe narrowing of the left posterior cerebral artery (PCA). Her exam improved in the emergency room and recombinant tissue plasminogen activator (rt-PA) was deferred. Brain MRI showed additional acute punctate left MCA infarcts, left MCA territory hypoperfusion (as indicated by increased oxygen extraction), and leptomeningeal collaterals (Figure 1). Digital subtraction angiography (DSA) demonstrated findings consistent with Suzuki stage 6 moyamoya disease (Figure 2). She was discharged without deficits on aspirin 325 mg. On 1 month follow up, she reported episodes of stuttering speech when yelling at her children, but otherwise showed no aphasia, weakness or other neurological deficits on examination. She is undergoing surgical evaluation for direct revascularization by neurosurgery.

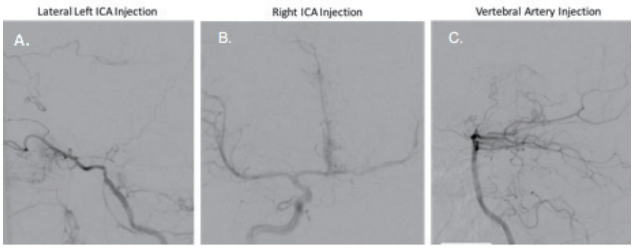

Figure 1. Axial DWI (left), FLAIR (center) and SWI (right) MRI Brain

A. Punctate acute left MCA territory infarct (arrow). Late subacute to chronic border zone infarcts (circle) B. Hyperintense sulcal vessels, presumed retrograde slow flow through leptomeningeal collaterals (FLAIR "Ivy Sign", arrows) C. Prominence of deep medullary veins draining the left MCA territory suggestive of increased oxygen extraction secondary to decreased cerebral perfusion (SWI "Brush Sign", circle).

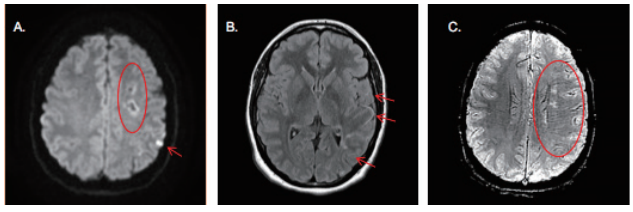

Figure 2. Digital subtraction angiography

A. Left Lateral ICA injection with occlusion of the ICA at the level of the ophthalmic artery. Enlarged ophthalmic artery with ethmoidal collaterals to ACA signifying this is a chronic process B. Right ICA injection with cross filling to the left MCA territory, but this is delayed and insufficient. Neovascularization of the left ACEA territory proximally C. Vertebral artery injection with engorged thalamic perforators.

DISCUSSION

Moyamoya syndrome involves progressive stenosis of intracranial internal carotid arteries with secondary collateral vessel proliferation in an attempt to maintain cerebral blood flow. It is strongly associated with previous head-neck radiation, neurofibromatosis type-1, vasculitic disorders, and sickle cell disease. Though a major gene locus on chromosome 17q25 has been identified for autosomal dominant moyamoya disease, the remainder are sporadic[1]. Moyamoya syndrome can clinically present with migraine, ischemia or hemorrhage. As in this case, classic radiographic features include slow flow on fluid-attenuated recovery MRI sequence ("Ivy Sign"), decreased perfusion/increased oxygen extraction on susceptibility weighted imaging ("Brush Sign"), and multifocal stenosis on DSA1.

The role of rt-PA in moyamoya syndrome poses a unique therapeutic challenge. In this case rt-PA was not given due to rapidly resolving symptoms and subacute infarcts. Moyamoya syndrome is not a contraindication to rt-PA, though there are no clear consensus guidelines. rt-PA's therapeutic effects are thought to be from lysis of intraluminal thrombus. Moyamoya syndrome symptoms typically result from transient changes in flow from baseline vascular hyperplasia rather than acute thrombus. Other possible mechanisms of moyamoya syndrome include induced hemodynamic low flow or distal embolization following stenosis. Given these many possibilities, it is unclear if rt-PA would be beneficial in Moyamoya syndrome from a mechanistic standpoint. There exists only one case report detailing intravenous rt-PA in moyamoya syndrome, the outcome being therapeutic and safe[2].

Our patient did not fit a classic Broca's aphasia given intact writing. The clinical syndrome is consistent

with aphemia, a rare apraxia of speech characterized by deficit in organization of articulatory and motor speech components with intact writing and comprehension[3]. It is a disorder of articulation rather than a motor aphasia. Small lesions in the dominant inferior frontal gyrus, as in this presentation, account for the majority of cases[3].

Finally, whole body cryotherapy (WBC) as an inciting event for moyamoya syndrome is unique. WBC is a specialized chamber in which the external body is exposed to -110 to -140 Celsius temperatures[4]. The systemic effects with cryotherapy are unclear yet it is increasingly seen to be used in sports and holistic medicine. Only local skin burns are reported as a side effect[4]. In our case of previously undiagnosed moyamoya syndrome, one mechanistic possibility for symptomatic infarction is that cryotherapy induced cerebral vasoconstriction, potentially through transient hyperventilation. There is a single report of reversible cerebral vasoconstriction syndrome following WBC[5], thus cryotherapy may cause cerebral autoregulation dysregulation or induced vasospasm in a yet to be determined mechanism.

CONCLUSION

Practitioners should be aware of possible rare neurologic sequelae with WBC, especially in patients with poor cerebrovascular reserve.

AUTHOR CONTRIBUTIONS

P.M. Chen drafting/revising the manuscript, data acquisition, study concept or design, analysis or interpretation of data. M.M. Chen drafting/revising the manuscript, data acquisition, study concept or design, analysis or interpretation of data. C. Chiang, drafting/revising the manuscript, data acquisition, study concept or design, analysis or interpretation of data. S.O. Data acquisition, critical review of manuscript. D.S.B. Data acquisition, critical review of manuscript. K.A . Data acquisition, critical review of manuscript.

REFERENCE

1. Scott RM and Smith ER. Moyamoya disease and moyamoya syndrome. N Engl J Med 2009; 360: 1226-1237. 2009/03/20. DOI: 10.1056/NEJMra0804622.
2. Yokoyama S, Manabe Y, Fujii D, et al. Intravenous tissue plasminogen activator therapy for an acute ischemic stroke patient with later diagnosed unilateral moyamoya syndrome. J Stroke Cerebrovasc Dis 2013; 22: 1190-1192. 2012/07/27. DOI: 10.1016/j.jstrokecerebrovasdis.2012.06.011.
3. Schiff HB, Alexander MP, Naeser MA, et al. Aphemia. Clinical-anatomic correlations. Arch Neurol 1983; 40: 720-727. 1983/11/01.
4. Patel K, Bakshi N, Freehill MT, et al. Whole-Body Cryotherapy in Sports Medicine. Curr Sports Med Rep 2019; 18: 136-140. 2019/04/11. DOI: 10.1249/jsr.0000000000000584.
5. Whole Body Cryotherapy Induced Reversible Cerebral Vasoconstriction Syndrome (P3.118). Neurology: P3.118.