A Curious Case of Moyamoya in an Adult

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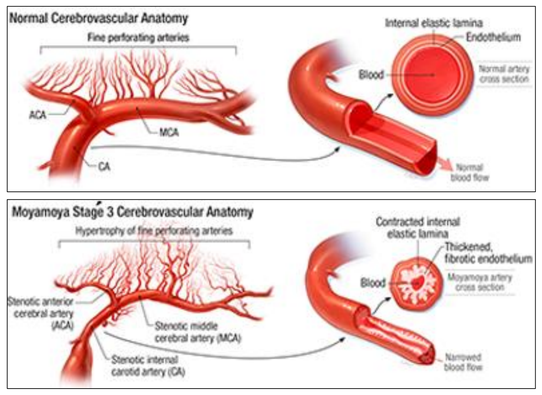
Introduction

- Moyamoya is a rare central nervous system (CNS) disease **primarily affecting Asians**, but can present in non-Asians.
- **Mortality rate is 10%** if left untreated necessitating early diagnosis. [1]
- It has association an with hereditary diseases such as NF-1, sickle cell, as well as, has a rare association with PKD.



From: Swedish Neuroscience Institution

Case Overview

HPI: 34-year-old Caucasian female presented with 5-day history of recurrent right-sided hemiparesis, dysarthria, and aphasia lasting 15-20 minutes with complete resolution of symptoms.

- History negative for fever, headache, neck stiffness, seizures, loss of consciousness, vision changes, nausea or vomiting.

PMH:

Polycystic Kidney Disease (PKD)
prior cocaine use (>2 years ago)
prior tobacco use
ADHD

FH:

Father and grandfather with PKD; grandfather passed from cerebral AVM rupture

Exam:

VS: unremarkable
NIH stroke scale was 1 for mild right upper extremity drift, but otherwise with no focal neurologic deficit.

Labs:

- CBC: WBC 8.4, Hb 14.2, Hct 41.2, Plt 194
- CMP: Na 140, K 4.4, Cl 102, CO2 24, BUN 14, Cr 0.8, Glu 99
- LDL: 96; HbA1C: 5.8
- UDS: normal

Clinical Course

- Emergent **CT head without contrast** showed no acute intracranial process.
- Treatment for ischemic stroke was initiated with full-dose aspirin and atorvastatin 20mg; thrombolytic was not administered as she was out of 3-hour window.
- **MRI brain with and without contrast** revealed multiple acute infarcts of left frontal and temporal lobes and severe bilateral middle cerebral artery (MCA) narrowing.
- At this point, there was a concern of vasculitis versus hypercoagulable state versus atypical infection in this young and relatively healthy patient.
- **Transthoracic ECHO: n**ormal
- Further work-up including **CRP, ANA, ANCA, HIV antibody, serum RPR, B-2 macroglobulin, and C3,/C4** compliment levels all unremarkable.
- On day 2 of admission, patient's symptoms completely resolved with repeat NIH stroke scale 0.
- **Catheter cerebral angiogram** showed complete occlusion of right MCA with marked irregular stenosis of left MCA and extensive collaterals from anterior and posterior cerebral arteries consistent with moyamoya.
- After discharge, she was seen at the neurosurgery clinic with a plan to have left MCA bypass surgery in 1 month followed by right MCA bypass surgery 6 months later.

Image Findings

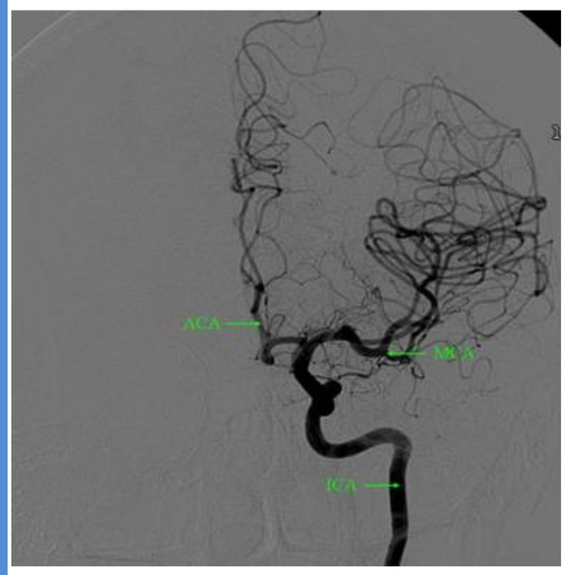


Fig 1. Normal cerebral angiogram- left

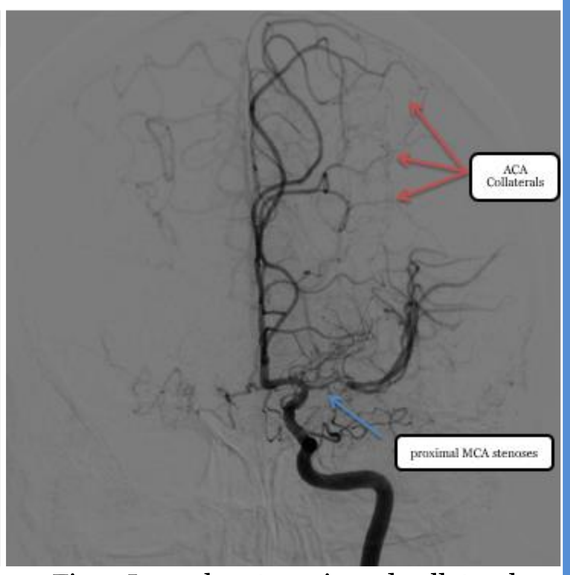


Fig 2. Irregular stenosis and collateral vessels in Moyamoya

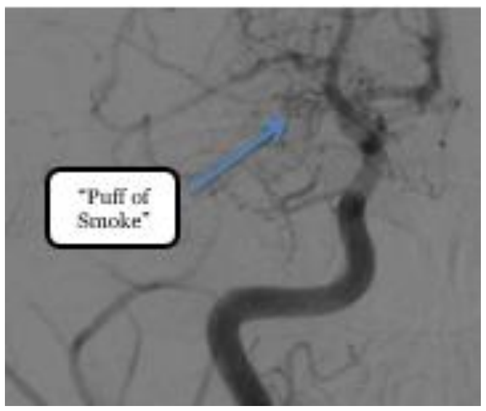


Fig 3. Cerebral angiogram showing classic "puff of smoke"

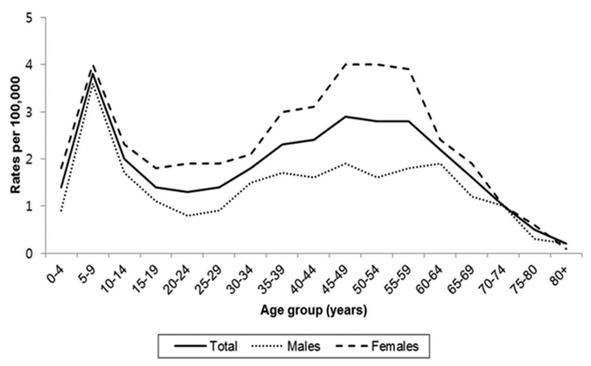


Fig 4. Bimodal age distribution of moymoya in Korea in 2007 – 2011

Discussion

- Moyamoya is a rare chronic, progressive, non-inflammatory occlusion of Circle of Willis arteries due to fibrocellular intimal thickening, forming collateral vessels termed **"puff of smoke"** in Japanese.
- Incidence in WA and CA: **0.086 per 100,000 people; 4 times more** in Asians than Caucasians.[2]
- Etiology unknown–believed to be autosomal dominant with incomplete penetrance. [3]
- Moyamoya syndrome: when associated with hereditary diseases such as sickle cell disease, NF-1. A **rare association with PKD** has been reported in two case reports.[4,5]
- **Bimodal age distribution** at 10 & 40-50 years old.

Adults	Children
• Headache • Hemorrhagic	• TIAs • Ischemic

- Diagnosis can be made with MR or CT angiogram, but **catheter angiography** is the **gold standard**. Criteria for diagnosis[6]: Bilateral stenoses of distal ICA or proximal ACA or MCA and abnormal vascular networks.
- Treatment: surgical bypass with superficial temporal artery to middle cerebral artery.
- Mortality rate in adults is reported at 10% if untreated, and **progressive neurologic deficit in 50-66%.**[1]

Teaching Points

- Moyamoya can occur in non-Asians and adults.
- Stroke-like presentation in an adult could be a sign of moyamoya.
- It has a rare association with polycystic kidney disease (PKD) – thus, obtaining family history is important.
- Swift diagnosis is crucial given 10% mortality rate in untreated adults.

References

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Fig 1: Virginia Brain and Spine Center; Fig 2, 3: from patient; Fig 4: Ahn IM, et al. Incidence, Prevalence, and Survival of Moyamoya Disease in Korea: A Nationwide, Population-Based Study. *Stroke* 2014;45:1090-1095