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.  
- It has association with hereditary diseases such as NF-1, sickle cell, as well as, has a rare association with PKD.

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: normal

- Further work-up including CRP, ANA, ANCA, HIV antibody, serum RPR, B-2 macroglobulin, and C3/C4 compliment levels all unrehearmal.
- 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.

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; HDL: 5.8
- UDS: normal

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.
- Etiology unknown—believed to be autosomal dominant with incomplete penetrance.
- 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.
- Bimodal age distribution at 10 & 40-50 years old.

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 moyamoya in Korea in 2007 – 2011

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

4. Przybyl JR, Massey JH. Moyamoya disease associated with polycystic kidney disease and cushingoid presentations. Department of Medicine, Duke University Medical Center, Durham, NC 27710.