Moyamoya Disease Presenting as an Intracranial Hemorrhage in a 69-year-old Man

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Background

- Moyamoya disease (MMD) is a very rare condition characterized by stenosis of large intracranial arteries with small collateral vessels at the base of the brain.[1,2]
- It is most common in women of East Asian populations and diagnosed based on radiographic evidence found with cerebral angiography.[3]
- Classic findings include small collateral vessels that appear as a “puff of smoke,” known as “moyamoya” in Japanese.
- The stenotic vessels and collaterals are prone to rupture or thrombosis, potentially resulting in either intracranial hemorrhage (ICH) or ischemic cerebral infarcts.[2]

Objective

To describe a case of MMD in an elderly Caucasian man presenting with first stroke and family history of ICH and aneurysms

Case Presentation

History:

- A 69-year-old man with past medical history of hypertension, type II diabetes mellitus, and rheumatoid arthritis was found unconscious by EMS after collapsing in public.
- He does not take blood thinners and had no history of stroke or cardiac disease. Family history was strong for prior ruptured aneurysms and ICH.

On exam:

- Upon arrival, his systolic blood pressure was in the 220s.
- He was awake, alert but was globally aphasic and had right-sided hemiplegia of upper and lower extremities with left-sided gaze preference and right homonymous hemianopsia.
- NIHSS was 22 on admission for left hemispheric syndrome.

Imaging:

- Initial non-contrast CT head showed left frontal ICH of 80 ccs with no midline shift nor hydrocephalus (Figure 1). Initial imaging also showed a severely stenotic left internal carotid artery (ICA) with terminal left ICA occlusion.

Pertinent labs:

- Serum ANA, c-ANCA and p-ANCA tests were unremarkable. CSF was not sampled due to hemorrhage.

Imaging

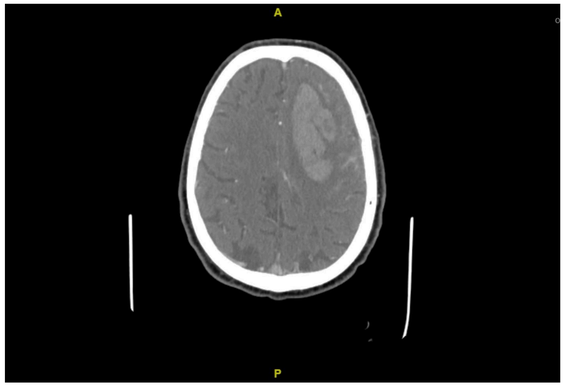

Figure 1. Non-contrast CT of the head demonstrating left ICH and leptomeningeal hyperintensities known as the “ivy sign.”

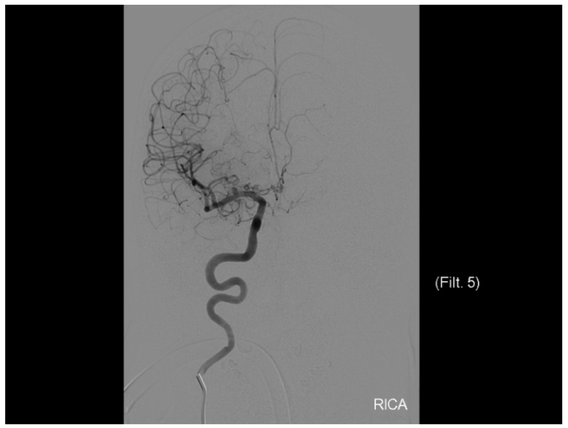

Figure 2. Right ICA cerebral angiography demonstrating stenosis with development of fine capillary network of anastomoses and hypoplasia of anterior cerebral arteries.

Disease Progression

- ICH remained stable and he progressively gained more strength in the right lower extremity.
- Digital subtraction angiography (DSA) was performed to further evaluate the left ICA stenosis, which identified a basilar apex aneurysm (measuring 6mm x 5.5mm), right ICA aneurysm (measuring 4.5mm x 3mm), severe narrowing of the left ICA with robust collateral flow to the left anterior cerebral circulation via the anterior communicating artery, the left posterior communicating artery, and robust pial-pial connections.
- The development of a fine capillary network at the right ICA terminus with hypoplasia of the bilateral anterior cerebral arteries and their branches was also noted, which prompted a full clinical picture of MMD (Figure 2).
- The patient was discharged a week after admission with close neurology outpatient follow up and annual neuro-interventional radiology for the multiple incidental aneurysms identified on DSA.
- The patient is still in recovery 3 months after.

Discussion + Conclusion

- MMD is becoming more frequently diagnosed in the U.S. despite associations with the East Asian population and with its bimodal age distribution
- Here we describe an elderly Caucasian man with MMD.
- His initial presentation with strong family history of aneurysms and hemorrhage, a spontaneous ICH, and a highly stenotic left ICA prompted additional imaging via DSA and led to the ultimate diagnosis of MMD.
- Long-term management and prognosis differ between spontaneous ICH and MMD; making the distinction is important.

References

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