Marianjoy Rehabilitation Hospital

part of Northwestern Medicine

A puff of smoke and a poor prognosis: A case report of Moyamoya disease

Beau Bigelow, MD; Aye Mon Win, DO; Dolly Devara, MD

Marianjoy Rehabilitation Hospital, Wheaton, IL

ROSALIND FRANKLIN UNIVERSITY of MEDICINE AND SCIENCE

Case Description

78-year-old woman of East Asian descent with hypertension and hyperlipidemia presented with headache, confusion, and fall with exam revealing right-sided hemiparesis, right neglect, and global aphasia. MRI demonstrated left PCA and MCA infarcts. CTA revealed Moyamoya anatomy in bilateral proximal MCAs. The patient was not deemed a candidate for tissue plasminogen activator (tPA) due to Moyamoya etiology of stroke. She was admitted for comprehensive stroke rehab. Due to barriers of global aphasia with minimal intelligible communication, severe oral apraxia, dysphagia, and dense right hemiparesis, she made minimal improvements in ADLs and ultimately required further subacute rehabilitation.

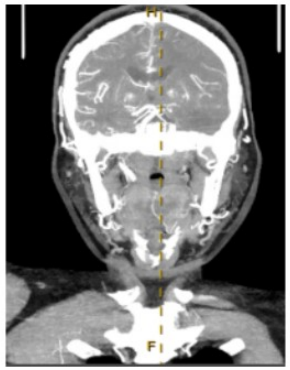

Figure 1. CT Head and Neck angiogram Coronal view. Shows Left PCA occlusion. Dashed Line corresponds to level of sagittal view in figure 2

Figure 2. CT Head and Neck angiogram Sagittal view. Shows left posterior lobe infarct and PCA occlusion Left side. Corresponds to dashed line in figure 1

Findings

Figure 3.CT Head and Neck angiogram of Left Internal carotid artery Curved AP view. There is occlusion of the left internal carotid artery at the carotid apex extending into the proximal left middle cerebral artery. (MCA not fully visualized)

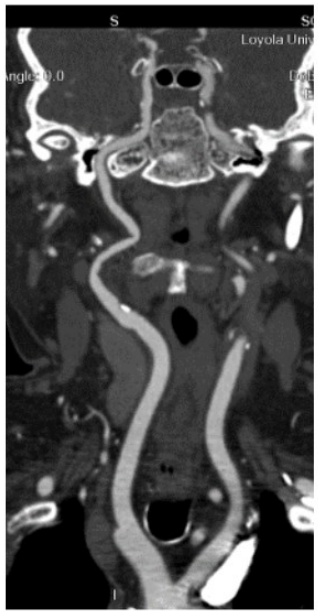

Figure 4. CT Head and Neck angiogram of Right Internal carotid artery Curved AP view. There is moderate to severe stenosis involving the supraclinoid portion of the right internal carotid artery with reconstitution of the caliber distally at the carotid apex

Discussion

Moyamoya disease is an often-inherited progressive cerebrovascular disease most commonly affecting Asian females. It is characterized by stenosis of the internal carotid arteries and Circle of Willis. Moyamoya, or "hazy puff of smoke" in Japanese, reflects the angiographic appearance of the collateral vessels. Resultant ischemia causes formation of abnormal vascular networks. In children, collateral pathway development is correlated with stage of disease; however, adults may lose the ability to form collateral networks. Optimal treatment is debated and there is no curative treatment. In the acute phase, tPA is contraindicated due to the high risk of bleeding in collateral vessels. Secondary stroke prophylaxis includes aspirin, calcium channel blockers, and possible surgical revascularization to improve blood flow abnormalities.

Conclusions

To our knowledge, there are few case reports of elderly patients who suffered from stroke secondary to Moyamoya disease in the United States. This case demonstrates the diagnosis of Moyamoya in a uniquely older patient who ultimately had a suboptimal acute rehabilitation course. Case reviews have demonstrated that that stroke patients with Moyamoya disease discharged from acute inpatient rehab had longer rehab courses with slower progress and overall lower FIM scores.

References

Newell D W., Abdu E (June 01, 2012) Moyamoya Disease: Current Concepts. Cureus 4(6): e47. doi:10.7759/cureus.47