CASE REPORT

NON-ATHEROSCLEROTIC OBLITERATION OF BILATERAL SUPRACLINOID INTERNAL CAROTID ARTERIES; A CLASSIC CASE OF MOYAMOYA DISEASE

Saad Ullah, Anas Ahmed Siddiqui*, Waseem Zaid Alkilani, Noman Ahmed Jang Khan, Hafiz Umair Siddiqui*

Department of internal medicine, Conemaugh Memorial Medical Centre, Johnstown, PA-USA, *Dow Medical College, Karachi-Pakistan

Moyamoya disease is an idiopathic progressive vasculopathy of distal internal carotid artery and circle of Willis which leads to the development of characteristic smoky appearance of the vascular collateral network on angiography. With the highest reported incidence among Japanese population, it has been under recognized as a cause of cerebrovascular accidents in Western countries. Here we report a case of a young 20-year-old Caucasian woman who presented to the emergency department with expressive aphasia, right arm weakness and numbness for three days. Imaging modalities confirmed Moyamoya disease.

Keywords: Internal carotid artery; Circle of Willis; Cerebral angiography

J Ayub Med Coll Abbottabad 2017;29(2):363–5

INTRODUCTION

Moyamoya disease is a progressive vaso-occlusive disease of the distal portion of Internal Carotid artery, and arteries of the circle of Willis.[1] The gradual evolution of stenosis in the arteries provides sufficient time for the formation of collateral vessels which gives an impression of a puff of smoke (Moyamoya is a Japanese word for a puff of smoke[2]) on angiography.[3] The disease predominantly affects Asian population but it has been reported in various ethnicities all around the world with varying age distributions and clinical manifestations.

CASE PRESENTATION

20-year-old Caucasian female with past medical history of mild aortic regurgitation, on combined oral contraceptive pills presented to the hospital with a three-day history of lethargy, expressive aphasia, right arm numbness and weakness. She denied any prior history of similar episode, seizure disorder, intravenous drug use, transient ischemic attack, strokes or tic disorder.

There was also no family history of neurofibromatosis, sickle cell disease or trait, and cerebrovascular disease besides a great aunt who had died from a stroke at the age of 70. On physical examination, she was alert and oriented to place and time, and could follow simple verbal commands. Expressive aphasia and naming impairment were evident. On neurologic examination, all cranial nerves were intact. Equal sensation and strength was noted except for 4/5 strength in her right upper and lower extremities. Deep tendon Reflexes were normal and symmetrical. Gait examination was normal as well. Computed Tomography of the head showed subtle areas of decreased attenuation within the deep white matter of the left frontal lobe (Figure-1). Magnetic resonance angiography (MRA) of head revealed obliteration of the bilateral supraclinoid internal carotid arteries with anterior cerebral arteries reconstituting via pial collaterals (Figure-2). A1, P1, and M1 segments of Anterior, posterior and middle cerebral arteries respectively could not be visualized on MRA establishing the diagnosis of Moyamoya disease. Her blood work including Echocardiography, ANA, complement level, rheumatoid factor and ESR was unremarkable. She received physical therapy and occupational therapy during the hospitalization. Subsequently she was discharged to a rehabilitation centre. At the time of discharge, she had residual right-sided weakness and expressive aphasia. Few months after presentation, superficial temporal artery - middle cerebral artery (STA-MCA) bypass surgery was performed bilaterally, one month apart. She currently has minimal residual expressive aphasia without other, neurological deficit.

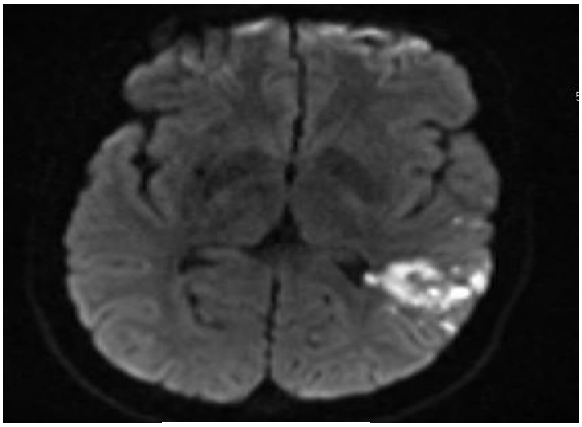

Figure-1: CT head, showing subtle areas of decreased attenuation within the deep white matter of the left frontal lobe.

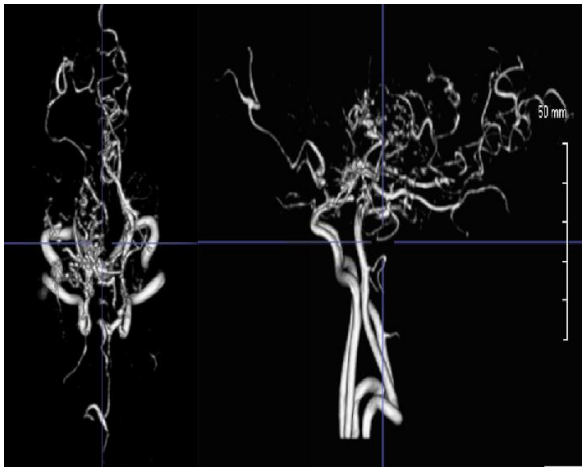

Figure-2: obliteration of the bilateral supraclinoid internal carotid arteries

DISCUSSION

Moyamoya disease was first reported in japan in 1957[1] and its incidence is reported to be the highest in Japan with females being affected almost twice as likely as males.[4] The Incident rate of 0.086/100,000 was reported in a study done in California and Washington, i.e., 4–13 times lower than its incidence in japan. Of note though, the incidence in Asian individuals in these states was comparable to the incidence rate in Japan. Our patient was a female Caucasian. The incidence of Moyamoya disease for the caucasian population is defined as 0.06 per 100,000[5]

Moyamoya disease has bimodal age distribution. The highest peak occurs in the late 40s and the second between 5 and 9 years of age[6] as compared to our patient who was 20 years old.

Arteries that are usually involved in the Moyamoya disease include distal carotid arteries and the proximal portion of Anterior and middle cerebral arteries. Posterior cerebral arteries and other arteries that provide circulation to the posterior part of the brain are rarely involved,[7] as was the case with our patient, Bilateral supra-clinoid internal carotid as well as Anterior Cerebral arteries were affected. Vasculature changes in renal, pulmonary, and coronary vessels has also been observed in this disease.[8]

The disease usually presents as a recurrent headache that is resistant to analgesics and other medical therapy with surgery being the only cure that alleviates the symptoms. The disease could also follow a silent course with stroke being the first presentation of the disease. Our patient presented with three-day history of lethargy, expressive aphasia, right arm numbness and weakness. Studies like CSF analysis and electroencephalogram do not aid in diagnosis.[9] Cerebral angiography, MRA and CTA are usually used to diagnose Moyamoya disease It is important to exclude the other conditions that cause atherosclerosis and vasculopathy before the diagnosis of Moyamoya disease is made. The coexistence of such diseases and typical features of Moyamoya disease in cerebral vessels lead to the diagnosis of Moyamoya syndrome instead of Moyamoya disease.[10]

Definitive treatment of Moyamoya disease does not exist yet. Different Medical therapies have been used to avoid the ischemic or haemorrhagic episodes associated with the disease but they have not provided any significant results.[11] However, surgical revascularization procedures have shown some symptomatic improvement in pediatric population.[12] Long-term trials suggest that patients who had undergone surgical revascularization process had a slower decline in cognitive function and fewer ischemic attacks.[13,14]

Our patient underwent superficial temporal artery - middle cerebral artery (STA-MCA) bypass surgery bilaterally, one month apart and she has minimal residual expressive aphasia without another neurological deficit.

CONCLUSION

Moyamoya disease, though rare, should be considered as a differential for patients with cerebrovascular disease who lack the usual risk factors and comorbidities.

CONFLICT OF INTEREST

There is no conflict of interest to be disclosed.

REFERENCES

1. Takeuchi K, Shimizu K. Hypogenesis of bilateral internal carotid arteries. No To Shinkei 1957;9:37–43.
2. Suzuki J, Takaku A. Cerebrovascular `moyamoya' disease: disease showing abnormal net-like vessels in base of brain. Arch Neurol 1969;20:288–99.
3. Kitamura K, Fukui M, Oka K. Moyamoya disease. In: Handbook of Clinical Neurology. Vol 2. Amsterdam, The Netherlands: Elsevier; 1989. p.293–306.
4. Wakai K, Tamakoshi A, Ikezaki K, Fukui M, Kawamura T, Aoki R, *et al*. Epidemiological features of moyamoya disease in Japan: findings from a nationwide survey. Clin Neurol Neurosurg 1997;99(Suppl 2):S1–5.
5. Uchino K, Johnston SC, Becker KJ, Tirschwell DL. Moyamoya disease in Washington State and California. Neurology 2005;65(6):956–8.
6. Baba T, Houkin K, Kuroda S. Novel epidemiological features of moyamoya disease. J Neurol Neurosurg Psychiatry 2008;79(8):900–4.
7. Yamashita M, Oka K, Tanaka K. Histopathology of the brain vascular network in moyamoya disease. Stroke 1983;14(1):50–8.
8. Ikeda E. Systemic vascular changes in spontaneous occlusion of the circle of Willis. Stroke 1991;22(11):1358–62.
9. Hoare AM, Keogh AJ. Cerebrovascular moyamoya disease. Br Med J 1974;1(5905):430–2.
10. Fukui M. Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis ("moyamoya" disease). Research Committee on Spontaneous Occlusion of the Circle of Willis (Moyamoya disease) of the Ministry of

Health and Welfare, Japan. Clin Neurol Neurosurg 1997;99(Suppl 2):S238–40.
11. Spittler JF, Smektala K. Pharmacotherapy in moyamoya disease. Hokkaido Igaku Zasshi 1990;65(2):235–40.
12. Fung LW, Thompson D, Ganesan V. Revascularization surgery for pediatric moyamoya: a review of the literature. Childs Nerv Syst 2005;21(5):358–64.
13. Fukui M. Current state of study on moyamoya disease in Japan. Surg Neurol 1997;47(2):138–43.
14. Imaizumi T, Hayashi K, Saito K, Osawa M, Fukuyama Y. Long-term outcomes of pediatric moyamoya disease monitored to adulthood. Pediatr Neurol 1998;18(4):321–5.

Received: 4 September, 2016	*Revised: 3 October, 2016*	*Accepted: 11 October, 2016*

Address for Correspondence:
Saad Ullah, Department of Medicine, Conemaugh Memorial Medical Centre, Johnstown, PA-USA
Tel: +1 917-975-5607
Email: Saadullah_98@hotmail.com