Could unilateral Moyamoya vasculopathy be a response to repetitive head trauma?

A Case of Subacute Subdural Hematoma with Chronic Middle Cerebral Artery Dissection and Infarction in a Young Soccer Player

By Poon, W.C.; Ho, L.Y.; Lee, W.Y.; Pang K.Y.; Chow Tom S.F.*
*Department of Neurosurgery, Pamela Youde Nethersole Eastern Hospital

Background

Moyamoya disease and moymoya syndrome[1]

- Discovered as a a new disease entity by Suzuki et. al. in 1963.
- A condition characterized by progressive stenosis of the terminal internal carotid artery and the proximal portions of the anterior cerebral and middle cerebral arteries, followed by the development of prominent collateral small vessels.
- These vessels produce a characteristic smoky appearance on angiography (also known as “moyamoya angiopathy”)
- Hence the name: “moyamoya” refers to a Japanese word meaning something hazy like a puff of smoke in the air.
- If moyamoya vasculopathy results from underlying disease, we call it Moyamoya syndrome; otherwise, it is moyamoya disease.

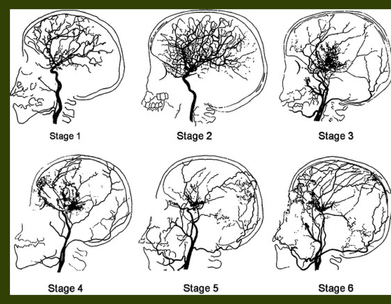
Usual presentation[2]

- ischemic complications: transient ischemic attacks, cerebral infarct
- haemorrhagic complications: intracranial haeomrrhage

Clinical approach

For patients presenting with ischemic or haemorrhagic complications due to moyamoya angiopathy confirmed with imaging studies, to ascertain the underlying etiology, diagnostic evaluation would be carried out, which included

- history taking
- physical examination
- blood biochemistry.



6 stages of moyamoya disease as proposed by Suzuki[1]

Treatment[2]

Currently the only well established treatment is various forms of revascularisation surgery to prevent future complications. Pharmacological treatment to prevent disease progression currently still lacks supportive evidence.

Case report

Known etiologies of moyamoya angiopathy included meningitis, atherosclerosis, radiation, neurofibromatosis, Down syndrome, sickle cell disease, and neoplasms.[3] Trauma was initally not recognized as a cause of moyamoya syndrome in the 1960s. Later, moyamoya angiopathy was observed to be associated with cranial trauma after 1970s. So far only a handful of case reports were published.[4] Cranial trauma was later recognized to be one of the cause of moyamoya vasculopathy.[3] The exact mechanism, however, remained obscure. We present here an unusual case where the likely cause of unilateral moyamoya vasculopathy is chronic repetitive head injury. This patient also has focal stenosis and dissection of M1 which may shed light on how trauma induces moyamoya angiopathy.

Patient description

- Patient Y. 21-year-old tall and thin young man.
- Frequent football player with frequent headers, playing football 3 times a week

History of presenting illness

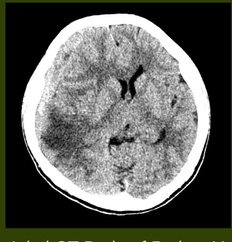
- Sudden onset mental slowing and subjective left sided weakness
- preceded by headache, dizziness and decreased appetite for 1 week
- no previous surgery or irradiation
- no childhood illness
- no family history of connective tissue disease
- no learning disability

Physical examination

- full Glasgow Coma Score
- power at grade 4 over the left side upper and lower limbs (according to Medical Research Council grading)
- no fever or neck stiffness
- no cafe-cu-lait spots or neurofibromata
- general examination and cardiovascular examination was unremarkable

CT Brain upon admission

- right chronic subdural haematoma with infarct.



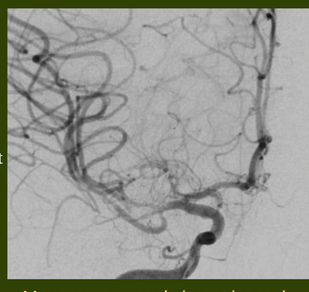
Inital CT Brain of Patient Y upon admission

Immediate treatment

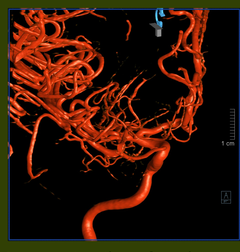
- Emergent Burr hole drainage of right chronic subdural haematoma
- Found established multiple layers of subdural membranes of different ages, implying the chronicity of the injury.
- The patient developed right epidural haematoma after Burr hole drainage, which was immediately evacuated with a craniotomy. The patient recovered well with mild residual left sided weakness at grade 4.

Digital subtraction angiography performed after Burr hole drainage

- stenosis and suspected dissection of the right M1
- unilateral moyamoya angiopathy (stage 2) over the right middle cerebral artery.



Moyamoya vessels in angiography of Patient Y



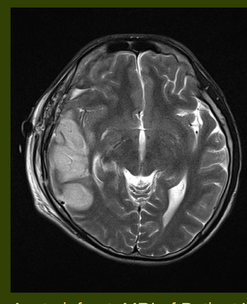
3D reconstruction of angiography of Patient Y showing vessels with puff of smoke appearance

Laboratory evaluation

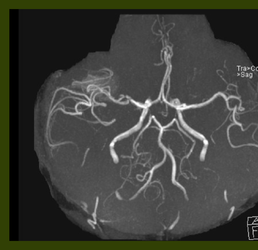
- normal haemoglobin level, white cell counts and platelet counts.
- normal liver and renal function tests
- normal clotting profile.
- negative immunological markers such as ANA, ANCA and RF
- inflammatory markers such as CRP and ESR were mildly elevated

Magnetic resonance angiogram

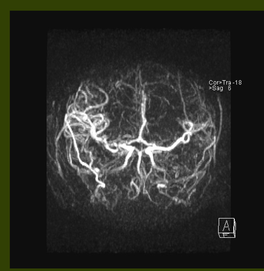
- performed to assess the degree of angiopathy
- showed middle cerebeal aretery (MCA) territory infarct.
- focal stenosis over the distal right M1 and origin of right M2
- suspicious puff of smoke appearance in adjacent cisterns
- no definite vessel wall enhancement to suggest active inflammatory activity.



Acute infarct, MRI of Patient Y



Multiple overlapping thin slap acquisition, MRA of Patient Y



Contrast-enhanced MRA of Patient Y

Discussion

Our proposed diagnosis: moyamoya syndrome induced by chronic repetitive head injury.

Reasons:
With young age, negative immunological markers, normal haemoglobin level, normal white cell count, and negative physical examination finding and clinical history, it was unlikely that our patient has other aforementioned known causes of moyamoya vasculopathy other than trauma. We would argue that trauma causes unilateral moyamoya phenomenon in this case.

We would like to make a hypothesis that multiple mild head injuries introduced shear forces to brain structures. Shear injury caused intimal injury to the MCA and silent dissection, which healed with intimal repair. Repetitive injuries lead to wall thickening and relative hypoperfusion leading to lenticulostriate hypervasculogenesis and early moyamoya syndrome. Patient’s subsequent decompensation could be related to the gradually expanding chronic subdural haematoma, distorting of stenotic MCA, new dissection, precipitation of critical stenosis and infarction.

Another possibility not completely excluded: patient has moyamoya disease that presented unilaterally. Repetitive head injury caused moyamoya vessels to rupture casuing subdural haematoma and dissection in the MCA. While it is likely for moyamoya disease to present unilaterally, it is rarely for the interncal carotid artery to remain uninvolved. It will be useful to repeat angiogram in 6 months.

Conclusion

Chronic repetitive head injuries could predispose to MCA dissection, resulting in infarct of the brain tissue it supplied and causing Moyamoya phenomenon of the affected vessels. It is advised that protective helmets should be worn during contact sports.

References:

1. Suzuki J, Kodama N. Moyamoya disease--a review. Stroke. 1983 Jan-Feb;14(1):104-9.
2. Cong Han, et. al. Moyamoya Disease in Asia. Neurology Asia 2012; 17(3) : 175 – 181.
3. Research Committee on the Pathology and Treatment of Spontaneous Occlusion of the Circle of Willis; Guidelines for diagnosis and treatment of moyamoya disease (spontaneous occlusion of the circle of Willis). Neurol Med Chir (Tokyo). 2012;52(5):245-66.
4. Fernandez-Alvarez E, et. al. "Moya-moya' disease caused by cranial trauma. Brain Dev. 1979;1(2):133-8.