

Headache Associated with Sexual Activity Unveiling Moyamoya Disease

Dear Editor,

Moyamoya angiopathy (MMA) is an intracranial vasculopathy characterized by bilateral progressive steno-occlusive changes in the intracranial portion of internal carotid arteries (ICA) and proximal portions of the anterior cerebral artery (ACA) and/or middle cerebral artery (MCA) with the development of compensatory abnormal vascular network at the base of the brain.^[1] MMA can present with a wide range of neurological symptoms, which can be less infrequently subtle and may lead to a delayed diagnosis of MMA.^[2,3] Headache in MMA is not uncommon, but frequently under-recognized.^[4] We herein report an extremely rare clinical scenario of headache associated with sexual activity unmasking a diagnosis of underlying MMA.

A 42-year-old man from rural India, without any comorbidity, presented to us with three episodes of sudden-onset severe, throbbing holocranial headache just before orgasm during sexual activity in the last 1 month. The severe headache would last for 10–12 minutes, followed by a milder-intensity throbbing headache that would gradually subside over hours of taking oral paracetamol tablets and sleep. There was no history of transient or fixed paresis, seizure, cognitive or behavioral symptoms, or similar episodes in the past. He had initially consulted his local physician, where he was only prescribed analgesics without pursuing brain imaging. The patient sought our consultation following the third episode of headache. Magnetic resonance imaging of the brain was performed, and it revealed a few scattered, nonspecific signal changes

in T2-weighted images. Magnetic resonance angiography revealed stenosis of the supraclinoid ICA, MCA and ACA with collateral formation suggestive of MMA [Figure 1]. Relevant investigations were conducted to rule out secondary causes of MMA.^[1] The patient was counseled to assume a more passive position during sexual activity. He was prescribed oral naproxen (500 mg) for acute symptomatic therapy and planned for revascularization surgery.

The symptom of headache, in association with or without other neurological deficits, is well documented in MMA. The prevalence of headache in MMA varies with ethnicity and geographical distribution, as evidenced by its frequency of 50% in Europe, 21.6–25% in South Korea, and 36.8% in India.^[1–6] Headache in MMA most commonly resembles migraine, either alone or combined with tension-type headache.^[3] However, headache phenotypically mimicking headache associated with sexual activity in MMA is very uncommon.

The pathophysiological basis for headache in MMA is largely speculative. The proposed hypothesis includes (I) stimulation of dural nociceptors by dilated leptomeningeal collaterals in MMA and (II) chronic cerebral hypoxia and resultant microvascular ischemia triggering cortical spreading depression.^[6] Das *et al.*^[1] classified headache associated with MMA as a transient perfusion-dependent neurological symptom. These transient perfusion-dependent neurological symptoms in MMA often occur in close temporal association with the occurrence of precipitating factors, including heavy exercises and heightened emotional states, both of which may occur during sexual activity.^[7,8] It has been long thought that orgasmic headache occur in the setting of trigeminovascular activation, leading to vasodilation from the release of inflammatory neuropeptide.^[9] A similar mechanism leading to the accentuation of dilation of leptomeningeal collaterals may be underpinning the headache associated with sexual activity in our patient, wherein the physical exercise and heightened emotions associated with sexual activity acted as a precipitating factor.

A lack of awareness regarding the subtle neurological symptoms of MMA on the part of the treating physician can often lead to delayed diagnosis.^[4,7] Our patient was initially considered as having a primary headache associated with sexual activity by his primary physician, and brain imaging was deferred. However, brain and intracranial vascular imaging is warranted in patients presenting with new-onset headache associated with sexual activity. This is to rule out grave neurological conditions, such as subarachnoid hemorrhage, arterial dissection, intracranial aneurysm, and reversible cerebral vasoconstriction syndrome (RCVS).^[9] However, MMA is almost never considered as a secondary etiology to headache associated with sexual activity, due to the rarity of the association between the two clinical conditions.

Headache remains an important and subtle neurological symptom in MMA, and it can mimic the phenotypes of varied primary headache. Though uncommon, MMA should be a rare

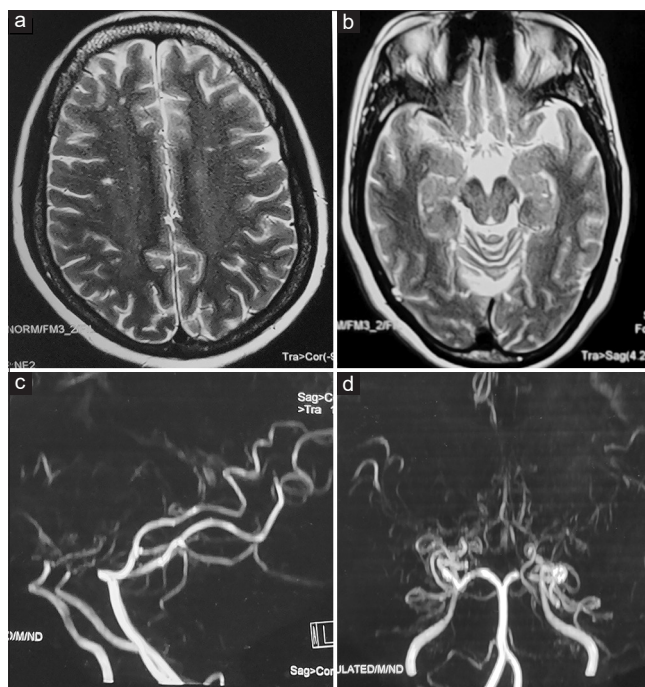


Figure 1: Magnetic resonance imaging of the brain T2-weighted sequence showing few scattered white matter hyperintensities (a) and attenuated anterior cerebral circulation at the level of the circle of Willis (b); and magnetic resonance angiography showing stenosis of distal ICA, thinned out MCA, and ACA along with collateral formation suggestive of Moyamoya angiopathy (c and d)

etiological consideration in headache associated with sexual activity.^[10]

Data availability

Data are not provided in the article because of space limitations but may be shared (anonymized) at the request of any qualified investigator for the purposes of replicating procedures and results.

Informed consent for participation in research study

The patient's legally authorized representative consented (written) to participate in the study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 19-Jul-2023 **Revised:** 30-Aug-2023 **Accepted:** 22-Sep-2023
Published: 29-Nov-2023

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DOI: 10.4103/aian.aian_637_23