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LETTER TO THE EDITOR

# Temporal lobe epilepsy with nocturnal wandering leading to discovery of Moyamoya Angiopathy

**Shambaditya Das**[1] · **Biman Kanti Ray**[1] · **Souvik Dubey**[1]

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Dear Editor,

Moyamoya angiopathy (MMA) is a chronic progressive occlusive intracranial vasculopathy characterized by angiographic findings of stenosis or occlusion at the terminal portion of the internal carotid artery (ICA) or proximal anterior cerebral artery (ACA) and/or middle cerebral artery (MCA) with compensatory collateral vessels at the base of the brain [1]. Previously considered to affect predominantly persons of Asian heritage (particularly Japanese), recent evidence suggests MMA is not uncommon, affecting individuals of many ethnic backgrounds throughout the world, including American and European populations. It is known to present with myriad of nonparetic symptoms. Seizures can account for 10.5% presenting symptom in MMA; various seizure types including focal seizure with or without secondary generalization, generalized seizure and febrile seizure may be seen. However, to the best of our knowledge, wandering in sleep is unheard of as a presenting and sole manifestation of MMA [2]. We herein present an index case of MMA in a young female unmasked during evaluation of walking in sleep.

A 16-year-female, initially evaluated by primary-care physician, sought our care with parents complaining of infrequent (2–3/year) episodes of wandering in sleep for last 6 years. On detailed query, the episodes lasted for 4–5 min with the patient getting up from sleep and walking around in the room, mumbling incomprehensible words with twitching around the angle of the mouth and finally returning to her bed, while regaining consciousness but remaining confused. This episodes used to occur at midnight or early morning, usually 4–5 h after falling asleep. There was no similar history in the family. Neurological examination revealed no significant abnormality.

Routine blood investigations were normal. Nocturnal video-electroencephalogram (EEG) and polysomnography (PSG) testing were unrevealing. Magnetic resonance imaging (MRI) brain showed multiple flow voids around the basal ganglia leading to a suspicion of chronic vaso-occlusive disease which was confirmed in MR angiography that revealed a bilateral terminal ICA steno-occlusive lesion with multiple collaterals suggestive of MMA. Positron emission tomography with computed tomography (PET-CT) indicated severe hypometabolism of bilateral mesial temporal lobe and hypermetabolic cingulate cortex (Fig. 1). Detailed evaluation for secondary causes of MMA was non-contributory.

Parents refused to any invasive investigation or therapy. Hence, the patient was managed conservatively with antiplatelet and anti-epileptic (oral oxcarbazepine) therapy, considering it to be an episodic nocturnal wandering in temporal lobe epilepsy. The patient has been under our follow-up for last one and half years without any reports of similar episodes or other new onset fixed or transient neuro-deficits.

Parasomnias and epileptic seizures have been known to cause abnormal paroxysmal events in sleep. NREM parasomnias like sleep-walking usually occur in the earliest sleep cycle, in NREM sleep, lasting for several minutes or longer, with the patient usually returning to bed at the end of the event. Episodic nocturnal wandering due to seizure, however, can occur at anytime throughout the night, often in stage 1/2 of NREM sleep, usually lasting less than 2 min, with the patient often not returning to bed at

✉ Souvik Dubey
drsouvik79@gmail.com

Shambaditya Das
drshambadityadas@gmail.com

Biman Kanti Ray
bimankantiray2019@gmail.com

[1] Department of Neurology, Institute of Post Graduate Medical Education and Research, Bangur Institute of Neurosciences, 52/1A Shambunath Pandit Street, Kolkata 700025, India

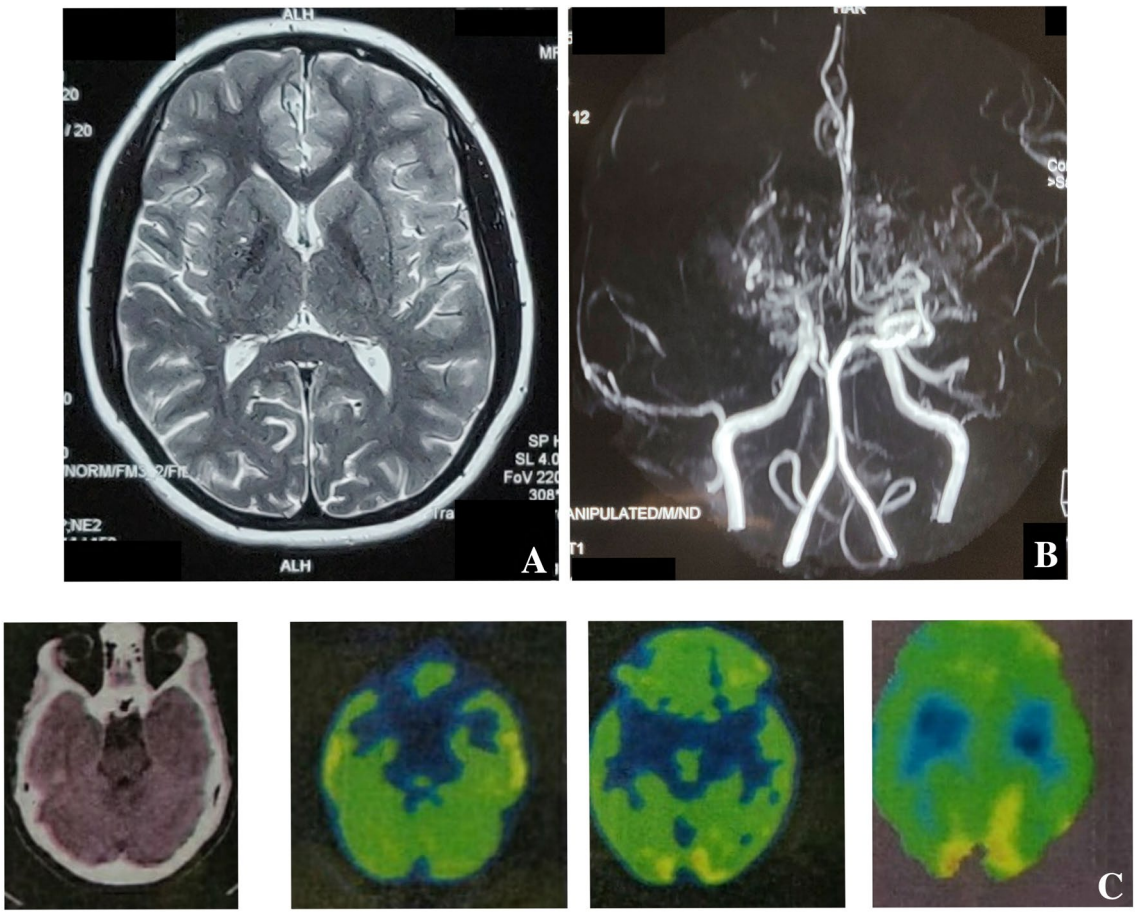


**Fig. 1** MRI brain T2 sequence shows multiple flow voids around the basal ganglia in both the cerebral hemisphere (**A**) and MR angiography shows bilateral terminal ICA stenosis with non-visualization of MCA, proximal ACA with multiple collaterals appearing as "puff of smoke" (**B**). PET-CT showing hypometabolism in bilateral mesial temporal lobe (**C**)

the end of the event. The presence of awakening, vocalization, automatism and post-ictal phenomenon is more commonly noticed in episodic nocturnal wandering than in sleep-walking [3, 4].

Both frontal and temporal origin seizures have been noted with episodic nocturnal wandering. In comparison to frontal lobe epilepsy, seizures in nocturnal temporal lobe epilepsy are less frequent, non-clustered and lack hyperkinetic activity, complex motor automatism with tonic contraction and have fewer positive family history [3]. Our patient with shorter duration, vocalization, automatism and confusion following ictus suggested a diagnosis of a seizure, further corroborated by responsiveness to anti-epileptic therapy. An infrequent, non-clustered attack, without complex automatism and tonic contraction pointed towards a temporal lobe origin nocturnal seizure in our case.

While video-EEG-PSG is considered the "gold standard" investigation, in situations of infrequent events, it may fail to generate conclusive evidence towards the diagnosis as was seen in our case. In such a setting, it is the elicitation of meticulous history that helps to arrive at a correct diagnosis [3].

The cingulate cortex as a part of functional network is known to modulate behaviors in emotional processes and, through the hippocampus and para-hippocampus, constitutes a bridge between the temporal and frontal lobe. It has been shown to be hyperperfused in both nocturnal frontal lobe seizure and sleep-walking, possibly responsible for the generation of the complex motor behavior during sleep [5]. Our case also demonstrated cingulate hypermetabolism possibly accounting for the wandering in sleep following the temporal origin ictus.

Moyamoya can present with a wide array of non-motor paretic symptoms, posing a great diagnostic dilemma to the physician [2]. A low index of suspicion and increased awareness of the rarer presentations of this rather uncommon disease can greatly aid in early diagnosis, management and favorable prognosis.

**Author contributions** CRediT Taxonomy. SD1: conceptualization—equal, data curation—lead, formal analysis—equal, investigation—equal, methodology—equal, resources—equal, supervision—equal, visualization—equal, writing-original draft—lead, and writing-review and editing—equal. BKR: conceptualization—equal, project administration—equal, supervision—equal, visualization—equal, writing-review and editing—equal. SD2: conceptualization—equal, formal analysis—equal, methodology—equal, project administration—equal, supervision—equal, visualization—equal, and writing-review and editing—equal.

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**Data availability** Data not provided in the article because of space limitations may be shared (anonymized) at the request of any qualified investigator for the purposes of replicating procedures and results. All data relevant to the study are included in the article or uploaded as supplementary information.

## Declarations

**Conflict of interest** The authors have stated explicitly that there are no conflicts of interest in connection with this article.

**Ethical approval** All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed consent for participation in research study** The patient's legally authorized representative consented (written) to participate in the study and approval was obtained from the ethics committee of Institute of Post-graduate Medical Education and Research, Kolkata, India (Ethics committee reference number-ECR/35/Inst/WB/2013/RR-16).

**Authorship agreement** I, Dr Souvik Dubey, take full responsibility for the data, the analyses and interpretation, and the conduct of the research. I have full access to all the data and have the right to publish any and all data, separate and apart from the guidance of any sponsor.

## References

1. Das S, Dubey S, Acharya M, Chatterjee S, Lahiri D, Das G et al (2019) Thalassemia and Moyamoya syndrome: unfurling an intriguing association. J Neurol [Internet] 266(11):2838–47 [cited 2020 Aug 30]. Available from: https://pubmed.ncbi.nlm.nih.gov/31422456/
2. Das DS, Dubey DS, Acharya DM, Ghosh DR, Chatterjee DS, Hazra PA et al (2020) The disease presentation of Moyamoya angiopathy in Eastern India. J Stroke Cerebrovasc Dis 29(8):104957
3. Derry CP, Davey M, Johns M, Kron K, Glencross D, Marini C et al (2006) Distinguishing sleep disorders from seizures: diagnosing bumps in the night [Internet], vol 63, Archives of Neurology. American Medical Association, p. 705–9 [cited 2021 Jun 11]. Available from: https://jamanetwork.com/
4. Hughes JR (2007) A review of sleepwalking (somnambulism): the enigma of neurophysiology and polysomnography with differential diagnosis of complex partial seizures [Internet], vol 11, Epilepsy and Behavior. Elsevier, pp 483–91 [cited 2021 Jun 11]. Available from: http://www.epilepsybehavior.com/article/S1525505007002995/fulltext
5. Bassetti C, Vella S, Donati F, Wielepp P, Weder B (2000) SPECT during sleepwalking. Lancet [Internet] 356(9228):484–5 [cited 2021 Jun 11]. Available from: http://www.thelancet.com/article/S0140673600025617/fulltext