A case report of Moya Moya disease presenting with seizure in a Caucasian adult.

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ABSTRACT

Moyamoya disease is a chronic cerebrovascular disease, more common in Japanese and Asian population. It is rare in European and American population. The typical presentation is seizure, Transient Ischemic Attack or Stroke and diagnosed radiologically with typical angiographic appearance of the vascular collateral network. We are reporting this unusual case of Moyamoya Disease in a Caucasian adult who presented to A&E with an episode of tonic colonic seizure. In past she had Recurrent Transient Ischemic Attacks so as a part of workup for stroke and seizure, all relevant workup was done. Carotid doppler showed an increase in end-diastolic flow velocity and a decrease in vascular resistance in both external carotid arteries. Subsequently, MRI Brain and MRA showed a rare Moyamoya disease. This disease should be considered an important differential in patients presenting with recurrent seizure episodes or neurological symptoms.

Keywords: Cerebro-vascular disease, Seizure, Transient ischemic attack, Stroke, Neurological symptoms, Moyamoya disease

How to Cite This:

Sajjad MM, Yousaf S. A case report of Moya Moya disease presenting with seizure in a Caucasian adult. Isra Med J. 2020; 11(3): 164-165.

INTRODUCTION

Moyamoya disease (MMD) is a chronic progressive disease of cerebral vasculature which was described in 1957 for the first time in Japan. It is of unknown aetiology with a higher incidence among the Japanese and Asian population. MMD is less frequently seen in North America and Europe. It has bi-modal age of presentation, with children at around five years and adults presenting around 40 year of age[1]. Transient ischemic attack or ischemic stroke is a typical clinical presentation in children, while in adults it typically presents as a haemorrhage stroke or a bleed. Less commonly, MMD may present with a headache or a seizure. It has an unknown aetiology, although RNF213 in the 17q25-ter region is recognized as an important susceptibility gene in East Asian populations for MMD[2]. CT scan and MRI Brain are useful for detection of ischemia or haemorrhage in MMD patients. Hemodynamic studies such as Trans-carotid Doppler can be used as initial non-invasive investigation and the findings can point towards distal stenosis which warrants further investigations for workup of MMD[3]. Angiographic studies such as MRI angiography and CT angiography are diagnostic for this condition with typical angiographic findings. We are reporting this unusual case of Moyamoya Disease in a Caucasian 47 year's old lady who presented to Emergency Department with an episode of tonic colonic seizure and past history of Recurrent Transient Ischemic Attacks

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Received for Publication: December 23, 2019
1st Revision of Manuscript: February 08, 2020
Accepted for Publication: September 02, 2020

CASE REPORT

We report a 43 years old lady who presented to the hospital with an episode of tonic clonic seizure lasting for 2,3 minutes with incontinence and no prodromal symptoms. It was followed by postictal confusion and drowsiness. She had history of 2 episodes of Transient Ischemic Attack 8 months back. Family history was significant for stroke on maternal side. She was a lifelong non-smoker with a healthy life style and dietary habits. Neurological examination was unremarkable. A CT Brain, Telemetry and Echo did not reveal any abnormality, Carotid duplex ultrasonography showed an increase in end-diastolic flow velocity and a decrease in vascular resistance in both external carotid arteries, subsequently an MRI Brain and MRA Brain was planned after the report of carotid Doppler. MRI Brain showed generalized atrophy with attenuation of left internal carotid artery (ICA) flow void and presence of multiple flow voids around left ICA. MR Angiogram Brain (a) showed multiple collaterals in the suprasellar cistern. DSA showed bilateral supraclinoid ICA stenosis with no visualization of circle of Willis, vascular collaterals, and enlarged extra cranial collaterals (b,c,f). Left proximal PCA (P1) was stenosed with hypertrophied perforators as seen on left vertebral artery injection (d,e).The diagnosis of Moya Moya disease was made and she was later on transferred to a specialized neurosurgical unit for further management.

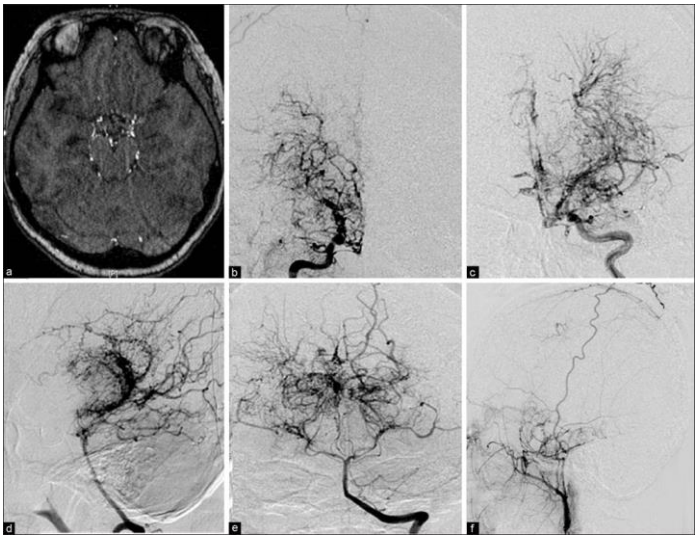

Figure-1: (a,b,c,d,e,f): MR Angiogram Brain (a) showed multiple collaterals in the suprasellar cistern.

DSA showed bilateral supra-clinoid ICA stenosis with no visualization of circle of Willis, vascular collaterals (b,c,f). Left proximal PCA (P1) was stenosed with hypertrophied perforators as seen on left vertebral artery injection (d,e).

DISCUSSION

Diagnosing MMD may be challenging. The clinical manifestations of moyamoya are variable and include transient ischemic attack (TIA), ischemic stroke, haemorrhagic stroke, and epilepsy. In our case although the initial presentation was a seizure, but keeping in mind past medical history of transient ischemic attacks so it was decided to investigate the patient according to the workup for TIA in addition to workup for seizure. Transient ischemic attacks, Intellectual decline, seizures, and involuntary movements are more common in paediatric population, and in contrast, adult patients present with intracranial haemorrhage[2]. In our case all the routine investigations including CT Brain, Telemetry and Echo was normal. The clue for further investigations was warranted by findings of carotid doppler. MRA and MRI Brain findings were consistent with diagnosis of MMD.

Neuroimaging plays an important role in the diagnosis of MMD. MRI Brain can detect lacunar ischemic zones while diffusion MR can be used for detecting acute ischemia. Magnetic resonance angiography (MRA) can determine stenosis in the distal internal carotid the arteries around the circle of Willis. MRA can also demonstrate the collateral "moyamoya vessels" in the basal region of the brain[4,5]. Transcranial Doppler ultrasonography (TCD) provides a noninvasive way to evaluate intracranial hemodynamics by measuring blood flow velocity in large intracranial vessels at the circle of Willis. It can be helpful in describing the flow across the carotids and can guide for further investigation as in or case.

The acute management for patients presenting with MMD is mainly symptomatic and is directed towards reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures.[6] The ultimate management is surgical revascularization of patients with moyamoya to reduce ischemic and haemorrhagic stroke rate and improve neurological and neuropsychological outcome[7]. Moyamoya disease has a progressive course and if left untreated can result in cognitive and neurologic decline due to recurrent ischemic or haemorrhagic strokes[8].

CONCLUSION

This case brings the importance of keeping in mind the differential of MMD in patients presenting with recurrent TIA's or seizures and also highlights the rare presentation of MMD in a Caucasian decent. It also highlights the importance of non-invasive investigations like carotid Doppler in suspected cases as an initial investigation for directing towards the diagnosis.

AUTHOR'S CONTRIBUTION

Sajjad MM: Conceived idea, Designed research methodology, Data analysis, Manuscript writing
Yousaf S: Manuscript drafting, Data compilation, Data analysis

Disclaimer: None.
Conflict of Interest: None.
Source of Funding: None.

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