



## Case Report

## Moyamoya Disease in an adult female from Nepal: A case report

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## ABSTRACT

**Introduction and importance:** Moyamoya is a progressive vasoocclusive disease of large intracranial arteries with characteristic collaterals formation. It has a bimodal distribution and more frequent in females compared to males.

**Case presentation:** We present a case of 38 years female who presented with a loss of consciousness following headache. She was investigated with a head CT, cerebral angiogram, and digital subtraction angiography which led to a diagnosis of Moyamoya disease. She was treated conservatively and scheduled for follow-up.

**Clinical discussion:** Although ischemic attacks are the common presenting symptoms in cases of Moyamoya disease, hemorrhagic forms are seen too especially among adults compared to children. Identifying Moyamoya disease can significantly affect the treatment options and give insight into managing the chronic nature of the disease to both the physicians and patients.

**Conclusion:** This case highlights the importance of searching for underlying cause in a young female presenting with intracranial haemorrhage for the first time and keeping Moyamoya disease as a differential.

## 1. Introduction

Moyamoya disease (MMD) is a cerebrovascular disease characterized by progressive narrowing of the large intracranial artery and the development of prominent small vessel collaterals. It produces a characteristic hazy appearance on angiography like a puff of smoke in the air and hence, been named Moyamoya, a Japanese term meaning the same [1,2]. Japan has the highest incidence of Moyamoya disease, where there are three cases per 100,000. Moyamoya is commonly seen in females and has a bimodal age of onset at around age 5 and at around age 40 [3]. The most common initial presentation of Moyamoya is ischemic stroke [3] [–] [7]. Transient ischemic attack (TIA) is also a frequent initial presentation and maybe recurrent [3,4]. Less commonly, Moyamoya disease may present with a headache or a seizure. Moyamoya disease affects bilateral internal carotid arteries, whereas a unilateral presentation of the same underlying pathology is known as Moyamoya syndrome and is associated with conditions like Down syndrome, neurofibromatosis type 1, and sickle cell disease. The study aimed to highlight a neglected but important cause of intracerebral haemorrhage in an adult Asian female.

The following case report has been reported in line with the SCARE 2020 criteria [8].

## 2. Presentation of case

A 38 years female presented with a chief complaint of headache followed by a loss of consciousness. She also had a history of two episodes of projectile vomiting containing digested food particles not stained with blood or bile. There was no associated fever, neck rigidity, abnormal body movements, dizziness, vision, auditory, speech, and gait abnormalities. She does not have a significant history of past medical illness, surgical intervention, drug allergies or family history of any similar illness. She did not report any positive family history for cerebrovascular diseases. She was taken to a medical college where a CT head was performed which revealed right intracranial haemorrhage with intraventricular extension. An emergency EVD was placed by an experienced neurosurgeon and postoperatively she was admitted to the ICU. She also had diabetes insipidus which was resolving with Desmopressin. PCR for SARS-Cov2 was negative. She was then brought to Kathmandu for further management.

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On presentation, she was delirious and hyperactive with GCS of E4V4M6 and the pupil was b/l equal and reactive to light. Neurological examination revealed the power of 5/5 in all four limbs with normal tone along with intact cranial nerves examination. Other physical examinations were unremarkable. Her vitals were also normal. Also, her blood reports were unremarkable. A repeat CT head was done which showed hematoma in the right lateral ventricle with extension to the left lateral, third and fourth ventricles. Therefore, CT Cerebral Angiogram was performed which showed nonvisualization of the right distal internal carotid artery and poor visualization of the M1 segment of the right middle cerebral artery and A1 and A2 segment of the bilateral anterior cerebral artery [Fig. 1]. Later, Digital Subtraction Angiography (DSA) was done which revealed nonvisualization of the right distal internal carotid artery and the right middle cerebral artery along with dense collaterals [Fig. 2]. All the findings were suggestive of Moyamoya disease.

She was treated conservatively with 1 g of intravenous ceftriaxone twice a day, 500 mg of intravenous levetiracetam twice a day, 4 mg of dexamethasone three times a day with subsequent tapering, and supportive treatment with pantoprazole and painkillers. A repeat CT head was done after two weeks which was normal. Her diabetes insipidus had resolved and she was stable and well oriented to time, place, and person. She was discharged and was advised for regular follow-up. On subsequent follow-ups in the hospital for six months following discharge, she did not complain of headache or focal neurological deficit. The patient was happy with her recovery and was asked to follow up every three months for one year.

### 3. Discussion

Moyamoya disease is common in Eastern Asia particularly in Japan, China, and Korea and there is a female predominance with a ratio of 1.9 [9]. It has a bimodal distribution with one of the peaks at 5–9 years and the other peak at 35–39 years of age [10]. Our patient was a 38-year-old

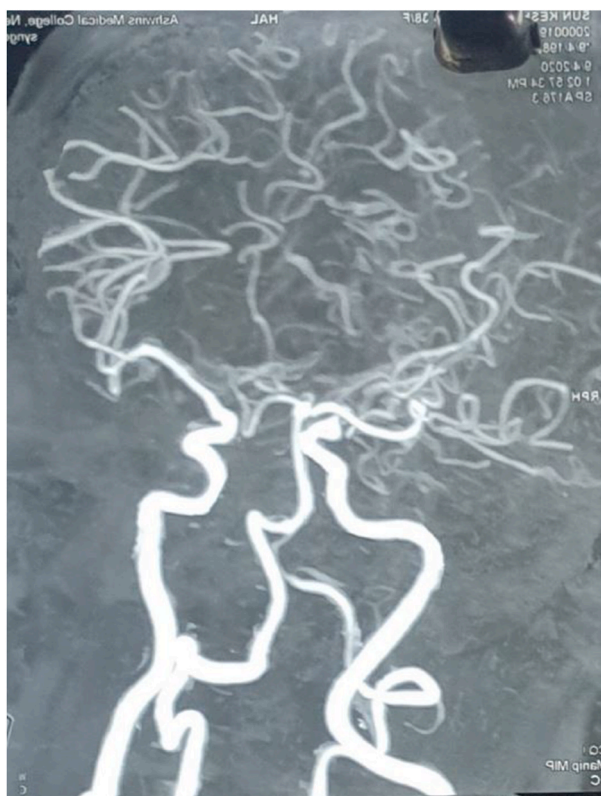


Fig. 1. CT Angiogram showing a puff of smoke.

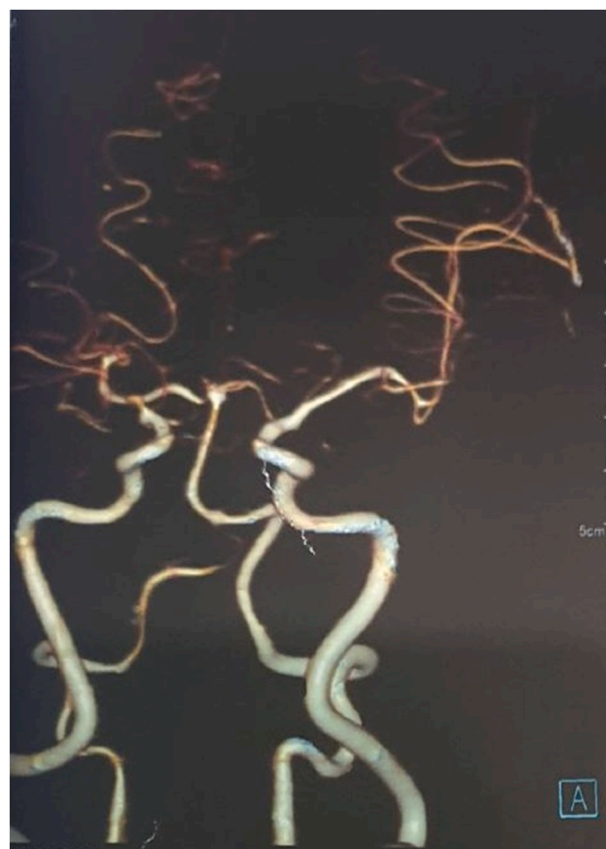


Fig. 2. DSA showing non visualization of the right distal ICA and the right MCA along with dense collaterals.

Asian female who is in line with previous studies. Although ischemic attacks are the common presenting symptoms in cases of Moyamoya disease, hemorrhagic forms are seen too especially among adults compared to children [3]. Moyamoya disease can present with features of stroke-like hemiparesis and speech impairment. They may also present with headaches or seizures. Seizures are less common usually. However, patients may be asymptomatic as well. Our patient presented with headaches, vomiting, and loss of consciousness. So, Moyamoya disease is one of the differentials that should be kept in mind in cases of females of age group 35–40 with a severe headache. Studies have found haemorrhagic presentation in 10% of adults compared to a mere 2.5% in cases of children [4,7]. Intraventricular haemorrhage was commonly found in Korea with or without intracerebral haemorrhage [11]. The presentation of our patient with right-sided intracranial haemorrhage with intraventricular extension was similar to the findings of studies done in neighboring countries China and Taiwan where intracerebral haemorrhage was found frequently [5].

Neuroimaging is important for the diagnosis of Moyamoya disease. The characteristic angiography finding is bilateral stenosis affecting the distal internal carotid arteries and proximal circle of Willis vessels, along with the involvement of prominent collateral vessels. Although conventional cerebral angiogram is the gold standard for the diagnosis of Moyamoya Disease, non-invasive imaging like CTA and MRA has taken over as initial imaging modality in multiple centers around the world [12]. Assessment of the severity of vascular abnormality through angiographic staging is important for the determination of future risks of ischemia or haemorrhage [4]. The digital subtraction angiography of our patient revealed the non-visualization of the right distal internal carotid artery and right middle cerebral artery with dense collaterals.

The treatment of acute episodes is mainly symptomatic with a focus on the reduction of elevated intracranial pressure, improvement of

cerebral blood flow, and prevention of seizures [13]. An external ventricular drain was placed to reduce the intracranial pressure in our patient who had an intraventricular haemorrhage. It is important to manage pain and avoid hypotension, hyperthermia, and hypocarbia. Secondary prevention is mainly based on surgical revascularization which may be direct or indirect [3,12]. Surgical revascularization is superior to conservative treatment with benefits with a great reduction in the risk of stroke even in a cluster of patients who had haemorrhage [14]. However, the superior technique for surgical revascularization in terms of effectiveness is still not backed up by proper data and research. Our patient has been discharged after improvement with conservative treatment and recovered well with no neurological deficits in the subsequent follow up visits.

#### 4. Conclusion

Moyamoya is a rare disease in the context of Nepal. It should be considered a differential diagnosis in patients presenting with recurrent headaches or neurological symptoms in the bimodal age distribution it is seen. This case highlights the importance of considering Moyamoya disease as a differential diagnosis in cases of young females presenting with intracranial bleed. Identifying Moyamoya disease can significantly affect the treatment options and give insight into managing the chronic nature of the disease to both the physicians and patients.

#### Patient perspective

The patient party were anxious before the operation. The operation was needed to stabilise the condition of the patient so the patient's family agreed to go through the surgery. The patient was delirious before the operation and recovered gradually. The patient party and the patient were eventually happy after successful recovery in their subsequent follow up visits.

#### Ethical approval

None declared.

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None declared.

#### Author contribution

All authors read and approved the final draft of the manuscript.

SG was involved in the concept of the case report, collection of case information, and manuscript revision. SG was involved in the concept of the case report, collection of case information, and manuscript revision. AB was involved in writing the manuscript and literature review. AKP was involved in the literature review and interpretation of clinical findings. PB did the literature review and carried out interpretation of clinical findings.

#### Research registration

None.

#### Guarantor

Suman Gaire, Medical Officer, Department of Emergency Medicine, Palpa Hospital, Palpa –32500, Nepal.

#### Consent

Written informed consent was obtained from the patient's husband for publication of the case report and the accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

#### Declaration of competing interest

None declared.

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#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2021.102424>.

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