

## Recurrent intracerebral hemorrhage in a young adult with Moyamoya Disease managed by Encephalo-duro-arterio-myosynangiosis: A Case Report

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### Abstract

#### Background

Moyamoya Disease is a chronic, progressive cerebrovascular disorder characterized by stenosis of intracranial arteries and formation of fragile collateral vessels. Adult patients commonly present with intracerebral hemorrhage.

#### Case Presentation

A 33-year-old female with a history of right periventricular intracerebral hemorrhage 3 months prior presented with headache and transient left-sided limb weakness. Magnetic Resonance Imaging (MRI) with angiography and Digital Subtraction Angiography (DSA) confirmed Moyamoya disease. Computed Tomography (CT) angiography of the brain and neck revealed marked luminal narrowing of bilateral internal carotid arteries from their origin, with significant narrowing and non-opacification of bilateral middle cerebral and anterior cerebral arteries. Multiple collateral vessels were noted around the circle of Willis, basal cisterns, and cortical regions, producing a characteristic “puff of smoke” appearance. These findings were consistent with Moyamoya disease.

The patient underwent right sided encephalo-duro-arterio-myosynangiosis (EDAMS). Postoperative management included antibiotics, antiepileptics, analgesics, and supportive care. The patient showed clinical improvement and was discharged on antiplatelet and adjunct therapy.

### Conclusion

Early diagnosis and timely surgical revascularization are crucial in preventing recurrent cerebrovascular events and improving outcomes in Moyamoya disease.

Informed Consent was obtained from the patient for publication of this case report and accompanying images.

### Keywords

Moyamoya Disease; Intracerebral hemorrhage; EDAMS; Cerebral revascularization; Stroke; Digital Subtraction angiography

### I. Introduction

Moyamoya Disease is a rare, chronic, progressive cerebrovascular disorder characterized by stenosis or occlusion of the terminal portions of the internal carotid arteries and the proximal branches, resulting in the formation of abnormal collateral vessels at the base of the brain<sup>(1)</sup>.

These collateral vessels produce the characteristic angiographic “puff of smoke” appearance, from which the disease derives its name<sup>(2)</sup>. The disease has a bimodal age distribution, commonly affecting children and young adults.

While pediatric patients typically present with ischemic symptoms, adults more frequently present with intracerebral hemorrhage due to rupture of fragile collateral vessels<sup>(3)</sup>. Early diagnosis using imaging modalities such as MRI and DSA is essential for appropriate management.

Current clinical guidelines emphasize the importance of early diagnosis and appropriate management strategies to improve patient outcomes<sup>(4)</sup>.

## II. Case Presentation

A 33-year-old female presented with complaints of headache and a history of left-sided limb weakness, which resolved spontaneously. The patient had a past history of right-sided periventricular intracerebral hemorrhage 3 months prior.

There was no significant family or social history. MRI of the brain with angiography, followed by DSA, confirmed the diagnosis of Moyamoya Disease.

The patient was admitted to a tertiary care center, and pre-anesthetic evaluation was performed. She subsequently underwent right-sided EDAMS as a revascularization procedure.

### In-Hospital Management

- Intravenous Fluids
- Ceftriaxone 2 g IV twice daily
- Diclofenac 75 mg IV
- Tramadol 50 mg IV
- Esomeprazole 40 mg IV
- Levetiracetam 500 mg IV
- Sodium citrate syrup at bed time

The postoperative period was uneventful, and the patient showed clinical improvement.

### Discharge Medications

- Cefuroxime 500 mg twice daily for 7 days
- Pantoprazole 40 mg twice daily for 7 days
- Phenytoin 100 mg three times daily for 1 month
- Levetiracetam 500 mg three times daily for 10 days
- Naproxen twice daily for 7 days
- Sodium citrate syrup at bedtime
- Aspirin + Atorvastatin once daily (continued)

## III. Discussion

Moyamoya Disease is characterized by progressive stenosis of intracranial arteries leading to the development of fragile collateral circulation, which predisposes patients to both ischemic and hemorrhagic strokes<sup>(5)</sup>.

In adults, hemorrhagic presentation is more common and is often associated with rupture of these abnormal collateral vessels. The present case highlights recurrent intracerebral hemorrhage in a

young adult, emphasizing the importance of early recognition and timely intervention.

DSA remains the gold standard for diagnosis, demonstrating characteristic vascular changes including stenosis and collateral vessel formation<sup>(6)</sup>. Surgical revascularization plays a crucial role in the management of Moyamoya disease. Indirect procedures such as EDAMS promote angiogenesis and improve cerebral blood flow over time<sup>(7)</sup>.

Studies have shown that revascularization significantly reduces the risk of recurrent stroke, particularly in patients with hemorrhagic presentation<sup>(8)</sup>.

From a pharmacological perspective, antiepileptic drugs such as Levetiracetam and phenytoin are commonly used to prevent postoperative seizures, while antiplatelet therapy and statins contribute to secondary prevention and vascular protection.

## IV. Conclusion

Moyamoya Disease should be considered in young patients presenting with recurrent intracerebral hemorrhage. Early diagnosis and timely surgical revascularization significantly improve clinical outcomes and reduce recurrence risk.

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