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Isolated Upper Motor Neuron Facial Palsy as a Rare Presentation of Cerebral Vasculitis Mimicking Moyamoya Disease: A Case Report

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Abstract

Moyamoya disease and cerebral vasculitis are rare but critical differential diagnoses in young patients presenting with cerebrovascular abnormalities [1]. We present a case of a woman in her 30 s who developed isolated upper motor neuron (UMN) facial palsy. CT angiography initially suggested Moyamoya disease due to narrowed cerebral vessels and prominent meningeal collaterals. However, magnetic resonance angiography (MRA) performed on Day 2 demonstrated a beaded appearance in the right middle cerebral artery, leading to a revised diagnosis of cerebral vasculitis. The patient was initially treated with aspirin, which was discontinued following tertiary review. Amitriptyline was initiated for neuropathic pain, resulting in marked symptomatic improvement. This case underscores the diagnostic challenge in differentiating these conditions and highlights the role of advanced imaging and multidisciplinary collaboration. are rare but critical differential diagnoses in young patients presenting with cerebrovascular abnormalities. **Case Presentation**: A woman in her 30 s presented with isolated right upper motor neuron (UMN) facial palsy. CT angiography (CTA) revealed narrowing of distal internal carotid and middle cerebral arteries with prominent meningeal vessels, initially suggesting Moyamoya disease. **Management and Outcome**: Magnetic resonance angiography (MRA) performed two days later showed a beaded appearance in the M1-M2 segments of the right middle cerebral artery, more consistent with cerebral vasculitis. Three neuroradiologists independently reviewed the imaging and supported the revised diagnosis. The patient was initially treated with aspirin, which was later discontinued after tertiary review. Amitriptyline was introduced for persistent neuropathic pain. Over four months, facial weakness

improved by 95%, though mild neuralgia persisted. **Conclusion**: This case underscores the diagnostic challenges in differentiating vasculitis from Moyamoya disease and highlights the critical role of advanced imaging modalities and multidisciplinary collaboration.

Keywords

Vasculitis, Moyamoya Disease, Case Report, Upper Motor Neuron Facial Palsy, Magnetic Resonance Angiography

1. Background

Moyamoya disease is a progressive cerebrovascular disorder involving stenosis or occlusion of distal internal carotid arteries (ICA) and proximal middle cerebral arteries (MCA), with the formation of fragile collateral vessels that resemble a "puff of smoke" on angiography [2] [3]. Although idiopathic in most cases, Moyamoya can be associated with conditions like Down syndrome and neurofibromatosis type 1 [4].

Cerebral vasculitis encompasses a range of disorders characterized by inflammation of cerebral vessels. It may be primary or secondary to systemic conditions, including autoimmune diseases and infections [5]. Imaging differentiation is crucial due to overlapping features between Moyamoya disease and vasculitis. In a comparative study by Kraemer and Berlit (2010), conventional angiography diagnosed all Moyamoya cases but only 76.5% of vasculitis cases, illustrating the diagnostic complexity [6].

Misdiagnosis is common. Graf *et al.* (2019) found that in 192 Caucasian patients with Moyamoya angiopathy (MMA), 62% were initially misdiagnosed, with vasculitis being the most common incorrect diagnosis (31%) [7]. Vessel wall MRI (VW-MRI) improves diagnostic precision, with concentric enhancement suggesting vasculitis and absent enhancement in classic Moyamoya [8].

This report describes a diagnostic challenge in a patient with isolated UMN facial palsy. Initial imaging suggested Moyamoya, but subsequent findings and expert review led to a final diagnosis of cerebral vasculitis.

2. Case Presentation

An Australian Caucasian woman in her 30 s presented to a regional hospital with acute onset of right facial drooping. She denied limb weakness, speech disturbance, headache, visual changes, or systemic symptoms. Past history included hypertension and pancreatitis. Family history was notable for cerebrovascular disease in multiple first-degree relatives. She was a non-smoker with no recent infections or medication changes.

On neurological examination, there was isolated right UMN facial palsy—evident by lower facial weakness with preserved forehead movement. Muscle strength,

reflexes, cerebellar function, and other cranial nerves were intact. No sensory deficits were identified.

3. Investigations

CT brain was unremarkable. CTA revealed small calibre distal ICA and MCA with prominent meningeal vessels, suggestive of Moyamoya disease. On Day 2, MRA (time-of-flight sequence) showed beading of the M1-M2 segments of the right MCA and scattered irregularities in anterior and posterior cerebral arteries without significant stenosis (Figure 1). These findings were more consistent with cerebral vasculitis.

Inflammatory markers included ESR 20 mm/h and a borderline ANA (1:40). ANCA, anti-dsDNA, ENA, antiphospholipid antibodies, and lupus anticoagulant were all negative. CSF analysis and brain biopsy were not performed due to low suspicion of systemic disease and patient preference.

Three neuroradiologists independently and blindly reviewed the imaging, confirming the findings supported cerebral vasculitis over Moyamoya disease.

4. Treatment

The patient was initially started on aspirin for presumed ischemic etiology. Following tertiary review and revised diagnosis, aspirin was discontinued. Amitriptyline was initiated for persistent retro-orbital and occipital neuralgia. No immunosuppressive therapy was commenced due to isolated presentation and improving symptoms.

5. Outcome and Follow-Up

Over the following four months, the patient reported 95% improvement in facial weakness. She experienced occasional episodes of transient blurred vision and persistent neuralgia but no new neurological deficits. She remains under follow-up with neurology and neuroradiology teams to monitor disease course and imaging progression.

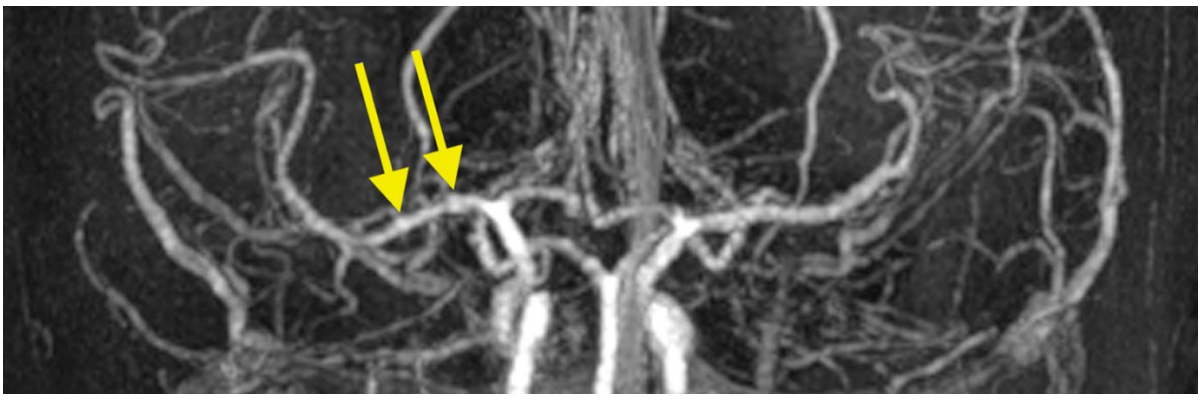

Figure 1. Time-of-flight magnetic resonance angiography (MRA) showing a beaded appearance of the M1-M2 segments of the right middle cerebral artery (yellow arrows), performed on Day 2 post-symptom onset. This appearance is more consistent with vasculitis than Moyamoya disease. Anatomical orientation markers included.

6. Discussion

This case presents a rare diagnostic dilemma of distinguishing cerebral vasculitis from Moyamoya disease in a patient with isolated UMN facial palsy (Table 1). Moyamoya disease typically shows bilateral stenosis with collateral formation, while vasculitis exhibits segmental beading without extensive collateralisation [9]. In this case, the absence of bilateral findings and presence of vessel irregularities without occlusion or collaterals favored vasculitis.

Three independent neuroradiologist reviews supported the vasculitis diagnosis. Similar diagnostic confusions have been reported [6] [7] [10]. VW-MRI could further support diagnosis but was not available in this case.

UMN facial palsy usually suggests a central lesion (stroke, tumour), and isolated presentation is rare. Focal small-vessel inflammation might explain the selective involvement seen here.

The absence of CSF or biopsy is a limitation, though clinical improvement and imaging supported a working diagnosis. This case underscores the utility of MRA, multidisciplinary evaluation, and caution against premature diagnoses in young patients with neurovascular symptoms [11].

Table 1. Distinguishing features of moyamoya disease vs. cerebral vasculitis [adapted from Kraemer & Berlit, 2010; Mossa-Basha *et al.*, 2016 [8]].

Feature	Moyamoya Disease	Cerebral Vasculitis
Age/Gender	Children, young adults, more common in females	Variable, often adults
Distribution	Bilateral ICA/MCA stenosis	Segmental, focal, or diffuse
Imaging	"Puff of smoke" collaterals	Beading, irregularity
Vessel Wall MRI	Typically no enhancement	Concentric enhancement
Association	Idiopathic, syndromic	Autoimmune, infectious
Management	Surgical revascularisation	Immunosuppression

7. Learning Points

- Isolated UMN facial palsy may be a sign of underlying central vasculopathy.
- Moyamoya disease and cerebral vasculitis can appear similar on CTA; MRA provides critical differentiating detail.
- Multidisciplinary and blinded radiology review strengthens diagnostic accuracy.
- Vessel wall MRI can aid in ambiguous neurovascular cases.
- Careful exclusion of systemic and autoimmune causes is vital when considering vasculitis.

Patient Perspective: "It's been an interesting journey. As someone from a medical background, I initially thought it was Bell's palsy. Being told it might be Moyamoya was terrifying. When they later diagnosed vasculitis, it was a relief but also raised many questions. I'm still dealing with pain and uncertainty, but I appreciate the thoroughness of the team in pursuing the correct diagnosis. The experience has taught

me the importance of second opinions and not settling for the first explanation—especially when something doesn't feel right. While some symptoms linger, I feel reassured knowing I'm under continuous care and monitoring."

Consent

Written informed consent was obtained from the patient in accordance with the journal's ethical standards and the CARE (CAse REport) guidelines for case reports. Informed consent was obtained from the patient for publication of this case report and the included images.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Chiu, D., Shedden, P., Bratina, P. and Grotta, J.C. (1998) Clinical Features of Moyamoya Disease in the United States. *Stroke*, **29**, 1347-1351. https://doi.org/10.1161/01.str.29.7.1347

[2] Takeuchi, K. and Shimizu, K. (1957) Hypogenesis of Bilateral Internal Carotid Arteries. *No To Shinkei*, **9**, 37-43.

[3] Scott, R.M. and Smith, E.R. (2009) Moyamoya Disease and Moyamoya Syndrome. *New England Journal of Medicine*, **360**, 1226-1237. https://doi.org/10.1056/nejmra0804622

[4] Omer, S., Zbyszynska, R. and Kirthivasan, R. (2018) Peek through the Smoke: A Report of Moyamoya Disease in a 32-Year-Old Female Patient Presenting with Ischaemic Stroke. *BMJ Case Reports*, **2018**, bcr-2017-221685. https://doi.org/10.1136/bcr-2017-221685

[5] González-Gay, M.A. and García-Porrúa, C. (2001) Epidemiology of the Vasculitides. *Rheumatic Disease Clinics of North America*, **27**, 729-749. https://doi.org/10.1016/s0889-857x(05)70232-5

[6] Kraemer, M. and Berlit, P. (2009) Primary Central Nervous System Vasculitis and Moyamoya Disease: Similarities and Differences. *Journal of Neurology*, **257**, 816-819. https://doi.org/10.1007/s00415-009-5425-7

[7] Graf, J., Schwitalla, J.C., Albrecht, P., Veltkamp, R., Berlit, P., Hartung, H., *et al.* (2019) Misdiagnoses and Delay of Diagnoses in Moyamoya Angiopathy—A Large Caucasian Case Series. *Journal of Neurology*, **266**, 1153-1159. https://doi.org/10.1007/s00415-019-09245-9

[8] Mossa-Basha, M., *et al.* (2016) Vessel Wall MRI for Differentiation of Vasculopathies: Improved Diagnostic Performance. *American Journal of Neuroradiology*, **37**, 323-330.

[9] Houkin, K., Abe, H., Yoshimoto, T. and Takahashi, A. (1996) Is "Unilateral" Moyamoya Disease Different from Moyamoya Disease? *Journal of Neurosurgery*, **85**, 772-776. https://doi.org/10.3171/jns.1996.85.5.0772

[10] Zhang, Y., *et al.* (2022) Moyamoya Disease as a Cause of Ischemic Stroke: A Case Study and Literature Review. *World Journal of Neuroscience*, **12**, 17-23.

[11] Lee, H., *et al.* (2023) Diagnostic Pitfalls of Moyamoya Disease in Adult Patients: A Case-Based Review. *Clinical Neurology and Neurosurgery*, **226**, Article ID: 107587.