Puff of Smoke: Moyamoya Disease Mimicking Acute Stroke

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Introduction

Stroke remains a leading cause of morbidity and mortality worldwide, with early recognition and intervention playing a crucial role in patient outcomes. Emergency physicians are trained to rapidly assess stroke presentations, activate stroke pathways, and initiate time-sensitive treatments such as thrombolysis or mechanical thrombectomy. However, not all strokes follow a predictable course.

Moyamoya disease, a rare and progressive cerebrovascular disorder, can present with ischemic strokes that mimic embolic large-vessel occlusion (LVO). Its pathophysiology and response to treatment differ significantly from traditional embolic strokes. In the emergency setting, recognizing Moyamoya disease as a potential stroke etiology is critical, as standard stroke management algorithms may not apply, and certain interventions can pose significant risks.

This case highlights a patient who presented with acute neurological deficits concerning for an LVO but was ultimately diagnosed with Moyamoya disease. It explores the diagnostic challenges, emergency medicine considerations, and the role of teleneurology and interfacility transfer decision-making in optimizing stroke care.

Case Presentation

A 60-year-old male with a past medical history of distal esophageal cancer post-chemoradiation, coronary artery disease status post-coronary artery bypass grafting, hypertension, hypercholesterolemia, and thrombocytopenia was admitted for worsening dysphagia, requiring J-tube placement.

His postoperative course was complicated by hypotension, which led to the development of new-onset atrial fibrillation. Due to his increased thromboembolic risk, he was initiated on therapeutic anticoagulation with enoxaparin.

On postoperative day three, the patient developed acute-onset left-sided weakness, with 1/5 strength in the lower extremity and 2/5 strength in the upper extremity, accompanied by facial droop. A code stroke was activated, and his NIH Stroke Scale score was assessed as 11. Given the acute presentation of focal deficits, a large-vessel occlusion was suspected.

A non-contrast head CT showed no acute hemorrhage, while CT angiography revealed occlusion of the right M1 segment of the middle cerebral artery. Given his recent initiation on anticoagulation, the patient was not a candidate for tenecteplase (TNK). Teleneurology was consulted and initially recommended urgent transfer to a thrombectomy-capable stroke center for possible mechanical thrombectomy.

During discussions with the receiving stroke center, concern was raised regarding atypical vascular findings on CT angiography. The occlusion pattern and collateral formation were not characteristic of a typical embolic large-vessel occlusion. This led to the suspicion of Moyamoya disease, prompting reconsideration of the appropriateness of thrombectomy.

Given the uncertainty of the diagnosis and unclear benefit of intervention, the initial transfer request was declined. The patient was ultimately transferred to a tertiary neurovascular center, where additional imaging, including repeat CT angiography and MRI, confirmed the diagnosis of Moyamoya disease with bilateral multifocal infarcts.

Discussion

Moyamoya Disease in the Emergency Setting

Moyamoya disease is characterized by progressive stenosis of the intracranial internal carotid arteries, middle cerebral arteries, and anterior cerebral arteries. As these vessels progressively narrow, fragile compensatory collateral vessels form, leading to the characteristic "puff of smoke" appearance on cerebral angiography. This abnormal collateralization develops in response to chronic cerebral hypoperfusion and is best visualized using digital subtraction angiography, although CT angiography and MRI can also suggest the diagnosis.[1]

Epidemiology and Risk Factors

Moyamoya disease has an incidence of 1 per 1.1 million people in the United States and is more common in East Asian populations, particularly Japan and Korea. The disease follows a bimodal age distribution, peaking in childhood and again in adults aged 30–50 years.[2] It can occur idiopathically or as a secondary condition associated with prior cranial radiation therapy, autoimmune diseases such as systemic lupus erythematosus and vasculitis, or hematologic disorders like sickle cell disease.[3] Because the disease progresses insidiously, many patients remain undiagnosed until they present with an ischemic or hemorrhagic event, highlighting the importance of early recognition in the emergency setting.

Stroke Patterns in Moyamoya Disease

Unlike embolic strokes, which typically involve a single vascular territory, Moyamoya disease can present with bilateral infarcts, watershed infarcts, and recurrent strokes despite anticoagulation or antiplatelet therapy.[4] The fragile collateral vessels increase the risk of both ischemic and hemorrhagic strokes, complicating acute management.[5]

Emergency Medicine Considerations

Moyamoya Can Mimic Large-Vessel Occlusion but Requires Different Management

Standard large-vessel occlusion treatment algorithms do not always apply to

Moyamoya disease. Key differences include:

- Thrombolysis: Patients with Moyamoya disease are often not candidates for thrombolytics due to the risk of hemorrhagic conversion from fragile collateral rupture.[6]
- Thrombectomy: Moyamoya-related occlusions may not be amenable to thrombectomy, as they result from chronic stenosis rather than acute embolic clot formation.[7]

Role of Teleneurology and Transfer Challenges

This case highlights the variability in stroke center acceptance criteria. Despite clear evidence of a large-vessel occlusion, the patient's imaging findings prompted concern for Moyamoya disease, leading to a transfer delay. Such cases emphasize the importance of rapid and informed decision-making in the emergency department. Emergency physicians must be prepared to recognize when stroke center capabilities are limited and advocate for appropriate transfers, ensuring that patients receive care at facilities with experience managing complex cerebrovascular disorders. Teleneurology consultations play a critical role in triaging stroke patients but can sometimes introduce challenges when different institutions have conflicting management approaches. Understanding these nuances can help emergency physicians balance the urgency of transfer with the need for a specialized neurovascular evaluation.

Recognizing When Stroke Management Needs to Deviate from Protocols

Stroke care is largely protocol-driven, but some cases require deviation from standard pathways. Moyamoya should be suspected in patients with recurrent strokes despite therapeutic anticoagulation or antiplatelet therapy, large-vessel occlusions without a clear embolic source, or stroke patterns involving bilateral or watershed infarcts. Identifying these red flags early can prevent unnecessary interventions and facilitate timely specialist involvement.

Conclusion

This case emphasizes the importance of considering Moyamoya disease in atypical stroke presentations, particularly when large-vessel occlusion is suspected but standard interventions may not apply. Emergency physicians must be prepared to identify stroke mimics, navigate treatment contraindications, and coordinate appropriate transfers to specialized centers when necessary.

While uncommon, Moyamoya disease is a critical diagnosis in emergency medicine, as failure to recognize it can lead to mismanagement, inappropriate interventions, or delays in definitive care. Increasing awareness of this condition allows emergency physicians to provide more tailored and effective stroke management, ultimately improving patient outcomes.

References

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