



Mania Secondary to Moyamoya Disease

Timeline and Approach to Treatment

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Introduction

- Moyamoya disease (MMD) is a progressive, rare cerebrovascular condition where stenosis and/or occlusion of intracranial arteries result in formation of compensatory collateral vessels.
- The most commonly affected vessels are the distal segments of the internal carotid arteries, along with the proximal middle and anterior cerebral arteries. This pathology increases the risk of hypoperfusion injuries such as transient ischemic attacks, strokes, and seizures.
- Neuropsychiatric manifestations occur in about half of patients with MMD; however, mania in the context of MMD is best understood as a rare clinical event.

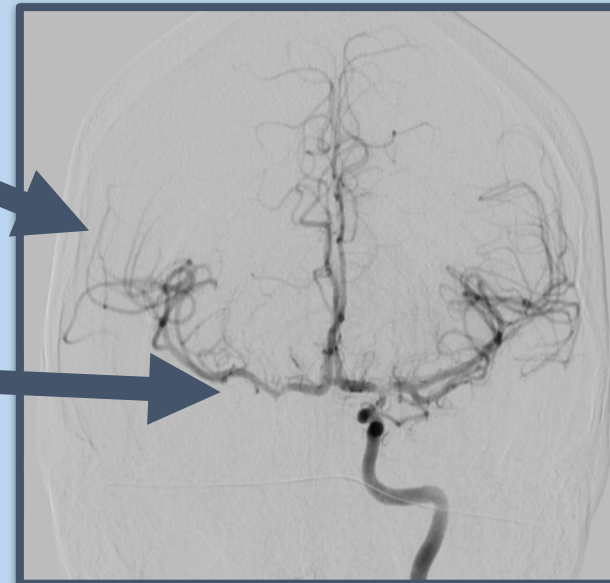
Case History

- 43-year-old female of Filipino descent with a history of Graves' disease and ischemic strokes (April 2025) in the setting of severe intracranial atherosclerotic disease, who underwent left encephalo-duro-arterio-synangiosis (EDAS) in June.
- In July, she was admitted for acute neurologic changes, as well as progressive behavioral changes, including agitation, impulsivity, and grandiosity.
- Imaging showed a left subdural hematoma (SDH) with midline shift.
- Psychiatry was consulted for management of agitation. Her exam was notable for severe aphasia, paranoia, disorganized thinking, and a lack of insight and judgement. Management included antipsychotics, benzodiazepines, and a mood stabilizer.
- After some improvements, her exam changed to apathy, mutism, and difficulty following commands.
- Imaging showed new left frontal and parietal infarcts, as well as enlargement of the SDH with increased midline shift, requiring evacuation by epidural drain.
- She was discharged on Depakote and stroke prevention. Later, mirtazapine was started and Depakote was self-discontinued.
- Following additional episodes of transient neurologic changes and periods of impulsive and irrational behaviors, Keppra was started for presumed focal seizures.

Imaging

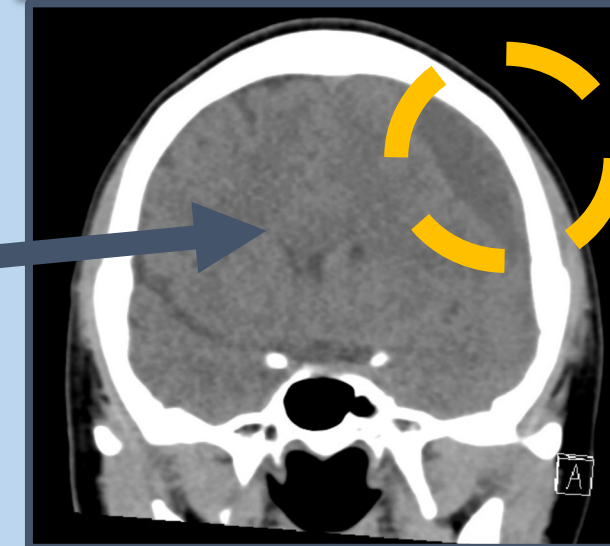
IR Cerebral Angiogram
June 21st, 2024

- Substantial collateral vascularization inconsistent with age
- Areas of low flow and stenotic features



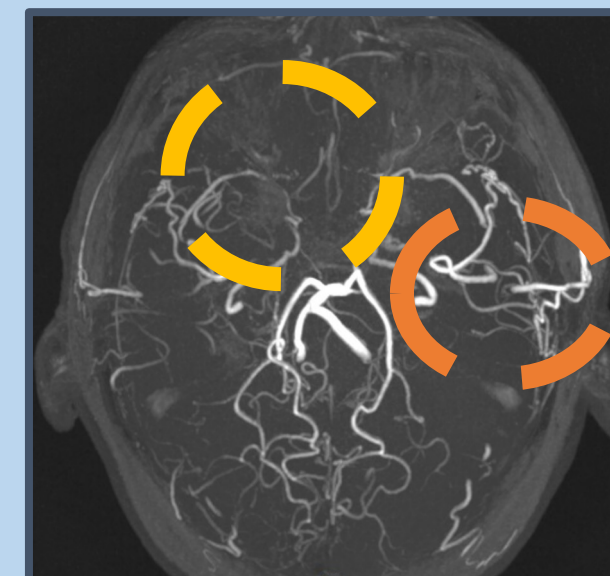
CT Head
June 28th, 2025

- Subdural hematoma
- Midline Shift



MRA Head
Dec 24th, 2025

- Radiographically difficult to identify anterior circle of willis vasculature due to ongoing stenotic changes
- EDAS related changes and improved arterial perfusion



Discussion

- While MMD is classically associated with ischemic and hemorrhagic strokes, chronic hypoperfusion affecting the frontal lobes and limbic system can precipitate severe psychiatric sequelae.
- The temporal relationship between this patient's multiple neurologic insults and the onset of acute mania with psychotic features strongly implicates organic cause rather than primary psychiatric etiology.
- In contrast to the common theory that left-sided lesions predispose to depression (and right-sided to mania), this patient exhibited mania in the setting of left-sided acute pathologies. This suggests that bilateral chronic stenosis might compromise global frontal circuits, resulting in a disconnect between emotional regulation and executive function.
- The use of antipsychotics, which can lower seizure threshold and increase mortality in stroke patients, requires careful consideration.
- In patients with MMD and potential seizure activity, Depakote serves as both a mood stabilizer and an anticonvulsant.
- Mirtazapine was chosen for depressed mood given its negligible impact on platelet aggregation and favorable side effect profile
- Future considerations: The patient's persistent lack of insight and judgment poses long-term ethical challenges regarding autonomy. This case supports the need for durable power of attorney or guardianship in patients with MDD with frontal involvement, as their cognitive decline may be fluctuating but progressively limiting.

References

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