MOYAMOYA DISEASE PRESENTING AS HEMICHOREA IN A FILIPINO ADOLESCENT: A CASE REPORT

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BACKGROUND

Moyamoya disease (MMD) is a chronic cerebrovascular disease characterized by idiopathic gradual stenosis or occlusion of the bilateral supraclinoid internal carotid arteries (ICA)[1]. In the pediatric population, typical presentations include transient ischemic attacks or ischemic strokes[2]. Movement disorders as presenting features in patients with MMD are **extremely rare**. Only **4.2%** of pediatric MMD present with **chorea**[3].

We report a case of a Filipino adolescent presenting with unilateral choreiform movements and was subsequently diagnosed with Moyamoya disease.

CASE PRESENTATION

- 13-year-old, right-handed, female
- **No known comorbidities**
- No trauma, recurrent throat infection, systemic illness, intake of medication

- **5-day history of involuntary movement of the right upper extremity** described as choreiform, irregular, nonrhythmic, intermittent, lasting <1min, disappeared during sleep
- No headache, seizures, transient ischemic attacks

- Given **Clonazepam** for 1 week with resolution of right hemichorea
- Maintained on **Aspirin** 3-5mkdose for secondary stroke prevention
- Possible **revascularization surgery**

- Intact higher cortical function
- Normal cranial nerve, sensory, and cerebellar functions; no meningeal irritation
- **Right upper extremity weakness (MMT 4/5)** and **pronator drift**
- **Choreiform movement of the right upper extremity**

- **Echocardiogram**: normal cardiac dimension and function for age
- **2-hr vEEG**: unremarkable findings
- **Cranial MRI**: 1) ivy sign, 2) chronic infarct in the right bilateral frontal lobes and body of the right caudate, 3) narrowing of the bilateral ICA with collateral vessel formation, 4) prominence of lenticulostriate and paracallosal arteries

NEUROIMAGING FINDINGS

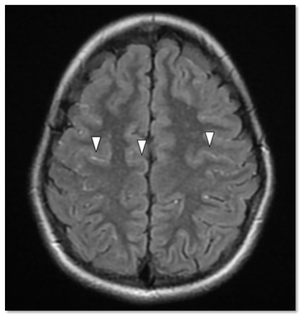

Figure 1. *MRI FLAIR sequence showing ivy sign (arrowhead) seen as linear high signals that follow a sulcal pattern (white arrowheads).*

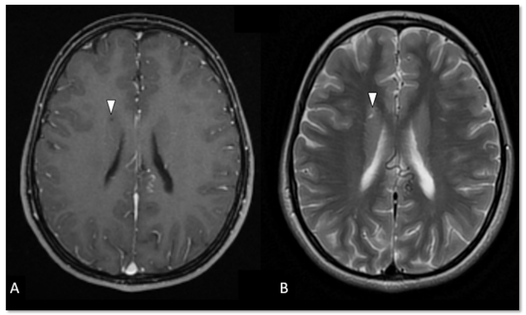

Figure 2. *A chronic infarct was seen as T1W-hypointense (A) and T2W-hyperintense (B) focus in the body of the right caudate nucleus (white arrowhead).*

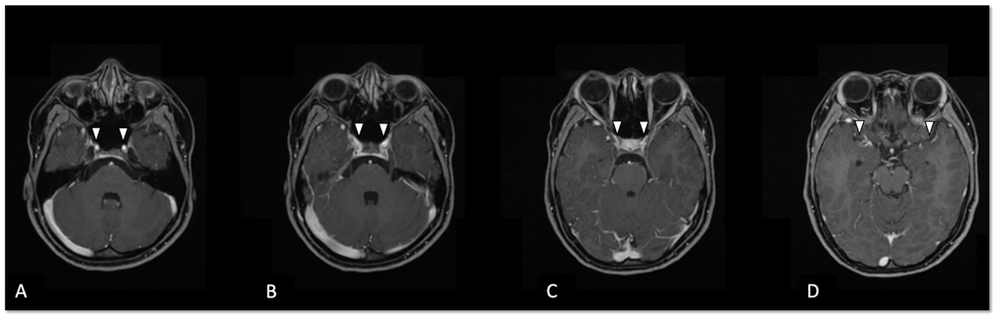

Figure 3. *There is narrowing of the distal cavernous/proximal clinoid segments of the bilateral internal carotid arteries as shown sequentially in the images from A to D (white arrowheads) with collateral vessel formation.*

DISCUSSION AND CONCLUSION

- Moyamoya disease must be considered in the **differential diagnosis** of a patient with **chorea**.
- Choreiform movements are caused by a **disruption in the fine balance between excitatory and inhibitory signals connecting the basal ganglia and cerebral cortex** via direct and indirect pathways.
- This is the **first Filipino female adolescent** at our institution diagnosed with MMD presenting as **hemichorea without an associated lesion in the contralateral basal ganglia**.
- Medical management was initiated with **Clonazepam** which addressed the choreiform movements and **Aspirin** to prevent subsequent ischemic events. Long term plan is to undergo **revascularization surgery**.

REFERENCES

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[3] Smith, E and Scott, R. (2015). Chapter 11 Causes of Nontraumatic Hemorrhagic Stroke in Children: Pediatric Moyamoya Syndrome. *Pediatric Neurosurgery*.

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