Moyamoya Syndrome and Cerebral Vascular Accident as a Presenting Manifestation of Neurosyphilis: A Case Report

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Abstract

Case Description: A 27-year-old right-hand-dominant Caucasian male presented with left-sided weakness. Four days prior to admission, he sought medical attention on an emergency room visit due to new-onset headache and fever. He had no history of hypertension, diabetes or other vascular risk factors. His Family Functional Independence Measures increased rapidly in comparison to other Moyamoya cases without consecutively deteriorating symptomology. Discussion: Historically, Moyamoya Syndrome is a unique etiology that has been associated with a number of diseases and conditions, including meningitis, Down syndrome, Sydenham’s chorea, Neurofibromatosis Type I, tuberculosis, head trauma, head irradiation, von Hippel-Lindau’s disease, polyarteritis nodosa, and even contraceptives use. To our knowledge, this is the first reported case of Moyamoya disease associated with neurosyphilis without concomitant immunodeficiency. In this case, a mechanism of cerebral vasculitis or inflammation is favored. On-going meningitis disease may allow for further Moyamoya collaterals and impending clinical deficits and debility unless invasive measures are taken to restore cerebral circulation. Conclusions: Given an increasing number of Moyamoya cases presenting to neurorehabilitation facilities, and the likelihood of an inflammatory role in Moyamoya Syndrome, treatment of underlying illness (if available) could potentially delay or negate need for invasive revascularization procedures and promote better (and possibly quicker) outcomes in an acute rehabilitation setting. Furthermore, it might be prudent to investigate associated diseases in efforts to improve standards of care.

Case Presentation

27-year-old right-hand-dominant Caucasian male presented with left-sided weakness. Four days prior to admission had painless rash in left hand that progressed to denude left hemiplegia, slurred speech, and a facial droop by presentation. Associated symptoms include headache, photophobia, difficulty walking, and recent insomnia.

Initial evaluation at the acute care hospital:
- National Institute of Health Stroke Scale (NIHSS) was 5.
- Diagnostic work-up:
  - Positive RPR titer (1:64), elevated Anticardiolipin IgG and IgM, as well as a positive VDRL.
- Thyroid stimulating hormone was marginally elevated at 5.03 with a normal free T4.
- CRP, ESR, B12, Folate, Protein S, Protein C, Factor V, and Factor VIII were normal.
- MRI: Edema and gliosis of right parieto-occipital lobes.
- CT-Angiogram showed irregular lumen of his right ICA, scaling on extensor surfaces of all four extremities; motor exam:  4/5 in the left lower extremity;  3/5 in proximal left upper extremity.
- Sacral and buttck area numbness, sciatica.
- CRP, ESR, B12, Folate, Protein S, Protein C, Factor V, and Factor VIII were normal.

Admission exam: Blood pressure: 142/89 mmHg; BMI = 35; integumentary exam revealed several psoriatic plaques with erythematous bases; and several papules were present on the right hand. New-onset headache and fever, and photophobia.

Final admission diagnosis:
- Neurosyphilis: Penicillin G 2.4 million units IV daily started 10 days after symptom onset. Motor recovery did not begin until 48 hours after treatment initiation.
- MMS: Aspin 8/1mg/day for secondary stroke prevention. Definitive treatment deferred until parastichy control.

Acute Rehabilitation:
- Administered on day 6/14 treatment days of Penicillin G. Completed 14 day course.
- Admission exam: Blood pressure: 128/88 mmHg; BMI = 35; integumentary exam revealed several psoriatic plaques with erythematous bases; and several papules were present on the right hand. New-onset headache and fever, and photophobia.

Follow-up:
- Repeat cerebral angiogram 2 months after onset of his original symptoms revealed increased severe critical stenosis of the supraclinoidal internal carotid artery with progression of pial and leptomeningeal collaterals.
- Right encephaloduroarteriosynangiosis (EDAS) performed and he has been free of new stroke symptoms.
- Unrelated ER visit one year after symptom onset revealed persistently positive RPR.

Management & Outcome

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Diagnosis: Acute right MCA and PCA watershed infarcts as well as subacute right internal capsule (genicul) infarct consistent with Moyamoya Syndrome (MMS). The positive VDRL CSF titer confirmed neurosyphilis.

Figure 3. CT-Angiogram showed irregular lumen of his right ICA. At right ACA, bilateral M1 segments of MCA.

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Discussion

Moyamoya disease (MMD) vs. Moyamoya syndrome (MMS):
- Chronic occlusive cerebrovascular disorder characterized by bilateral and symmetric stenosis of the internal carotid arteries (mainly supraclinoidal portion) with compensatory enlargement of the perforating vessels at the base of the brain.
- Collateral circulation develops around the blocked vessels which are small, friable, and prone to hemorrhage. This results in the pathognomonic “puff of smoke” appearance on cerebral angiography.
- In advanced disease, anterior choroidal artery, posterior communicating artery, and ophthalmic artery can become affected.
- Complications are dependent on area of ischemia or hemorrhage and similar to those of traditional strokes.

Etiology:
- Tumors, trauma, and infectious etiologies. However, despite extensive research, a clear mechanism is still unknown.
- Autoimmune disorders have been reported to be related to Moyamoya phenomenon, including anti-phospholipid antibodies syndrome, systemic lupus erythematosus, Graves’ disease, and HLA-As for HLA-Ab.
- The positive VDRL CSF titer confirmed neurosyphilis.

Conclusions

- This case demonstrates MMS as a sequelae of untreated neurosyphilis.
- To our knowledge, this is the first reported case of MMS associated with neurosyphilis without concomitant immunodeficiency.
- An etiology of cerebral vasculitis or inflammation is favored in this case given the history and presentation.
- Early diagnosis of MMS associated diseases could prevent the need for invasive procedures.
- It is important to note that his RPR continues to be positive one year after initial event and adequate treatment.
- Centers for Disease Control and Prevention (CDC) guidelines recommend a patient with tertiary syphilis undergo screening every six months post treatment, and in some cases undergo repeat CSF evaluation, to ensure decreasing cell counts and resolution of disease.
- On-going insidious neurosyphilis may contribute to Moyamoya collaterals which would nullify initial EDAS and place patient at risk for additional strokes.

Acknowledgements

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References


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