Moyamoya Disease – A Vasculopathy and an Uncommon Cause of Recurrent Cerebrovascular Accidents

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ABSTRACT

Moyamoya disease is a very rare chronic cerebrovascular disease of unknown etiology characterized by recurrent ischemic or hemorrhagic strokes. Initially diagnosed in Japan and named after finding puff of smoke like collateral blood vessels around the occluded blood vessels of circle of Willis. With increase awareness this disease is now diagnosed more often. Medical and surgical treatment have been used to treat the disease, with surgical treatment been mostly experimental. Special attention should be given to the surgical treatment which has shown to have an edge over the medical treatment in some clinical trials especially in young patients with recurrent strokes to prevent progressive cognitive decline and to improve their quality of life. In our patient, who is a young man, the diagnosis was picked up late and when surgical evaluation was performed, it was considered to be fruitless with findings of nonviable brain tissue on MRI imaging.

CASE REPORT

PRESENTATION:

A 42 year old African American male presented to the Emergency department with four days history of localized left lower extremity weakness and pain. His medical history was significant for hypertension (HTN) and he had recently been admitted to a nearby hospital for left sided weakness and dysarthria. At that time he was found to have right frontoparietal stroke with diminished flow in the left internal carotid artery. He was subsequently discharged to a rehabilitation facility on Aspirin 325 mg po q day, lisinopril 5 mg q day and Amlodipine 10 mg q day. His social history was significant for drinking 1 pint of vodka per day for the past 8-9 years, occasional tobacco use and occasional cocaine use (last use one month prior). His family history was significant for Diabetes Mellitus and hypertension (no history of premature coronary artery disease or cerebrovascular accidents was reported).

ASSESSMENT:

The patient was of thin stature. Upon arrival his blood pressure was 146/96 mm Hg. Rest of the vital signs were within normal limits. The neurologic examination was positive for a depressed affect and late response with moderate cognitive impairment. Strength on the left upper and lower extremities was 3/5 and he had positive left sided Babinski's sign. The neck was supple and the rest of the systemic examination was unremarkable. Laboratory data revealed normal comprehensive blood picture and basic metabolic panel.

The non-contrast CT scan of the head showed an area of decreased attenuation within the right centrum semiovale with the suspicion of an evolving ischemic event. There was evidence of old ischemic strokes in the left frontal area and bilateral lacunar infarcts involving the basal ganglia. No associated intracranial hemorrhage was noticed. (Figures 1-4). At that time Aggrenox 1 capsule BID was added to his medications along with Pravastatin 40 mg po q hs. The coagulation workup for Anti-thrombin III, Homocysteine along with immunologic studies with Complement level C3 and C4 were within normal limits. Urine toxicology failed to reveal evidence of any substance abuse. Lumbar puncture was performed to rule out possibility of any infectious process in the brain. Cerebrospinal fluid analysis including VDRL and RPR was normal. Liver enzymes as well as Hepatitis and Human Immunodeficiency Virus (HIV) serology was negative. As part of work up for stroke in young people, 2 D echocardiogram was performed which failed to reveal any shunt or intra arterial or intra-ventricular clots.

DIAGNOSIS:

During the patient's stay in the hospital further brain imaging was performed. A brain MRI demonstrated an area of bright T2 and FLAIR signal in the anterior right frontal lobe and right centrum semiovale area as well as restricted diffusion in the right centrum semiovale. These were compatible with acute or subacute areas of infarction (Figures 5-8) Old infarcts of the right frontal lobe and left frontal lobe were seen (Figures 5-9). No evidence of hemorrhage was identified on T1-weighted imaging. T2-weighted and FLAIR images demonstrate an old tiny left caudate head lacunar infarct shown by an area of T2-weighted bright signal (Figure 9). Edematous change in the area of the right parietal lobe, creating some mass effect on the ventricular system and 5-mm right-to-left shift of the midline at the level of the lateral ventricles was noticed (Figure 10).

To look for the cause of these multiple strokes, bilateral carotid artery duplex scan was performed. It showed moderate 16-49% stenosis at bilateral bifurcation site. Magnetic Resonance Arteriography (MRA) revealed an abnormality at the level of the circle of Willis. Small filling defects were seen throughout the right and left middle cerebral artery and their branches suspicious of arterial occlusive disease involving the anterior aspect of the circle of Willis and the middle cerebral arteries (Figures 11-14). In lieu of these findings conventional cerebral angiogram was recommended. On the third hospital day, the cerebral angiogram confirmed findings suggestive of a rare disease 'Moyamoya disease' with vascular occlusion of the intracranial internal carotid artery and multiple collaterals supplying the anterior aspect of the brain. Most of the blood supply to the brain was derived from the posterior circulation (Figure 15).

MANAGEMENT:

The patient was sent to a tertiary care centre for possible surgical consideration. A brain tissue SPECT was done which showed nonviable brain tissue and the decision was made not to operate upon the patient. He was subsequently discharged to a rehabilitation centre.

DISCUSSION

Moyamoya disease is a chronic cerebral vasculopathy first described in Japan in 1957 (1). It was observed that the collateral vessels after severe bilateral stenosis or occlusion of the arteries around the circle of Willis give the appearance of a puff of smoke on arteriography and the name "moyamoya" (Japanese: puff of smoke) was given to the disease. The etiology remains unknown with some studies showing familial tendency as well as some association with Sickle cell disease, Neurofibromatosis type 1 (NF1), Fibromuscular dysplasia, Down's syndrome, and the use of oral contraceptive pills.

When moyamoya disease is associated with any of the above etiology it is called 'moyamoya syndrome'. The differential diagnosis of moyamoya includes: multi-infarct dementia, radiation induced cranial arteritis and vasculitis of cerebral arteries. The overall incidence of moyamoya in Japan is 0.35 per 100,000 (nationwide survey) (2).

Very few cases of moyamoya disease were reported in the past in the Unites States but now with increased awareness more cases are being diagnosed and it is becoming possible to study the etiological, diagnostic and treatment trends in this disease (3, 4, 5, 6, 7, 8, 9). Considering the racial trend of this disease, in USA far less cases are seen in African Americans as compared to Caucasians and others ethnicities (0.13 per 100,000 according a study) (4). Brain tissue sample of patients with moyamoya disease usually reveals evidence of prior stroke, and the risk of subsequent strokes may be as high as 10% per year. The usual progression of Moyamoya Disease is worsening vascular stenosis and increased collateral formation, ultimately leading to a hemorrhagic stroke, which is the most common finding on autopsy.

Since the cause of moyamoya disease is unknown, treatment in the acute phase is mainly symptomatic. Anticoagulants, antiplatelet drugs, and corticosteroids have been administered without obvious benefit. Surgery is recommended in most patients, particularly in light of the ineffectiveness of medical treatment and is intended to restore the circulation for the ischemic brain area to prevent further infarcts and transient ischemic attack (TIA) (10). Although many patients undergo surgical revascularization procedures (direct and indirect) the relative safety and efficacy of surgical revascularization is not proven in adults. Surgical treatment in hemorrhagic Moyamoya remains controversial, as it may increase the risk of hemorrhagic events (11).

TEACHING POINT

Moyamoya disease is an uncommon chronic cerebral vasculopathy of an unclear etiology and causes recurrent cerebro-vascular accidents, with intracerebral hemorrhage

being the most common cause of death. Although many patients undergo surgical revascularization procedures, the relative safety and efficacy of this treatment is unproven in adults.

ABBREVIATIONS

HTN- Hypertension
DM- Diabetes Mellitus
CVA- Cerebro-vascular accident
MRA- Magnetic Resonance Angiography
CT- Computed Tomography
MRI- Magnetic Resonance Imaging
MCA- Middle Cerebral Artery
TIA- Transient Ischemic Attack
VDRL- Venereal Disease Research Laboratory
RPR- Rapid Plasma Reagent
HIV- Human Immunodeficiency Virus
SPECT- Single Photon Emission Computed Tomography
NF1- Neurofibromatosis type 1

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FIGURES

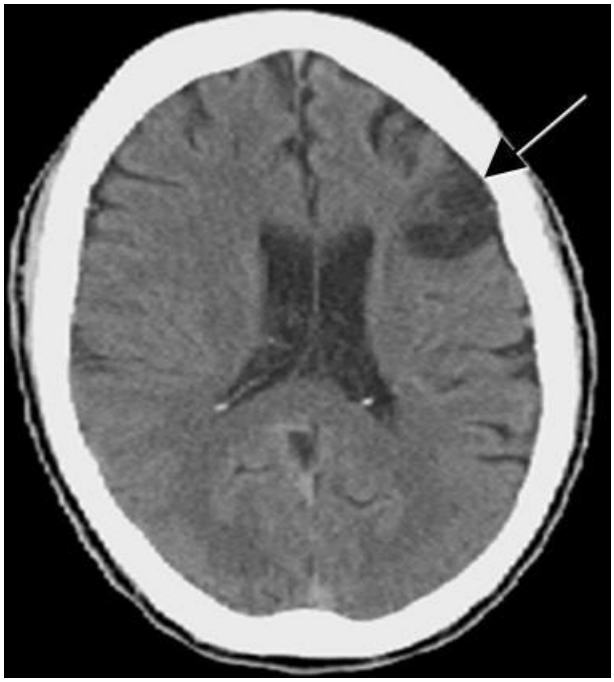

Figure 1: Axial CT scan of brain without contrast. Area of old ischemic infarction involving the left frontal region (arrow).

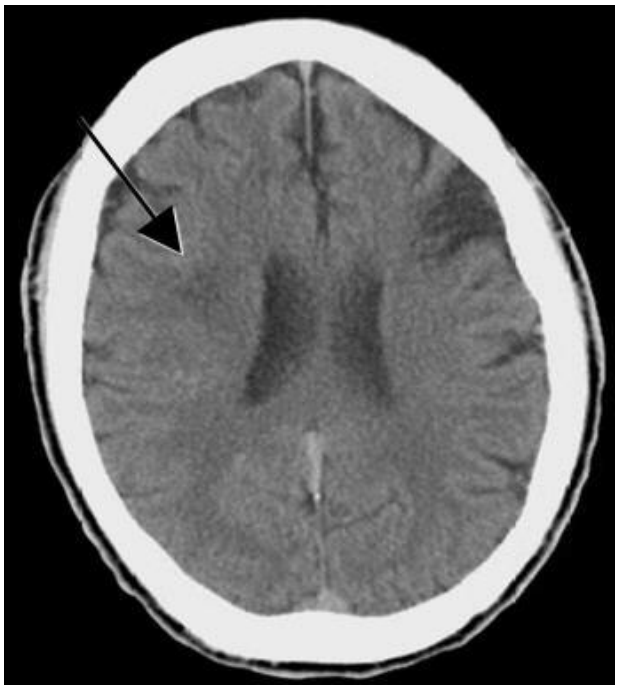

Figure 2: Axial CT scan of brain without contrast. Area of decreased attenuation within the right centrum semiovale (arrow), no associated intracranial hemorrhage.

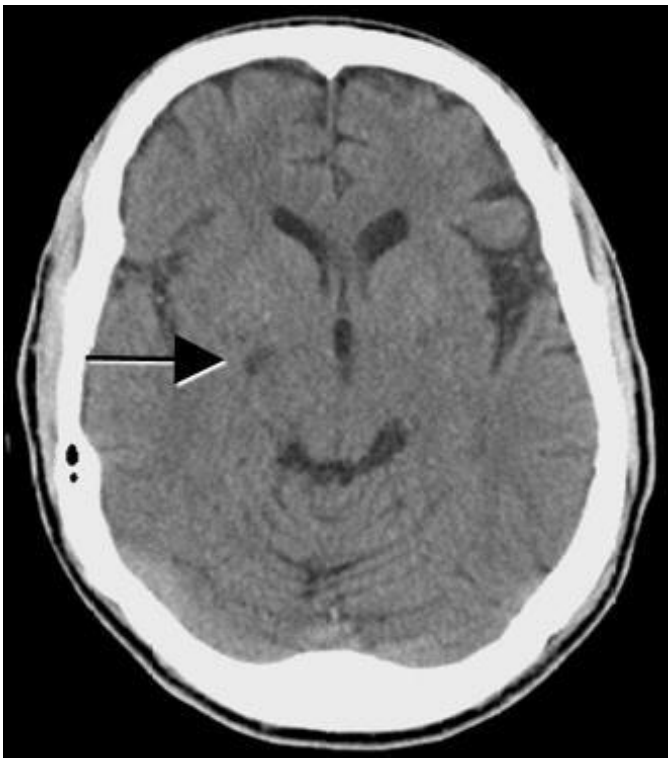

Figure 3: Axial CT scan of brain without contrast. Small lacunar infarct involving the right basal ganglia (arrow).

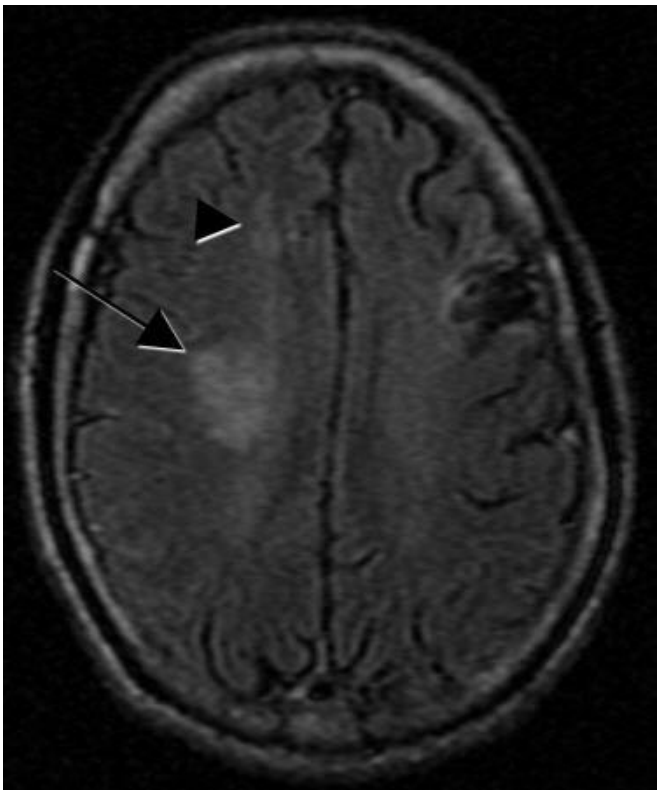

Figure 5: Brain MRI without contrast (axial Flair sequence): Acute or subacute right centrum semiovale infarct (arrow) with a small focal area of probable lacunar infarct in the right frontal lobe (arrowhead).

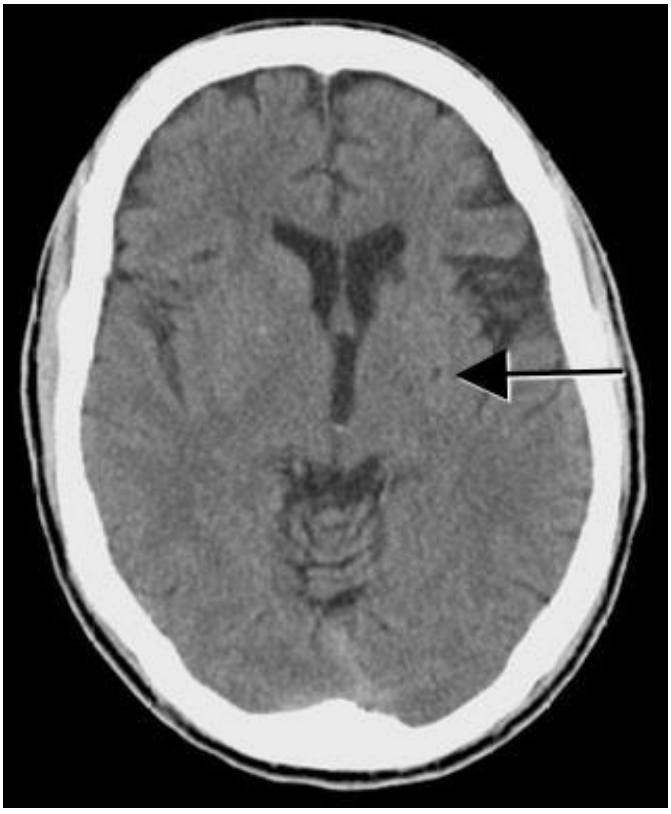

Figure 4: Axial CT scan of brain without contrast. Small lacunar infarct involving the left basal ganglia (arrow).

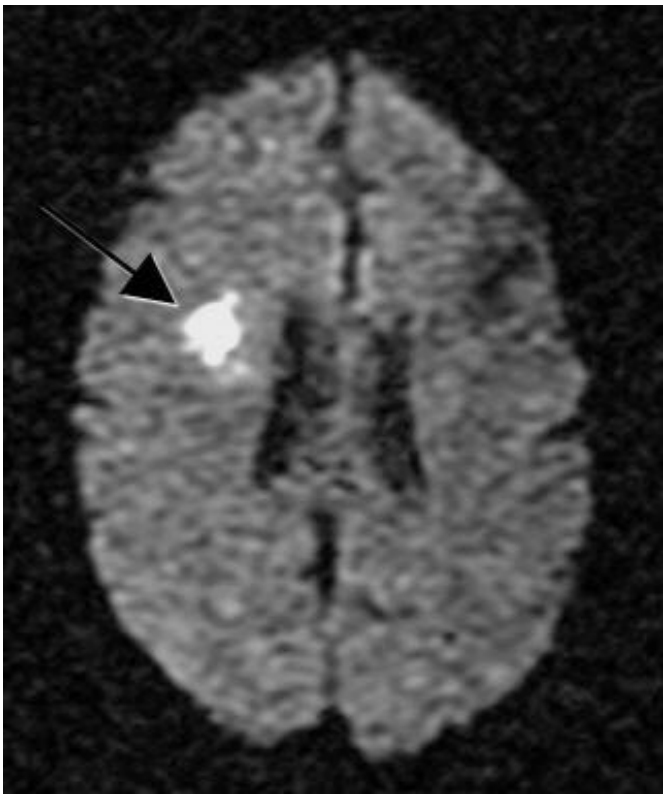

Figure 6: Brain MRI without contrast (axial DWI sequence): Showing bright signal in right parietal and right centrum semi-ovale (arrow).

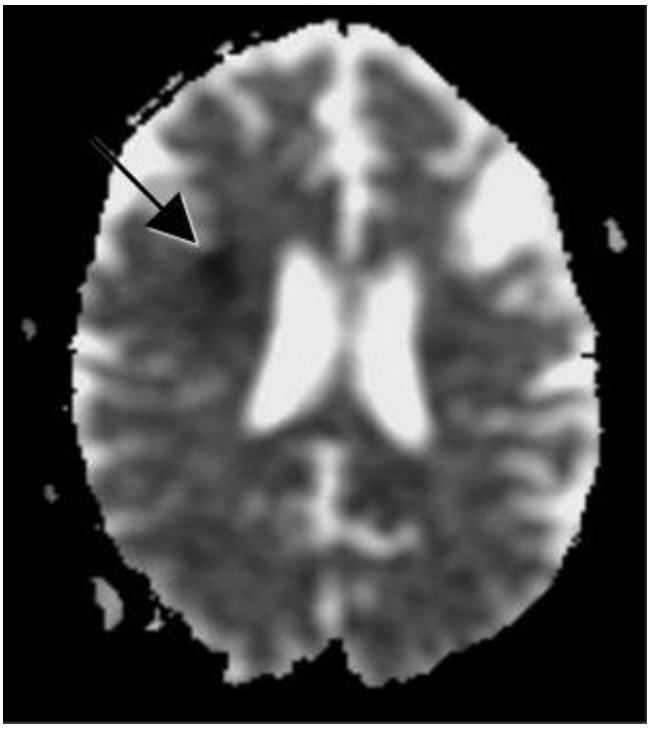

Figure 7: Brain MRI without contrast (ADC map): Showing infarct in right frontal lobe (arrow).

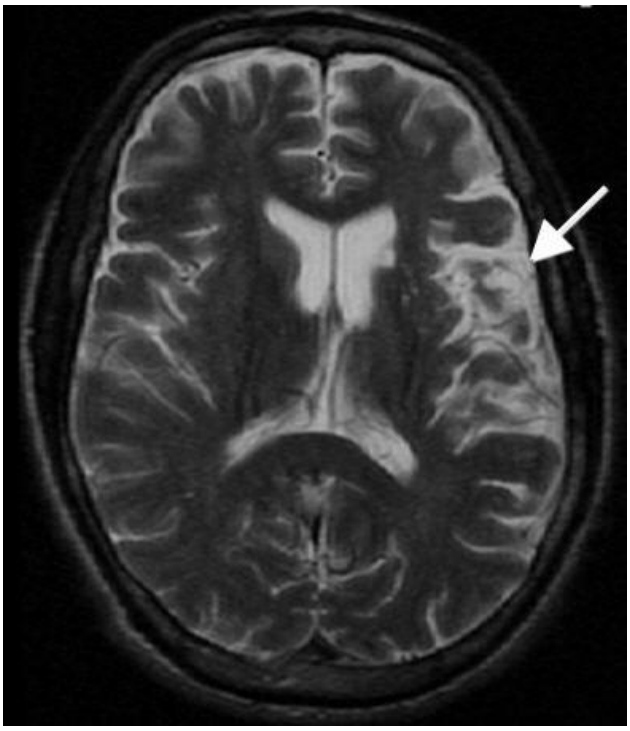

Figure 9: Brain MRI without contrast (axial T2 weighted sequence): Old infarct in left parietal region (arrow).

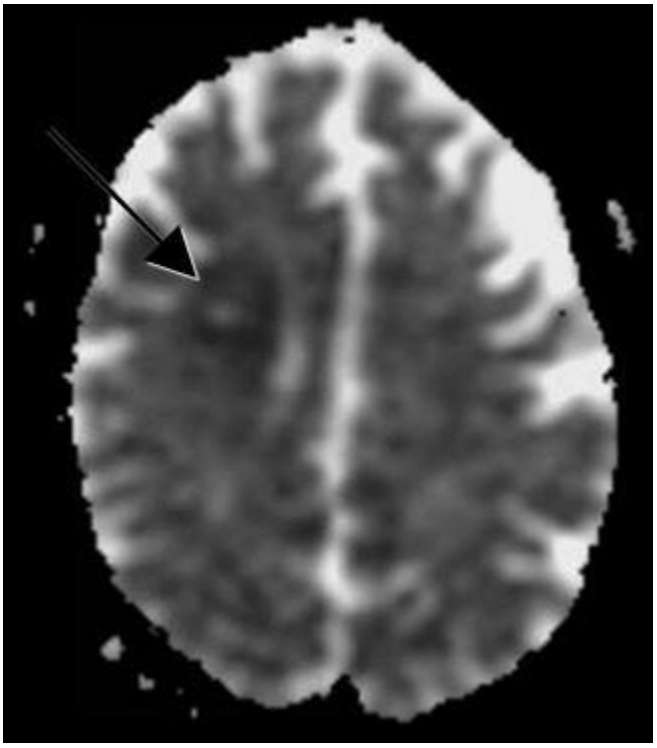

Figure 8: Brain MRI without contrast (ADC map): Showing infarct in the right centrum semiovale (arrow).

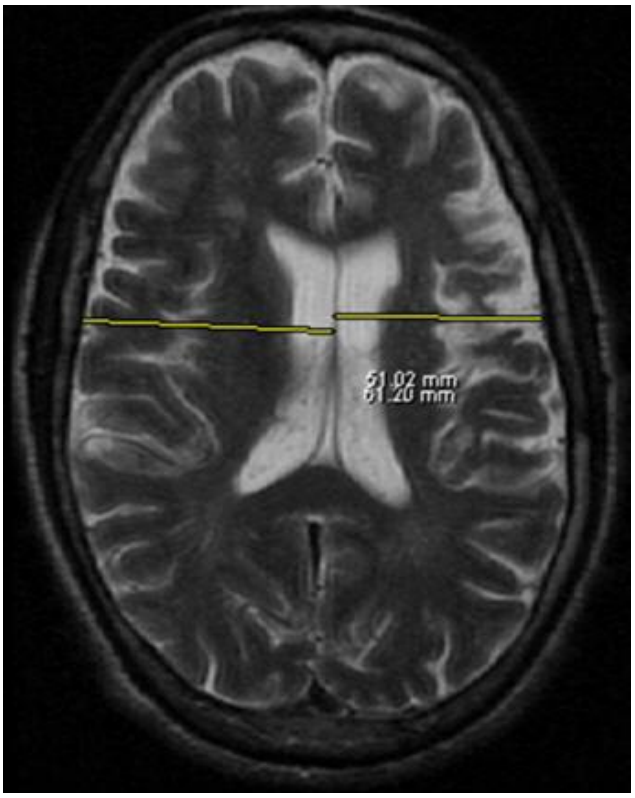

Figure 10: Brain MRI without contrast (axial T2 weighted sequence): Edematous changes in right parietal lobe with 5 mm right to left shift (marked by measurement lines). Mild generalized atrophy.

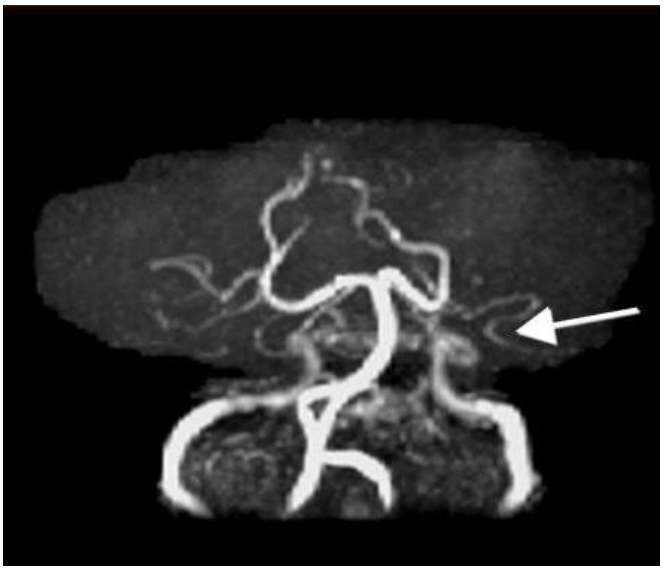

Figure 11: Brain MR angiogram with contrast (3D time of flight imaging). Abnormality was seen at the level of circle of Willis. Very minimal filling defects are seen throughout the right and left middle cerebral artery and their branches. Arterial occlusive disease appears to be present involving the anterior aspect of the circle of Willis and the middle cerebral arteries.

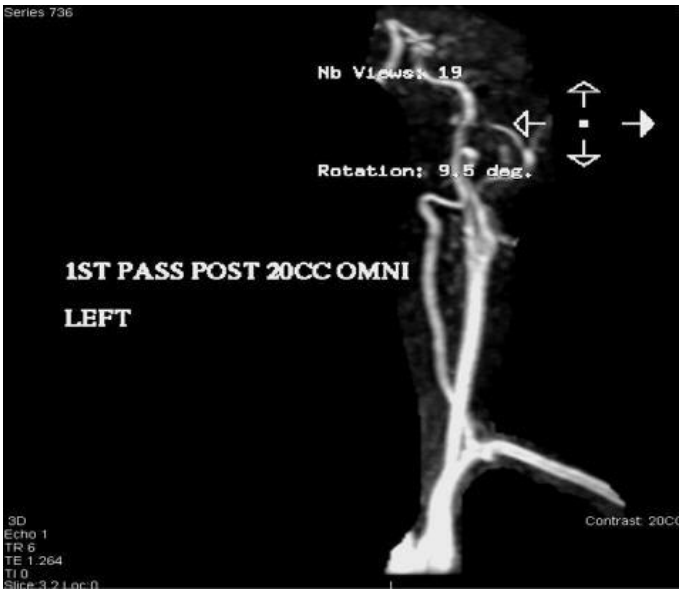

Figure 12: Brain MR angiogram with contrast (3D time of flight imaging) showing minimal filling defects throughout the left middle cerebral artery and their branches.

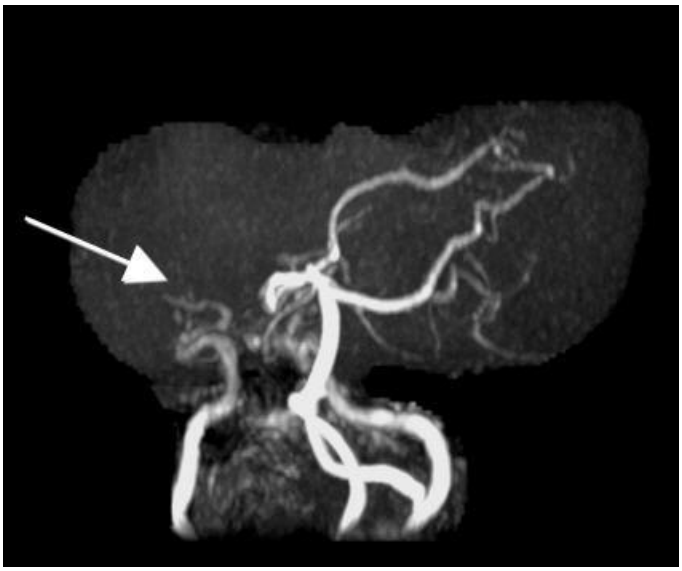

Figure 13: Brain MR angiogram with contrast (3D time of flight imaging) showing minimal filling defects throughout the right and left middle cerebral artery and their branches.

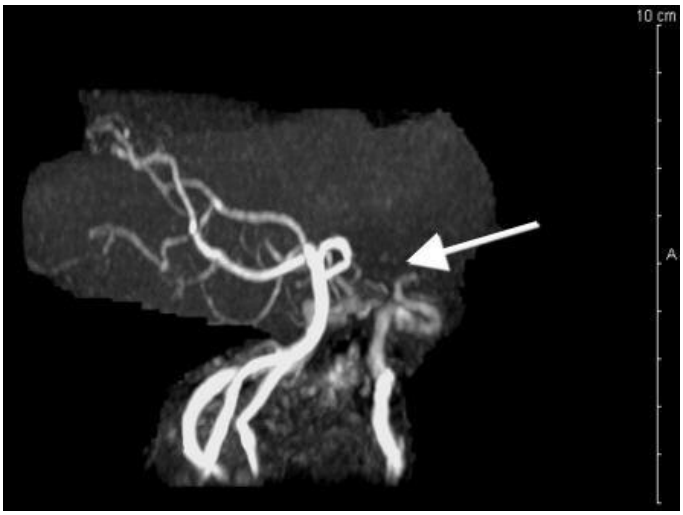

Figure 14: Brain MR angiogram with contrast (3D time of flight imaging) showing minimal filling defects throughout the right and left middle cerebral artery and their branches.

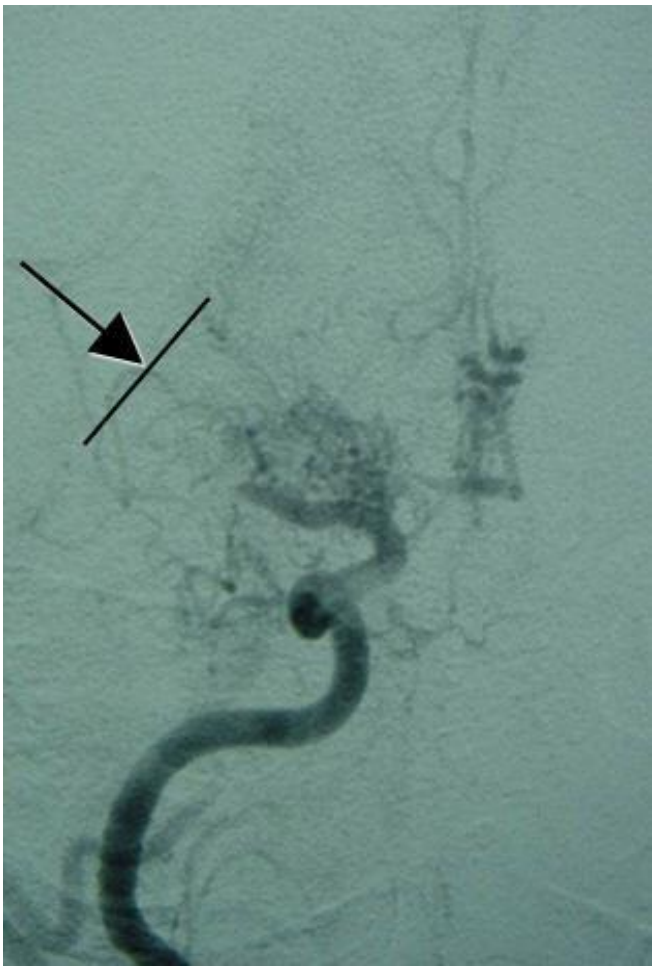

Figure 15: Conventional cerebral angiogram. Moyamoya disease with vascular occlusion of the intracranial internal carotid artery with multiple collaterals supplying the anterior aspect of the brain (arrow). Most of the supply is from the posterior circulation as shown on MRA.

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