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Imaging of Moya Moya in Young Adult: About 3 Cases:

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Abstract:*A persistent non-inflammatory, non-atherosclerotic occlusive vasculopathy in is known as Moya Moya angiopathy (MM). It is typified by progressive occlusion or stenosis at the apices of the intracranial the internal carotid artery (ICA), which include the middle and proximity anterior cerebral arteries. This can result in hemorrhagic or ischemic stroke, which has a high risk of mortality as well as morbidity* ***[1].*** *Imaging is essential to diagnosis, treatment planning, and post-treatment. Arteriography is used to confirm the diagnosis. It demonstrates the pathognomonic growth and dilatation of the lenticulostriate arteries, which is characterized by stenosis in the ICA branches and resembles a puff of smoke. The three angiographic criteria for a diagnosis of moyamoya are as follows: (1) bilateral observations of vascular alterations; (2) stenosis of the distal ICAs together with portions of proximal ACA and MCA; and (3) dilated baseline collateral arteries.* ***[2]****. There aren't many options for treatment right now, but surgical revascularization might stop ischemic episodes and maintain quality of life. The rarity of this pathology has led us to present three examples of Moya-Moya disease in young people in this review.*

Keywords: Moya-Moya disease, Stroke disease, Imaging.

Background:

An uncommon and progressive cerebrovascular disease known as moyamoya is brought on by clogged arteries in the basal ganglia, a region of the brain near the base of the brain. The Japanese term "moyamoya," which translates to "puff of smoke," refers to the tangled look of tiny vessels that are making up for the obstruction. [**3]**

Compared to persons in the Western Hemisphere, residents of East Asian nations like Korea and Japan have a comparatively higher prevalence of MM for an unexplained cause. **[4]**

This medical condition may show in a variety of ways, mostly as a stroke. Young people who have had a hemorrhagic stroke and pediatric patients who have had a vascular accidents transitory ischemic cerebral stroke. **[5]**

Imaging is essential to a successful diagnosis and to the ongoing treatment of the condition. The gold standard for assessing a disease's anatomy is still angiography, although MRI, a non-invasive, non-irradiating imaging technique, has a role in follow-up, particularly in pediatric patients.

Here, we document three instances of young adult moya moya illness, two of which were hemorrhagic stroke cases and one of which was ischemic. Every patient had an MRI, CT scan, and just a single intervention arthroplasty. We'll research patient outcomes and management.

Case Presentation:

45-year-old male patient who is right-handed who was taken to the emergency room due to a transient ischemic attack (TIA) that caused abrupt onset weakness in his left hemibody and facial asymmetry that resolved in 20 minutes.

The patient's arterial blood pressure was elevated upon admission (160/95 mmHg), but his heart rate and respiration were normal. He was also aware and stable.

A neurological evaluation revealed no impairment of the nervous system. The NIH score was not positive.

Negative Babinski sign.

Both the pulmonary and cardiovascular exams were normal.

After the patient's MRI, which revealed a little hematoma in the internal capsule (Figure 1).

Given the patient's young age and the need to rule out a vascular abnormality, a CT vasculogram was carried out. The results revealed a bilateral intracranial internal carotid artery stenosis that spread to the anterior and middle cerebral arteries.(Figure 2)

As a result, we suspected moya moya and carried out a conventional angiogram, which verified the diagnosis and revealed the same results at the CT vasculogramm as well as swollen lenticulostrial arteries that resembled a "puff of smoke." (Figure 3)

The patient was administered aspirin and simvastatin among other medications. Surgery was scheduled as a result of repeated symptoms (dysarthria, worsening of hemorrhaging) during hospitalization.

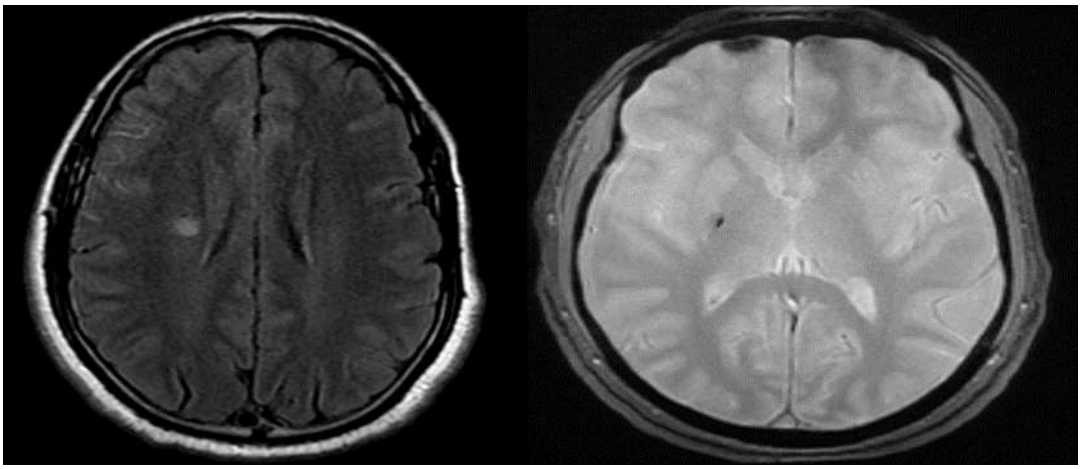

Figure 1: An MRI of the brain confirmed the existence of a hemorrhagic stroke in the internal capsule's posterior arm.

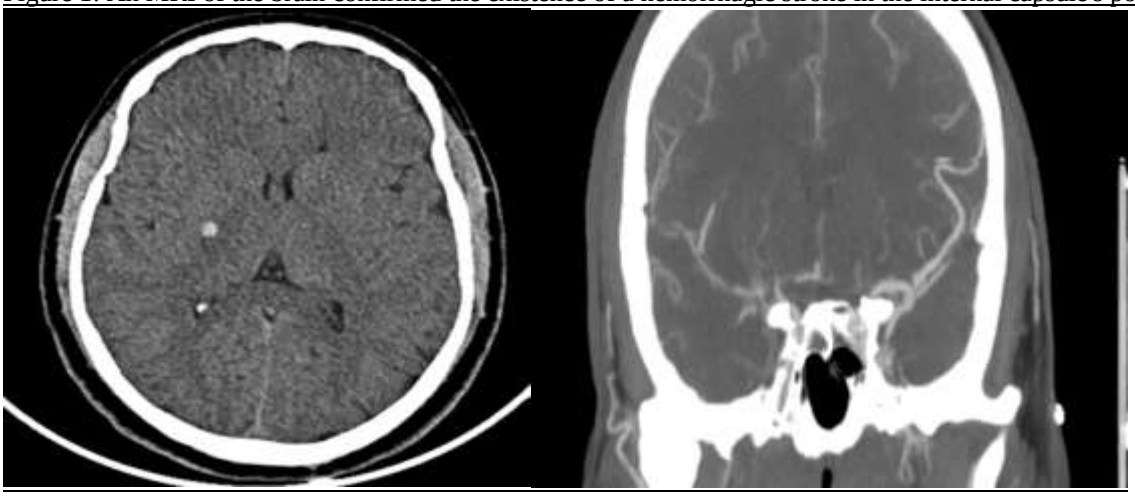

Figure 2: Angio-PW aims for stenosis of the right proximal ACA and MCA, and a brain CT scan confirmed the existence of a hemorrhagic stroke of the right posterior arm of the internal capsule.

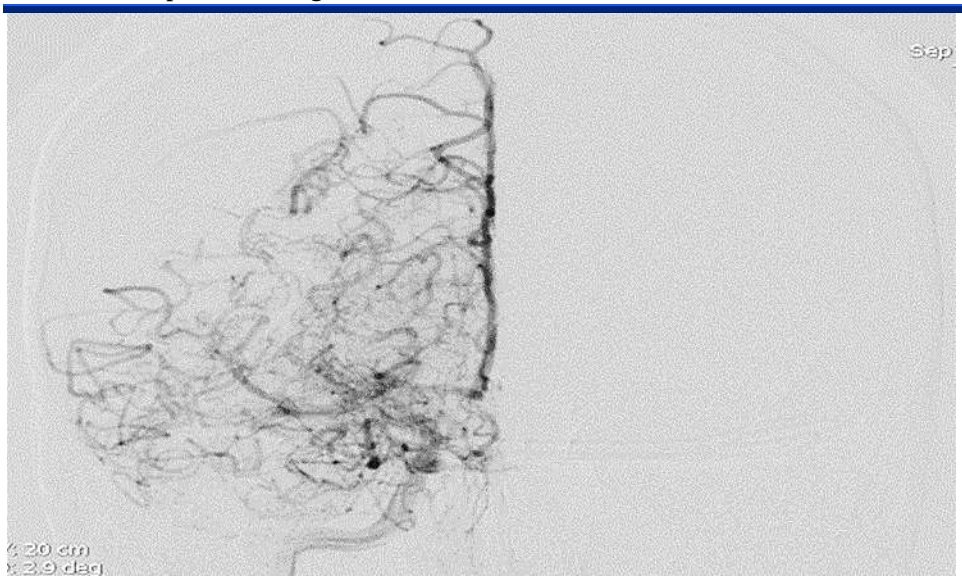

Figure 3: "Puff of smoke" caused by stenosis of the right proximity MCA and ACA, with dilated basal collateral arteries compensating for the occlusion".

Case Presentation:

A 47-year-old woman's symptoms began two years ago with a brief bout of left weakness in the face that was not looked into. Actually hospitalized due to left hemibody weakness.

Upon clinical assessment, the patient was steady and completely conscious. Language and speech functions were perfectly normal. His blood pressure at the time was 120/80 mmHg, which is normal.

At the time of admission, he had a normal neurological evaluation and a zero National Institutes of Health Stroke Scale (NIHSS) score.

No indication of an arrhythmia, such as atrial fibrillation, was seen using Holter-ECG.

Lab testing included a complete bloodstream are counted, the function of the liver tests, and a thorough ionogram were all normal.

After the patient's CT scan, a right Middle Cerebellar Artery (MCA) infarction was discovered.(Figure 4)

A CT scan of the vasculature revealed a bilateral proximal branches of the ACA and intracranial internally carotid arteries were stenotic, with tiny aberrant net-like capillaries proliferating and producing the recognizable "puff of smokes." (Figure 5)

An examination of the blood and CSF revealed no anomalies, which helped to rule out the diagnosis of autoimmune vasculutis.

The patient had been diagnosed with moyamoya illness since no cause could be identified.

For the patient's continued stroke prevention, aspirin and a statin were given. The patient fully recovered clinically, and there was no change in the vascular status at the next follow-up. The secondary preventive treatment regimen was lowered to aspirin as a single medication after three months. His programming called for surgery.

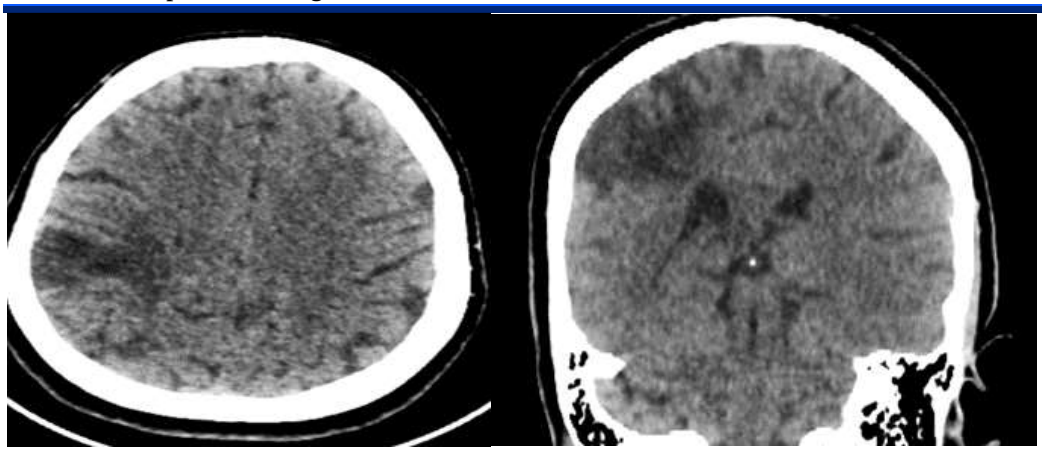

Figure 4: Infarction of the right Medium Cerebellar Artery (MCA). Hypoattenuating foci are seen across the right side white matter and sulcal effacement in the right MCA area on axial and coronal non-enhanced computed tomography, which are consistent with infarction.

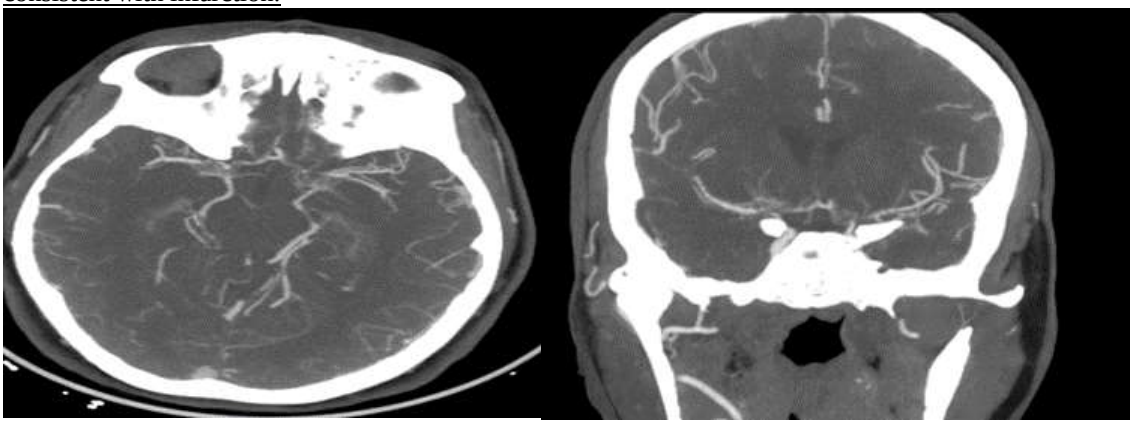

Figure 5: Angio-CT: proximity branch of the ACA and intracranial the internal carotid arteries stenosis on both sides linked to tiny aberrant net-like vessels proliferate, producing the recognized "puff of smoke".

Case Presentation:

A 40-year-old patient who had a remarkable history of diabetes type 2 was brought into our department due to an unexpectedly severe headache.
Upon assessment, he had a Glasgow, for instance Coma Score of 12, which indicated confusion.
No specific neurological deficiency was present. The funduscopy revealed no signs of retinal haemorrhage or papilloedema. 97% of the air's saturation was oxygen, and the respiratory rate was 20. At 110 bpm, he was tachycardic but apyretic. He had a consistent high blood pressure reading of 110/70 across all arms. No extra cardiac sounds were audible. There were no lumps in his soft, non-tender abdomen.
An immediate brain CT scan was carried out based on the patient's clinical history; the results revealed a diffuse subarachnoid accompanied with intraventricular hemorrhage, rated as a Fischer 4. (Figure 6)

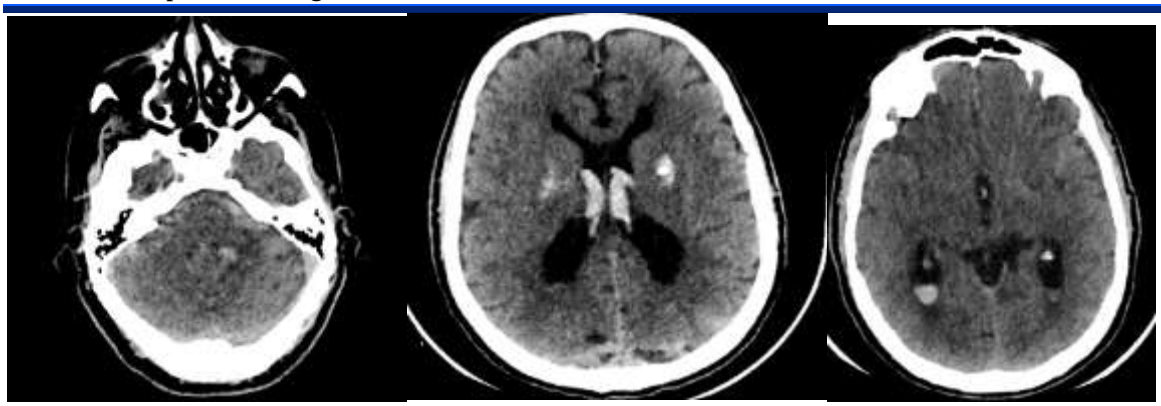

Figure 6: Subcortical and intraventricular hemorrhaging are seen by axially non-enhanced Computed Tomography (CT), Fischer 4.

Angio-CT revealed no aneurysm, but it did objectify a stenosis of the intracranial internal coronary arteries that extended to the proximal branches of the MCA and ACA on both sides. (Figure 7)

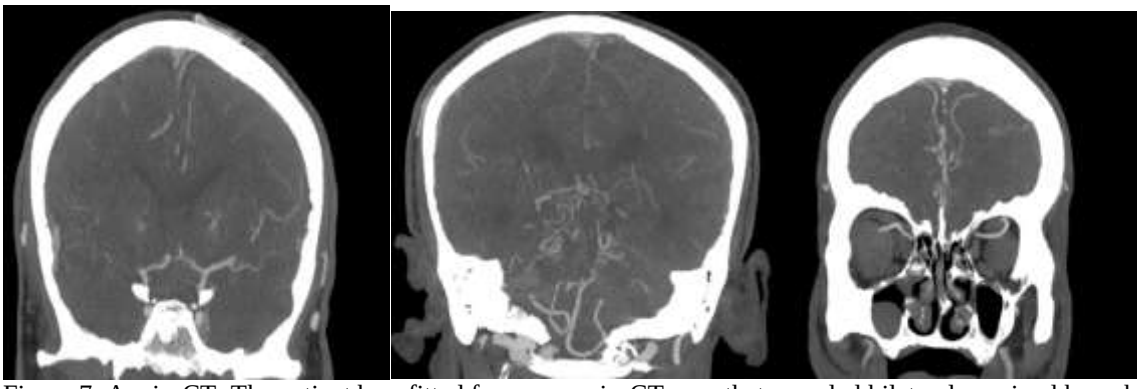

Figure 7: Angio-CT: The patient benefitted from an angio-CT scan that revealed bilateral proximal branches of MCA and ACA and intracranial internal carotid artery stenosis, which are linked to the proliferation of tiny aberrant net-like vessels that give rise to the recognizable "puff of smoke."

We came to a decision that Moya Moya was aggravated by active hydrocephalus, subarachnoid hemorrhage, and intraventricular hemorrhage, which was graded Fischer 4.

Following a discussion with the neurosurgeon team, the patient had external ventricle derivation therapy for his active hydrocephalus, and he is now scheduled for surgery.

Discussion:

MOYA MOYA:

A very rare secondary circulation is affected by MM, an intrinsic primary disease process that results in bilateral progressive stenosis of the anterior intracranial circulation, extending from the proximal segments of the intracranial Internal Carotid Artery (ICA) to the proximal portions of the anterior cerebral artery (ACA) and the middle cerebral artery (MCA) **[6]**. Due to a compensatory reaction, many smaller vessels, including the lenticulostriate arteries, start to grow and multiply. This results in the angiographic appearances of a "Puff of Smoke," or "Moyamoya" in Japanese".

Children and young adults are susceptible to moyamoya, which has a bimodal age distribution:

1. Early Childhood: peak ~4 years of age (two-thirds)
2. Middle Age: 30-40 years of age (one-third)

Nomenclature (disease vs syndrome):

Individuals who exhibit the distinctive moyamoya vasculopathy and also possess established concomitant conditions are classified as having moyamoya syndrome. Research has linked various conditions, including sickle cell anemia, neurofibromatosis-1, Down's

syndrome, congenital cardiac defects, antiphospholipid syndrome, a condition renal artery stenosis, and thyroiditis, to **moyamoya disease**.

On the other hand, those who do not have any identified risk factors are considered to have moyamoya disease. **[7]**

Epidemiology:

Based on epidemiological research and surveys carried out in Japan, the yearly incidence for 100,000 individuals was estimated to be between 0.35 and 0.94 with two peaks, one occurring between the ages of 5 and 9 and the other between 45 and 49. **[4]**

With about 0.086 cases newly diagnosed per 100,000 people annually—roughly one case per million—MM is an uncommon condition in the United States. **[4]**

With the exception of a few isolated instances that have been recorded, no epidemiologic statistics have been created in our nation. However, certain cases have resulted in manuscripts. **[23, 24]**

Pathology:

An uneven internal elastic lamination with luminal constriction, hyperplasia of the tunica the media, and thickening of the intimal region with vacuolar degradation in the tunica media's smooth muscle cells are among the histological findings. Angiogenesis, which results in the growth and multiplication of smaller collateral for blood arteries to enhance blood flow to under perfused parts of the brain, compensates for decreasing blood supply. **[6]**

Clinical Presentation:

There are four clinical types of moyamoya illness, according to the Japanese Health Ministry's classification: ischemic, hemorrhagic, epileptic, and "other." Children are more likely to get the ischemic type, but adults are more likely to contract the hemorrhagic form. **[8]** Transient Ischemic Episodes (TIAs) or lacunar strokes are a kind of the moyamoya illness that cause mental impairment in children. Adults may have subarachnoid hemorrhages as well as other intracranial hemorrhages.

As a result of the gradual stenosis of the ICA and its major branches, changes in cerebral blood flow are thought to be the cause of the symptoms and indicators of MMD. Two main categories might be used to characterize symptoms:

a) As a result of brain ischemia (stroke, TIA, epileptic episodes, etc).
b) As a result of the negative effects of using compensatory mechanisms to prevent persistent ischemia (headache from dilation of transdural the collateral, cerebral hemorrhage from rupturing delicate dilated capillaries, etc). **[13, 14]**

Imaging Finding:

The anatomical and functional characteristics provide the basis for the diagnosis of MM. CT and MRI scans are examples of first-line imaging exams.

Obviously, a **CT scan** is all that is needed to detect an ischemic or hemorrhagic stroke while the condition is still advanced (9). Ischaemic foci may occur periventricularly and subcortically in the white matter and basal ganglia.**(10)**

When big cerebral veins show occlusions or stenoses on an **Angiography**, together with collateral vasculature, this should raise suspicions of the illness, particularly in young patients. **[9]**

MRI displaying bleeding or ischemic lesions; bleeding lesions will show up as a continuous T2* hyposignal and a varied signal in T1 and T2, whereas ischemic lesions will be seen as a hyposignal T1, hypersignal T2, and variable in diffusion.

The **FLAIR** sequence is particularly helpful in diagnosing the "ivy sign," which is an increase in signal intensity throughout the cerebral hemispheres' gyri and fissures that is most likely caused by a decrease in cortical flow. **[11]**

For the purpose of detecting stenosing lesions, **Angio-MRI (MRA)** at time of flight (3D TOF) with superior special resolution is recommended. It is also a helpful noninvasive method for screening and monitoring patients with moyamoya disease in particular youngsters.

As a result, conventional **angiography** is still the preferred test for confirming the diagnosis and staging. Indeed, MM remains definitively diagnosed based on its findings, including the presence of collateral for vasculature in brain's based regions without causal disease, or stenosis or occlusion as of the proximal parts of the anterior and middle arteries and the distal sections of the intracranial internal carotid arteries. **[1]**

As demonstrated by Table 1, angiography also makes it possible to stage the illness using methods like the Suzuki classification.

TABLE 1. THE SUZUKI STAGING SYSTEM[1,36]	
Stage	**Angiographic Findings**
I	Narrowing of the carotid fork (i.e., ICA bifurcation)
II	Initiation of the moyamoya: continued narrowing of the ICA; dilation of the ACA and MCA; initial moyamoya blush
III	Intensification of the moyamoya: loss of proximal ACA and MCA; leptomeningeal collateralization from the PCA; increase in moyamoya blush
IV	Minimization of the moyamoya: progressive occlusion of ICA reaching origin of PCA; reduction in moyamoya blush
V	Reduction of the moyamoya: complete loss of ICA, ACA, and MCA; increased collateral supply from ECA; further reduction in moyamoya blush
VI	Disappearance of the moyamoya: disappearance of blood supply from ICA; blood supply exclusively from ECA; disappearance of moyamoya vessels
Abbreviations: ACA, anterior cerebral artery; ECA, external carotid artery; ICA, internal carotid artery, MCA, middle cerebral artery; PCA, posterior cerebral artery.	

Treatment:

Conservative treatment: It is unknown what causes the disease's therapy. Treatment for mild cases is cautious. Medical treatment, such as antiplatelet and anticoagulant medication, which are seldom used owing to the danger of bleeding, may be used to prevent ischemia and strokes.

Acetylsalicylic acid treatment is highly advised to avoid ischemia attack recurrence; if acetylsalicylic acid treatment fails to be tolerated or is unsuccessful, clopidogrel or alternative thienopyridine may be given. **[15]** As a result, the goal of pharmaceutical therapy is to aggressively avoid future neurovascular events, and there is no approved gold standard medication regimen for hemorrhagic or ischemic sequelae. Furthermore, it is also advised to strictly manage other risk factors as diabetes, hypertension, and dyslipidemia. **[16, 15]**

Surgical treatment: Surgery, including both indirect and direct anastomoses, is recommended in extreme situations. Usually, anastomoses are made between the middle cerebral artery and the superficial temporal artery. One of the indirect treatments is synangiosis, which involves inserting vascularized tissue in the cortex of the brain to encourage the growth of new blood vessels. Recently, a young lady with moyamoya syndromes and recurrent symptoms of TIA was successfully treated by having a stent implanted in her internal carotid artery. **[17]**

Japanese Adults Moyamoya (JAM) trial regulations:

The most current moyamoya guideline in Japan supports immediate revascularization surgery for individuals with MMD presenting as cerebral ischemia symptoms.
Considering hemorrhagic-onset patients, there has been a disagreement regarding whether revascularization surgery has a possible role in lowering the risk of re-bleeding. **[18].**
Causal treatment of the disease is not known. Cases of mild clinical course are normally treated conservatively. In severe cases, it is indicated to carry out surgery [13]. It is also indi-cated to administer antiplatelet medicines. Anticoagulative therapy is rarely used, due to the risk of bleeding [12].
Surgery includes direct anastomoses, indirect procedures, and combined therapies. Anastomoses are usually per-formed between the superficial temporal artery and the Causal treatment of the disease is not known. Cases of mild clinical course are normally treated conservatively. In severe cases, it is indicated to carry out surgery [13]. It is also indi-cated to administer antiplatelet medicines. Anticoagulative therapy is rarely used, due to the risk of bleeding [12].
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Outcome:

Surgical therapy of MMD with different revascularization procedures has been shown to be safe and successful in lowering ischemia events in children as well as adults. **[19, 20]** Post-revascularization angiography and MRI investigations often demonstrate a decrease in lenticulo-strial collaterally, which closely tracks improvements in symptomatic in both adult and pediatric populations. **[21, 22].**

Conclusions:

Moya-Moya disease remains an uncommon occurrence in our nation, therefore radiologists should be aware of it, since imaging, particularly conventional angiography, is critical to the diagnosis. If surgical was the sole effective form of treatment, radiographic interventional methods such as stenting are now more often employed. Permitting an efficient the revascularization and preventing patients operation problems with a gratifying result in most circumstances.

List of abbreviations :

CT: Computerized Tomography
MM: Moya Moya
TIA: Transient ischemic attack
NIHSS: National Institutes of Health Stroke Scale
ICA: Internal Carotid Artery
ACA: the anterior cerebral artery
MCA: the middle cerebral artery

Declarations:

The authors do not declare any conflict of interest

Ethics approval and consent to participate

Not applicable

Consent for publication

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

The data sets are generated on the data system of the CHU Hassan II of Fes, including the biological data.

Competing interests

The authors declare that they have no competing interests

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Author's contribution

Saddouki Fatima is the corresponding author, he participated in the organization and writing of the article and studied the cases. Professor **EL Bouardi Nizar** supervised working and validated the figures.

Professor **EL Bouardi Nizar** and chief of the department of radiology read and allowed the article for publication.

All authors read and approved the final manuscript.

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