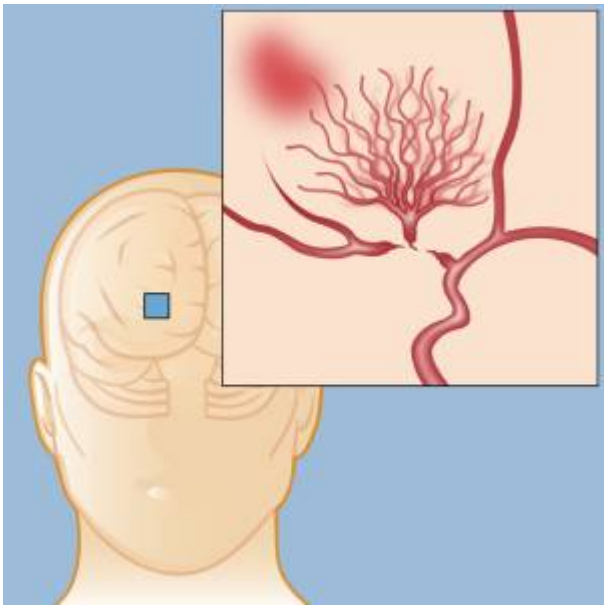
Moyamoya Disease Diagnosis

- The role of systemic inflammation in the formation and rupture of intracranial aneurysms in moyamoya disease: a retrospective cohort study
- Moyamoya disease in a 10-year-old male patient in the Middle East with the outcome of the surgery: A case report and literature review
- Analysis of risk factors for the occurrence of rebleeding following surgery for haemorrhagic moyamoya disease
- The Orbital Grading system yields higher precision than the Matsushima grading system in assessing angiographic outcomes after EDAS for Moyamoya disease: an interrater reliability analysis
- Long-Term Outcomes in Patients With Hemorrhagic Moyamoya Disease Combined With Hypertension After Encephaloduroarteriosynangiosis
- Association of intravascular enhancement sign on 3D T1- weighted TSE sequences with cerebral perfusion and infarction events in moyamoya disease
- Blurred by a "Puff of Smoke"-A Case-Based Review on the Challenging Recognition of Coexisting CNS Demyelinating Disease and Moyamoya Angiopathy
- Moyamoya syndrome in a 6-year-old beta-thalassemia major patient: A case report



Diagnosis of Moyamoya disease requires bilateral symmetrical stenosis or occlusion of the terminal portion of the internal carotid arterys (ICA)s as well as the presence of dilated collateral vessels at the base of the brain [1]. (If unilateral, the diagnosis is considered questionable, [2] and these cases may progress to bilateral involvement).

Other characteristic findings include:

1. stenosis/occlusion starting at the termination of ICA and at origins of ACA and MCA

2. abnormal vascular network in the region of BG (intraparenchymal anastomosis).

3. transdural anastomosis(rete mirabile), AKA “vault moyamoya.”Contributing arteries: anterior falcial, middle meningeal, ethmoidal, occipital, tentorial, STA

4. moyamoya collaterals may also form from the internal maxillary artery via ethmoid sinus to the

forebrain in the frontobasal region.

CT

Work-up in suspected cases typically begins with a non-enhanced head CT. Up to 40% of ischemic cases have normal CT. Low-density areas (LDAs) may be seen, usually confined to cortical and subcortical areas (unlike atherosclerotic disease or acute infantile hemiplegia which tend to have LDAs in basal ganglia as well). LDAs tend to be multiple and bilateral, especially in the PCA distribution (poor collaterals), and are more common in children.

Magnetic resonance imaging

Magnetic resonance imaging for Moyamoya Disease Diagnosis.

Angiography

In addition to helping to establish the diagnosis, angiography also identifies suitable vessels for revascularization procedures and unearths associated aneurysms. The angiography-related complication rate is higher than with atherosclerotic occlusive disease. Avoid dehydration prior to and hypotension during the procedure. Six angiographic stages of MMD are described by Suzuki and Takaku [3] that tend to progress up until adolescence and stabilize by age 20.

1 stenosis of suprasellar ICA, usually bilateral

2 development of moyamoya vessels at the base of the brain; ACA MCA & PCA dilated

3 increasing ICA stenosis & prominence of moya-moya vessels (most cases diagnosed at this stage); maximal basal moyamoya

4 entire circle of Willis and PCAs occluded, extracranial collaterals start to appear, moyamoya vessels begin to diminish

5 further progression of stage 4

6 complete absence of moyamoya vessels and major cerebral arteries.

EEG

Non-specific in the adult. Juvenile cases: high-voltage slow waves may be seen at rest, predominantly in the occipital and frontal lobes. Hyperventilation produces a normal buildup of monophasic slow waves (delta-bursts) that return to normal 20–60 seconds after hyperventilation. In >50%of cases, after or sometimes continuous with buildup is a second phase of slow waves (this characteristic finding is called “rebuild up”) which are more irregular and slower than the earlier waves, and usually, normalize in ≤10 minutes [4].

Cerebral blood flow (CBF) studies

CBF is decreased in children with MMD, but relatively normal in adults. There is a shift of CBF from the frontal to the occipital lobes [5] probably reflecting the increasing dependency of CBF on the posterior circulation. Children with MMD have impaired autoregulation of CBF to blood pressure and CO2 (with more impairment of vasodilatation in response to hypercapnia or hypotension than vasoconstriction in response to hypocapnia or hypertension) [6]. Xenon (Xe-133) CT can identify areas of low perfusion. Repeating the study after an acetazolamide challenge (which causes vasodilatation) evaluates the reserve capacity of CBF and can identify areas of “steal” which are at high risk of future infarction.

Ultrasound

Ultrasound parameters are independently correlated with ipsilateral cerebral stroke in patients with Moyamoya disease (MMD). Ultrasound provides a new way to identify stroke in MMD patients. Future prospective cohort studies are needed to verify the clinical value of ultrasound in identifying patients with MMD at high risk of stroke [7].

18F-FDG PET

Vascular cognitive impairment (VCI) is a critical issue in moyamoya disease (MMD). However, the glucose metabolic pattern in these patients is still unknown. This study aimed to identify the metabolic signature of cognitive impairment in patients with MMD using 18F-2-fluoro-2-deoxy-D-glucose positron emission tomography (18F-FDG PET) and establish a classifier to identify VCI in patients with MMD. One hundred fifty-two patients with MMD who underwent brain 18F-FDG PET scans before surgery were enrolled and classified into nonvascular cognitive impairment (non-VCI, n = 52) and vascular cognitive impairment (VCI, n = 100) groups according to neuropsychological test results. Additionally, thirty-three health controls (HCs) were also enrolled. Compared to HCs, patients in the VCI group exhibited extensive hypometabolism in the bilateral frontal and cingulate regions and hypermetabolism in the bilateral cerebellum, while patients in the non-VCI group showed hypermetabolism only in the cerebellum and slight hypometabolism in the frontal and temporal regions. In addition, we found that the patients in the VCI group showed hypometabolism mainly in the left basal ganglia compared to those in the non-VCI group. The sparse representation-based classifier algorithm taking the SUVr of 116 Anatomical Automatic Labeling (AAL) areas as features distinguished patients in the VCI and non-VCI groups with an accuracy of 82.4%. This study demonstrated a characteristic metabolic pattern that can distinguish patients with MMD without VCI from those with VCI, namely, hypometabolic lesions in the left hemisphere played a more important role in cognitive decline in patients with MMD [8].

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