Moya Moya syndrome: how to diagnose?

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Learning objectives

To learn more about Moya Moya disease we propose the following objectives:

· Explain the Moyamoya disease, a rare disease in our territory for whom diagnosis is essential the imaging technics.

· Identify the imaging features that help differentiate the moyamoya disease from other vascular lesions and the methods of imaging we need to use to characterize the disease.

· Discuss the imaging findings that should be included in radiology reports in patients with moyamoya disease.

· Compare between the different diagnostic methods, from the most invasive ones like the conventional angiography to other less invasives like MRI.
Background

Pathogenie:

Moya moya disease is a chronic and progressive narrowing of the internal carotid arteries at the base of the brain where they divide into middle and anterior cerebral arteries.

The walls of the arteries become thickened, which narrows the inside diameter of the vessel. The narrowing can eventually result in complete blockage and stroke.

To compensate for the narrowing arteries, the brain creates collateral blood vessels in an attempt to deliver oxygen rich blood to deprived areas of the brain. These tiny collateral vessels, when seen on an angiogram, have a hazy, filmy appearance.

(The Japanese were the first to describe the condition, and they named it puff of smoke).

The tiny moya moya collaterals can break and bleed into the brain, causing hemorrhages.

It is often accompanied by aneurysms, and it affects both sides of the brain.

Etiology

The cause of moyamoya disease is not known. The disease is believed to be hereditary. Fukui reported a family history in 10% of patients with the disorder. Moreover, Mineharu suggested that familial moyamoya disease is autosomal dominant with incomplete penetrance that depends on age and genomic imprinting factors.

Associated diseases.

Although moyamoya disease may occur by itself in a previously healthy individual, many disease states have been reported in association with moyamoya disease, including the following:

- Immunologic diseases - Infections - Leptospirosis and tuberculosis
- Hematologic disorders - Aplastic anemia, Fanconi anemia, sickle cell anemia, and lupus anticoagulant
Congenital syndromes - Apert syndrome, Down syndrome, Marfan syndrome, tuberous sclerosis, Turner syndrome, von Recklinghausen disease (Neurofibromatosis and Hirschsprung disease)

Vascular diseases - Atherosclerotic disease, coarctation of the aorta and fibromuscular dysplasia, cranial trauma, radiation injury, parasellar tumors, and hypertension

**History**

The natural history of moyamoya is variable. Disease progression can be slow, with rare, intermittent events, or fulminant, with rapid neurologic decline.

However, regardless of the course, moyamoya inevitably progresses in the majority of patients.

**Diagnosis:**

**Clinical sign:**

There are a multitude of symptoms associated with moyamoya disease, including TIAs, ischemic strokes, intracranial hemorrhages, seizures, headaches, choreiform movements, and cognitive deficits. In contrast to adults who often present within the setting of intracranial hemorrhage (#46%) , children affected with moyamoya disease typically exhibit signs and symptoms of cerebral ischemia secondary to TIAs and/or cerebral infarctions.

- **Ischemic Symptoms**

Symptoms of cerebral ischemia in moyamoya are typically associated with the regions of the brain supplied by the internal carotid arteries and middle cerebral arteries; these regions include the frontal, parietal, and temporal lobes. Hemiparesis, dysarthria, aphasia, and cognitive impairment are common.

Patients may also have seizures, visual deficits, syncope, or personality changes that can be mistaken for psychiatric illness. Ischemic symptoms may be transient or fixed.

Ischemic sx of a posterior cerebral artery can be seen .

- **Hemorrhage**
Intracranial hemorrhage is common in adults with moyamoya, but it has also been described in children.

The location of the hemorrhage can be intraventricular, intraparenchymal (frequently in the region of the basal ganglia), or subarachnoid.

Historically, bleeding has been attributed to rupture of fragile collateral vessels associated with moyamoya as progressive stenosis of the internal carotid artery occurs.

Shifting circulatory patterns at the base of the brain have been implicated in the development of cerebral aneurysms (usually at the apex of the basilar artery and posterior communicating artery, areas of increased shear stress in moyamoya); this may be another cause of hemorrhage in moyamoya.

- Epileptic: Epileptic seizures (20 to 30%) Children++

Associated sign:

### Headache is a frequent presenting symptom in patients with moyamoya (dilatation of meningeal and leptomeningeal collateral vessels may stimulate dural nociceptors).

Development of choreiform movements (Dilated moyamoya-associated collateral vessels in the basal ganglia = disk with concomitant retinovascular anomalies).

Moyamoya disease should be taken into consideration and worked up in any child presenting with ischemic symptoms, especially in the setting of hyperventilation, crying, and/or physical exertion. A suspected diagnosis of moyamoya disease is confirmed with radiological studies.
Findings and procedure details

Moyamoya disease is a progressive vasculopathy leading to stenosis of the main intracranial arteries. The incidence of moyamoya disease is high in Asian countries; in Europe and North America, the prevalence of the disease is considerably lower. Clinically, the disease may be of ischaemic, haemorrhagic and epileptic type. Cognitive dysfunction and behavioral disturbance are atypical symptoms of moyamoya disease.

Characteristic angiographic features of the disease include stenosis or occlusion of the arteries of the circle of Willis, as well as the development of collateral vasculature.

in this work we illustrate mains imaging signs of Moya Moya syndrome;

**Computed tomography (CT)**

CT angiography can show the intracranial stenoses seen in moyamoya.

Thus, CT angiography should be considered when magnetic resonance imaging (MRI) is not readily available and a diagnosis of cerebral occlusive vasculopathy is being considered.

Head computed tomography Workup of a child with moyamoya disease typically begins with a head computed tomography (CT) to assess for more common pathology including tumors and/or hydrocephalus.

Classically, findings on a head CT in pediatric patients with moyamoya include:

* hypodensities suggestive of prior infarctions in watershed areas (especially in the distribution of the middle cerebral artery), basal ganglia, deep white mater, and periventricular regions.

* hemorrhagic pathology, including intracerebral, intraventricular, subarachnoid, and subdural hemorrhages (rare in the pediatric population).

* cerebral atrophy and encephalomalacia may also be detected (Depending upon the severity of prior infarctions,)

However, the CT scan can be normal.
CT angiography is useful in both

*the diagnosis of moyamoya disease showing stenosis or occlusion of the arteries of the circle of Willis, as well as the development of collateral vasculature. Fig. 1 on page 9, Fig. 2 on page 9

*And to evaluate neovascularization after surgical bypass

**MRI Findings.**

The widespread availability of MRI and magnetic resonance angiography has led to the increasing use of these methods for primary imaging in patients with symptoms suggestive of moyamoya.

*An acute infarct is more likely to be detected with the use of diffusion-weighted imaging, whereas a chronic infarct is more likely to be seen with T1- and T2-weighted imaging.

*Intracerebral, intraventricular, subarachnoid, and subdural hemorrhages Fig. 4 on page 13

*Diminished cortical blood flow due to moyamoya can be inferred from fluid-attenuated inversion recovery (FLAIR) sequences showing linear high signals that follow a sulcal pattern.

*Cerebral atrophy Fig. 3 on page 10, Fig. 11 on page 11, Fig. 12 on page 12

*The finding most suggestive of moyamoya on MRI is reduced flow voids in the internal, middle, and anterior cerebral arteries coupled with prominent flow voids through the basal ganglia and thalamus from moyamoya-associated collateral vessels. These findings are virtually diagnostic of moyamoya.

Fluid attenuated inversion recovery MRI which demonstrates linear high signal intensity following a sulcal pattern (ivy sign) has been used to infer cortical ischemia and is felt to represent slow flow in the poorly perfused cortical circulation in children with moyamoya.

*characterization of both the stenosis and basal collateral formation associated with moyamoya disease.

The MR findings most suggestive of moyamoya disease remains diminished flow voids in the ICA, ACA, and MCA bilaterally, with concurrent large flow voids in the basal ganglia and thalamus representing collateral moyamoya vessel formation. Recently, MRI/MRA has been suggested as a reliable alternative to conventional angiography for diagnosis
of moyamoya, based on the ease and fewer procedural associated risks in the pediatric population

MRA is beneficial in diagnosing larger basal ICA, leptomeningeal, and transdural collaterals, whereas smaller collateral remain underestimated.

**Angiography**

Formal angiography should consist of a full five-vessel or six-vessel study that includes both external carotid arteries, both internal carotid arteries, and one or both vertebral arteries, depending on the collateral patterns seen.

The definitive diagnosis is based on a distinct arteriographic appearance characterized by stenosis of the distal intracranial internal carotid artery, extending to the proximal anterior and middle arteries.

Disease severity is frequently classified into one of six progressive stages that were originally defined in 1969.

Development of an extensive collateral network at the base of the brain along with the classic "puff of smoke" appearance on angiography is seen in the intermediate stages of the Suzuki grading system.

Imaging of the external carotid arteries is essential to identify any preexisting collateral vessels so that surgery, if performed, will not disrupt them. Fig. 5 on page 14

Aneurysms, as well as the rare arteriovenous malformation known to be associated with certain cases of moyamoya, are also best detected by means of conventional angiography. Fig. 9 on page 18, Fig. 8 on page 17, Fig. 10 on page 19

Conventional angiography remains the gold standard for both the diagnosis and surgical planning for patients with suspected moyamoya disease.
Fig. 2

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Fig. 1

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Fig. 3: Right hemispheric atrophy No recent hemorrhagic or ischemic injury

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Fig. 11

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Fig. 12: presence of intra parenchymal bleeding stigmata, intra ventricular and subarachnoid.

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Fig. 4: left Thalamic hematoma

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**Fig. 5:** Bilateral carotid occlusion with development of a large arterial network of collateral circulation.

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Fig. 10

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Conclusion

In patients with Moya Moya disease the CT, MRI and Angiographic findings are very specific and they can readily diagnose and evaluate the extent of the disease.

Due to its high resolution and contrast MRI was found to be more informative than CT in detection of abnormal vessels (MMV).

Angiography has been the most accurate method for detection of Moya disease but as it is an invasive procedure,

It has inherent risks. Due to its non-invasive quality, MRI and MR angiography are now preferred methods of investigation for MoyaMoya disease.
References


