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Recognizing The 'Ivy Sign': A Radiologic Hallmark in Pediatric Moya Moya Disease

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Abstract **Case Report**

Moya Moya disease is a rare, progressive cerebrovascular disorder characterized by stenosis or occlusion of the terminal portion of the internal carotid arteries and the development of abnormal vascular networks. The "Ivy sign" is a characteristic Magnetic resonance imaging (MRI) finding frequently encountered in patients with Moya Moya. It is due to the formation of leptomeningeal collateral development and increased numbers of pial vascular webs. We report a case of a 4-year-old female who presented with experienced generalized tonic-clonic seizures and had a history of ischemic stroke in 2024 and was diagnosed to have Moya Moya disease, including the presence of the "Ivy sign".

Keywords: Moya Moya disease, MRI, Ivy Sign, leptomeningeal collaterals.

INTRODUCTION

Moya Moya disease is an idiopathic condition characterized by progressive stenosis of the intracranial internal carotid arteries, leading to the formation of a compensatory, fragile vascular network at the base of the brain [1]. This network resembles a "puff of smoke" on angiographic imaging, from which the disease derives its name. The condition predominantly affects children and young adults, often presenting with ischemic events such as strokes or transient ischemic attacks. Early diagnosis is crucial to prevent disease progression and associated neurological deficits. The "ivy sign," a radiological finding on FLAIR MRI sequences and in post contrast T1-weighted images, appears as diffuse leptomeningeal hyperintensity and enhancement and has been associated with decreased cerebral vascular reserve in Moya moya disease. Recognizing this sign is vital for clinicians, especially when evaluating pediatric patients with neurological symptoms [2].

CASE REPORT

A 4-year-old girl presented with generalized tonic-clonic seizures. Her medical history was significant for an ischemic stroke in 2024. Given her clinical presentation and history, an MRI of the brain was performed.

On T1, T2-weighted (T2W) and Fluid attenuated inversion recovery (FLAIR) (Figures 1 and 2.), there was cortico-subcortical atrophy with gyriform signal abnormalities in the bilateral fronto-parietal cortical ribbon, hypointense on T1, hyperintense on T2/FLAIR, with intense leptomeningeal contrast enhancement on T1 post contrast sequences (Figure 3), forming the "Ivy sign", which is indicative of leptomeningeal collateral circulation.

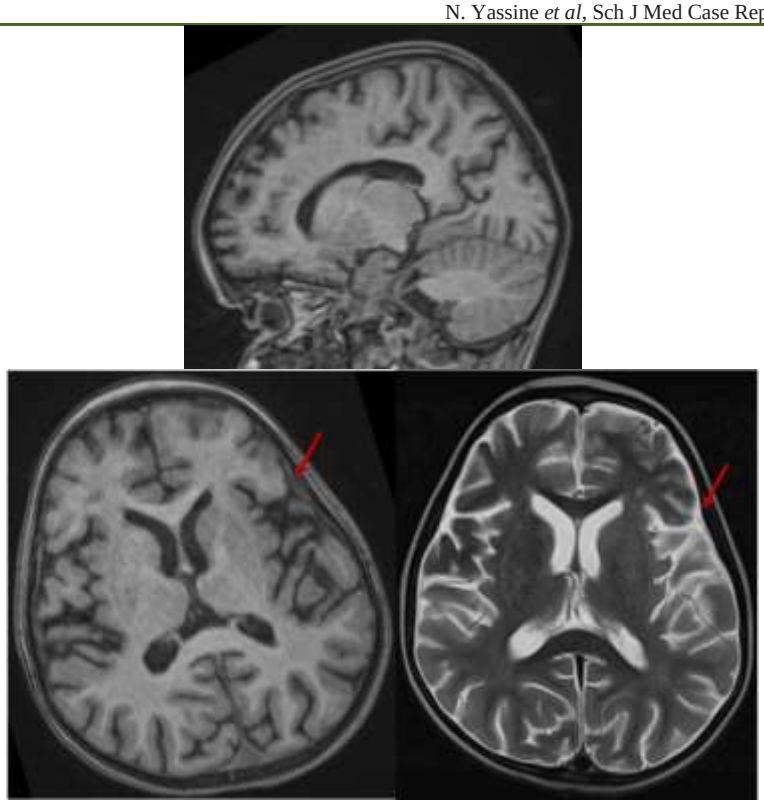

Figure 1: T1 (axial and sagittal), T2 (axial) sequences: Cortico-subcortical atrophy with gyriform signal abnormalities in the bilateral fronto-parietal cortical ribbon (red arrows)

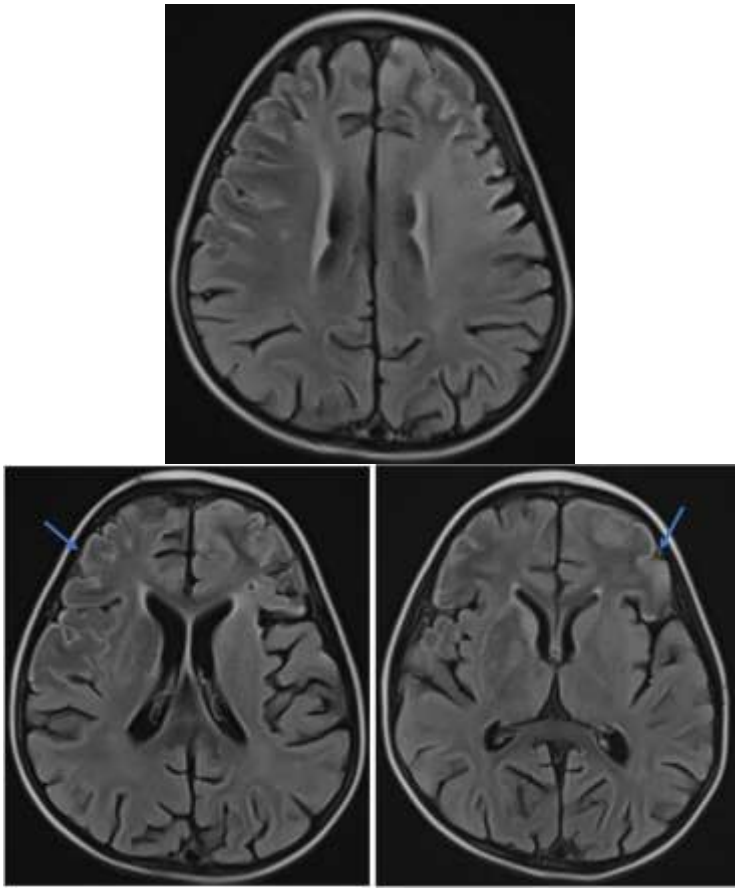

Figure 2: FLAIR axial sequences: Cortico-subcortical atrophy with gyriform signal abnormalities in the bilateral fronto-parietal cortical ribbon (blue arrows)

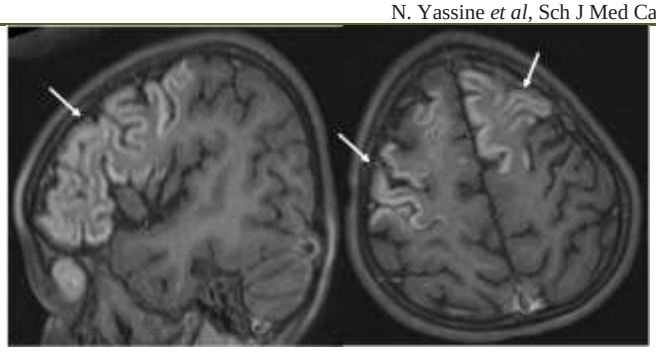

Figure 3: T1 post contrast sequences (sagittal and axial) : Bilateral intense florid curvilinear enhancement in the fronto-parietal cortical ribbon (white arrows)

In addition, signal abnormality was observed in the right pallidum, internal part of the head of the caudate nucleus, genu, and posterior limb of the right internal capsule, extending to the ipsilateral cerebral peduncle and left frontal cortex, appearing isointense on T1, hyperintense on T2/FLAIR, hyperintense on diffusion with ADC restriction, suggesting a recent ischemic event (Figure 4).

On T1 post contrast sequences, intense contrast enhancement of the putamen and the external portion of the head of the right caudate nucleus was also observed, likely due to compensatory hypervascularization. (Figure 4).

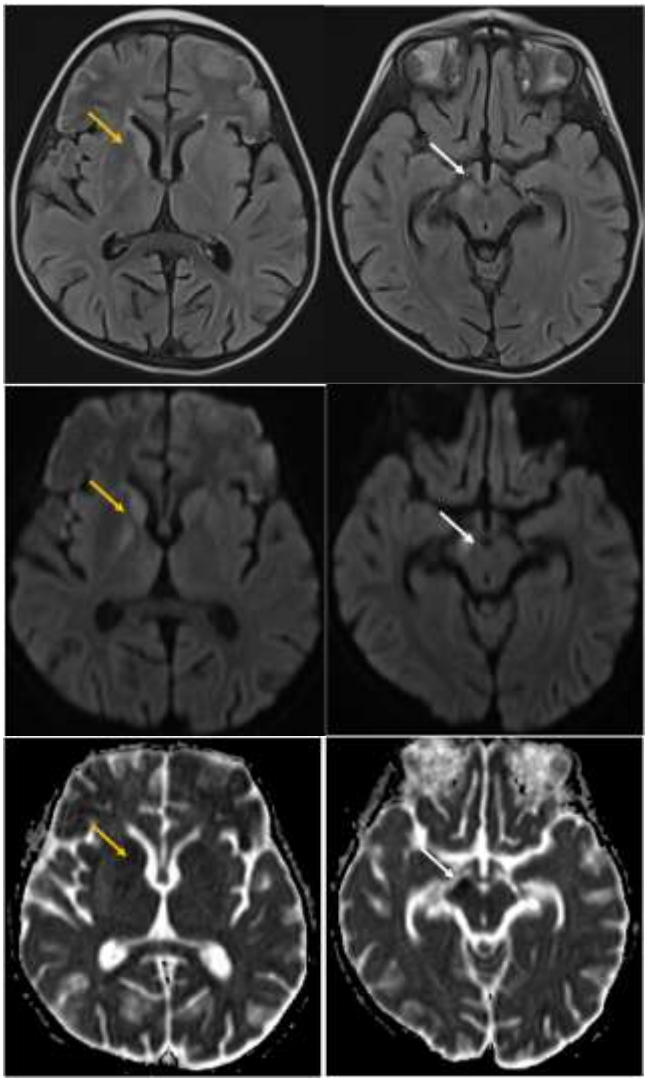

Figure 4.1: FLAIR (axial), diffusion sequences (ADC): recent ischemic stroke of the right pallidum and internal part of the head of the caudate nucleus (yellow arrows), extending to the ipsilateral cerebral peduncle (white arrows)

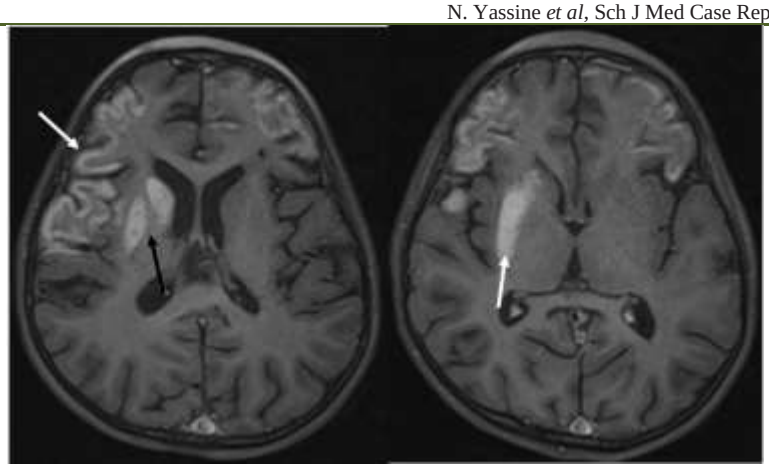

Figure 4: T1 post contrast sequences (axial): Intense contrast enhancement of the putamen and the external portion of the head of the right caudate nucleus (yellows arrows): compensatory hypervascularization

On MR angiography, occlusions were present in both supra-clinoid internal carotid arteries, with flow reduction at their terminations and along the entire course of both middle cerebral arteries (MCAs), as well as the A1 segments of the anterior cerebral arteries (ACAs) and anterior communicating arteries. Lenticulostriate collateral circulation were observed at the basal cisterns, Sylvian valleys, and parafalcine regions bilaterally, appearing as signal voids on T2. (Figure 5 and 6).

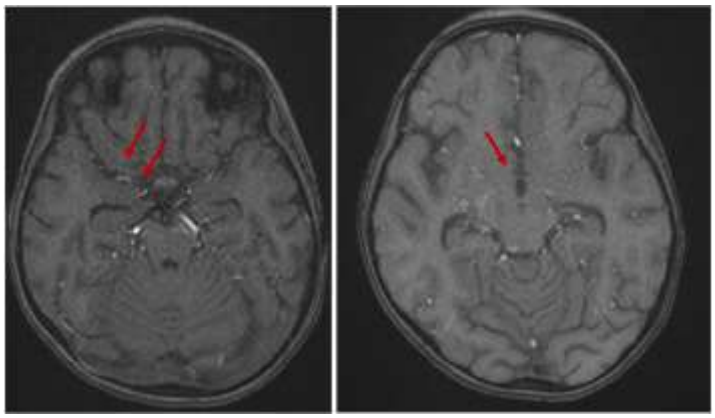

Figure 5: MR angiography: bilateral supra-clinoid internal carotid artery carotid, MCAs and ACAs (A1 segments) stenosis (red arrows).

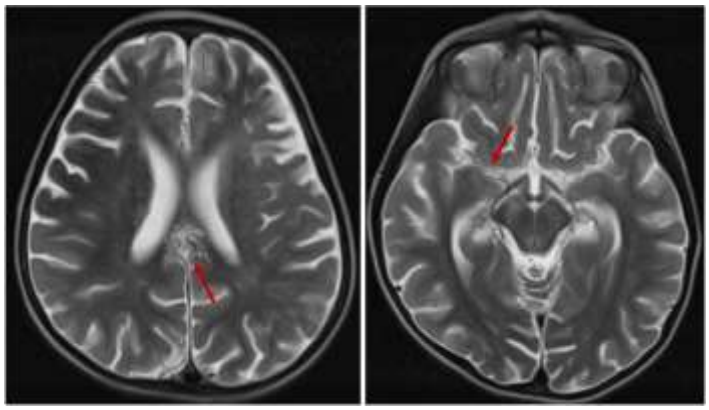

Figure 6: T2 sequences (axial): supra and infra tentorial collateral circulation: "puff of smoke" pattern (red arrows)

We concluded to recent ischemic strokes affecting the deep territories of the right MCA and PCA, secondary to bilateral terminal carotid and MCA stenosis and associated leptomeningeal, lenticulostriate, and basal cisternal collateral circulation, consistent with Moya Moya disease.

Digital subtraction angiography (DSA) was performed and revealed severe, extensive stenosis of the supra-clinoid portions of both internal carotid arteries, extending to the pre-communicating segments of the anterior cerebral arteries (ACAs) and the middle cerebral arteries (MCAs) throughout their entire course.

Prominent arterial collateral network at the lenticulostriate level, with retrograde pial anastomoses between the anterior and posterior circulations, both superficial and deep was also noted, forming a "puff of smoke" pattern, characteristic of Moya Moya disease (Figure 7).

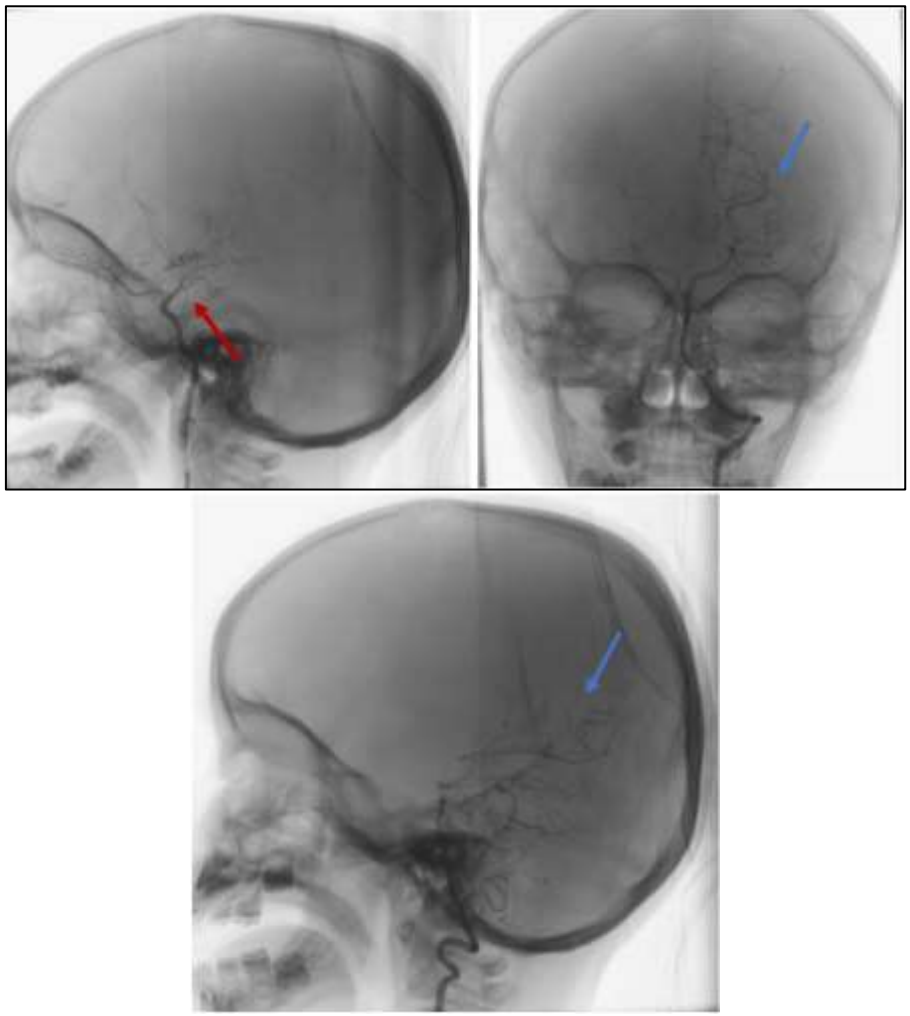

Figure 7: DSA examination: severe, extensive stenosis of the supra-clinoid portions of both internal carotid arteries, extending to the pre-communicating segments of the anterior cerebral arteries (ACAs) and the middle cerebral arteries (MCAs) throughout their entire course (red arrow) and prominent collateral network (blue arrows)

DSA concluded to severe stenosis of the supra-clinoid internal carotid arteries, with marked stenosis of the pre-communicating segments of the ACAs and MCAs throughout their length, associated with arterial collateral circulation. Consequently, all these findings were evaluated as being strongly suggestive of idiopathic Moya Moya disease.

This case underscores the importance of recognizing the “Ivy sign” in pediatric patients, as it serves as a critical imaging marker for Moya Moya disease and can guide timely diagnosis and intervention.

DISCUSSION

Moya Moya disease is a progressive, non-atherosclerotic occlusive arteriopathy affecting the distal internal carotid artery as well as the proximal segments of the middle and anterior cerebral arteries [1]. It is characterized by the development of multiple collateral vessels. The term *Moyamoya*, meaning “hazy” or “puff of smoke” in Japanese, describes the angiographic appearance of the abnormal cerebral vasculature. The disease is bilateral but often asymmetric, as observed in our patient. Although unilateral involvement occurs in approximately 18% of pediatric cases, it typically progresses to bilateral involvement within two years [3].

Moya Moya disease exhibits a bimodal age distribution, with a peak incidence at around 5 years of age and a second, lower peak around 40 years. It is twice as common in females as in males [4]. Our patient, a 4-year-old girl, falls within the younger affected population.

The symptoms of Moya Moya disease vary between children and adults. In children, it most commonly presents as transient ischemic attacks (TIAs) with infarcts predominantly in the internal carotid artery

(ICA) territory. Associated neurological deficits may include monoparesis, hemiparesis, aphasia, and dysarthria. In adults, subarachnoid and/or intraparenchymal hemorrhage is the predominant clinical manifestation [3].

The golden standard method for the diagnosis of Moya Moya disease is cerebral angiography. However, since cerebral angiography is invasive and not easy to perform, particularly in pediatric age groups, the primarily preferred method is non-invasive MRI and MR angiography (MRA) [5].

Thus, (MRA) is the primary imaging modality for diagnosing Moya Moya disease, with a sensitivity of 73% and a specificity of 100%. When combined with MRI brain, sensitivity increases to 92% [5].

- MRA reveals occlusion and narrowing of the distal ICA and proximal cerebral arteries, along with the development of collateral circulation.
- MRI can demonstrate ischemic infarcts in the ICA territory as well as the "Ivy sign", both of which were identified in our patient [5,6,7].
- The "Ivy sign", first described in 1995, was initially observed on post-contrast MR sequences and later on FLAIR images. It results from the formation of leptomeningeal collaterals between the external and internal carotid arteries, which appear in approximately 70% of Moya Moya patients. The name "Ivy sign" comes from its resemblance to ivy creeping on stones. This sign represents diffuse leptomeningeal and cortical enhancement in post-contrast MR images and leptomeningeal and cortical hyperintensity in FLAIR imaging [7,8]. This sign is believed to result from slow retrograde flow in engorged pial collateral vessels formed in response to arterial stenosis, leading to decreased cerebral vascular reserve. Studies have demonstrated a significant correlation between the presence of the ivy sign and reduced cerebral vascular reserve, highlighting its diagnostic value [8].

Several collaterals develop in these patients in order to compensate the stenosis occlusion. The first collateral is known as the basal Moya Moya. It includes abnormal dilatation of the lenticulostriate and thalamo-perforating arteries in the thalamus and basal ganglia (Moya Moya vessels). The second collateral includes dilatation of the anterior coroidal and posterior pericallosal arteries. The third collateral is known as the ethmoidal Moya Moya. It includes the abnormal dilatation of the anterior and posterior ethmoidal arteries that supply blood to the ACA branches from the ophthalmic arteries. The last collateral path is between the dural arteries and the pial arteries and is called “vault Moya Moya” [9].

The gold standard for diagnosing Moya Moya disease and planning surgical intervention is conventional cerebral angiography, particularly for identifying small collateral vessels. However, due to its invasive nature, radiation exposure, and challenges in pediatric patients, MRI and MRA remain the preferred non-invasive imaging methods [10].

The diagnosis of Moya Moya disease can be made when all other causes of cerebrovascular disease have been eliminated. If diseases such as Down Syndrome, NFM type-1, Sickle Cell Anemia, radiation treatment, glycogen storage disease type-1a, hereditary spherocytosis, tuberculosis meningitides and SLE are associated with the Moya Moya vessels, this is called Moya Moya syndrome [9].

Early diagnosis is essential, as timely surgical interventions like revascularization procedures can improve cerebral perfusion and reduce the risk of recurrent ischemic events [11].

CONCLUSION

The diagnosis of Moya Moya disease is now easily achievable with modern neuroimaging techniques (MRI and MRA), especially in children. This case underlines the importance of recognizing the “Ivy sign” in pediatric patients. The presence of this sign should prompt consideration of Moya Moya disease and lead to further diagnostic evaluations. Early identification and intervention are crucial in managing Moya Moya disease, as timely surgical revascularization can significantly improve outcomes and reduce the risk of future ischemic events.

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