

Rare case of childhood cerebral arterial disease

Published on 13.06.2023

DOI: 10.35100/eurorad/case.18172

ISSN: 1563-4086

Section: Neuroradiology

Area of Interest: CNS Head and neck

Imaging Technique: CT-Angiography

Case Type: Clinical Cases

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Patient: 6 years, female

Clinical History:

A 6-year-old female child presented with the complaint of recurrent stroke-like symptoms since her infancy. She had power of 3/5 in bilateral lower limbs 3 years back when she had one of the episodes. It was gradually improving. Her developmental milestones were slower compared to the child of the same age. 6 months back she developed sudden upper limb weakness following loss of consciousness. Now she complains of headaches and is slow to react towards the noxious stimuli. Because of multiplicity of the episodes she was suspected of vasculitis and was sent for cerebral angiographic imaging.

Imaging Findings:

Plain CT head demonstrate no obvious abnormalities. Cerebral CT Angiogram along with head and neck CT Angiography shows moderate to severe stenosis of the right internal carotid artery in the cervical, petrous, laceral and cavernosal segments. The supraclod segment shows near complete occlusion. The terminal branches of the ICA i.e. ACA and MCA are reformed from the normal contralateral side. There are tiny innumerable tortuous cortical and penetrating vessels, which are seen supplying the cerebral cortex and the deep white mater as well as the grey mater nucleus. Bilateral MCA shows loss of smooth luminal contour. On sagittal and coronal MIP images the reformed collaterals due to compensatory mechanisms shows puff of smoke appearance, which are seen arising from the deep arterial territory to the cortical superficial aspect. These vessels are seen fanning out from the centre to the periphery.

Discussion:

MoyaMoya disease is a chronic childhood progressive cerebrovascular disease of unknown origin. If there is know cause i.e. Neurofibromatosis, Down's syndrome, thyroid disorder and vasoocclusive disease like sickle cell anemia it is termed Moya Moya syndrome. There is predilection for the ICA, which is stenosed causing recurrent stroke-like symptoms. The literal meaning of "Moya Moya" is puff of smoke. The name is suggested due to its characteristic appearance in cerebral angiographic images. The child may present with recurrent stroke-like symptoms, headache, recurrent episodes of seizure, cognitive decline and altered sensorium[1,2,5]. There is bimodal occurrence of the disease i.e. among the 5-6 years and 30-40 years of age. The condition is more prevalent in the Asian population especially in Japan and China with incidence up to 0.35 in 100000 in Japan [2,3,4].

Various imaging modalities can be used for imaging of the disease namely conventional catheter angiography, CT angiography and MR angiography. Conventional catheter angiography is considered the gold standard. However, recently CT and MR angiography are considered the first line of imaging due to less invasive nature and also due to betterment in their spatial resolution and their ability to reformat images in multiple planes as desired. Brain parenchyma changes are also well visualized with the help of cross-sectional imaging, which is a limitation for the conventional angiography. Imaging findings in case of the disease are symmetrical stenosis of the bilateral ICA and their subsequent branches without evidence of atheromatous changes. There may be absent formation of the major

The hallmark of the disease are formation of multiple variable-sized tortuous collaterals from the pial circulation or from the penetrating vessels. These small collaterals are the cause for characteristic “puff of smoke” appearance. MR angiography may show serpentine pial vessel which is otherwise known as “Ivy sign”. Angiographic grading of the disease is known as Suzuki grading, which consist of six grades and ranges from narrowing of the carotid fork in the least severe spectrum to the disappearance of the ICA and establishment of the collateral supply from the ECA or vertebral arteries[3,4,5].

Differential Diagnosis List: Moyamoya disease

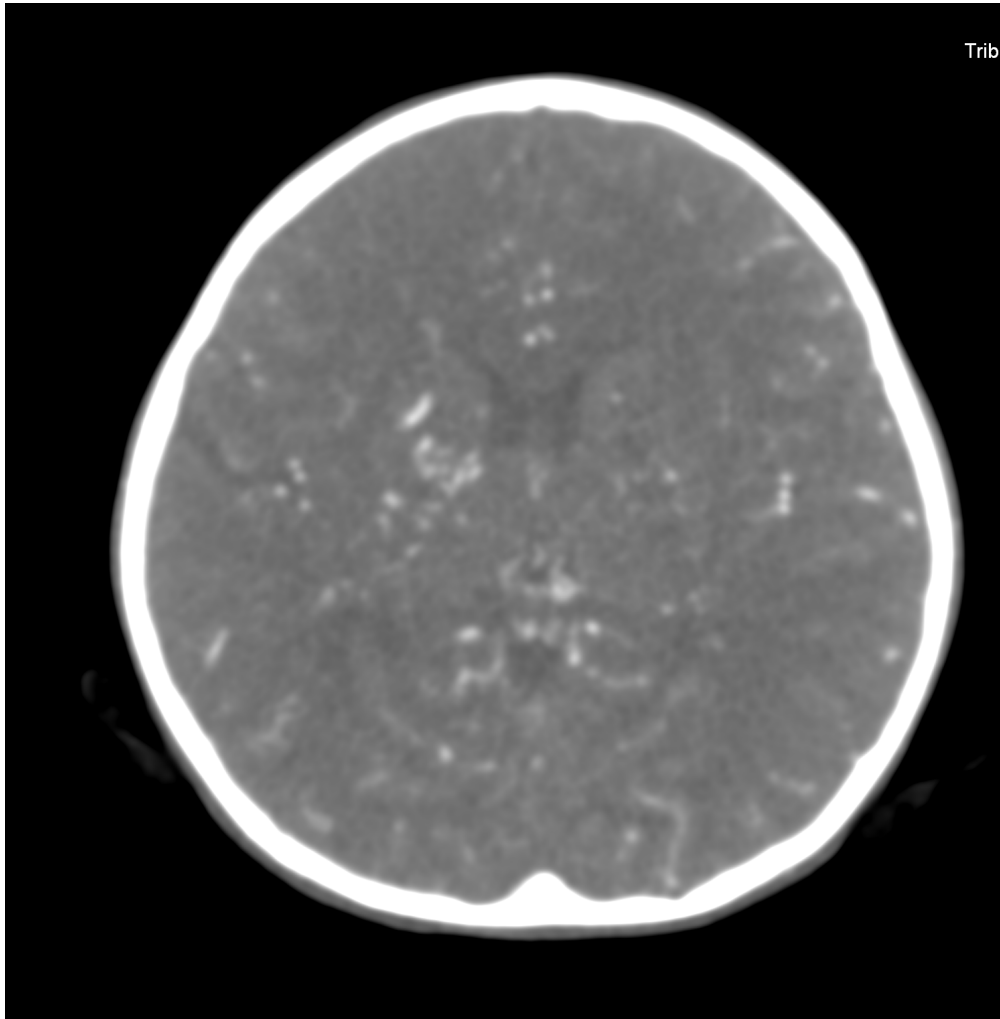
Final Diagnosis: Moyamoya disease

References:

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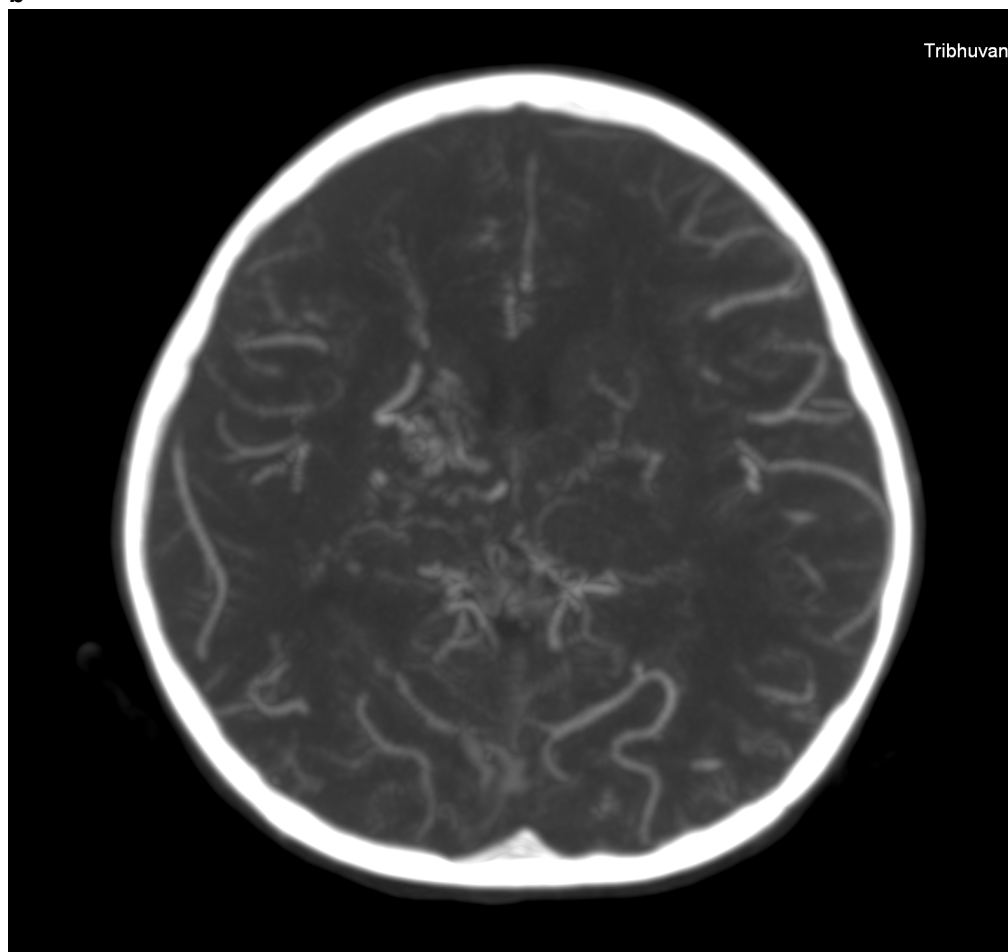
Figure 1

a



Description: Axial CT-Angiographic images demonstrates the prominent perforating vessels and cortical vessels **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

b



Description: Axial MIP images depicting the tiny tortuous perforating vessels in the deep gray matter nuclei region and also the cortical branches; features of compensatory changes to chronic occlusive pathology **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

Figure 2

a



Description: Axial MIP images shows the prominent perforating vessels coursing through the deep cerebral cortex towards the periphery in a characteristics puff of smoke appearance **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

b



Description: More lateral Saggital MIP images demonstrating the prominent perforating branches and puff of smoke appearance **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

c



Description: Coronal MIP images demonstrates the irregularities in the bilateral MCA, A1 segment of ACA and prominent vessels in the cerebral cortex. Notice the attenuation and caliber difference between the right and left ICA. Right ICA is attenuated in caliber and diffuse luminal narrowing
Origin: Department of Radiology TUTH, Maharajgunj, Nepal

Figure 3

a



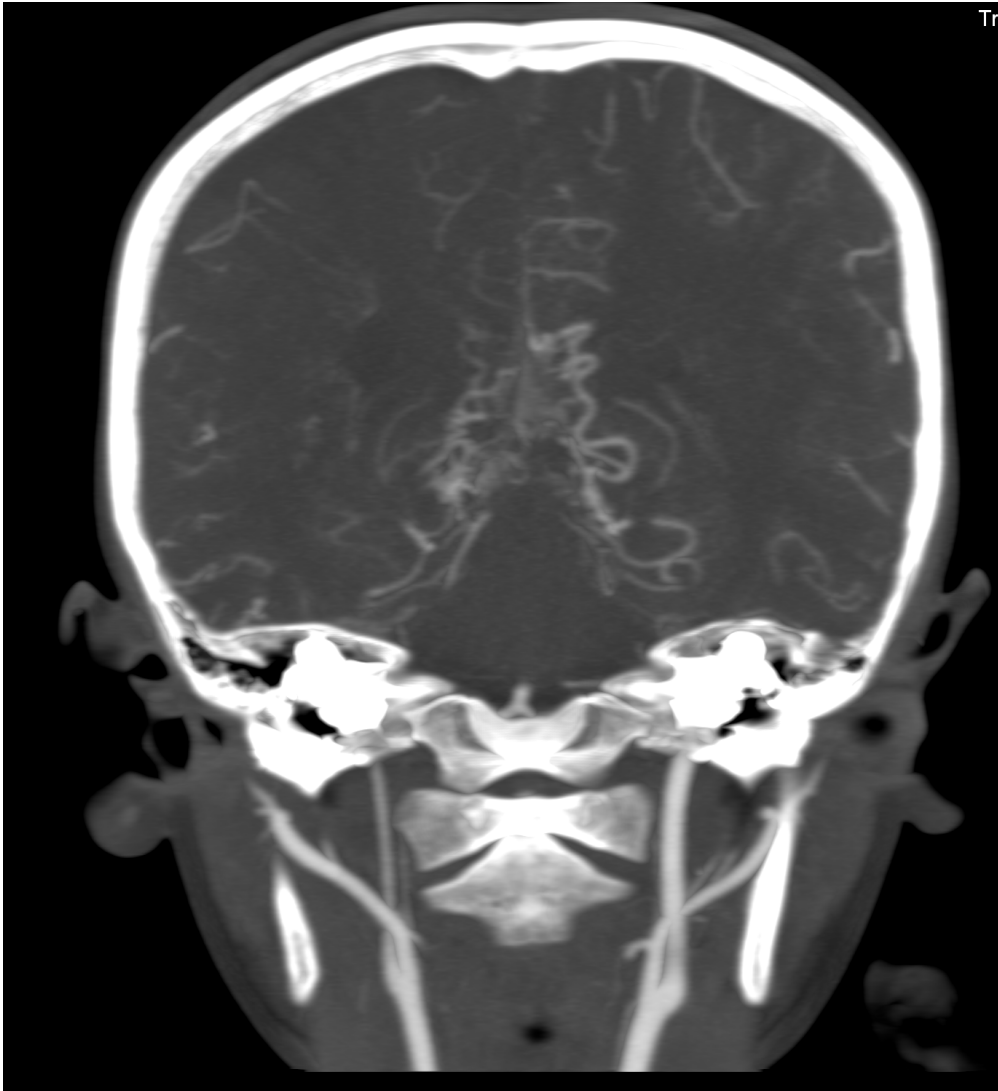
Description: Saggital reformat image showing the diffuse narrowing of the cervical and petrosal segment of the right ICA **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

b



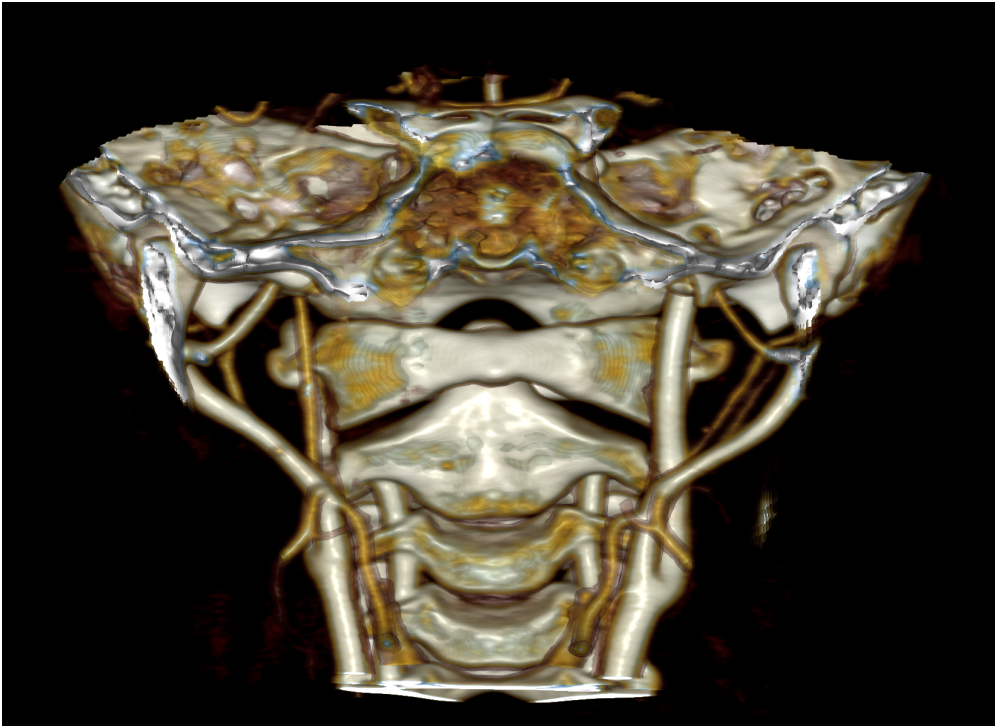
Description: Sagittal reformat image of the left ICA which shows normal caliber of the left ICA **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

c



Description: Coronal MIP images show the diffuse narrowing and low attenuation of the right ICA and normal caliber and left ICA **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal

d



Description: Coronal 3D image shows shows the narrow caliber of the right cervical ICA **Origin:** Department of Radiology TUTH, Maharajgunj, Nepal