Moyamoya disease

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Citation

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Abstract

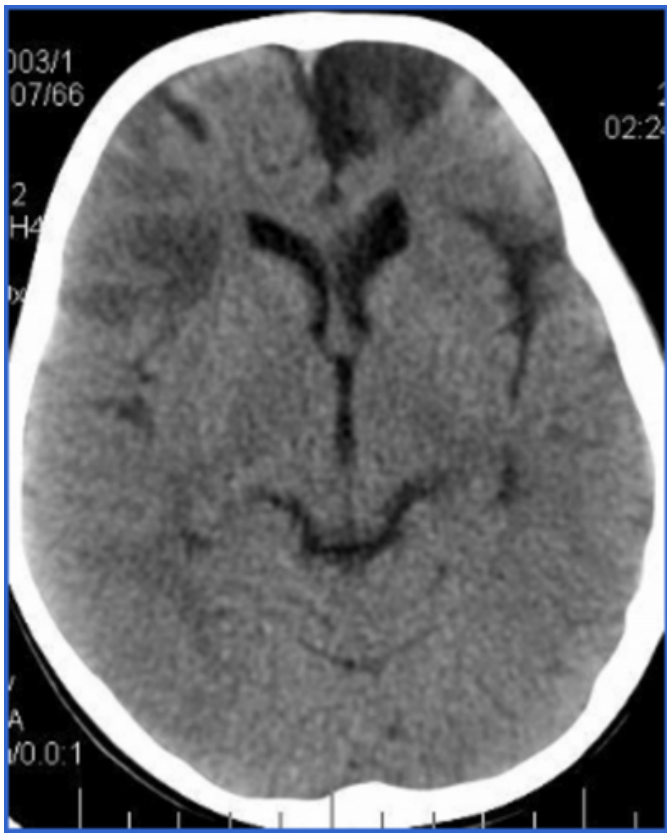
We present here CT and MR images of a childhood case of Moyamoya disease with its classic MR angiographic picture and also the 'ivy' sign. Although the catheter angiographic 'puff of smoke' appearance has been exctensively documented in the literature, the corresponding MR angiographic appearance has been sort of often neglected. We here document a case of Moyamoya disease with extensive collaterals demonstrated at MR angiography.

CASE REPORT

A 6year old male child born of non-consanguinous marriage who presented with complaints of acute onset left sided weakness. Bleeding time,clotting time ,prothrombin time , protein C & S were normal. Sickling test was negative CT scan brain was performed which showed infarcts in right fronto-temporal & left anterior frontal regions.(Fig 1).

Figure 1

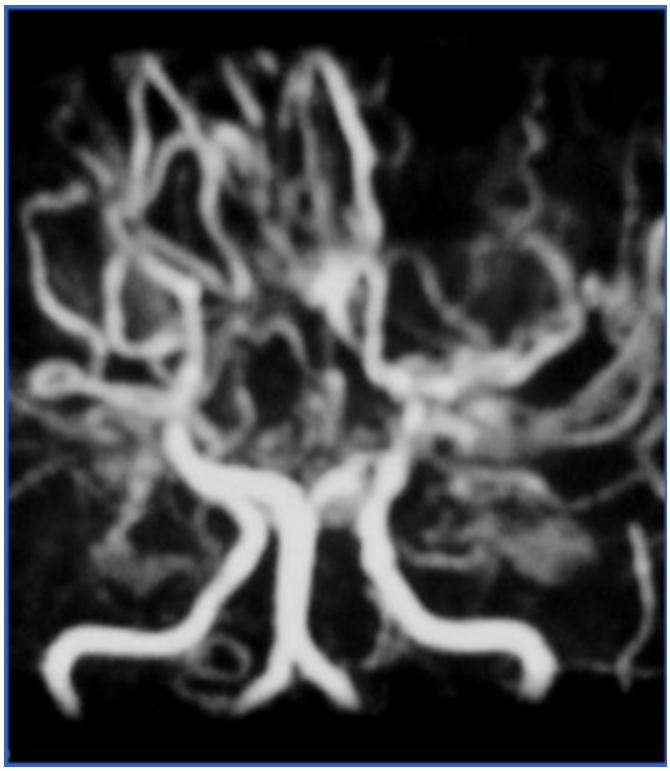
Figure 1. Axial plain CT of the brain showing infarcts in right fronto-temporal & left anterior frontal regions.



MRI was done for further evaluation which showed subacute, non-hemorrhagic infarcts in the areas described above. MR angiography showed multiple collateral vessels in the base of the brain & in the most parts of anterior circulation along with non-visualisation of the distal internal carotid arteries & proximal vessels of circle of Willis(figure2).

Figure 2

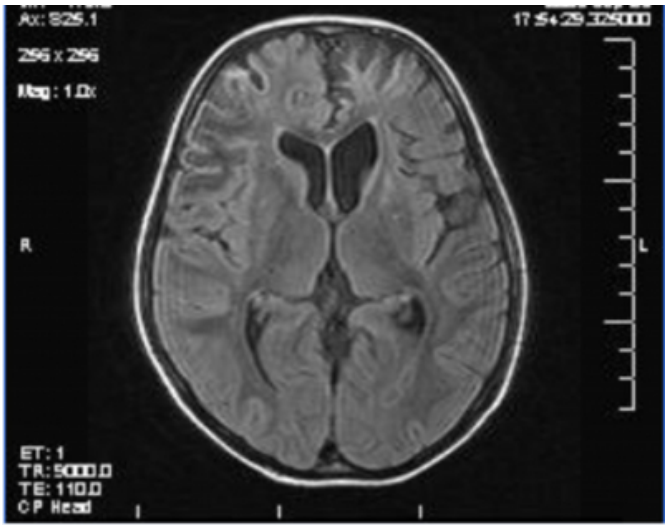
Figure 2. 2D –Time of Flight MR angiogram. Non-visualisation of bilateral supraclinoid internal carotid arteries & the proximal portions of both anterior & middle cerebral arteries. Extensive collateral formation is seen in the base of



Posterior circulation appeared normal. Also the leptomenigeal “ivy sign” was demonstrated on FLAIR(Fig 3)

Figure 3

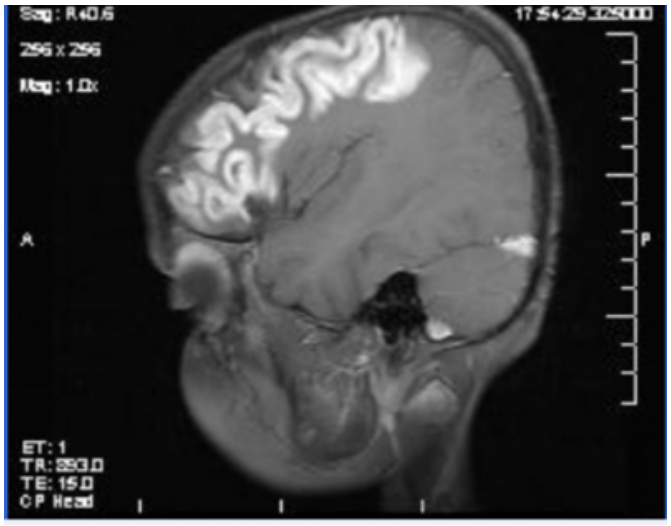
Figure 3. Ivy sign. Transverse unenhanced FLAIR MR image shows subtle high signal intensities along leptomeninges in bilateral frontal regions.



& contrast enhanced “ivy sign” on post contrast studies(fig 4).

Figure 4

Figure 4. Post contrast ivy sign.Far right lateral saggital gadolinium-enhanced T1 MR image shows florid curvilinear enhancement in frontal region .



A diagnosis of moyamoya disease was made.

DISCUSSION

Moyamoya disease was first described by Suzuki in 1963. It is a idiopathic progressive occlusive cerebrovascular disorder of unknown etiology characterized by progressive narrowing or frank occlusion of distal internal carotid arteries (ICA) & proximal circle of Willis vessels with secondary collateralization (1). In addition to the enlarged lenticulostriate and thalamoperforator arteries, multiple leptomeningeal and transdural anastomoses develop between the cortical arteries and those of dura mater, scalp, and orbit (2). It is more common in females & shows bimodal age peaks: 6 years and beyond 35 years (1).

Similar phenomenon can be caused by occlusion of bilateral supraclinoid ICA by atherosclerosis, sickle cell disease , dissection and other etiologies and in that case the phenomenon of extensive collateral formation at the base of the brain is termed Moyamoya phenomenon.

Children with the idiopathic form or Moyamoya disease typically present with cerebral ischemia versus hemorrhage in adults. Most common symptoms include transient ischemic attacks, alternating hemiplegia, headache or just developmental delay. CT scans show multiple infarcts in over 80% with abnormal enhancement and cerebral atrophy. The anterior circulation is most frequently affected. The collateral vessels are seen as multiple punctate dots on contrast CT and as the flow voids in the region of basal gangliaon MRI. On angiography (conventional or MR) the characteristic ‘puff of smoke’ (moya moya in Japanese languauage) is seen which represent the lenticulostriate and thalamostriate collaterals. Sometimes slow flowing engorged pial vessels and thickened arachnoid membranes give rise to appearance of bright sulci on FLAIR also called the leptomeningeal ‘ivy sign’. Also leptomeningeal enhancement is seen after giving intravenous contrast, also called the contrast enhanced ‘ivy sign’(3).

Prognosis depends on the age and stage at diagnosi, the rapidity and degree of collaterals developed. After a progressive course for many years, the disease frequently stabilizes with reszidual disability.

Many operations have been developed for the condition , but currently the most favoured are the indirect procedures EDAS (encephaloduroarteriosynangiosis), EMS (encephalomyosynangiosis), and the direct procedure STA-MCA bypass. Direct superficial temporal artery (STA) to middle cerebral artery (MCA) bypass is considered the treatment of choice, although its efficacy, particularly for hemorrhagic disease, remains uncertain (4).

References

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