

Characteristic MRI signs of Moyamoya disease in a child

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Abstract

Moyamoya disease is a cerebrovascular occlusive disease which may present with headache, stroke and recurrent transient ischemic attacks. It has a bimodal age distribution, in the 1st and 3rd-4th decade of life. It is more common in women. The incidence of moyamoya disease is higher in Asian countries as compared to North America and Europe [1,4,5].

Keywords

cerebrovascular; occlusive; stroke; Asian.

Introduction

Presenting symptoms of moyamoya can include hemiplegia, sensation disturbances, aphasia, headache, vertigo, seizures and involuntary movements [2]. Associated risk factors include neurofibromatosis, down syndrome, thyroid disease, HCV, cryoglobulinemia, sickle cell anemia and cranial irradiation. The disease is of two main types- ischemic and hemorrhagic. The former is more common in children, while the latter, in adults [2]. Progressive stenosis at the distal portions of internal carotid arteries leads to typical collateral artery formation, which can be demonstrated on imaging. Digital subtraction angiography (DSA) is considered as the golden standard.

Case Presentation

This presentation is regarding a sixteen years old female with progressive left-sided weakness. Her past medical history was unremarkable and she had not undergone any previous surgery or radiotherapy. On neurological examination, tone was decreased over left side, power was 3/5 over left upper and lower limb, deep tendon reflexes were exaggerated, and planter was extensor over left side. Blood and CSF examinations were normal. MRI of the brain without contrast along with TOF (Time of flight) MRA (Magnetic Resonance Angiography) was performed.

Prominent deep medullary veins on susceptibility-weighted images (SWI) known as 'brush sign' (Figure 1, arrows) can predict the severity of the disease. Basal collateral vessels (Figure 2, arrows) deve-

lop due to steno-occlusion of the intracranial portion of the internal carotid and proximal middle cerebral arteries. This is characterized by 'puff of smoke' sign on MR angiography (Figure 3, arrow).

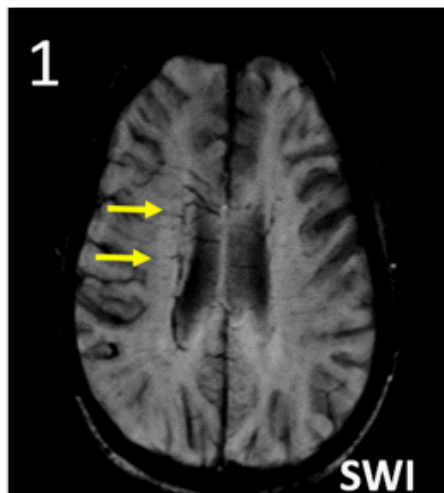


Figure 1: Prominent deep medullary veins on axial susceptibility-weighted images (SWI) are seen in the region of right corona radiata.

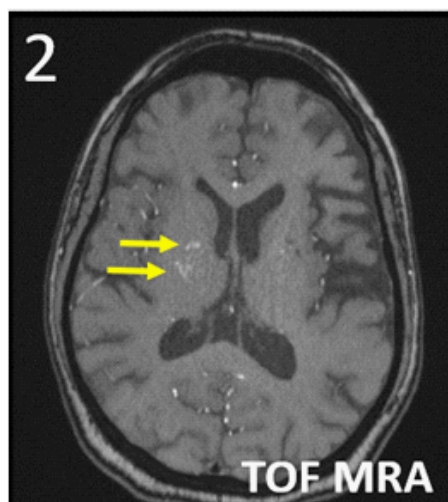


Figure 2: Multiple small, collateral vessels are seen in the basal ganglia on axial MR time of flight angiography image.

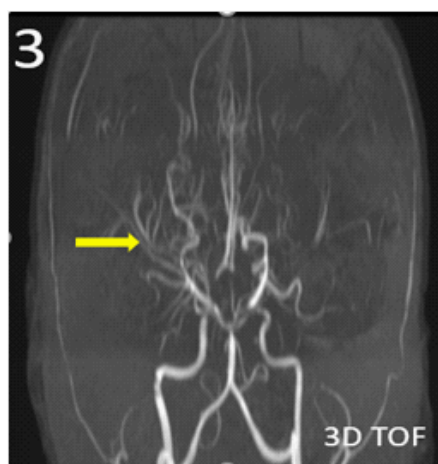


Figure 3: Multiple collateral vessels are seen distal to the ICA occlusion, giving rise to a 'puff of smoke' appearance on angiography.

Discussion

Various characteristic signs of moyamoya have been described in literature. Ivy sign refers to the appearance of the brain on postcontrast T1-weighted or FLAIR images where prominent leptomeningeal collaterals with slow blood flow and post contrast enhancement give an ivy type appearance [5]. This sign can be used to indicate the hemodynamic status of the leptomeningeal collateral pathways, which is more prominent in hemispheres with poorer visualization of the cortical branches of the middle cerebral artery on Magnetic Resonance Imaging [2]. Advanced MR imaging has enabled evaluation of small venous structures and iron in the brain. In particular, SWI has been used to evaluate deep venous flow in acute or chronic ischemia and to demonstrate increased oxygen extraction in cerebral ischemia [3]. This is a useful indicator as it does not require contrast. Hypertrophy of perforating arteries, which produces the puff of smoke sign, is seen on angiography [4].

Digital subtraction angiography can be used for angiographic grading of severity as well as the assessment of the postoperative changes. Other imaging modalities, including SPECT, PET, xenon-CT, and dynamic perfusion CT have been applied to predict the patients with severe hemodynamic impairments. Advanced, new MR imaging-based methods such as DSC-weighted bolus tracking MR imaging and ASL have been used recently for quantitative hemodynamic analysis [3].

Conclusion

Moyamoya disease should be taken into consideration as a possible cause of ischaemic strokes in the young population and this case illustrates the same. The characteristic signs on different MR sequences help in diagnosis, as shown in the above presentation.

References

1. Tarasow E, Kułakowska A, Łukasiewicz A, Kapica-Topczewska K, Korneluk-Sadzyńska A, Brzozowska J, Drozdowski W. Moyamoya disease: diagnostic imaging. *Polish journal of radiology*. 2011; 76(1): 73.
2. Li J, Jin M, Sun X, Li J, Liu Y, Xi Y, et al. Imaging of moyamoya disease and moyamoya syndrome: current status. *Journal of computer assisted tomography*. 2019; 43(2): 257.
3. Horie N, Morikawa M, Nozaki A, Hayashi K, Suyama K, Nagata I. "Brush sign" on susceptibility-weighted MR imaging indicates the severity of moyamoya disease. *American journal of neuroradiology*. 2011; 32(9): 1697-1702.
4. Ortiz-Neira CL. The puff of smoke sign. *Radiology*. 2008; 247(3): 910-911.
5. Savolainen M, Pekkola J, Mustanoja S, Tyni T, Hernesniemi J, Kivipelto L, Tatlisumak T. Moyamoya angiopathy: radiological follow-up findings in Finnish patients. *Journal of neurology*. 2020; 267(8): 2301-2306.

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