Imaging abnormalities on MRI SWI in a 9 year old girl with Hemiplegic Migraine

L.McCarron[1], A.Griffith[1], E.Phelan[2], D.Webb[1]

1.Department of Paediatric Neurology, Our Lady's Children's Hospital, Dublin, Ireland

2.Department of Radiology, Our Lady's Children's Hospital, Dublin, Ireland

Introduction

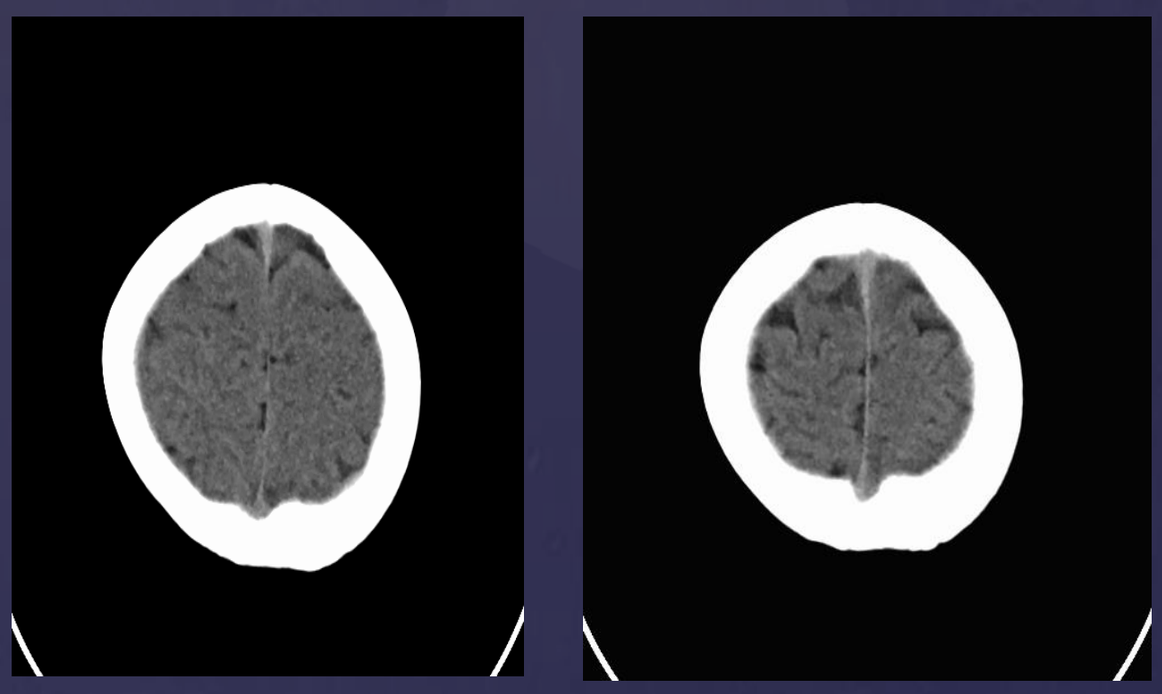
Hemiplegic Migraine is a rare form of migraine typically associated with an aura of hemi-sensory and or motor symptoms. Familial hemiplegic migraine (FHM) is inherited in an autosomal manner while sporadic forms also occur and the severity of symptoms varies considerably.

Case History

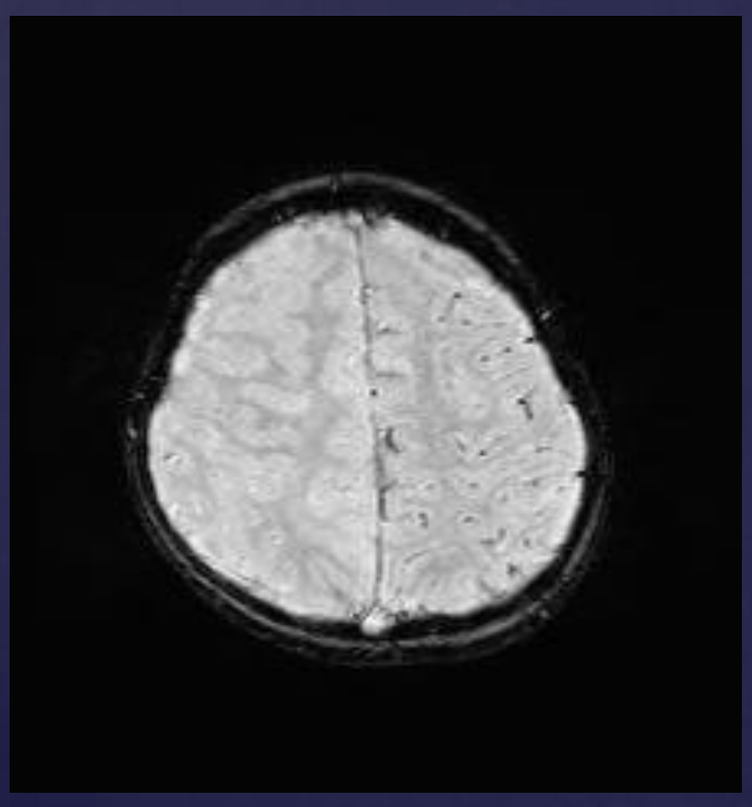
A previously healthy 9 year old girl with normal neurodevelopment presented following a sudden onset of right sided weakness. She had been well the previous day and woke with right sided arm and leg weakness and an expressive dysphasia. There was a strong family history of migraine with aura. She was pallid with normal vital signs including blood pressure. She was orientated, but experienced difficulty completing sentences. She had a right sided hemi-paresis with anti gravity limb movements (grades 3-4/5). Her cranial nerve examination was normal including fundoscopy. Headache was not reported at presentation but developed 5 hours later. Her symptoms resolved over 4 hours.

Imaging

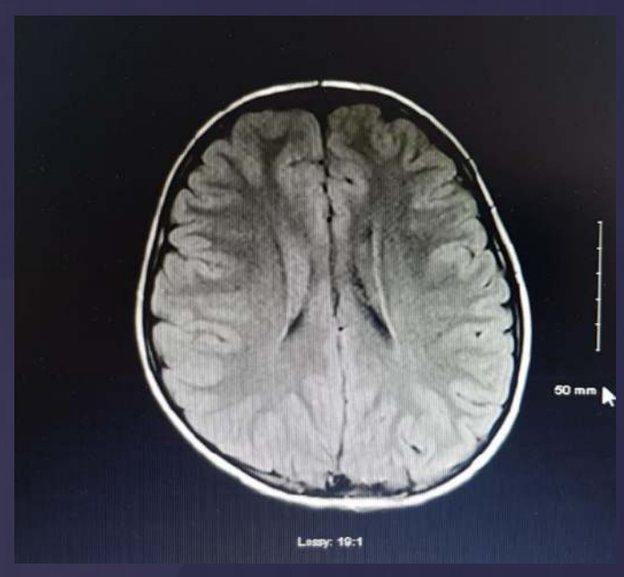
A CT brain 1.5 hours after onset of symptoms demonstrated effacement over the superior surface of the left hemisphere.



A SWI MRI brain study 5 hours after onset of symptoms demonstrated dilated venous vessels throughout the left hemisphere. See image below.



Repeat MRI four months later demonstrated complete resolution prior findings. See image on the right.



Discussion

This case presents radiological evidence specifically SWI SWI abnormalities during the Aura phase of hemiplegic Migraine in the paediatric population. SWI imaging is used in paediatric and neonatal neuroimaging for the evaluation of hemorrhage, ischemic stroke, vascular malformations, traumatic brain injury, brain tumors, congenital infections and neurodegenerative disorders.

The migraine aura is generally indicative of a reversible cerebral cortical dysfunction that is most probably caused by cortical spreading depression - defined by neuronal excitability associated with transient oligaemia and then followed by hyperaemia in the cortex contralateral to the hemiplegia and resulting in prolonged inhibition of neuronal activity.

Mutations in CACNA1A, ATP1A2 and SCN1A have demonstrated an increase in neuronal excitability and reduction in the threshold for cortical spreading depression. Glutamate release is known to activate neuronal N-methyl-d-aspartate (NMDA) and a-amino-3- hydroxy-5-methylisoxazole-4-propionic acid (AMPA) receptors leading to neuronal hyperactivity and hypermetabolism. During this phase, neuronal tissue requires higher oxygen consumption resulting in higher oxygen extraction fraction (OEF) and prominent, hypointense veins on SWI as evident on our patients MRI.

Molecular and cellular mechanisms are also thought to contribute via vasogenic leak and oedema. This can be seen on evaluating ADC values on MRI. Functional MRI studies have been employed more recently in order to establish the role of perfusion in migraine. In one case cerebral hypoperfusion during motor weakness and headaches was shown to be associated with decreased CBF and increased MTT. The area of hypoperfusion correlated with the area of prominent hypointense sulcal veins.These changes were not evident on follow up imaging 24 hours later.

A dual threshold for cerebral hypoperfusion was proposed many years ago with perfusion below the first threshold resulting in a functional failure of neurons and an associated neurological deficit. The neurons however remain viable in this instance. In hypoperfusion below the second threshold, irreversible ischaemic injury ensues. In the latter, diffusion changes will be evident and thus useful in differentiating between acute ischaemia and hemiplegic migraine[4]

Current literature suggests that neuronal hyper excitability is the predominant inciting mechanism with vascular changes as a secondary cause. There are limited MRI studies in children during a hemiplegic migraine aura. Our case identifies SWI abnormalities in the aura phase of hemiplegic migraine.

References

1. Goadsby PJ. Migraine pathophysiology. *Headache*. 2005 Apr. 45 Suppl 1:S14-24.
2. Cutrer FM, O'Donnell A, Sanchez del Rio M. Functional neuroimaging: enhanced understanding of migraine pathophysiology. *Neurology*. 2000. 55(9 Suppl 2):S36-45.
3. Eur J Pediatr 2016 Feb;175(2):295-8. doi: 10.1007/s00431-015-2609-2. Epub 2015 Aug 7.
Pediatric hemiplegic migraine: Role of multiple MRI techniques in evaluation of reversible hypoperfusion, Thangamadhan Bosemani1, Vera J Burton2,3, Ryan J Felling2,Richard Leigh4, Christopher Oakley2, Andrea Poretti1 and Thierry AGM Huisman, Cephalgia:2013
4. Russell MB and Ducros A. Sporadic and familial hemiplegic migraine: pathophysiological mechanisms, clinical characteristics, diagnosis, and management. Lancet Neurol 2011; 10: 457–470.