Cerebral cavernous malformation diagnosis

Cerebral Cavernous malformation (CCM) diagnosis occurs more frequently than some years ago, due to the increased diffusion of magnetic resonance imaging. Progress in knowledge on genetical and molecular pathogenesis may change the management strategy of these patients allowing more tailored approaches [1)].

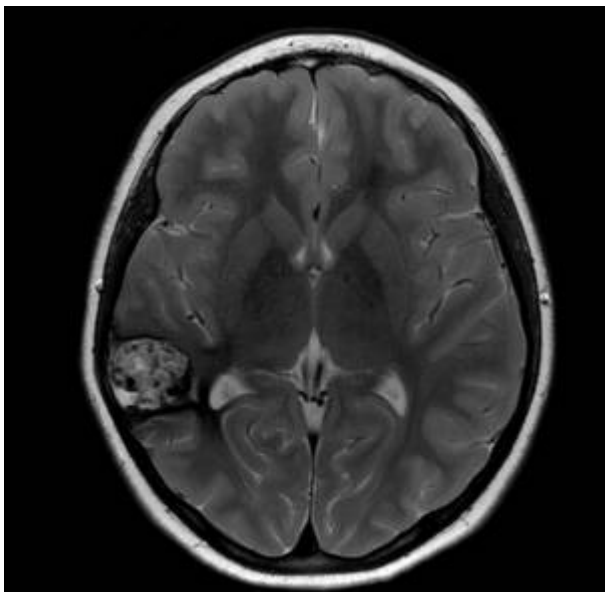
Macroscopic calcifications of cerebral cavernomas were found only in 18 cases (11%). Cerebral angiography was done in 31 cases (18.9%). In 9 cases angiography was totally normal, and in 11 cases the cavernoma presented only as an avascular mass. In the remaining cases there was no conformity in the angiographic appearance of cerebral cavernous haemangiomas. Operative extirpation is the treatment of choice if a solitary lesion is favourably located. In addition to our patient there are now 21 cases (12.8%) in which cavernomas were treated successfully by operative extirpation [2)].

MRI

With the advent of MRI in the 1980s, Cavernous malformations were characterized as pathological entities with pathognomonic findings including a “popcorn” appearance, hemosiderin rings, and mixed signal intensities [3)].

Cavernous malformations can be grouped into four types based on MRI appearances using the Zabramski classification.

MRI is the modality of choice, demonstrating a characteristic “popcorn” or “berry” appearance with a rim of signal loss due to hemosiderin, which demonstrates prominent blooming on susceptibility weighted sequences.



T1 and T2 signal is varied internally depending on the age of the blood produces and small fluid-fluid levels may be evident.

Gradient echo or T2* sequences are able to delineate these lesions better than T1 or T2 weighted images. In patients with familial or multiple cavernous angiomas GRE T2* sequences are very important in identifying the number of lesions missed by conventional Spin echo sequences.

The SWI sequence, being more sensitive to substances which distort the local magnetic field than the GRE T2*W sequence, showed a higher sensitivity in identifying cerebral cavernous malformations. Thus, routine clinical neuroimaging protocol should contain SWI sequences to evaluate patients with (or suspected) cerebral cavernous malformations [4].

Susceptibility weighted imaging (SWI) may have sensitivity equal to that of GRE in detecting these capillary telangiectasias in the brain. SWI is also highly sensitive in detecting calcification as compared to T1 and T2 images.

If a recent bleed has occurred then surrounding oedema may be present.

The lesions generally do not enhance, although enhancement is possible.

Quantitative Susceptibility Mapping (QSM) MRI allows accurate assessment of iron content in cerebral cavernous malformations (CCM), and a threshold increase by 6% in QSM has been shown to reflect new symptomatic hemorrhage (SH) in previously stable lesions.

It is unclear how lesional QSM evolves in CCMs after recent SH, and whether this could serve as a monitoring biomarker in clinical trials aimed at preventing rebleeding in these lesions.

In 16 CCM patients who experienced a SH within the past year, whose lesion was not resected or irradiated.

The data acquisition was performed using QSM sequence implemented on a 3T MRI system ASSESSMENT: The lesional QSM assessments at baseline and yearly during 22 patient-years of follow-up were performed by a trained research staff including imaging scientists.

Biomarker changes were assessed in relation to clinical events. Clinical trial modeling was performed using two-tailed tests of time-averaged difference (assuming within-patient correlation of 0.8, power = 0.9 and alpha = 0.1) to detect 20%, 30% or 50% effects of intervention on clinical and biomarkers event rates during two years of follow-up.

The change in mean lesional QSM of index hemorrhagic lesions was +7.93% per patient-year in the whole cohort. There were 5 cases (31%) of recurrent SH or lesional growth, and twice as many instances (62%) with a threshold (6%) increase in QSM. There were no instances of SH hemorrhage or lesional growth without an associated threshold increase in QSM during the same epoch [5].

1)
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3)
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