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Imaging Appearances of Spinal Cavernous Malformations

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PURPOSE

Cavernous malformations (CMs) of the spinal cord are rare in the general population but have a higher incidence in autosomal dominant CNS cavernous malformation populations. Acute hemorrhage, pediatric cases, and CMs in nerve roots can be confusing. The literature is inconsistent regarding genetic cases and MRI. We present the range of imaging findings and pitfalls, based on a large genetic population.

EDUCATIONAL OBJECTIVES

- ♦ Learn the variety of appearances of spinal cord CMs.
- ♦ Understand the special challenges of diagnosing these, especially in the setting of acute hemorrhage.
- ♦ Learn optimal techniques for imaging spinal CMs.

INTRODUCTION: WHAT ARE THEY?

CNS cavernous malformations (CMs) are vascular malformations comprised of sinusoidal, endothelial-lined spaces without vascular wall elements or intervening parenchyma. (Fig. 1) They are well known in the brain, but they can also occur in the spinal cord and even nerve roots. Most CMs worldwide are sporadic, with about half of sporadic CMs in the brain associated with developmental venous anomalies, but about 20% of brain CMs are the result of autosomal dominant mutations in one of 3 genes (CCM1, CCM2, and CCM3). In familial CMs, large numbers, even hundreds, may occur in the brain. In contrast to AVMs, CMs are low flow lesions that have a tendency to repeated hemorrhage and can grow. Spinal cord CMs are seen much less commonly than brain CMs, and the MRI appearance may be much less familiar to radiologists.

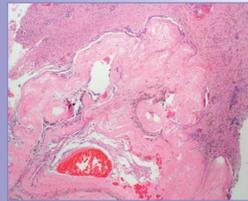


FIGURE 1 Photomicrograph of a brainstem cavernous malformation, showing back to back dilated vessels with a rim of gliotic brain containing numerous hemosiderin-laden macrophages (4x mag).

METHODS:

We reviewed MRI findings from 32 patients with cavernous malformations of the spinal cord and present representative cases of the range of findings. Our institution serves an unusually large population with autosomal dominant CCM1 with the Common Hispanic Mutation. We present selected cases from this population.

TYPICAL APPEARANCE

Medium-size lesions have internal mixed-signal intensity and peripheral hemosiderin deposition. Unlike brain CMs, there is a typical pattern of hemosiderin in a longitudinal distribution in the spinal cord. Most are peripheral in location in the spinal cord. (Figs. 2, 3)

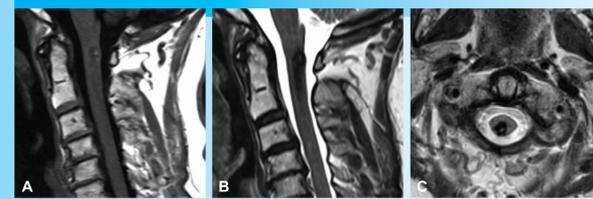


FIGURE 2 66 year old. Sagittal T1 (A) and T2-TSE (B), axial T2-TSE (C). Classic appearance with T1 hyperintense signal centrally and peripheral hypointense signal, longitudinal pattern seen on sagittal T2.



FIGURE 3 30 year old imaged because of back pain, thoracic spine MRI. Sagittal T1 (A) and T2 TSE (B). Typical central T1 and T2 hyperintensity with peripheral hemosiderin deposition oriented longitudinally.

Small lesions have signal characteristics consistent with hemosiderin only and may be subtle. Typical TSE or FSE techniques for spinal imaging are relatively insensitive for small CMs, and gradient-based techniques are important in evaluating suspected spinal CMs. (Fig. 4; also Fig. 8)

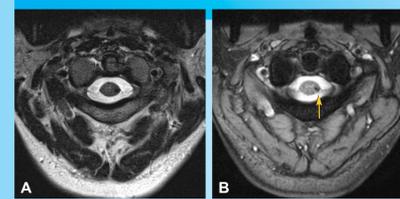


FIGURE 4 MRI cervical spine of 41 year old. The small CM in the left side of the spinal cord is very subtle on axial TSE T2 (A) and much more conspicuous on 2D MEDIC (B, arrow).

NERVE ROOT CMs

These are rare, with an article in 2006 finding only 12 in the literature. One of our patients with multiple spinal cord CMs also had 2 nerve root CMs. (Fig. 7)

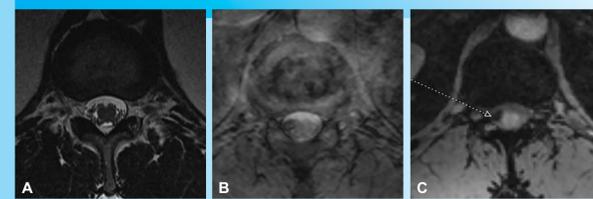


FIGURE 7 57 year old imaged for back pain. Axial TSE T2 (A), 2D MEDIC (B), and 3D MEDIC (C) show a nodular right ventral nerve root with susceptibility effect on gradient-based images. No enhancement (not pictured). Multiple CMs were also present within the thoracic spinal cord and in the brain. Proven CCM1.

ACUTE HEMORRHAGE

CMs that present with acute hemorrhage can be particularly confusing. Patients often present with acute myelopathy, and the lesions can be very complex on MRI, with areas of both T1 and T2 hyperintensity and low T2 signal intensity. Blood tends to accumulate in a longitudinal fashion. Features may include blood layers, hematomyelia, and edema. Enhancement is variable but often not a prominent feature. Use of gradient echo techniques, attention to use of fat saturation, and MRI of the brain including gradient-based techniques can be very helpful. (Figs. 5, 6) Blood may extend around or distal to the CM (caudal extension shown in Fig. 6).



FIGURE 5 17 year old who presented with acute onset of sensory loss and left hemiparesis. MRI cervical spine sagittal T1 (A) and TSE T2 (B) shows heterogeneous lesion with fluid-fluid layer and edema. No prior history was known. Brain MRI (C) TSE T2 shows a brain CM in the left frontal lobe, and subsequent genetic testing confirmed CCM1 mutation. Spinal cord CM confirmed surgically.



FIGURE 6 5 year old child presented with acute onset low extremity paresis, no known prior history of CM. Sagittal T1 (A) and TSE T2 (B, C) shows central, longitudinal T1 hyperintensity extending centrally within the distal spinal cord and a complex appearance just superior (C), including fluid layers. Brain MRI followed (D), which showed a right frontal lobe focus of susceptibility change (arrow). Genetic testing of both the patient and a parent confirmed previously unknown CCM1. Surgery confirmed that the complex lesion (C) was a CM.

ISOTROPIC GRADIENT BASED IMAGING

As you would expect, gradient based imaging increases sensitivity for the detection of spinal CMs compared to spin echo sequences. (Fig. 4). Interestingly, 3D MEDIC appears to have greater sensitivity in some cases compared to 2D MEDIC, with the added benefit of being isotropic, facilitating multiplanar reconstruction. (Fig. 8) We expect that comparable techniques from other manufacturers would have similar advantages.

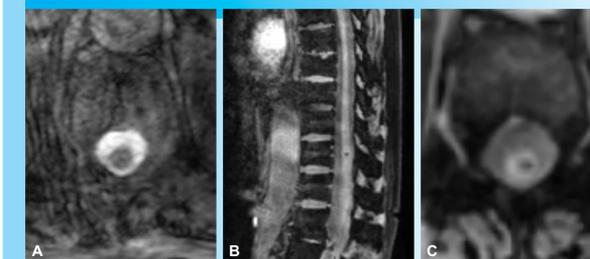


FIGURE 8 Thoracic spine MRI. Central hypointensity is very subtle on axial 2D MEDIC (A), much more obvious on 3D MEDIC (B, sagittal, C, reconstructed axial).

PERILS OF FAT SATURATION

The use of fat saturation on post-contrast T1 sequences may suggest an erroneous appearance of enhancement. Thus, if fat saturation is used, one may easily confuse a spinal CM with a tumor. Lack of enhancement is readily demonstrated if post-contrast T1 sequences are performed without fat saturation. (Figs. 9, 10) CMs sometimes enhance, but T1 hyperintensity without enhancement is most likely due to methemoglobin.

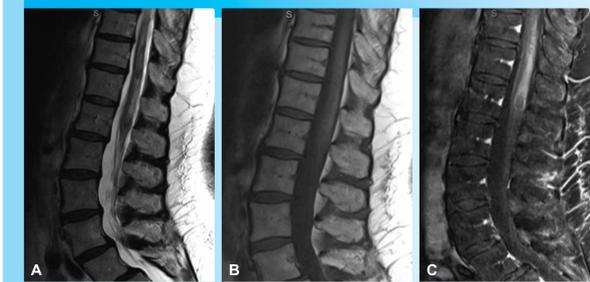


FIGURE 9 64 year old who presented with 3 week history of numbness of both lower extremities, several episodes of urinary incontinence. Sagittal TSE T2 (A) shows mixed signal intensity in the distal spinal cord. Sagittal precontrast T1 (B) demonstrates subtle hyperintensity, and postcontrast T1 with fat saturation (C) appears to show much more impressive hyperintensity, simulating enhancement, and the patient was referred from another institution because of presumed tumor. However, repeat sagittal T1 precontrast (D) and postcontrast without fat saturation (E) shows minimal to no enhancement; the T1 hyperintensity instead is mostly due to methemoglobin. Addition of sagittal T2 GR (F) further clarified the presence of blood. MRI brain (G, axial T2 GR) showed numerous CMs. Spinal CM confirmed surgically.

LOOK AT THE BRAIN

In several cases illustrated here, the patient presented with acute spine symptoms. The diagnosis of a spinal cavernous malformation prompted imaging of the brain, which revealed additional cavernous malformations. Subsequent genetic testing confirmed the diagnosis. The presence of multiple lesions suggests familial rather than sporadic disease and has profound implications for the patient's family and genetic counseling. Additionally, finding more lesions in the brain can increase your confidence in the diagnosis of a spinal cavernous malformation. (Fig. 10; also Figs. 5, 6, 9)

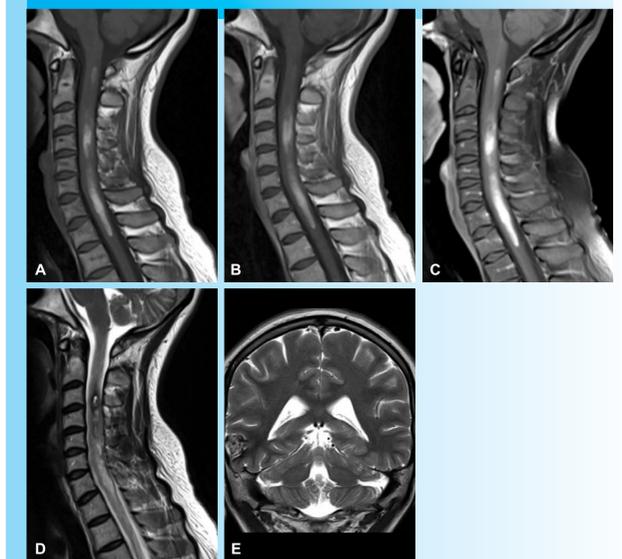


FIGURE 10 Sagittal T1-weighted MRI of a 31 year old patient. Precontrast (A) and postcontrast without (B) and with (C) fat saturation shows multiple foci of T1 hyperintensity in the cervical spinal cord. Altered cord-to-background contrast with fat saturation could be misleading. Sagittal T2 (D) shows heterogeneous signal intensity. Brain MRI (E, coronal TSE T2) demonstrated typical right temporal lobe CM.

CONCLUSIONS—LEARNING POINTS:

- ♦ Spinal CMs are probably more common in genetic CM disease than reported in the literature.
- ♦ The possibility of familial or genetic CMs should be considered with hemorrhagic spinal cord lesions.
- ♦ The appearance of spinal CMs with acute hemorrhage with edema can be confusing.
- ♦ Beware of fat saturation on post contrast imaging.
- ♦ Gradient based sequences add sensitivity to the detection of spinal CMs.
- ♦ The diagnosis of spinal CMs should prompt imaging of the brain.

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