



Cavernous Malformation

Last Updated: October 1, 2018

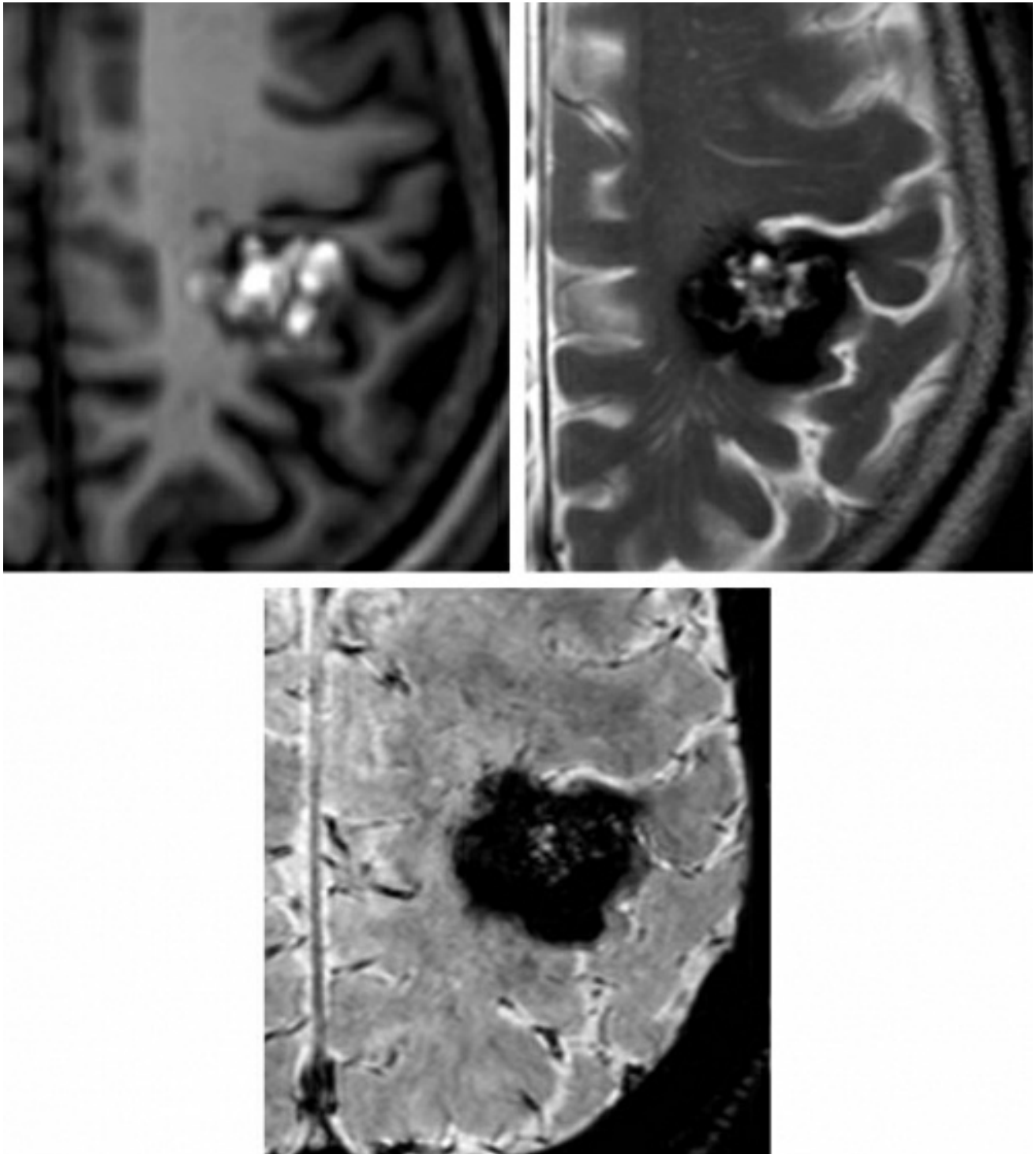


Figure 1: Axial T1, T2, and SW images reveal a “popcorn” lesion within the left postcentral gyrus demonstrating heterogenous T1 and T2 signal accompanied by hypointense rim of blooming

artifact (hemosiderin) typical of a cavernous malformation.

Overview

Cavernous malformations are benign vascular hamartomas consisting of dysplastic immature vascular channels with variable mass effect depending on size. Though nearly always angiographically occult, these lesions may be dynamic in their behavior over time with evidence of hemorrhagic products of mixed stages, and may present with acute parenchymal hemorrhage. Presentation may include seizure (up to 50%), focal neurologic deficit (25%), and asymptomatic/incidental (20%).

Imaging

- General Characteristics
 - Masses of immature, cavern-like vascular channels or blood pools, of variable complexity
 - Loculated intralesional hemorrhages with different stages of blood product evolution
 - Complete hemosiderin rim usually surrounds lesion
 - Lesions classically described as having a “popcorn” appearance on MRI
 - Dynamic behavior on serial imaging including enlargement, regression, and de novo formation
 - Location
 - Occurs throughout CNS
 - Brain parenchyma (common)
 - Spinal cord (rare, more common in patients with multiple cavernous malformation syndrome)
 - Extraaxial (rare)
 - Can originate within any venous sinus (cavernous sinus most common site)

- Size
 - May attain large size before becoming symptomatic
 - Vary from microscopic to giant (> 6 cm)
 - Majority fall in the range of 0.5 - 4.0 cm
- Multiple cavernous malformations
 - Multiple (familial) CM syndrome
 - Autosomal dominant, variable penetrance
 - Nonsense, frame-shift, or splice-site mutations consistent with 2-hit model
 - Radiation-induced
- Associated abnormalities
 - Developmental venous anomaly
 - Superficial siderosis
 - Cutaneous abnormalities
 - Café au lait spots
 - Hyperkeratotic capillary-venous malformations
- **Differential Diagnoses**
 - Arteriovenous malformation
 - Hemorrhagic neoplasm
 - Calcified neoplasm
 - Hypertensive microbleeds
 - Amyloid angiopathy
- **Nonenhanced CT**
 - **Technique**
 - Axial 5mm collimation is typical
 - Reformatted images in coronal/sagittal planes can be made available in difficult cases, though are generally not needed
 - **Findings**

- Negative in up to 50% of cases without recent parenchymal hemorrhage
- Well-delineated round/ovoid hyperdense lesion, usually < 3 cm
- Partially calcified in some cases
- No evidence of surrounding hypodense edema or mass effect (unless recent hemorrhage)

- **CT Angiography**

- **Findings**

- Minimal to no enhancement, usually negative (angiographically occult)

- **MRI**

- **Technique**

- Multiple pulse sequences available to evaluate various tissue properties as well as dynamic properties (flow, velocity)

- **Findings**

- Most commonly identified as a popcorn-like mass lesion with mixed signal on T1, T2, FLAIR reflecting mixed blood-containing loculations
- T1 signal
 - Variable depending on blood product evolution
- T2 signal
 - Variable depending on blood product evolution
 - T2 hypointense hemosiderin rim
 - May see loculations of blood with fluid-fluid levels

- FLAIR
 - Again, variable signal dependent on age of blood products
 - May reveal perilesional parenchymal edema if recently hemorrhagic
- T2* GRE, SWI
 - Prominent hypointense blooming from susceptibility artifact
 - Very sensitive for detection, particularly in cases of multiple lesions, familial syndrome
- DWI
 - Usually normal
- T1WI C+
 - Minimal or no enhancement (may show associated developmental venous anomaly)
- MRA
 - Usually normal (angiographically occult)
- **Pitfalls**
 - Large acute hemorrhage may obscure more typical features
- **Digital Subtraction Angiography**
 - **Findings**
 - Usually normal (angiographically occult vascular malformation)
 - Extradural lesions may be hypervascular
 - Slow intralesional flow without AV shunting may be seen

- Avascular mass effect if large or acute hemorrhage
- Associated other malformations, most commonly DVA
- Generally, there is no current role for endovascular therapy
- **Staging, Grading, & Classification**
 - *Zabramski classification: associated with risk of re-hemorrhage*
 - Type 1 = subacute hemorrhage (hyperintense on T1WI; hyper- or hypointense on T2WI)
 - Highest risk of re-hemorrhage
 - Type 2 = mixed signal intensity on T1, T2WI with degrading hemorrhage of various ages
 - Type 3 = chronic hemorrhage (hypointense to isointense on T1, T2)
 - Type 4 = punctate microhemorrhages (T2*GRE and SWI sequences most sensitive)

For more information, please see the corresponding chapter in [Radiopaedia](#), and the [Cavernous Malformation](#) chapter within the [Brain Tumor Mimics](#) sub-volume in the Neurosurgical Atlas.

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DOI: <https://doi.org/10.18791/nsatlas.v2.03.03.04.04>

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