Pediatric Spinal Cavernous Malformation

- Abdominal Pain Mimicking a Neurological Disorder: A Case Report of Spinal Cavernous Malformation in a Pediatric Patient
- Pediatric Central Nervous System Vascular Malformation : Pathological Review with Diagram
- Clinical and genetic characteristics of 9 rare cases with coexistence of dual genetic diagnoses
- Giant Cavernous Malformation Mimicking an Infiltrative Intracranial Neoplasm in Children-Case Report and Systematic Review of the Literature
- Multiple spinal intramedullary cavernous angiomas with bleeding episode mimicking an intramedullary tumor
- Extradural hemorrhagic spinal cavernous angioma in a paucisymptomatic child: A rare case with review of the current literature
- Spinal involvement in pediatric familial cavernous malformation syndrome
- Natural History of Spinal Cord Cavernous Malformations: A Multicenter Cohort Study

A **Pediatric Spinal Cavernous Malformation (SCM)** is a rare vascular lesion of the spinal cord in children, composed of dilated, thin-walled vascular channels without intervening normal neural tissue.

Pathophysiology

SCMs consist of:

- Compact clusters of sinusoidal capillaries
- Thin walls lacking smooth muscle and elastin
- No arterial supply; low-flow lesion
- Often associated with microhemorrhages and hemosiderin deposits

Epidemiology

- Extremely rare in children
- More common in adults (peak: 30–50 years)
- Pediatric cases may present earlier due to larger lesions or genetic predisposition

Etiology

- **Sporadic**: Most pediatric cases
- **Familial**: Linked to *CCM1*, *CCM2*, or *CCM3* gene mutations (often multiple lesions)
- May coexist with cerebral cavernous malformations (CCM)

Clinical Presentation

- Progressive **myelopathy**
- Sudden neurological decline due to **hemorrhage**

- Pain (back or radicular)
- Weakness, sensory loss
- Bowel/bladder dysfunction
- Sometimes incidental finding

Diagnosis

- **MRI** with and without contrast:
  - “Popcorn” or “mulberry” appearance
  - Mixed signal intensity (T1/T2) with hemosiderin ring (T2 hypointensity)
  - Gradient echo or SWI: sensitive to hemorrhage
- **No enhancement** or mild enhancement post-Gd

Differential Diagnosis

- Ependymoma
- Astrocytoma
- Hemangioblastoma
- Spinal arteriovenous malformations (AVMs)
- Lipomas or dermoids

⚖ Management

- **Observation** if asymptomatic or mild stable symptoms
- **Surgical resection**:
  - Indicated for progressive deficits or recurrent hemorrhage
  - Best done with neurophysiological monitoring
  - Gross total resection is curative
- Radiosurgery: not typically used due to spinal cord risk

Prognosis

- Favorable with early diagnosis and complete resection
- Risk of rebleed if untreated (especially in symptomatic cases)
- Long-term monitoring recommended (especially in familial forms)

References

- Gross BA, Du R. Spinal Cavernous Malformations: Clinical features and surgical outcomes. *Neurosurg Focus*. 2010.
- Labauge P et al. Genetics of cavernous angiomas. *Lancet Neurol*. 2007.
- Wang X, et al. Pediatric spinal cavernous malformations: A review. *Childs Nerv Syst*. 2020.