Type 3 Vascular Malformation

- Synonyms:
 - Juvenile AVM; intramedullary extramedullary AVM
- Juvenile-type AVM (large, complex, intramedullary extramedullary AVM)
- Rarest of spinal AVMs (7%)
- Age
 - 1st-3rd decades
- Gender
 - Slight male predominance for Cobb syndrome
- Epidemiology
 - Intramedullary (types 2, 3):15-20% of spinal AVM; type 4: 10-20%
 - AVMs account for < 10% of spinal masses
- Large complex nidus, multiple feeding vessels; may be intramedullary and extramedullary, and even extraspinal

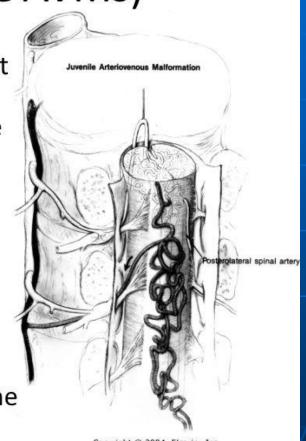
Type III (Juvenile AVMs)

 Arise in single or multiple adjacent somites → thus intradural and extradural, may involve soft tissue and bone in addition to the cord

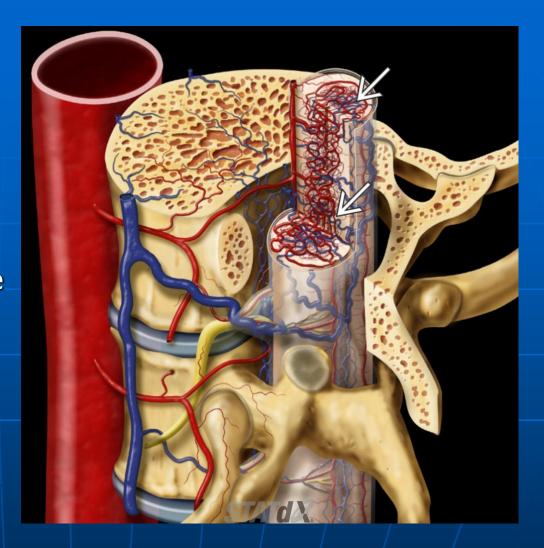
 Diffuse shunts with normal spinal cord existing between loops of abnormal vessels

No distinct nidus

 Metameric: tissue derived from the entire somite



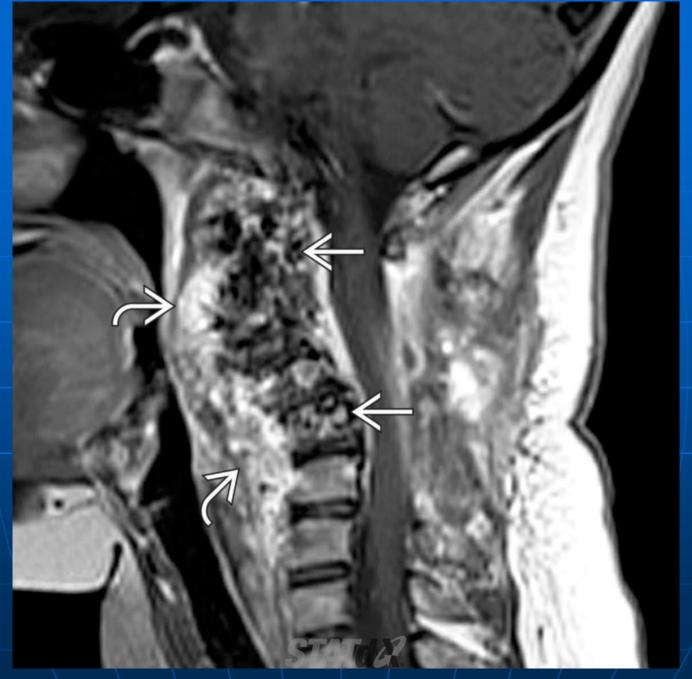
- Progressive neurologic decline (weakness), subarachnoid hemorrhage
- Pain in lower extremities or back; motor deficit
- Poor prognosis for type 3 (juvenile) AVM



Sagittal oblique graphic of the thoracic cord shows a large complex intramedullary nidus of a type 3 arteriovenous malformation (AVM) (white solid arrow) with multiple feeding vessels.



Sagittal T2WI MR exhibits extensive flow voids (white solid arrow), involving a long segment of the cervical cord, seen throughout the subarachnoid space. These lesions are fed by multiple arteries at multiple vertebral levels. An apoplectic presentation from SAH or intramedullary hemorrhage is strongly associated with intramedullary AVMs.



Sagittal T1WI C+ MR displays a vascular lesion composed of multiple flows in the cervical spine, involving the vertebral bodies (white solid arrow) and prevertebral muscles (white curved arrow).

Check List

Consider

- Dynamic enhanced MRA is first-line vascular imaging choice
- Consider other types of spinal vascular malformation if evidence of intramedullary flow voids
- Rarely, cervical type 1 dAVF may drain intracranially with high flow, varix formation, and SAH
- Posterior fossa dural fistula may mimic cervical spinal type 1 fistula when intracranial fistula has caudal intraspinal venous drainage

Image Interpretation Pearls

 Imaging and clinical findings are frequently subtle or nonspecific; early diagnosis requires high level of suspicion

- Progressive neurologic decline (weakness), subarachnoid hemorrhagePain in lower extremities or back; motor deficit
- Poor prognosis for type 3 (juvenile) AVM