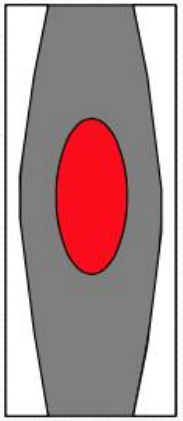
Intramedullary Tumors

- Primary tumors – much more common
  - Ependymoma
  - Astrocytoma

  } ~ 90-95% of all intramedullary tumors
  - Hemangioblastoma
  - Subependymoma, Oligodendroglioma, Ganglioglioma (rare)
- Secondary tumors – metastases
  - Lung, breast, renal, gastric
  - Lymphoma, leukemia
  - Melanoma



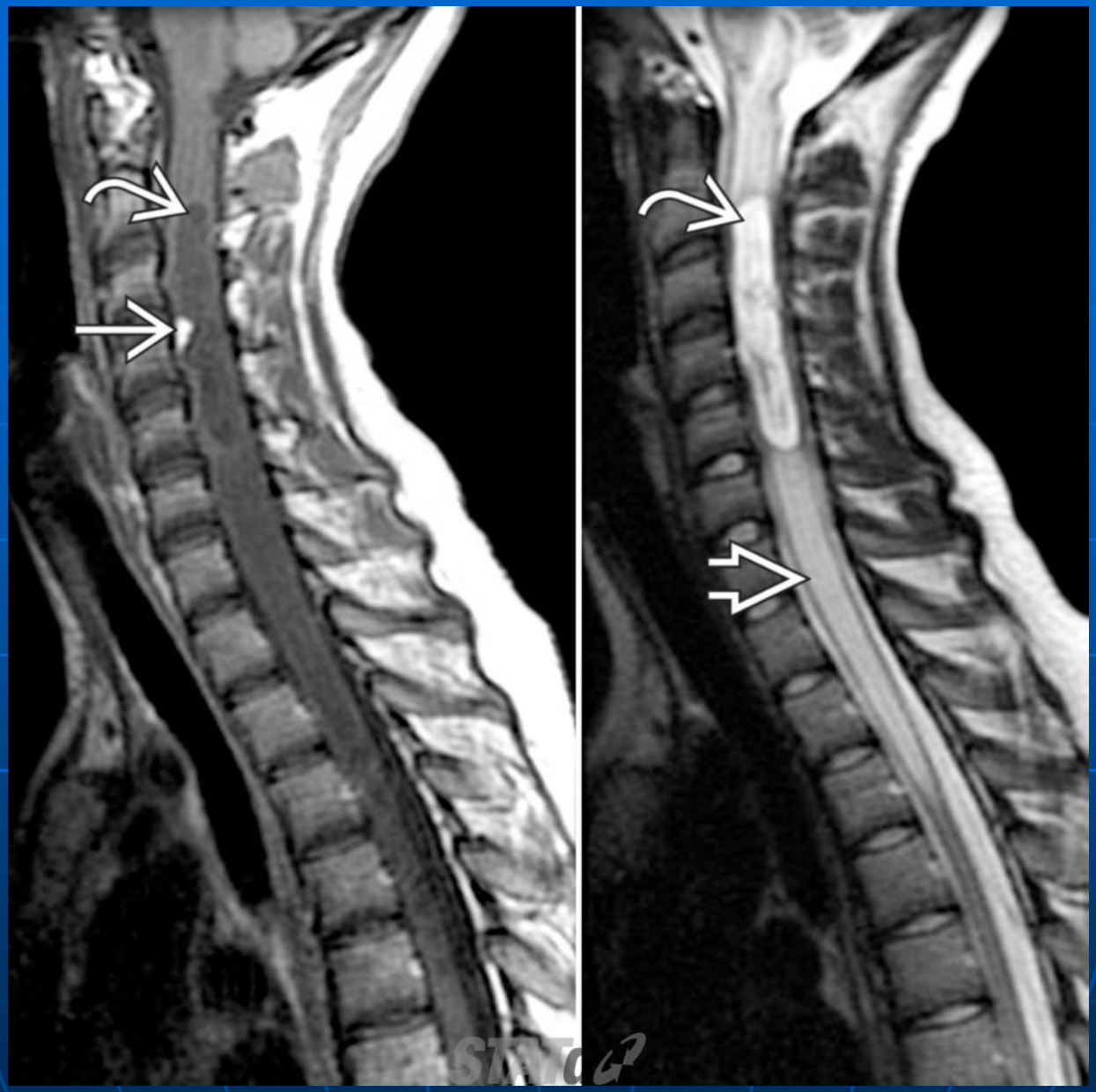
Hemangioblastoma

- Low-grade, capillary-rich neoplasms of cerebellum and spinal cord that occur sporadically or in setting of von Hippel-Lindau (VHL) syndrome.
- WHO grade I
- No malignant degeneration
- 68% of spinal HBs are sporadic (32% VHL-associated)
  - Multiple HBs indicate VHL syndrome
- Patients with suspected HB/VHL should undergo contrast-enhanced MR imaging of entire neural axis to exclude multiple lesions

Imaging

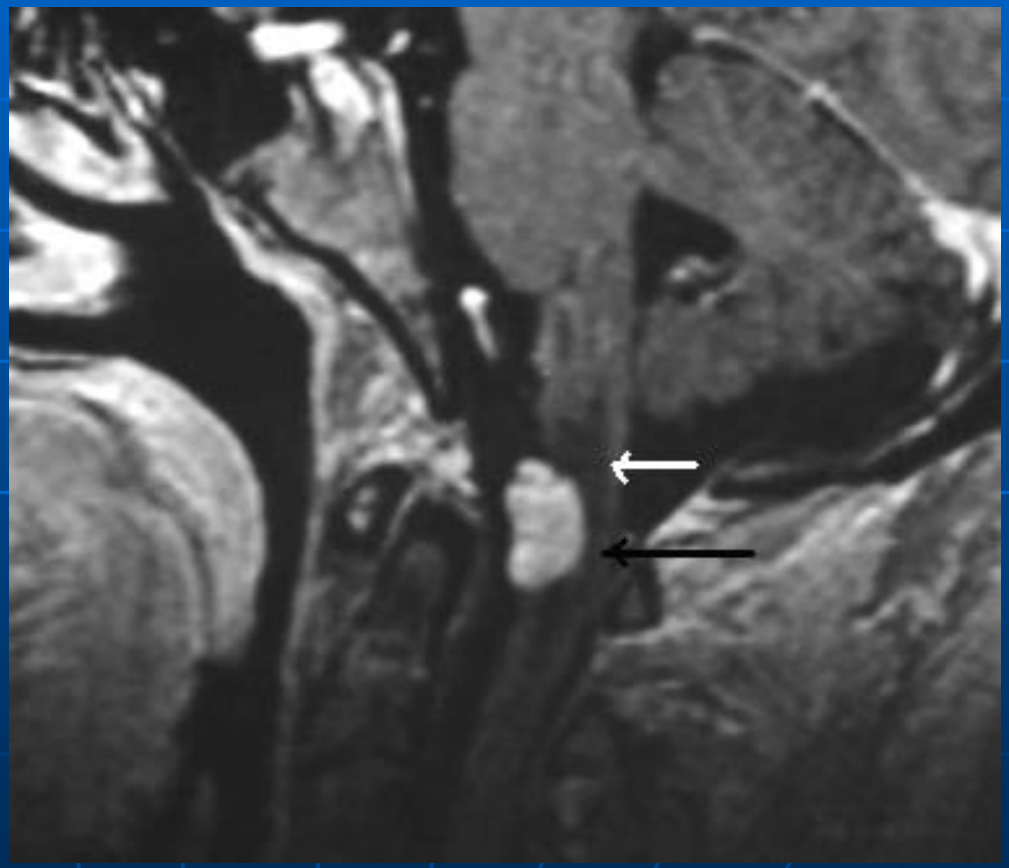
- Thoracic > cervical > lumbar, sacral
- Subpial and posterior aspect of spinal cord
- Usually shows uniform contrast enhancement
- Large lesions have vessel flow voids
- Often associated with intraspinal cyst
- Extensive syrinx suggests hemangioblastoma (HB)
- Spinal cord thickening remote from tumor and without syrinx

Sagittal graphic depicts a focal intramedullary mass (white solid arrow) in the cervical cord with prominent feeding vessels, adjacent cysts (white open arrow), and cord expansion with edema.



Sagittal T1WI C+ MR (left) demonstrates an enhancing nodule (white solid arrow) in the ventral cord with an associated cyst (white curved arrow). Sagittal T2WI MR (right) reveals the heterogeneous cyst (white curved arrow) and extensive surrounding edema (white open arrow).

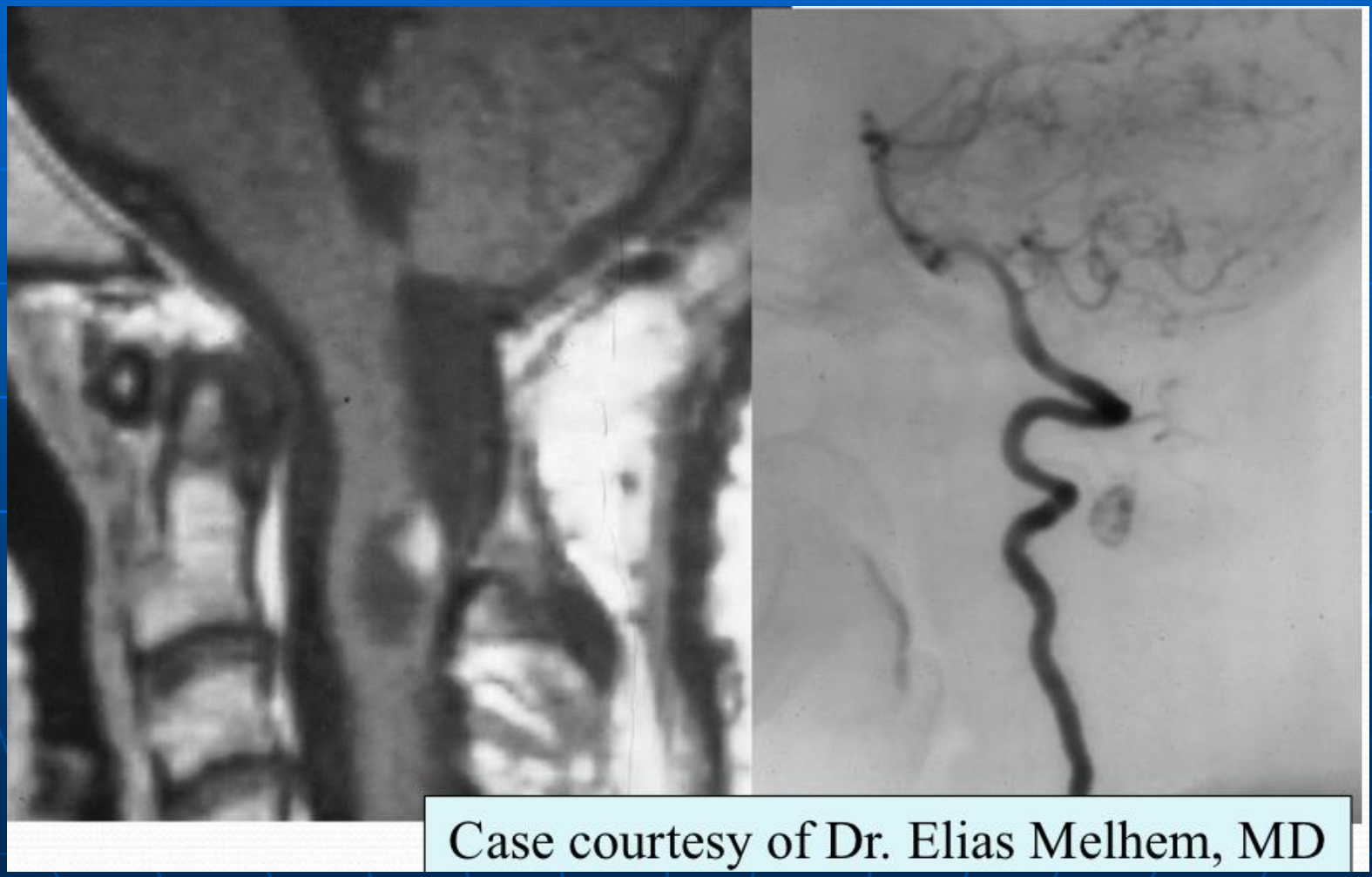
Hemangioblastoma



Hemangioblastoma

- Thoracic spinal cord as the most common location
- Densely enhancing solid tumor nodule within a large "syrinx" cavity and associated "feeding" vessels is highly suggestive, if not diagnostic, of a hemangioblastoma
- Can bleed with devastating consequences
- Can be extramedullary and exophytic
- May have lots of flow voids

Another Hemangioblastoma



DDx:

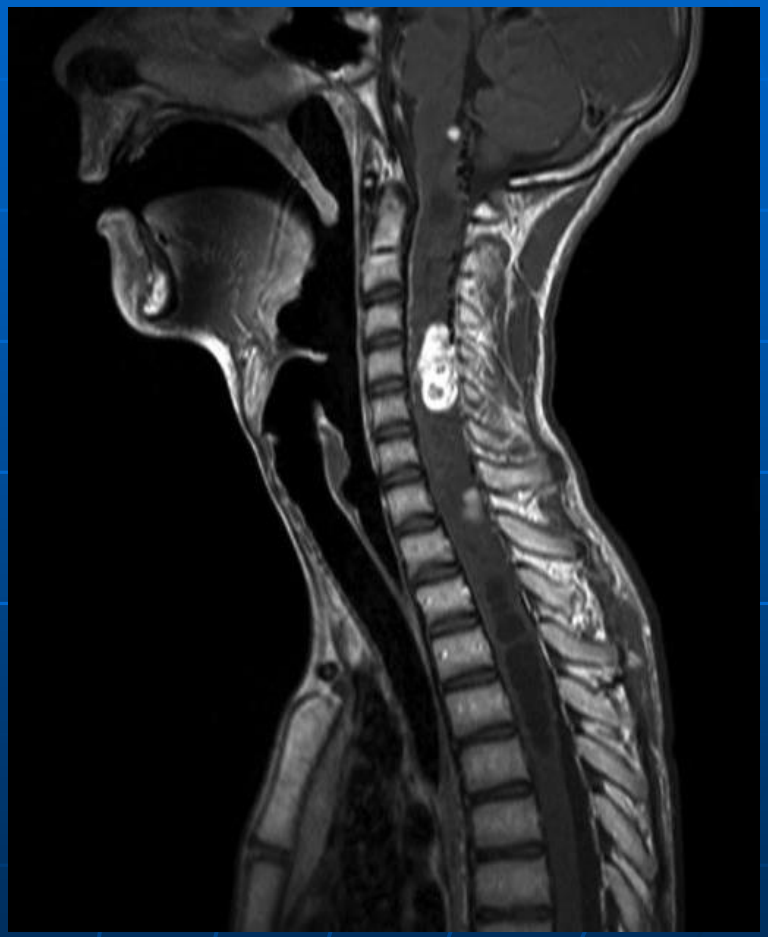
- Arteriovenous Malformation (AVM)
  - Cord often normal/small, gliotic
  - Syrinx, focal enhancing nodule absent
- Cavernous Malformation
  - Mottled or speckled pattern of prior hemorrhage, hemosiderin rim
  - Minimal enhancement
  - Hypervascular Cord Neoplasms
- Ependymoma
  - Centrally located mass
  - Intense enhancement with well-defined margins and occasional central cavitation
  - ± syrinx
- Astrocytoma
  - Heterogeneous enhancement
  - Usually not hypervascular
  - Peritumoral edema common
- Vascular metastasis (known primary, e.g., renal cell carcinoma)
- Intradural Extramedullary Tumors
  - Meningioma
  - Schwannoma
    - Rarely associated with syrinx, uncommon flow voids
  - Paraganglioma
    - Filum much more common than spinal cord

Von Hippel-Lindau disease

- Multi-system disorder
- characterized by the development of numerous benign and malignant tumors (at least 40 types), as well as several non-malignant lesions.

CNS manifestations

- CNS hemangioblastoma(s)
  - cerebellar (~75%)
  - spinal (~25%)
- Retinal hemangioblastoma(s)
- Choroid plexus papilloma (CPP)
- Endolymphatic sac tumors (ELST)



Non CNS manifestions

- pheochromocytoma(s)
- numerous pancreatic lesions (may be the earliest manifestation [3])
  - pancreatic cysts
  - pancreatic islet cell tumours
  - microcystic adenoma(s) of pancreas
  - pancreatic adenocarcinoma (rare)
- renal lesions
  - renal cell carcinoma(s): usually of the clear cell type [7]; can occur in up to 40% of cases and is one of the most feared complications; RCCs present at an earlier age in those with vHL
  - renal cysts: can occur in up to 75% of cases [5]; often tend to be bilateral and multiple
  - renal angiomyolipoma(s)
- liver cysts
- papillary cystadenoma(s) of the epididymis

Von Hippel-Lindau Disease

- Autosomal dominant, gene VHL (3p25-p26)
- Incidence 1:36.000

- Retinal angiomas (70%)
- Hemangioblastomas of the brain (50%) and spinal cord
- Renal cell carcinoma (40%)
- Pheochromocytoma
- Endolymphatic sac tumors (10%)
- Cysts of the pancreas, kidney, liver and epididymis.