

# Vascular Tumors

Updated: April 24, 2010

|   |   |
|---|---|
| HEMANGIOBLASTOMA .....                    | 1 |
| HEMANGIOPERICYTOMA .....                  | 3 |
| SPINAL HEMANGIOBLASTOMA → see p. Onc50 >> |   |

## HEMANGIOBLASTOMA

- rare benign vascular neoplasm that arises almost exclusively in CNS.

- classified by WHO as **meningeal tumors of uncertain origin**.  
*primitive endothelial cells around 4<sup>th</sup> ventricle?*
- 1-2.5% of all intracranial neoplasms, but:
  - 8-12% of **posterior fossa** tumors! (most common **primary adult intraaxial posterior fossa tumor**).  
Cerebellar hemangioblastomas = **LINDAU tumors** (*Lindau first described them in 1926*)
  - 3-7% of **spinal cord** tumors. see p. Onc50 >>
  - extremely rare in supratentorial compartment, optic nerve, brainstem, peripheral nerves.
- male-to-female** ratio ≈ 2:1.
- usual age at diagnosis - **third ÷ fifth decades** (rarely affect children).

20-25% hemangioblastomas are part of **von HIPPEL-LINDAU (VHL) disease** - **autosomal dominant** deletion of **VHL** (tumor suppressor gene on 3p) with variable penetrance and delayed expression:

- retinal angiomas** (*von Hippel's disease*) - usually in peripheral retina (vision is unaffected; exudation in region of angiomas may lead to retinal detachment);  
H: photocoagulation / cryocoagulation.
- cerebellar hemangioblastomas** (*Lindau's syndrome*); usually multiple; some may produce erythropoietin-like substances → asymptomatic **polycythemia**.
- various **visceral tumors** - kidneys [renal cell carcinoma, cysts, angiomas], adrenal glands [pheochromocytoma], pancreas [cysts], epididymis [papillary cystadenomas, cysts], liver [angiomas, cysts].

**Renal carcinoma** is most common cause of death!

- classified as phakomatosis, although it does not include any cutaneous manifestations!
- often presents at younger age.
- incidence - 1 in 30,000-40,000.
- tumor-suppressor gene is located on 3p25-26 (encodes protein pVHL that inhibits "elongation" step during RNA synthesis).
- perform **screening of all family members** (*retinal examination* including children during first 2 years of life).  
Retinal angioma is indication for MRI!

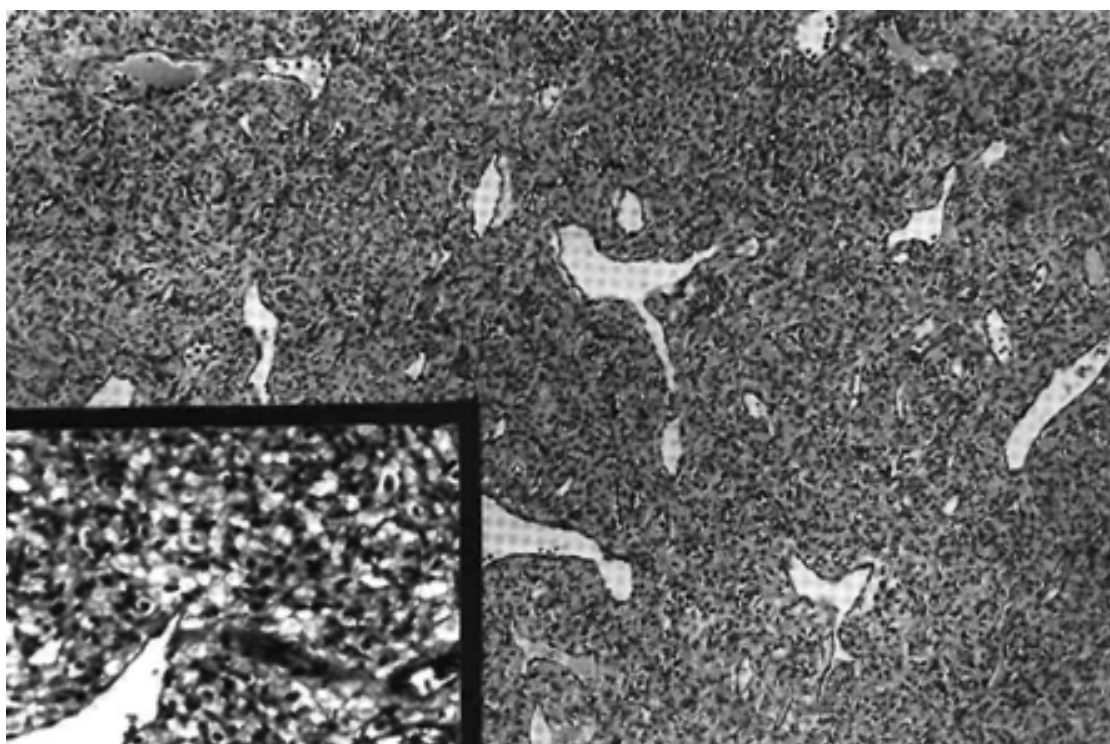
### **PATHOLOGY**

- cherry-red in color (highly-vascular - **may simulate AVM!!!**).
- ≈ 30% are **solid**.
- ≈ 70% are as **mural nodule** and **cyst** that contains yellow proteinaceous fluid; cyst wall is glial nonneoplastic reaction to secreted fluid; mural nodule is touching pial surface of cerebellum.
- grows **inside parenchyma attached to pia mater** (gets rich vascular supply from pial vessels), but no dural attachment.
- not invasive (but border does not contain any particular membrane or capsule).
- no calcification.
- extramedullary* and *extradural* hemangioblastomas have been described.
- subarachnoid dissemination is extremely rare, tumor enlarges extremely slowly.

Histology - primitive vascular channels with 3 types of cells:

- relatively normal-appearing **endothelial cells** that line capillary spaces.
- small, perivascular, **pericytes** with dark compact nuclei and sparse cytoplasm.
- stromal cells** with multiple vacuoles and granular eosinophilic cytoplasm **rich in lipids\***; some nuclear pleomorphism; represent abnormally differentiating mesenchymal cells of angiogenic lineage, with some morphological features of endothelium, pericytes, and smooth-muscle cells.

\*can be mistaken for **XANTHOCHROMIC ASTROCYTOMAS**



### **CLINICAL FEATURES**

Long history (≈ 1 yr) of minor neurological symptoms → sudden exacerbation.

**Cerebellar lesions** → cerebellar dysfunction, ICP↑ (due to hydrocephalus).

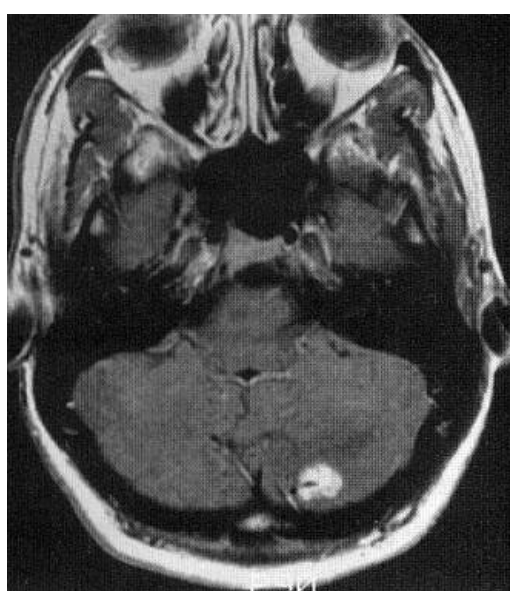
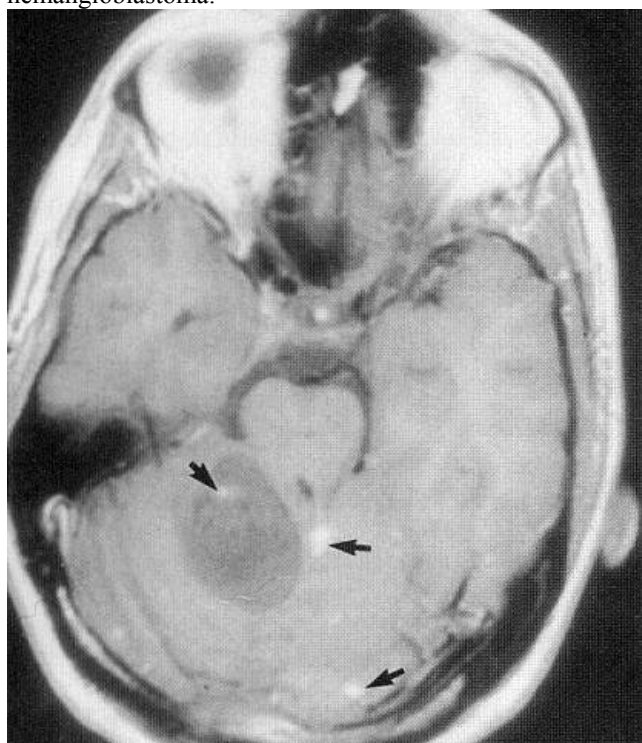
**Spinal cord lesions** → pain, progressive spinal cord compression.

### **DIAGNOSIS**

- complete neural axis imaging:
  - CT / MRI** - clearly delineated **intensely enhancing mass** or uniform intense enhancement of tumor **nodule with adjacent cyst**.
  - Angiography** (usually done before surgery) - highly vascular tumor blush, enlarged feeding arteries and draining veins.

- detect VHL disease complex:
  - 1) **ophthalmologic evaluation:** aneurysmal dilation of peripheral retinal vessels (earliest characteristic feature) → tortuous vessels marked by afferent arteriole and venule leading to raised retinal lesion.
  - 2) **abdominal CT / ultrasound**

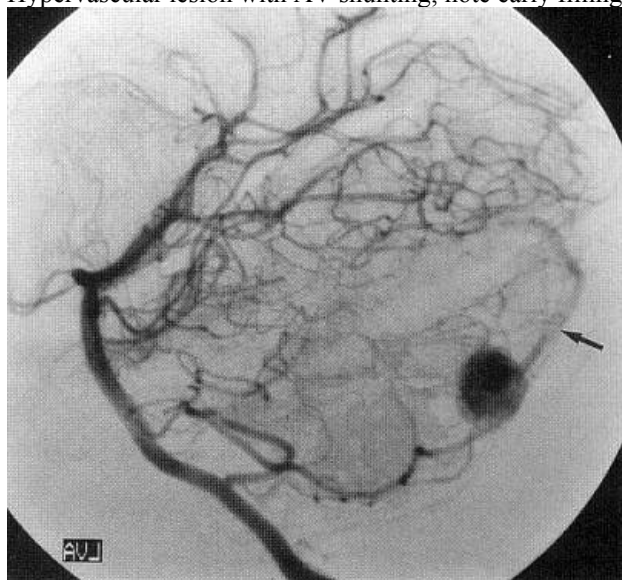
Multiple hemangioblastomas (*arrows*) in von Hippel-Lindau disease (T1-MRI); large cyst associated with right cerebellar hemangioblastoma:



Contrast CT - cystic mass in left cerebellar hemisphere with enhancing mural nodule (*arrowhead*) and surrounding edema (*arrow*):



Hypervascular lesion with AV shunting; note early filling vein (*arrow*) indicative of hemangioblastoma:



### TREATMENT

*Asymptomatic lesions* may be safely **observed** with MRI to rule out tumor enlargement.

*Symptomatic lesions* → **surgical removal:**

**cerebellar lesions** - via suboccipital craniectomy;

**spinal lesions** - via laminectomy.

Hydrocephalus → external ventricular drain prior to tumor resection!

- **coagulate tumor surface** with wide bipolar forceps (avoid penetration of tumor itself due to its extreme vascularity and difficulties with hemostasis).
- **dissect tumor circumferentially** by careful coagulation and cutting small feeding vessels and adhesions between tumor and surrounding tissue by putting cottonoid strips into developing plane to avoid direct pressure on brain or spinal cord tissue.
- identify **feeding vessels** → coagulate and cut (arterial feeders prior to draining veins!)
- gently **pack resection cavity** with wet cotton balls → blood oozing stops after few minutes.
- **need for permanent shunt** is determined by response to external ventricular drain clamping.

Other options - **endovascular embolization** of solid component (tumor vascularity↓), **stereotactic radiosurgery**.

Indications for **radiotherapy** (at least 50 Gy):

- 1) unresectable
- 2) incompletely excised
- 3) recurrence

**PROGNOSIS**

- local recurrences < 25% (esp. in VHL disease, younger age, multiple hemangioblastomas).

## HEMANGIOPERICYTOMA

- rare *dural* tumor from **perivascular pericytes** (high percentage in torcular region); also occurs *elsewhere in body*.

- homozygous **deletions of CDKN2/p16 gene** are common.
- some classifying it as subtype of meningioma (but do not have mutations of NF2 gene).
- grossly - smooth, lobulated, well-encapsulated, very vascular.
- histology (similar to angioblastic meningioma and hemangioblastoma) - elongated pericytes with processes which wrap around thin-walled vascular channels of single layer of endothelial cells (i.e. cells lie external to vascular endothelium); cells are arranged in whorls or pinwheels, with rich investment of reticulin.
- locally aggressive, may metastasize (far more aggressive than ordinary meningiomas).
- imaging - lobulated (vs. meningioma - spherical) dural based mass, no calcification, no hyperostosis, multiple areas of flow void (reflect high vascularity).
- treatment – **surgery** → **radiotherapy** and/or **chemotherapy**.

BIBLIOGRAPHY for ch. “Neuro-Oncology” → follow this [LINK >>](#)