Brigham and Women's Hospital

Harvard Medical School

Rad-Path Conference: Intraventricular Neoplasms

September 23, 2013

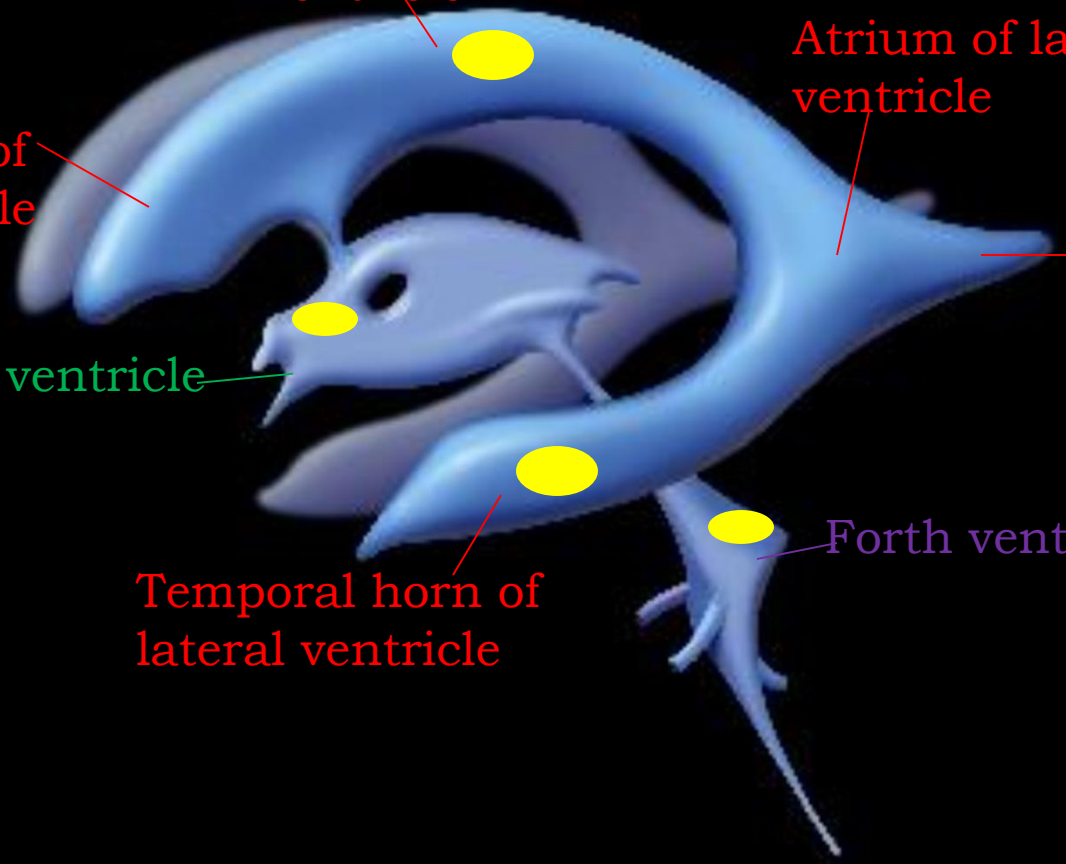
Thani Chansakul, MD

Tejus Bale, MD

Objectives

- Review ventricular anatomy and histologic structure
- Review differential diagnosis of intraventricular neoplasms
- 5 Unknown cases
- Discussion of clinical and imaging features of the cases

Ventricular anatomy



Choroid plexus

Histologic structure of the ventricles

- The epithelial lining of the ventricles is composed of ependymal cells → Ependymoma
- Subjacent to the ependymal lining is a layer of subependymal plate composed of glial cells → Subependymoma
- The septum pellucidum is also lined by glial cells and residual neuronal precursor cells → Central neurocytoma

Histologic structure of the ventricles

- The choroid plexus develops from invagination of primitive pia-arachnoid and vessels into these vesicles, creating the choroidal fissures
- The choroid plexus → Choroid plexus neoplasms
- Rich vascular supply in CP → Deposition of metastases
- Arachnoid cap cells may be trapped within the CP → Meningioma

Differential diagnosis

Based on cell origins:

- *Ependymoma* (ependymal linings)
- *Subependymoma* (subpendymal layer)
- *Choroid plexus neoplasms including choroid plexus papilloma, atypical papilloma and carcinoma* (choroid plexus)
- *Central neurocytoma* (neuronal precursor cells)
- Meningioma (arachnoid cap cells trapped in choroid plexus)
- *Metastasis*

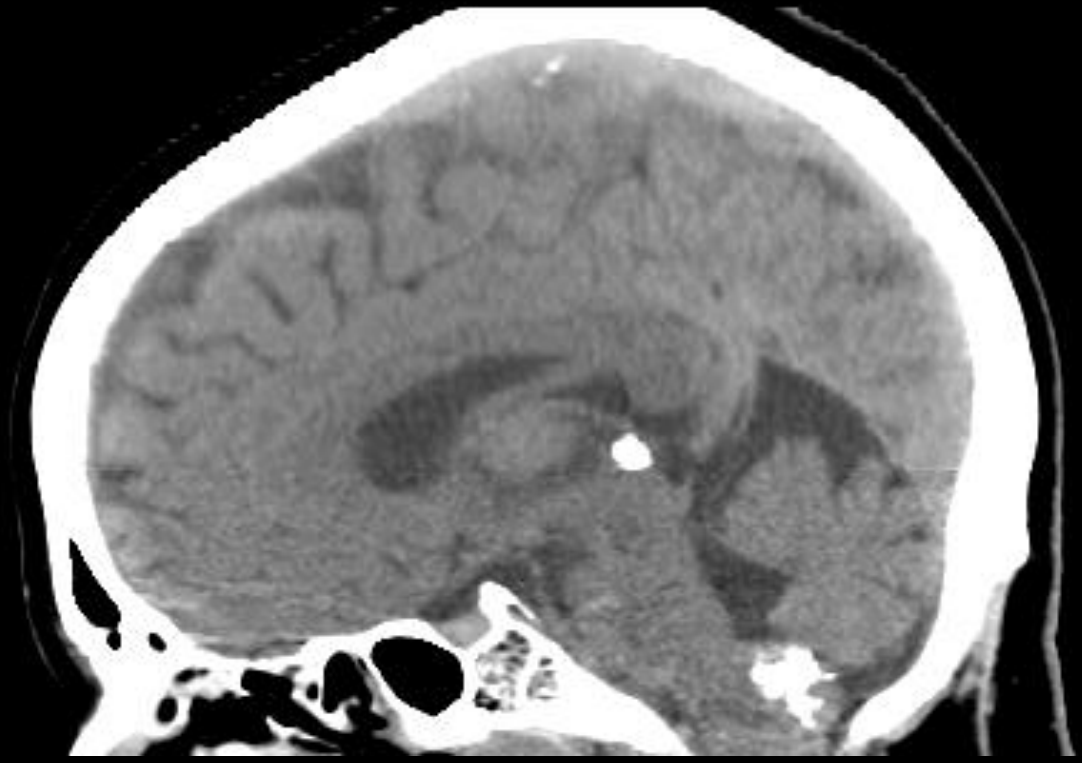
Miscellaneous:

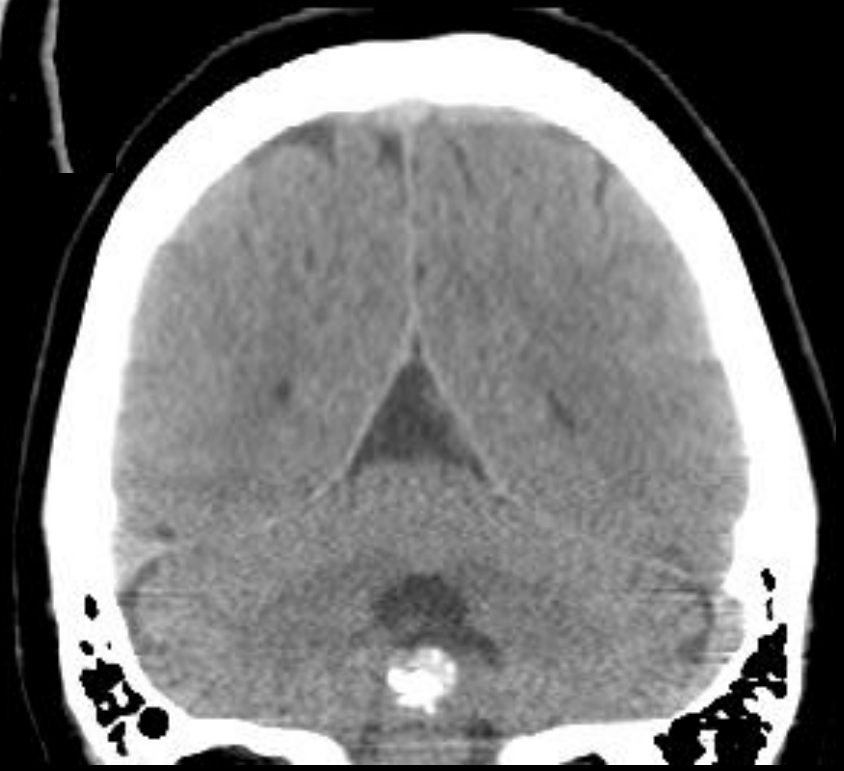
- *Subpendymal giant cell tumor* (history of TS)
- *Chordoid glioma* (rare, uncertain cell origin, classically involving anterior third ventricle and hypothalamus)
- *Rosette-forming glioneuronal tumor* (rare, WHO grade I, mostly in fourth ventricles)

Case 1

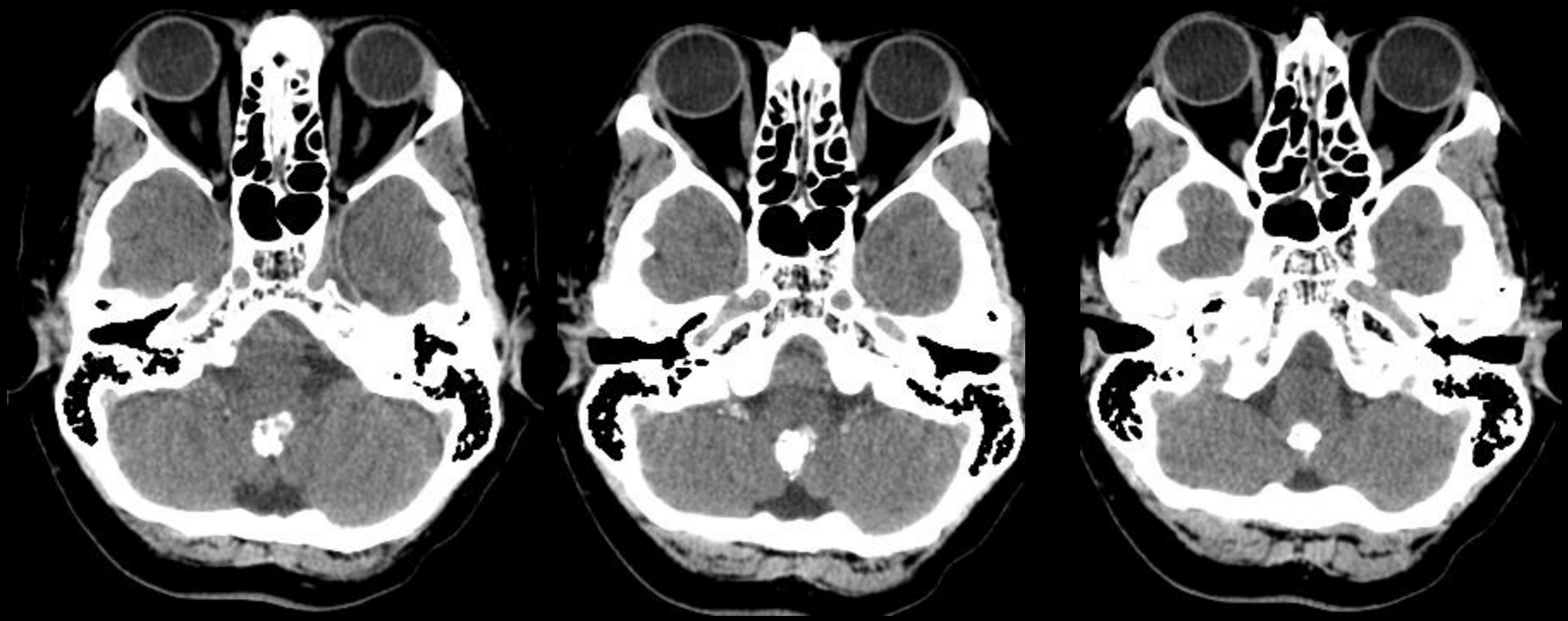
56-year-old female with history of migraine who woke up on the day of presentation with headache that is different from her usual migraine headache, described as posterior and bilateral, accompanied with nausea.

Case 1

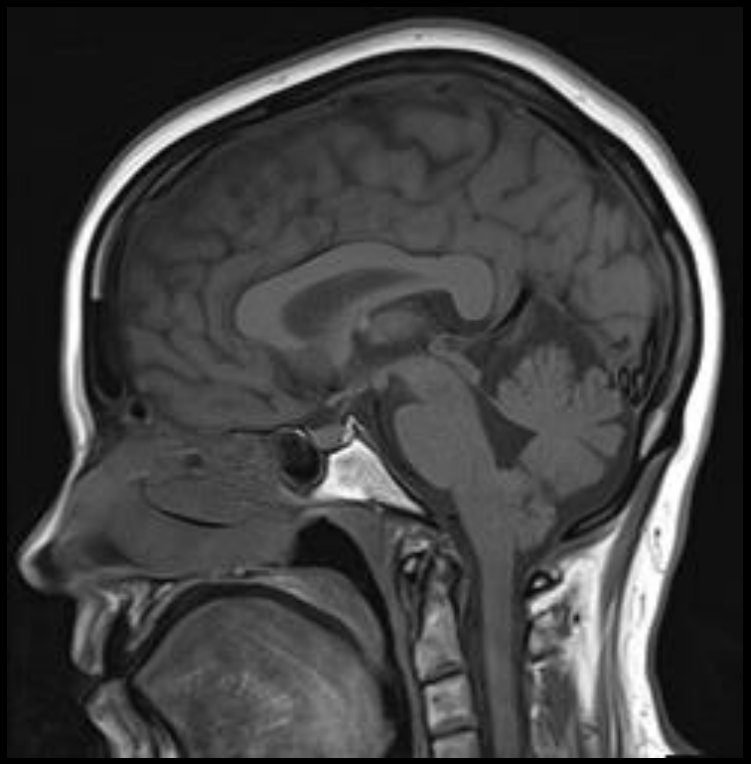




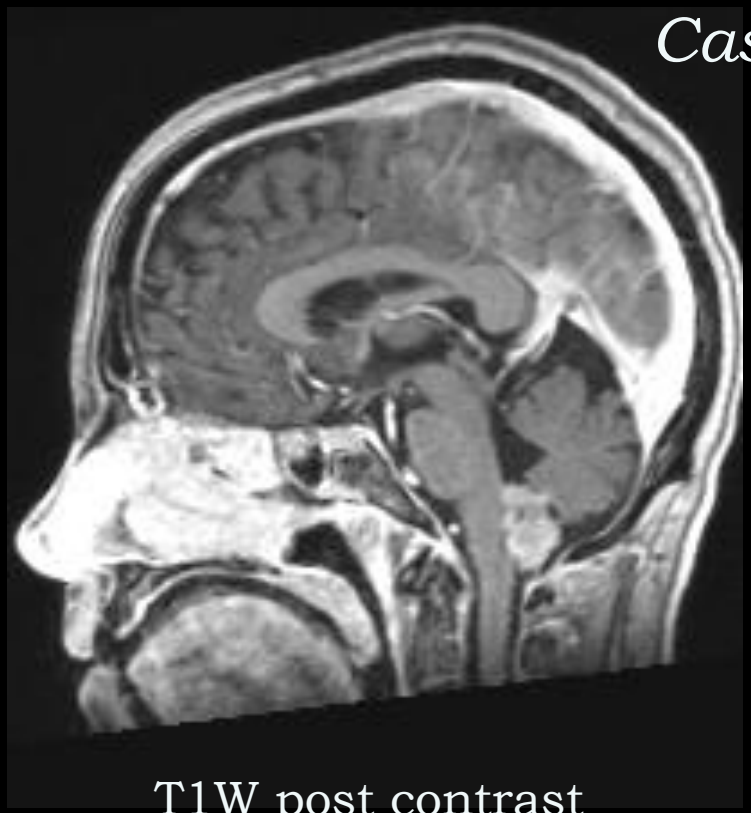
Case 1



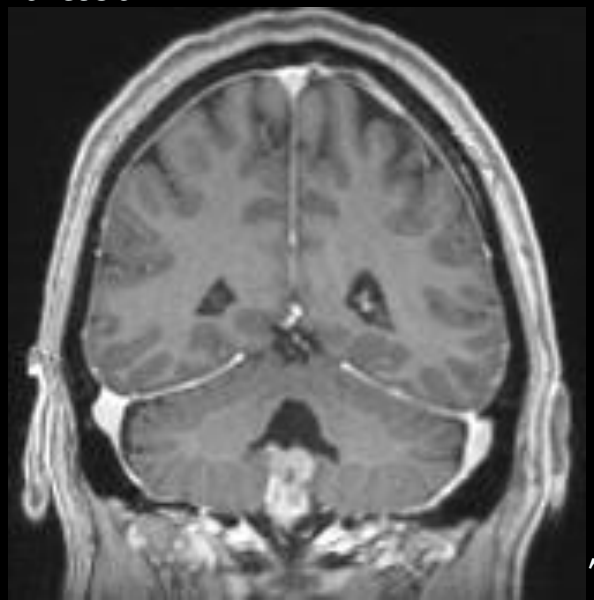
Case 1



T1W pre contrast

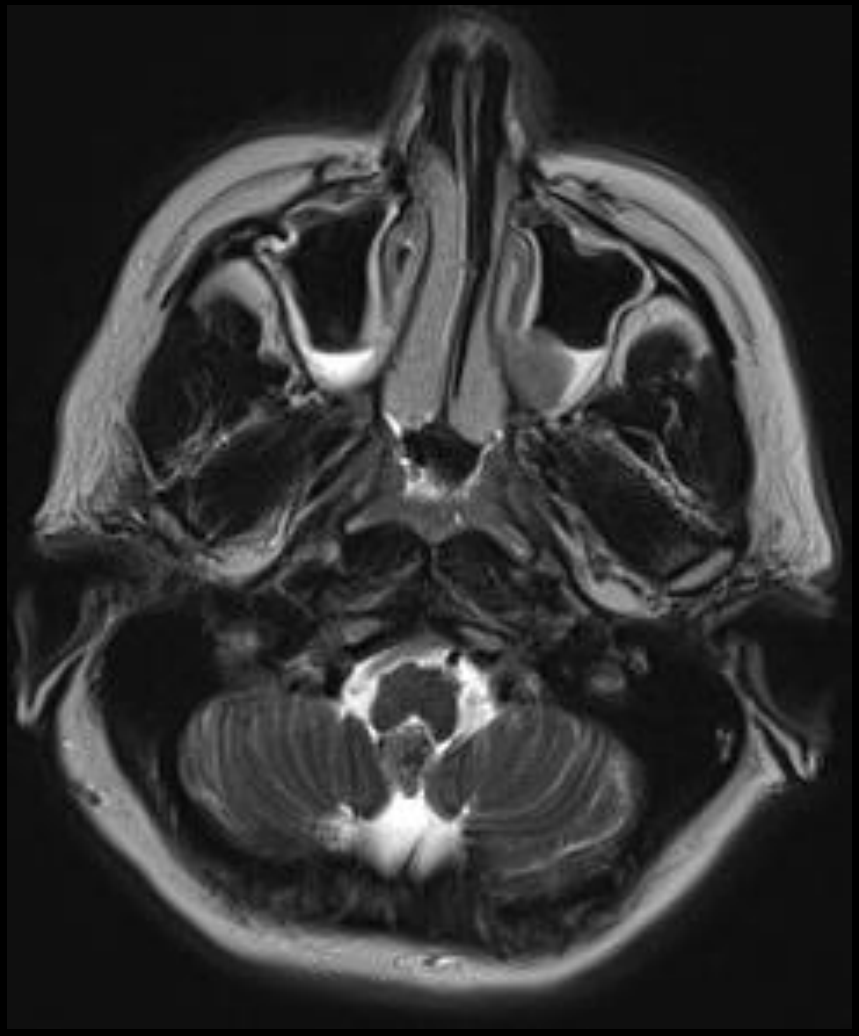


T1W post contrast

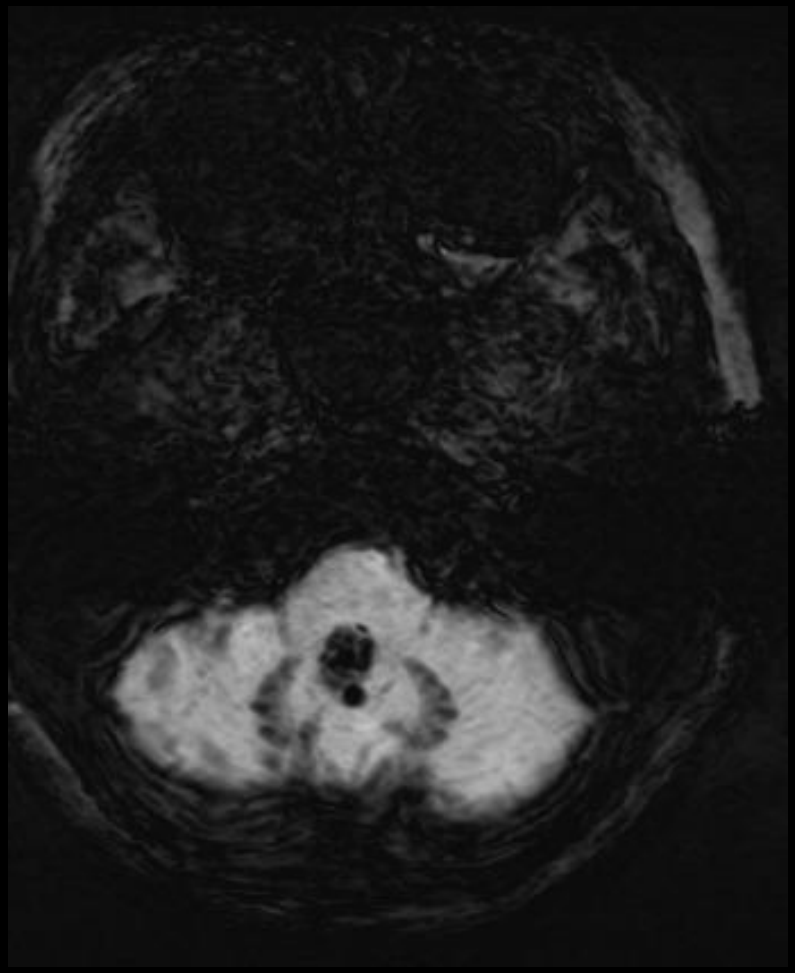


T1W post contrast

Case 1

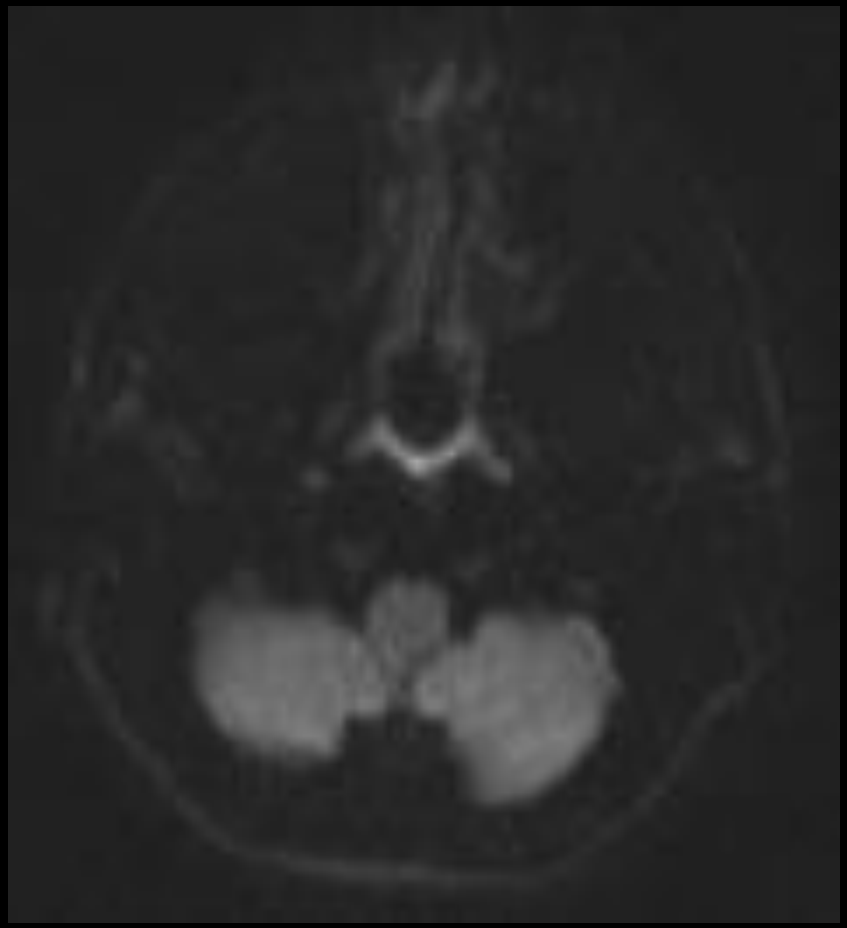


T2W

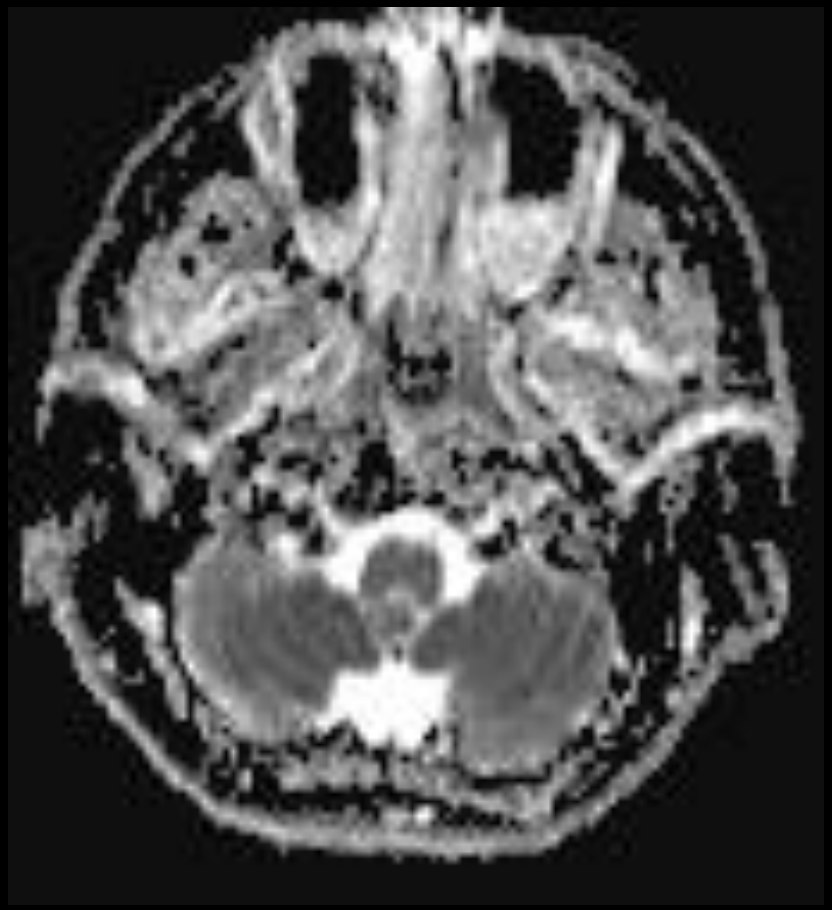


GRE

Case 1



DWI



ADC

Case 1: Summary of findings

- Noncontrast CT head:

  - Heterogeneous, partially calcified mass arising from the floor of the fourth ventricle and extending out into the foramen of Magendie without evidence of hydrocephalus.

- MRI brain:

  - Enhancing mass inferior to the fourth ventricle, in the midline.

  - Susceptibility associated with this lesion likely represents calcification.

  - Foci of restricted diffusion within this lesion.

Case 1: Differential diagnosis

Based on cell origins:

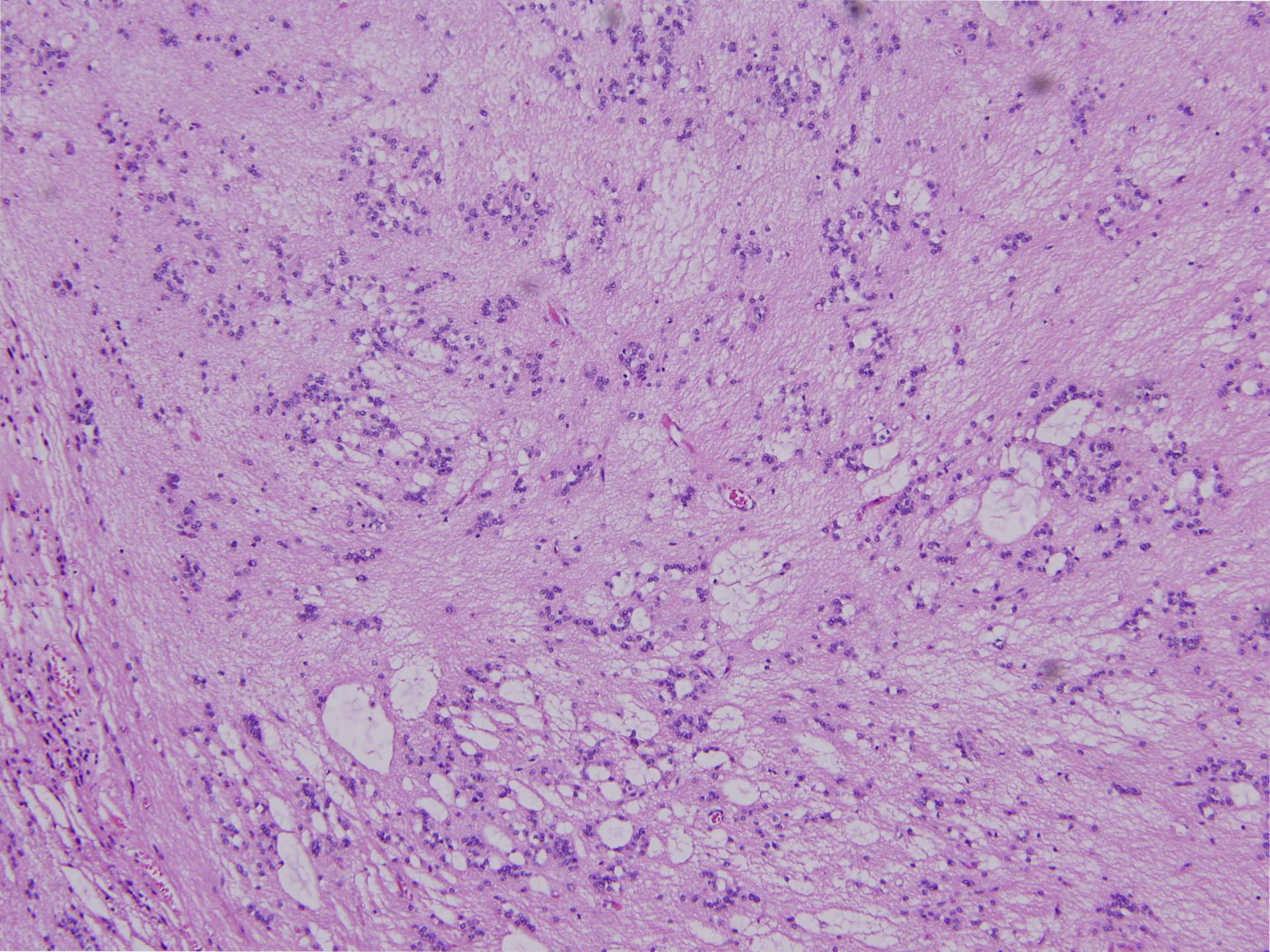
- *Ependymoma*
- *Subependymoma*
- *Choroid plexus neoplasms*
- *Central neurocytoma*
- Meningioma
- *Metastasis*

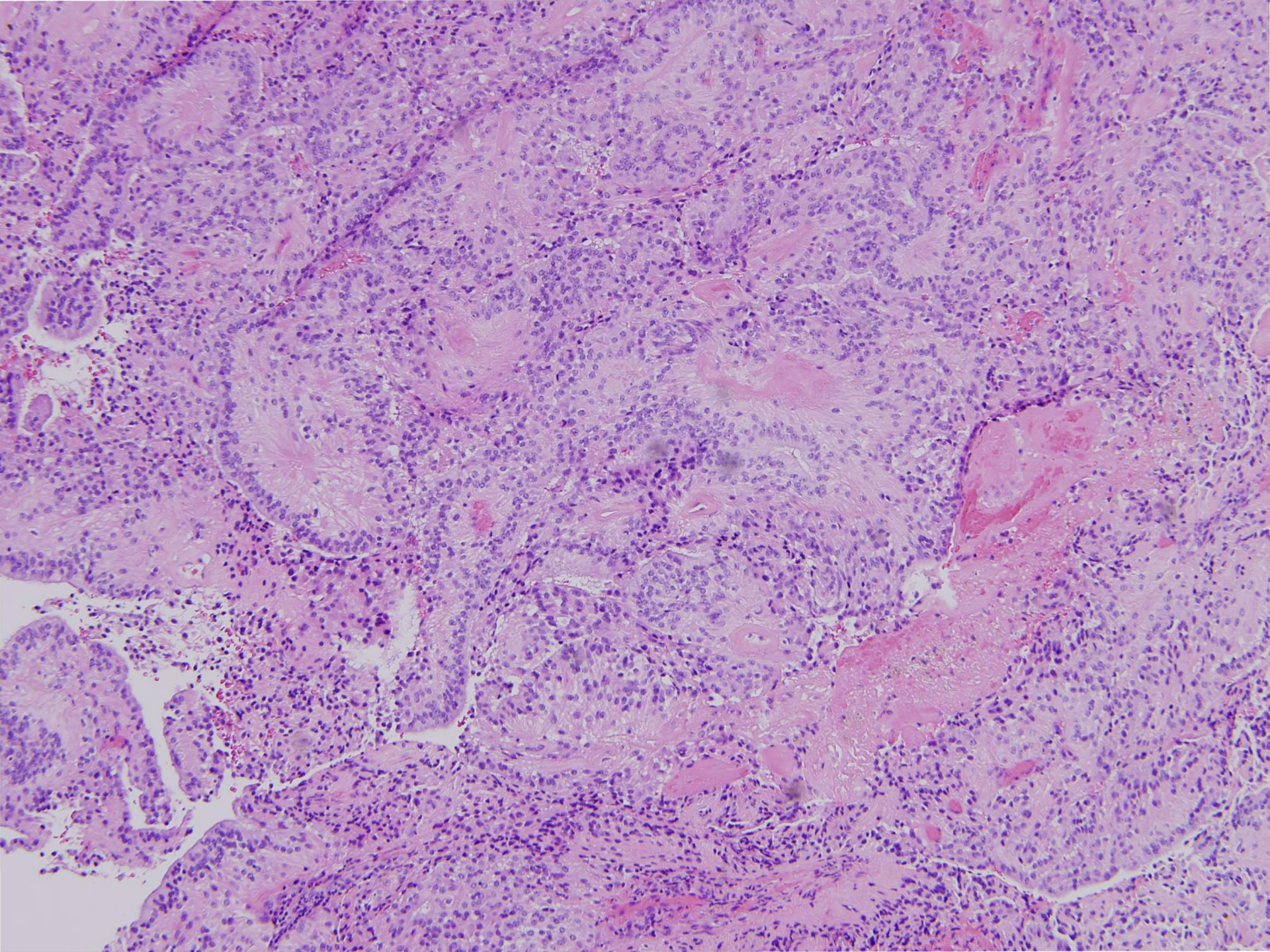
Miscellaneous:

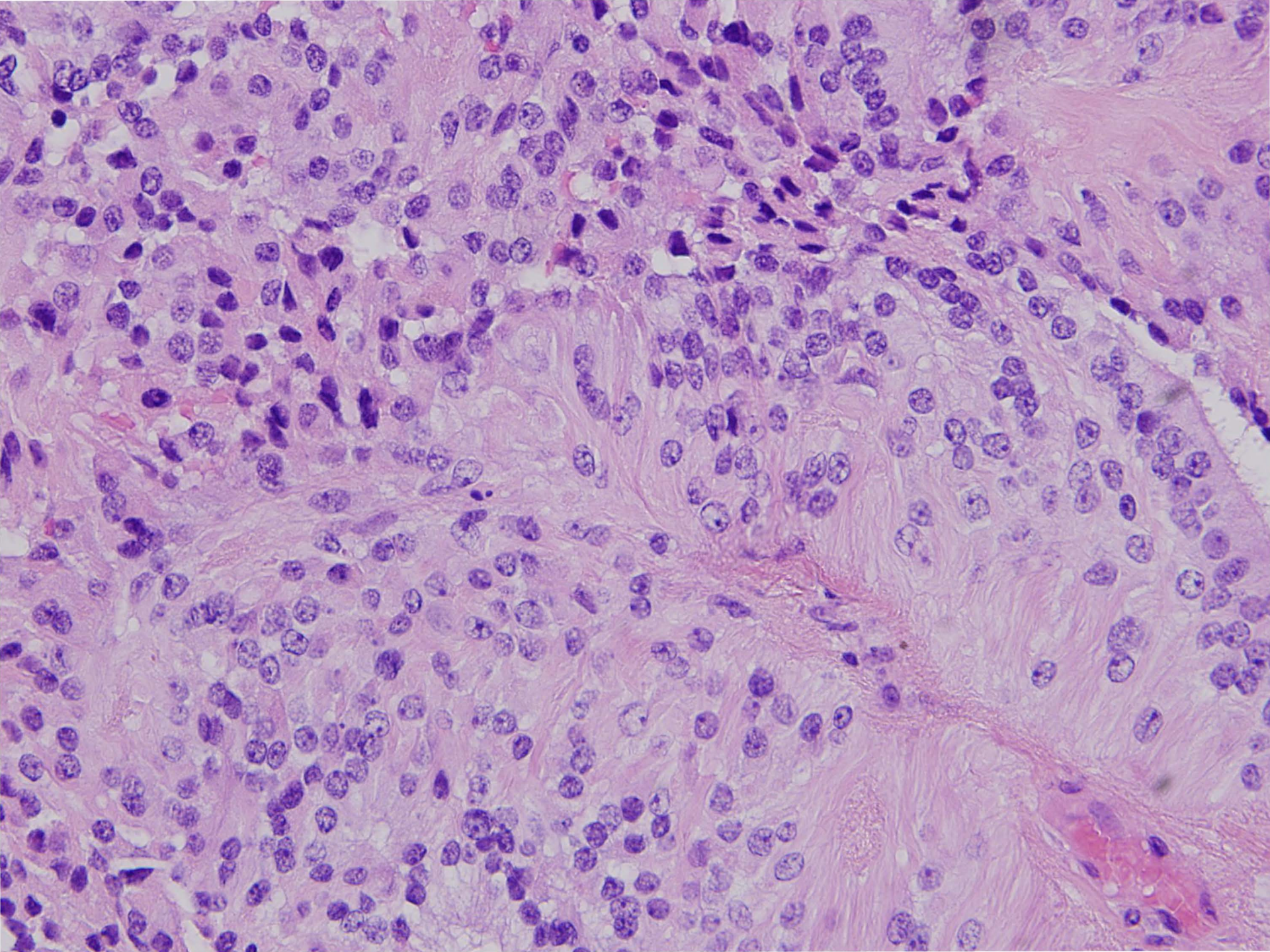
- *Subpendymal giant cell tumor*
- *Chordoid glioma*
- *Rosette-forming glioneuronal tumor*

Case 1: Pathology

- MRN 28107217, BS-13-16615







Ependymoma

SPECIMEN DESIGNATED "4TH VENTRICULAR MASS"

- EPENDYMOMA, W.H.O. Grade II

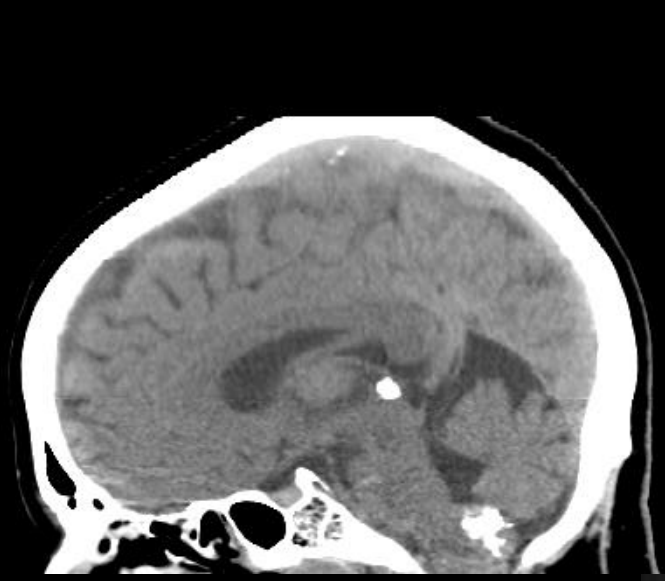
- The tumor involves choroid plexus.
- The estimated MIB-1 proliferation index is 3% (block A3).
- The PAS stain demonstrates the pseudorosette architecture.
- The Reticulin stain highlights the vascular structures.

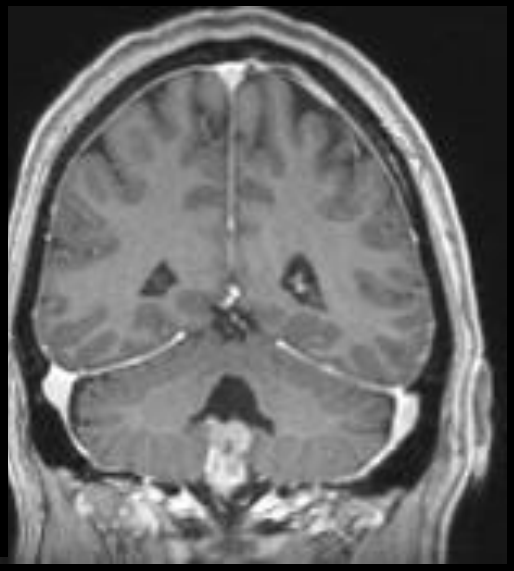
- Immunohistochemistry performed at BWH demonstrates the following staining profile in lesional cells:

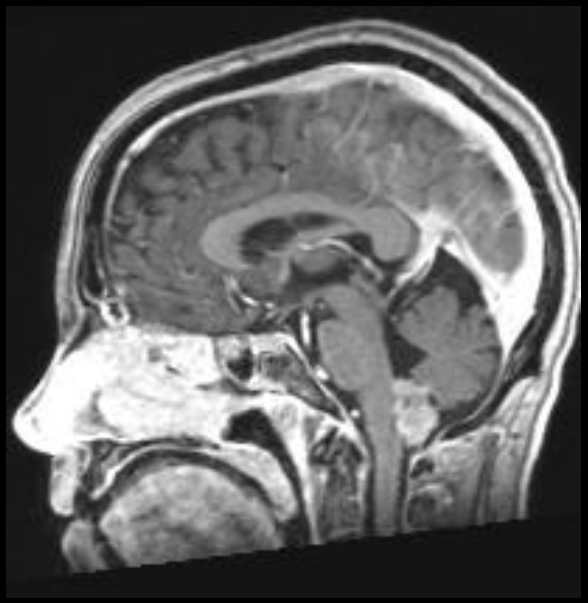
- Positive - GFAP, AE1/AE3 (focal)
- Negative - Olig2

Case 1: Discussion

EPENDYMOMA





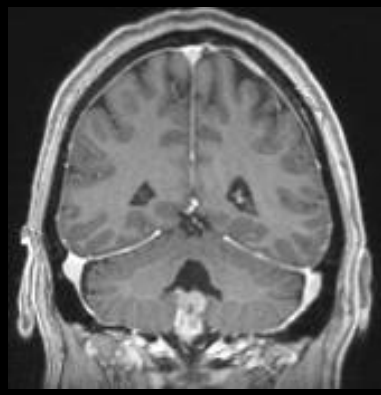


Ependymoma: Clinical features

- Arises from the ependymal cells, WHO grade II - III
- 3-5 % of intracranial neoplasms
- 60% infratentorial, 40% supratentorial (most of supratentorial ependymomas are intraparenchymal)
- Infratentorial ependymomas are more common in children (mean age, 6 years)
- Supratentorial ependymomas, mean age, 18-24 years
- Presenting symptoms:
    - -Infratentorial: increased intracranial pressure, ataxia, paresis
    - -Supratentorial: headache, focal neurologic deficit, seizure
- 5-year survival rate 50-75%, total resection is often difficult due to infiltrating nature
- Small risk of CSF spread → imaging of entire neuroaxis

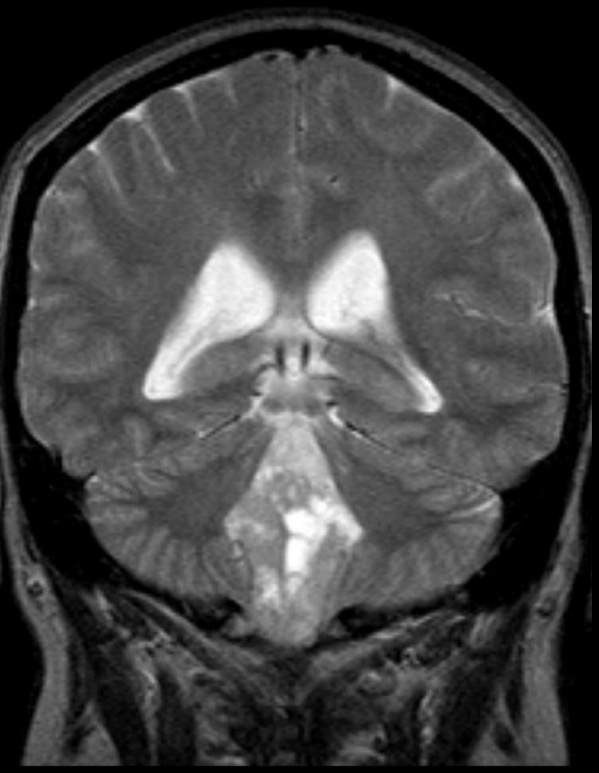
Ependymoma: Imaging features

- Frequently demonstrate cystic components and calcifications, occasional intratumoral hemorrhage
- Intraventricular lesions may extend into adjacent brain, causing vasogenic edema
- Tumors in 4th ventricle tend to fill the ventricle—like a plaster cast or toothpaste—and may extend through the foramen of Luschka, Magendie and magnum
- CT: hypo- to isoattenuating, calcifications
- MR:
  - Iso- to hypointense on T1W, iso- to hyperintense on T2W
  - Heterogeneous enhancement
  - Blooming if calc or hemorrhage present
  - Restricted diffusion may be seen in soft tissue component due to higher cellularity

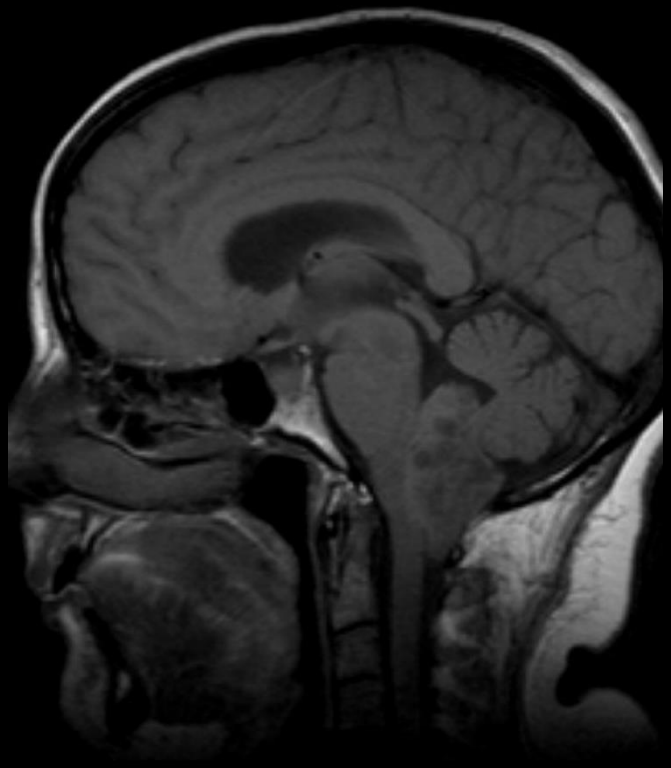


Ependymoma: Imaging features

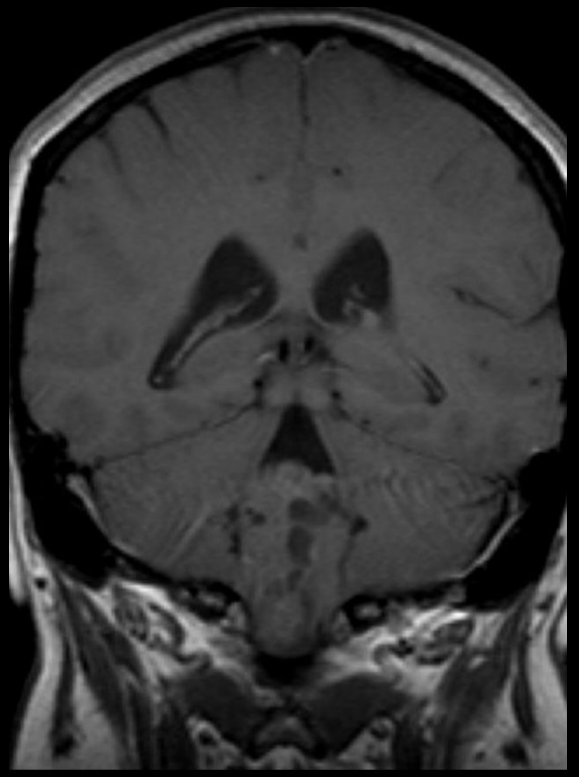
Companion Case



T2W



T1W

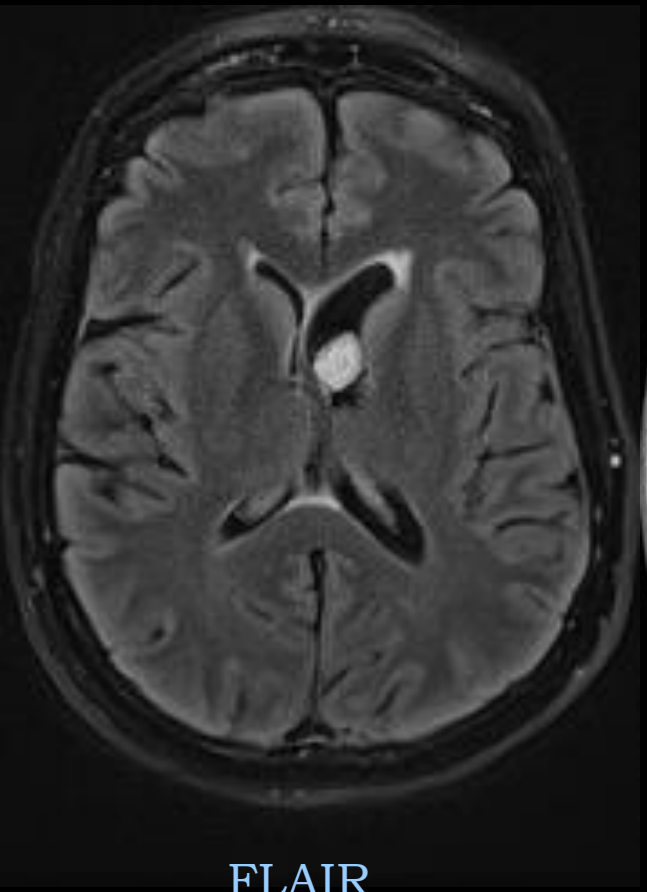


T1W

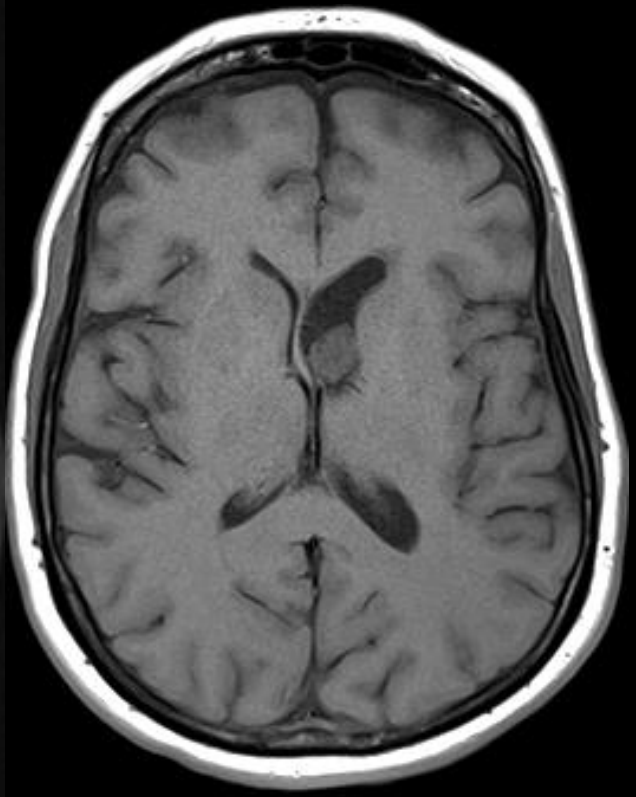
Case 2

52-year-old female with known intraventricular mass first noted 5 years ago now presenting with several weeks of positional headache (worse when lying down and with Valsalva), dizziness, lack of coordination and “wobbly” gait.

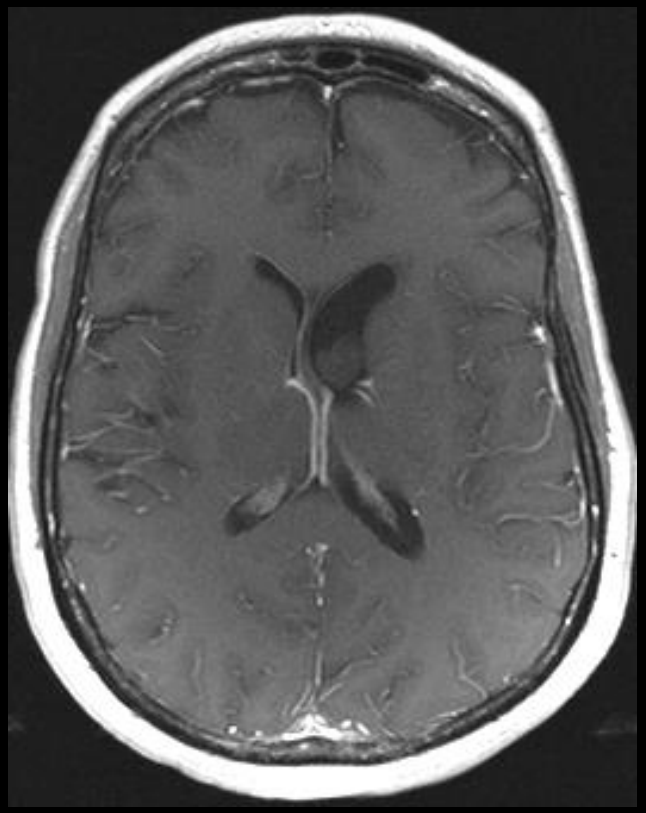
Case 2



FLAIR

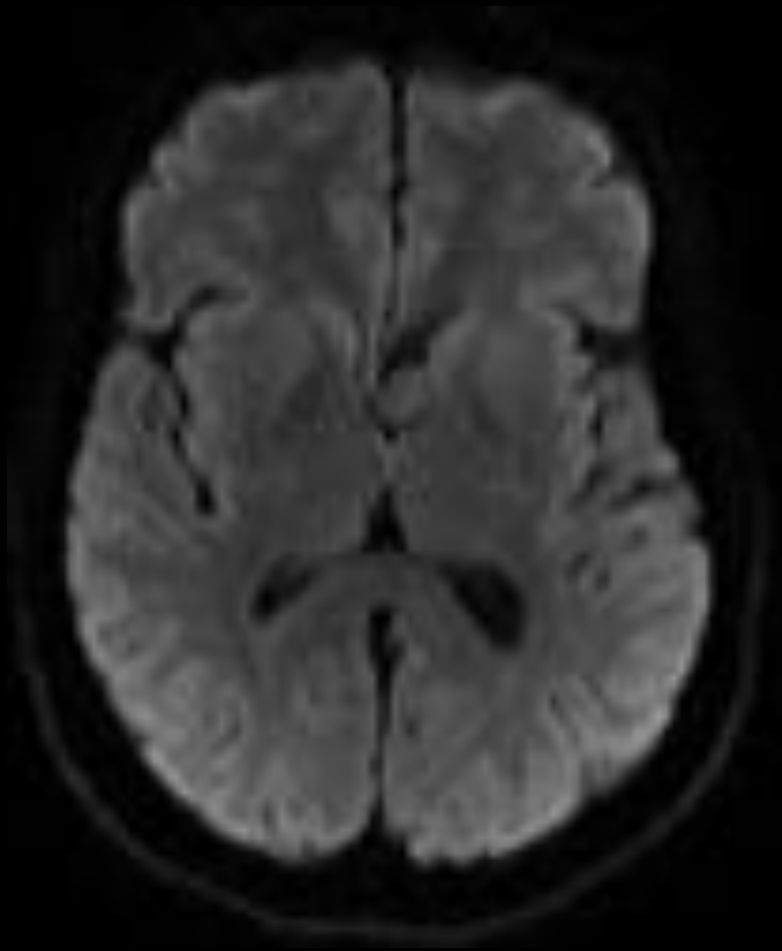


T1W pre contrast

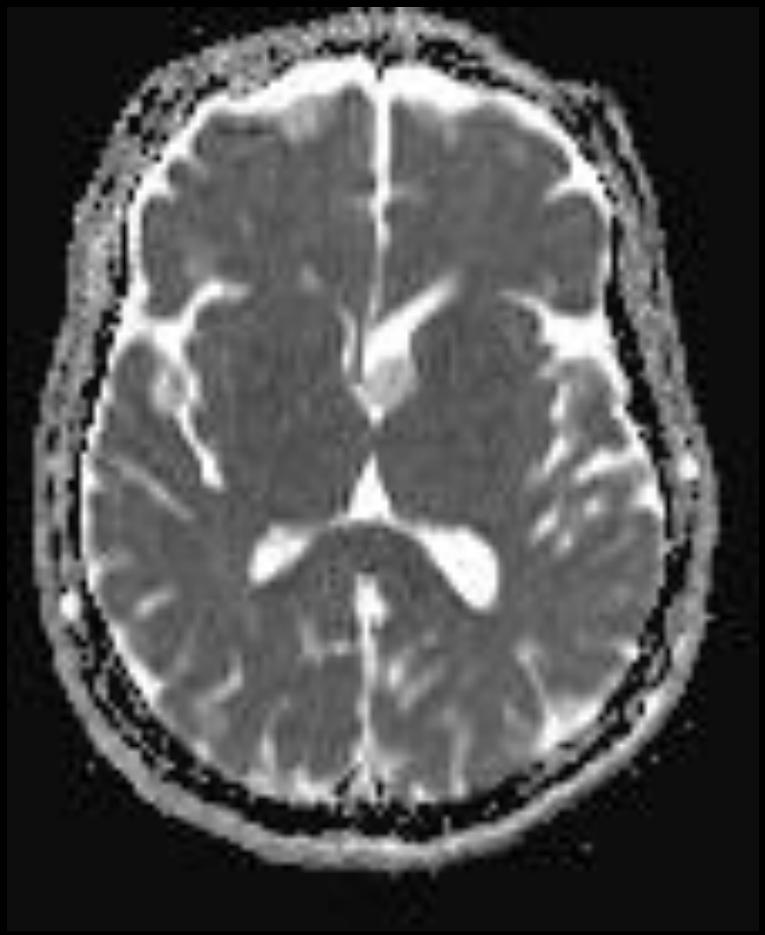


T1W post contrast

Case 2



DWI



ADC

Case 2: Summary of findings

- MRI brain:

  - Non-enhancing, well-circumscribed mass in the left lateral ventricle protruding into the foramen of Monro with mild dilatation of left lateral ventricle.

  - No restricted diffusivity.

Case 2: Differential diagnosis

Based on cell origins:

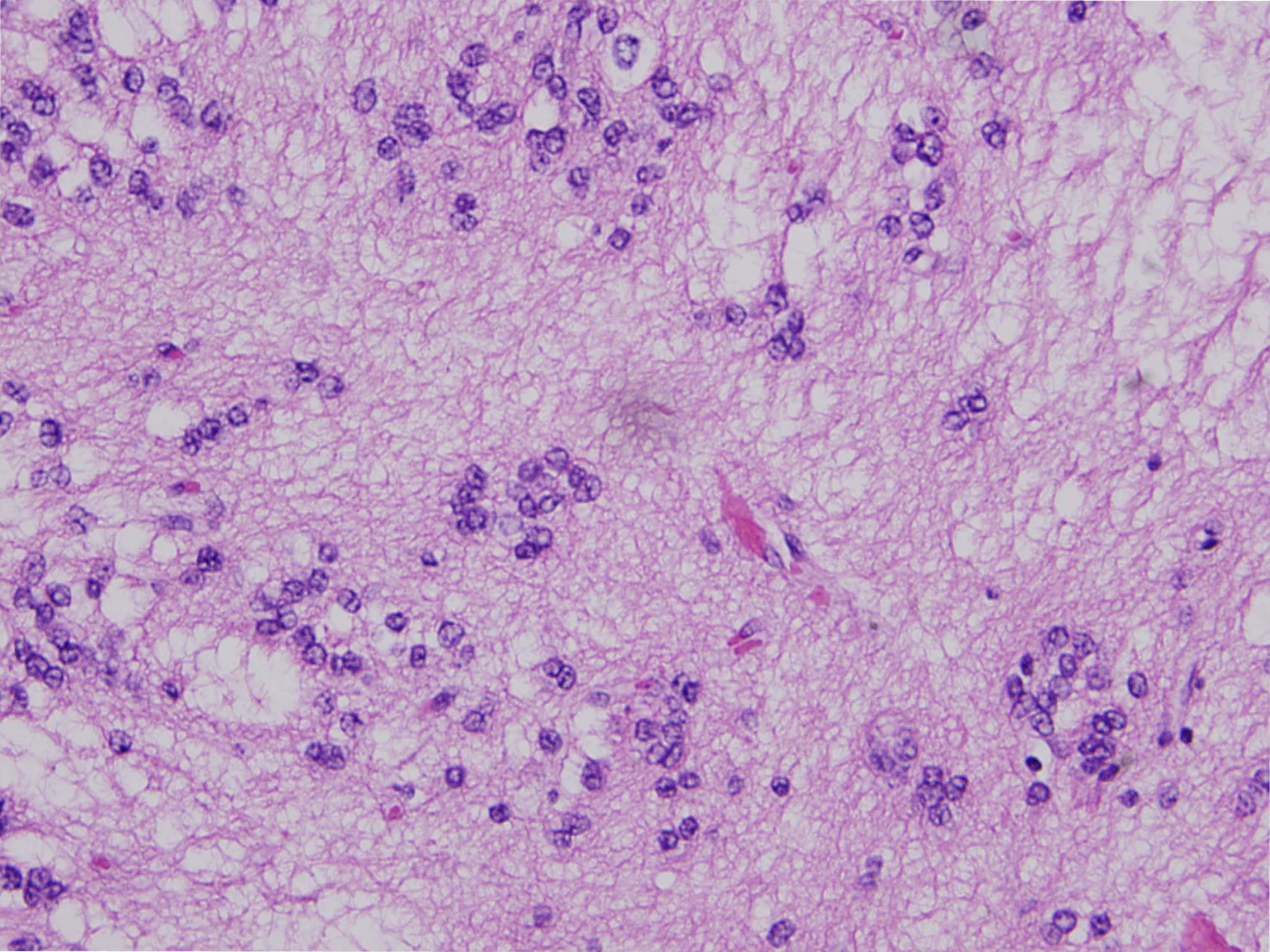
- *Ependymoma*
- *Subependymoma*
- *Choroid plexus neoplasms*
- *Central neurocytoma*
- Meningioma
- *Metastasis*

Miscellaneous:

- *Subpendymal giant cell tumor*
- *Chordoid glioma*
- *Rosette-forming glioneuronal tumor*

Case 2: Pathology

- MRN 19194810, BS-11-35950



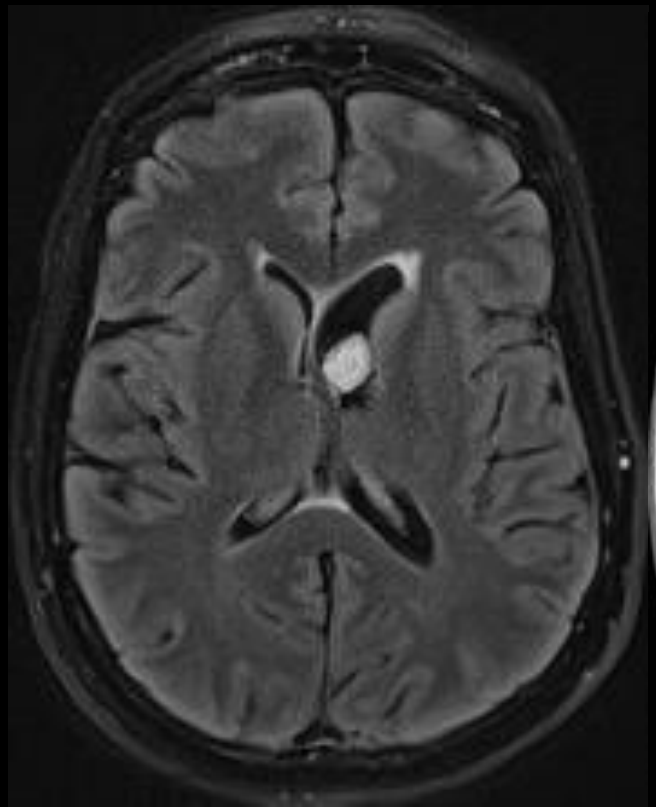
Subependymoma

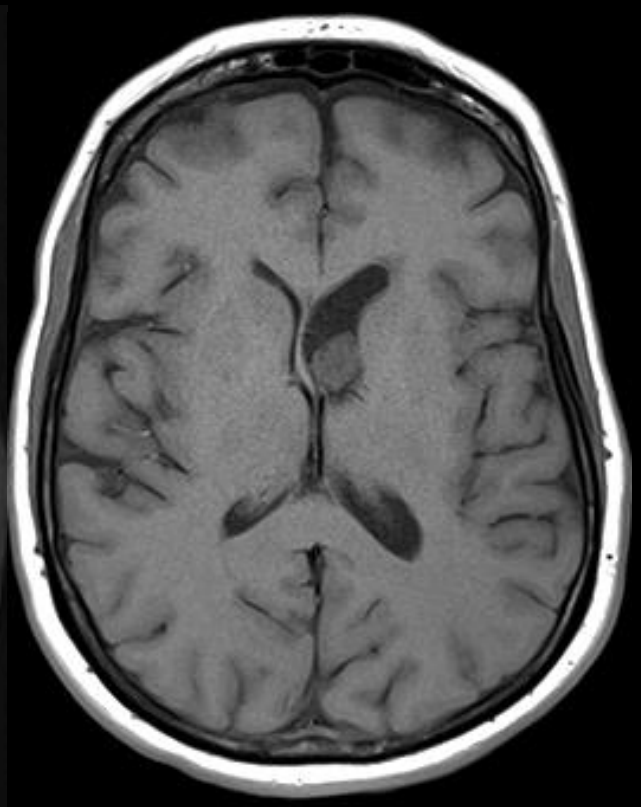
SPECIMEN LABELED "VENTRICULAR MASS":

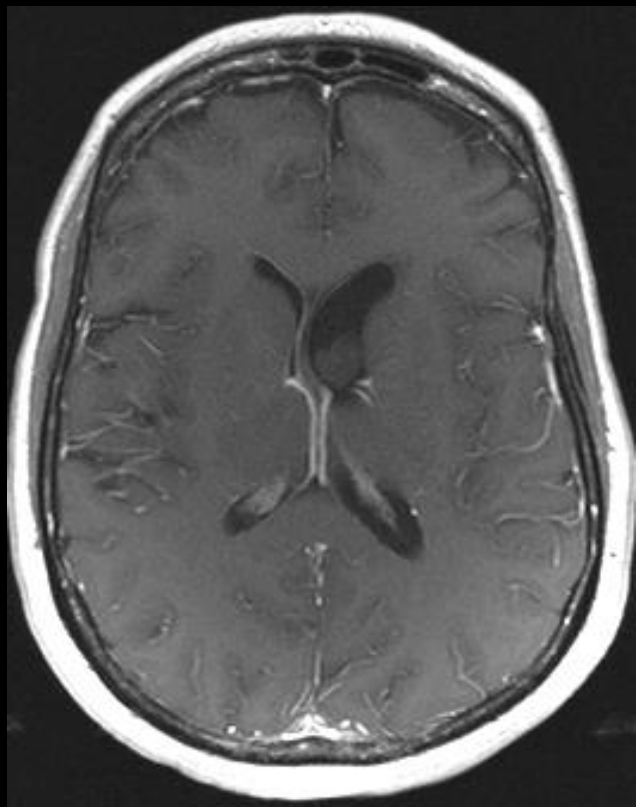
- SUBEPENDYMOMA, W.H.O. Grade I

Case 2: Discussion

SUBEPENDYMOMA





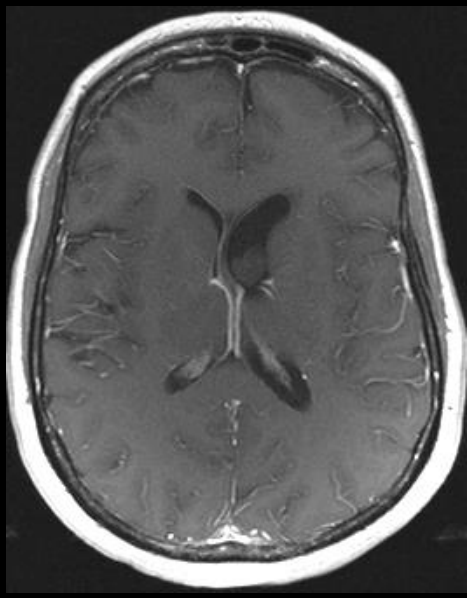


Subependymoma: Clinical features

- Arises from subependymal glial layer, WHO grade I
- 0.2 – 0.7% of intracranial neoplasms
- Most common in 4th ventricle (50-60%) and lateral ventricle (30-40%).
- 82% occur in patients older than 15 years
- Male predominance, 2.3:1 male-to-female ratio
- Presenting symptoms: typically incidental finding in asymptomatic patient but when large can cause symptoms related to increased intracranial pressure
- Good prognosis, rare recurrence

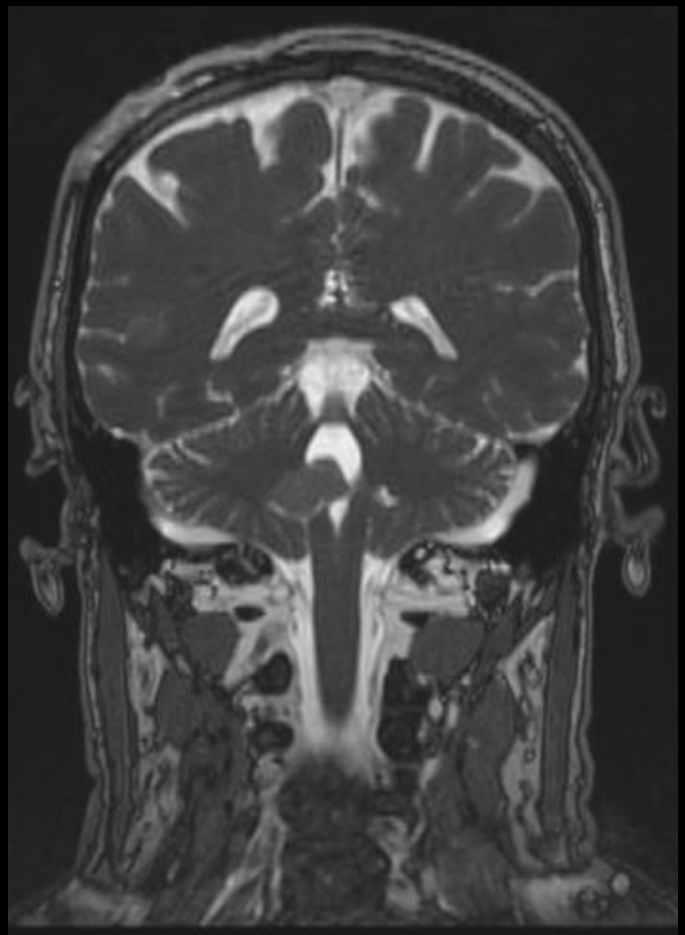
Subependymoma: Imaging features

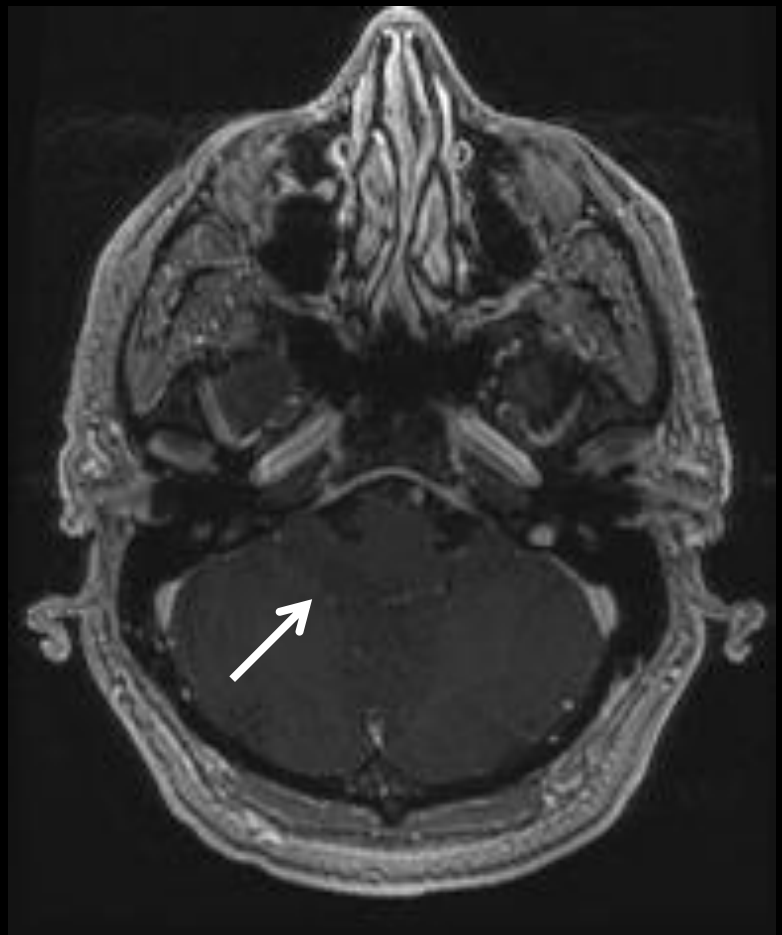
- Cystic degeneration is common, calcifications and hemorrhage may be seen
- No invasion of brain parenchyma, no edema in the adjacent white matter
- No CSF dissemination
- MR:
  - Hypo- isointense on T1W, hyperintense on T2W
  - Variable enhancement but most demonstrate no or minimal enhancement



Subependymoma: Imaging features

Companion Case



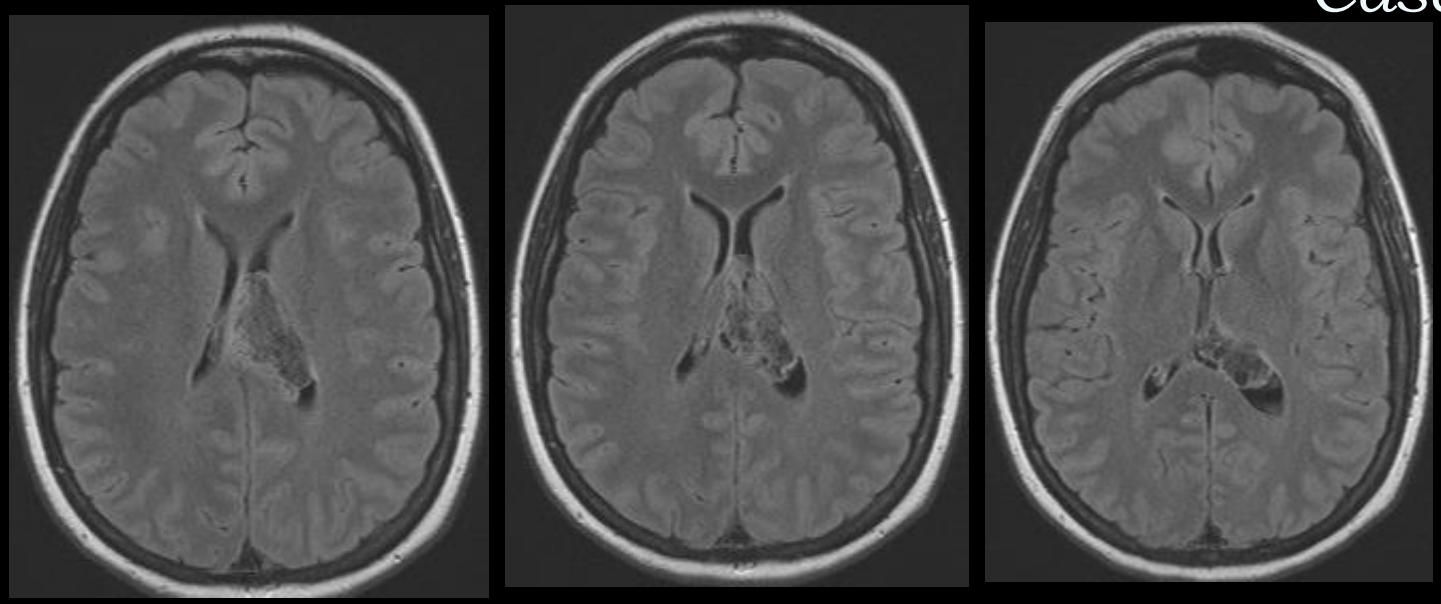


Case 3

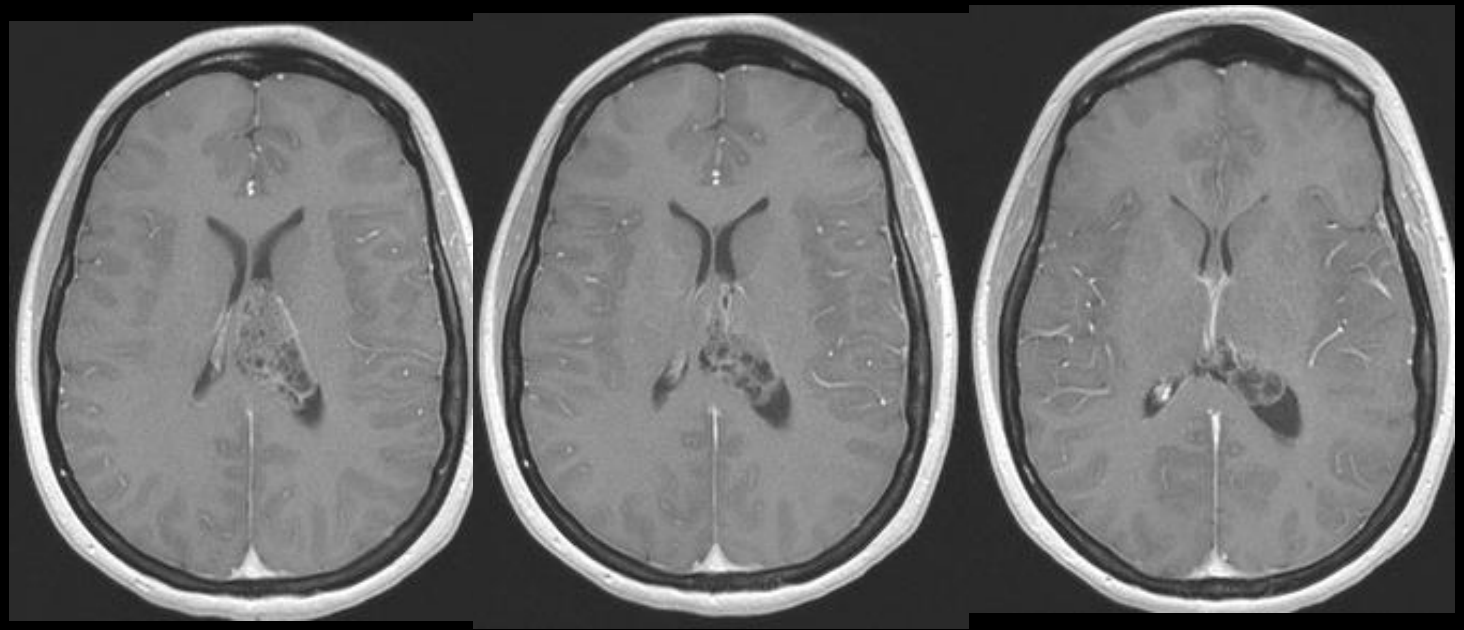
27-year-old female who volunteered for an experimental MRI study

Case 3

FLAIR



T1W post contrast

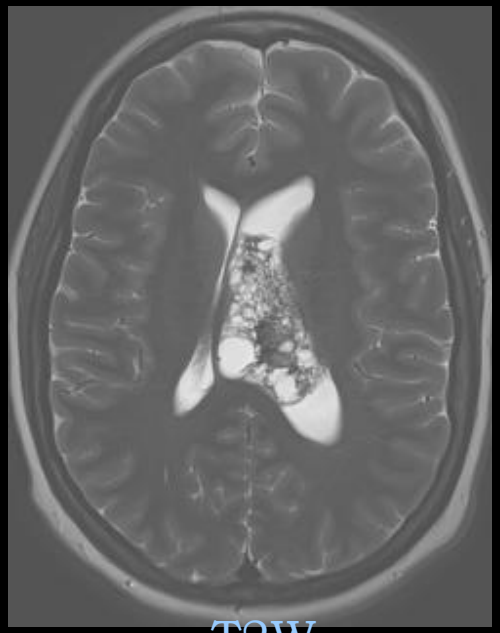


Case 3

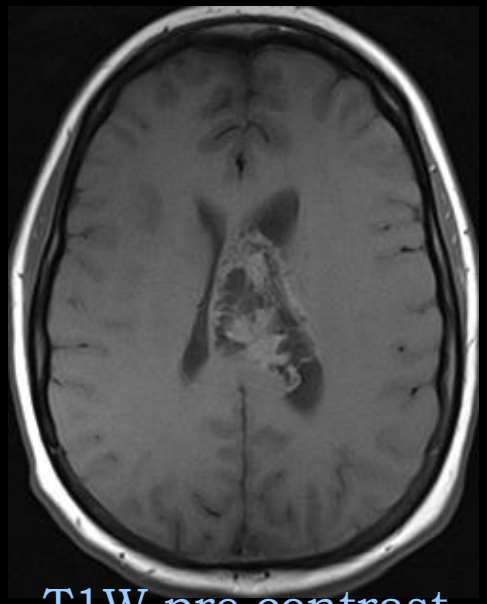
The patient presented for follow-up two years later. Patient reports mild chronic headache and subtle word-finding difficulty.

2 years later

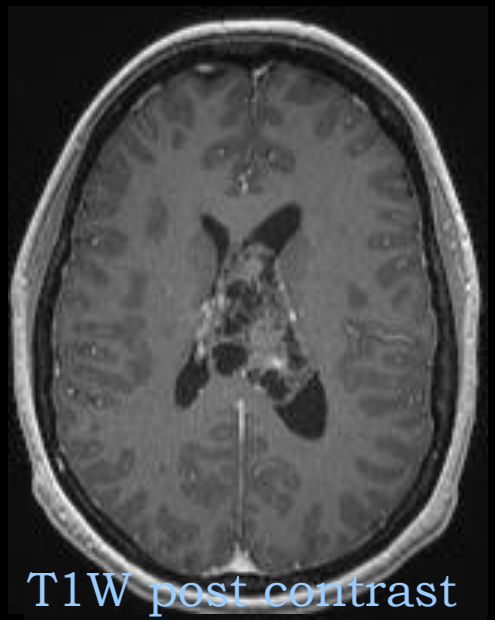
Case 3



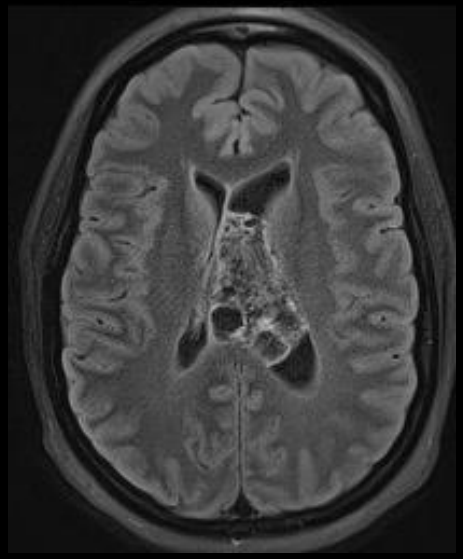
T2W



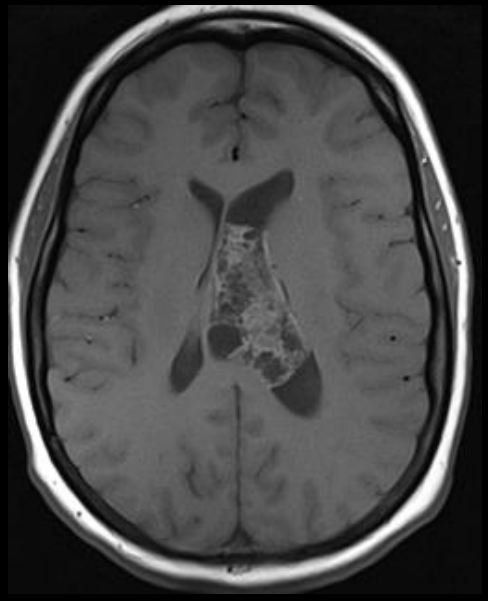
T1W pre contrast



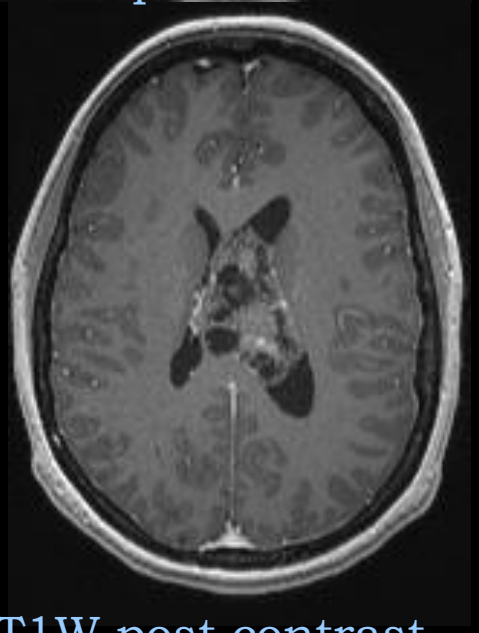
T1W post contrast



FLAIR

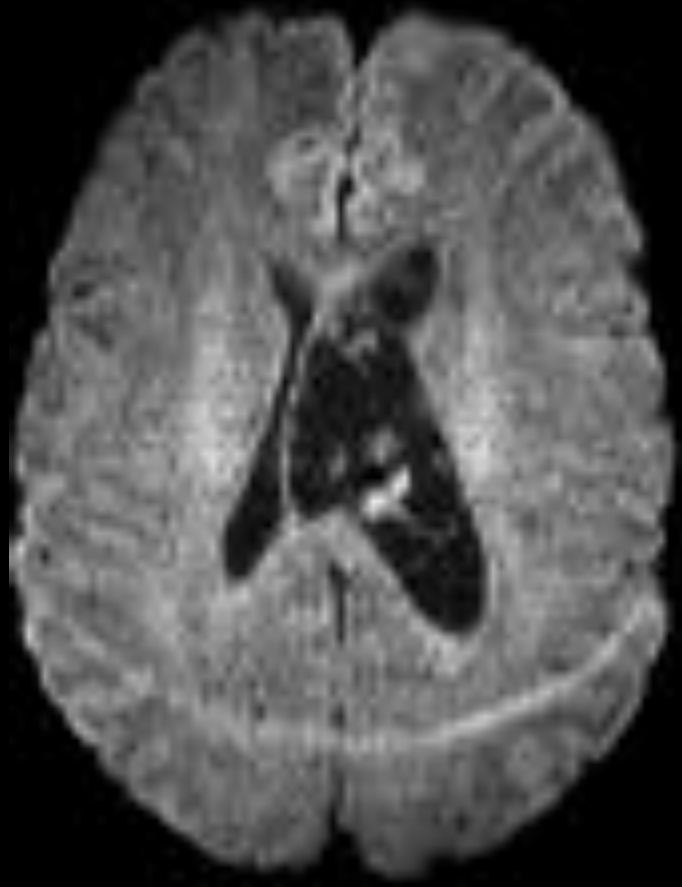


T1W pre contrast

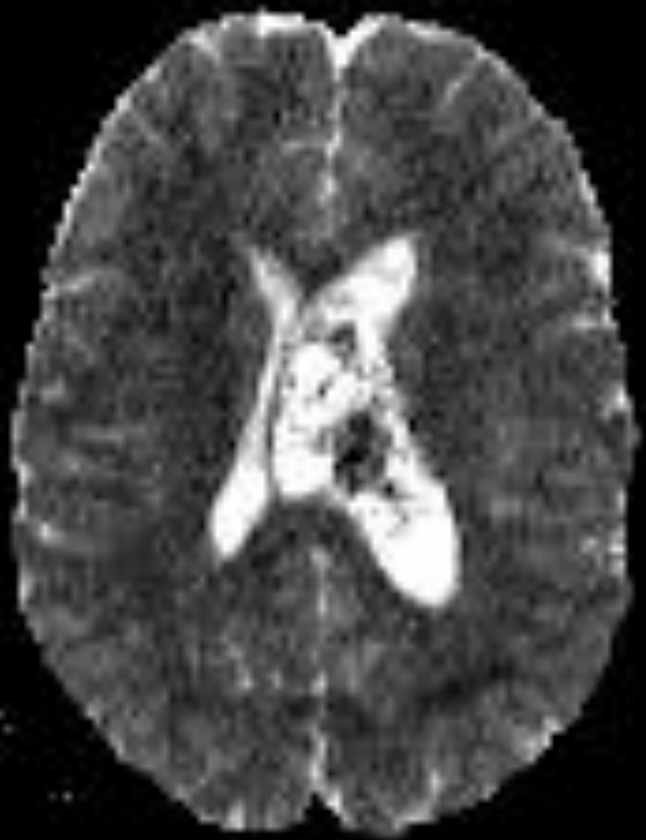


T1W post contrast

Case 3



DWI



ADC

Case 3: Summary of findings

•MRI brain:

-A multiseptated predominantly cystic lesion with some solid component that fills the left lateral ventricle and displaces the septum pellucidum to the right.

- The lesion slightly expands the body of the left lateral ventricle.

- The majority of these cysts have a high diffusivity similar to CSF. The solid components demonstrate lower diffusivity suggesting that these are cellular.

- There is mild enhancement of the solid component.

- The overall size of lesion appears to have enlarged over the past 2 years. Much of the enlargement is related to enlargement of the cystic spaces.

Case 3: Differential diagnosis

Based on cell origins:

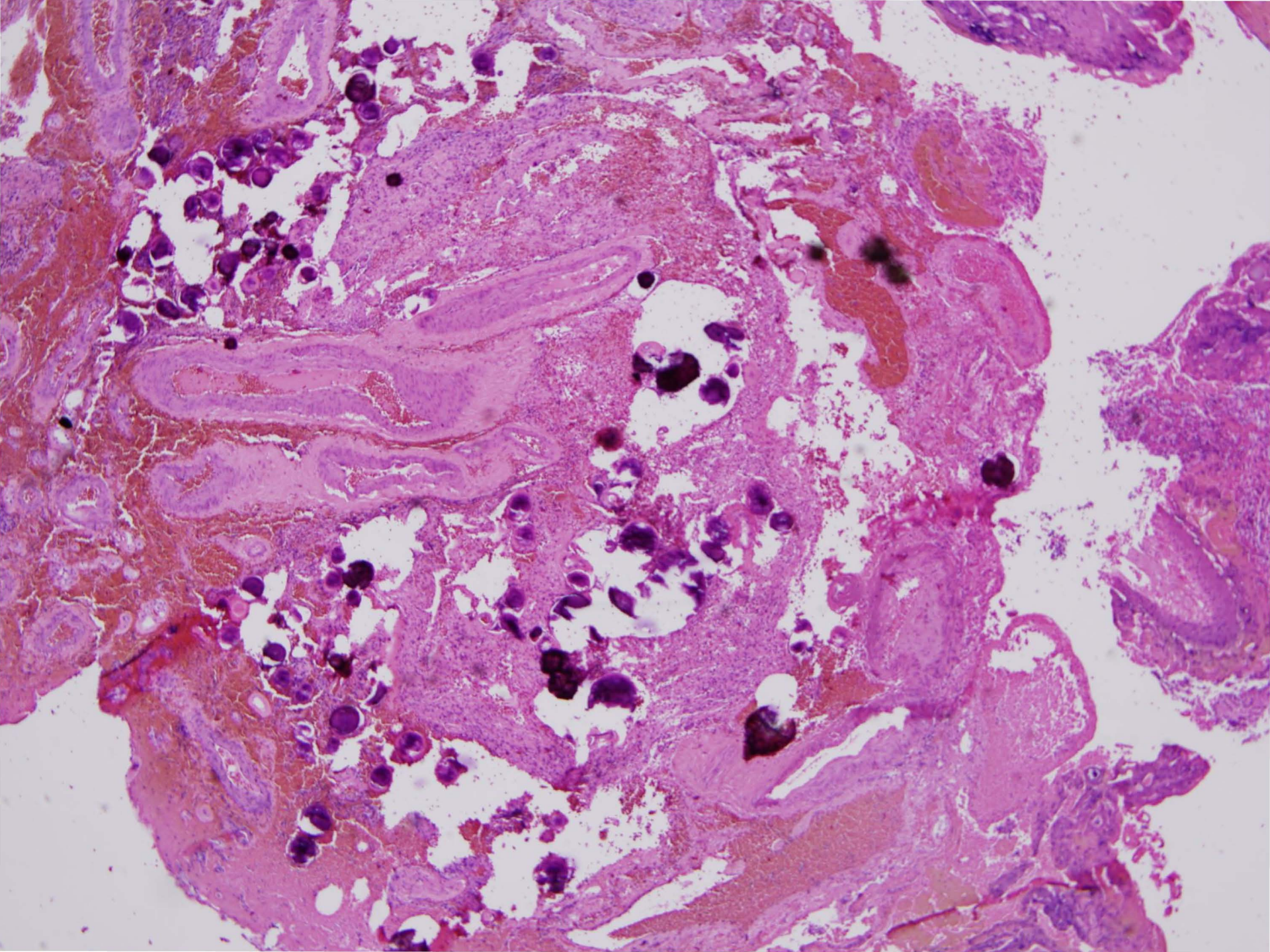
- *Ependymoma*
- *Subependymoma*
- *Choroid plexus neoplasms*
- *Central neurocytoma*
- Meningioma
- *Metastasis*

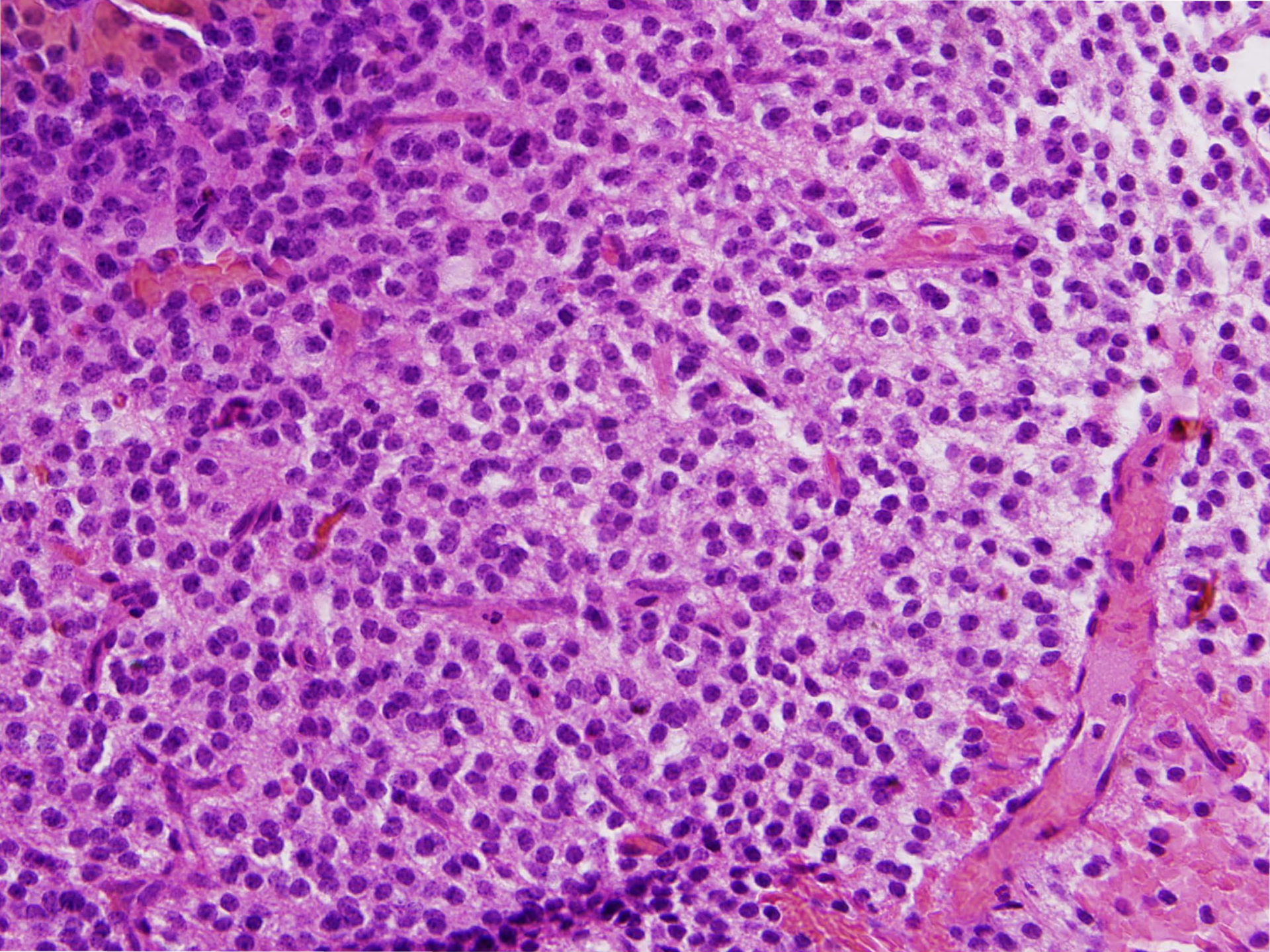
Miscellaneous:

- *Subpendymal giant cell tumor*
- *Chordoid glioma*
- *Rosette-forming glioneuronal tumor*

Case 3: Pathology

- MRN 24142325 , BS-09-59936





Central Neurocytoma

SPECIMEN LABELED “Intraventricular tumor”,:

- - CENTRAL NEUROCYTOMA, WHO GRADE II, ICD-O 9506/1. See NOTE.

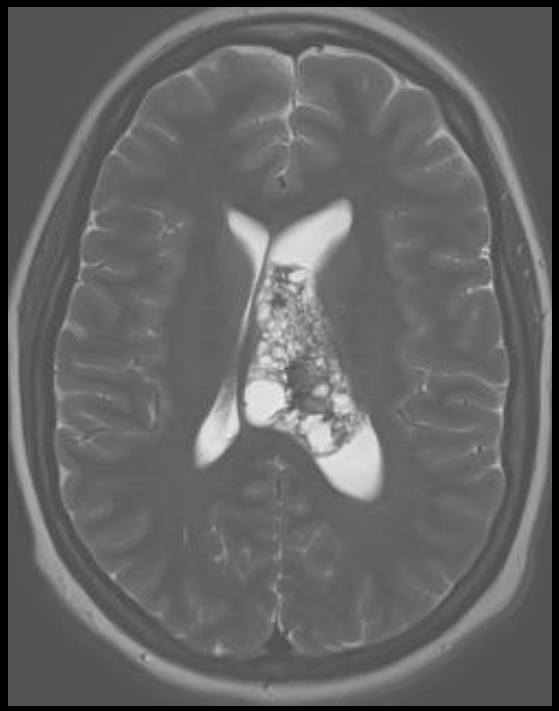
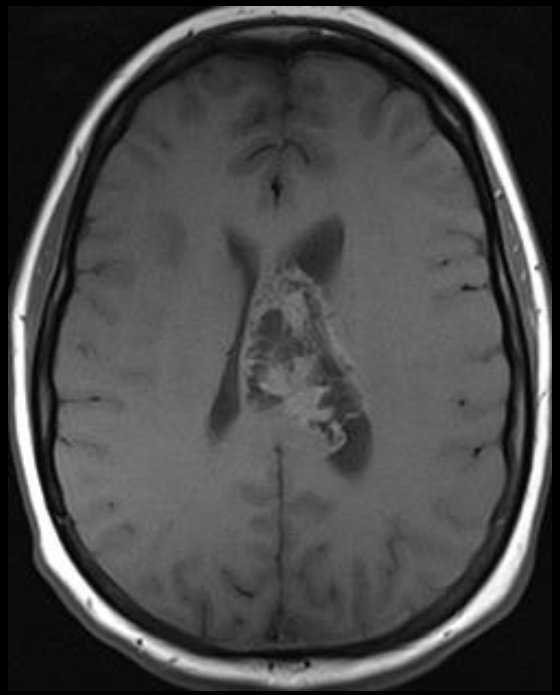
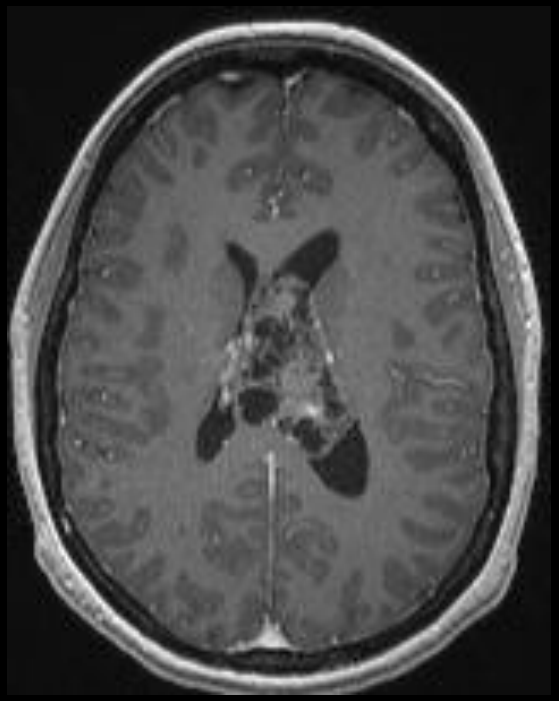
NOTE:

- The tumor is composed of uniform, round cells embedded in a fine fibrillary background. Mitosis, necrosis and microvascular proliferation are not observed.
- Immunohistochemistry performed in the BWH shows following staining pattern in the tumor cells:
- GFAP: Negative
- Synaptophysin – Positive
- Neu N – Focally positive

- Proliferative activity (MIB-1) is estimated 2-4 %

Case 3: Discussion

CENTRAL NEUROCYTOMA

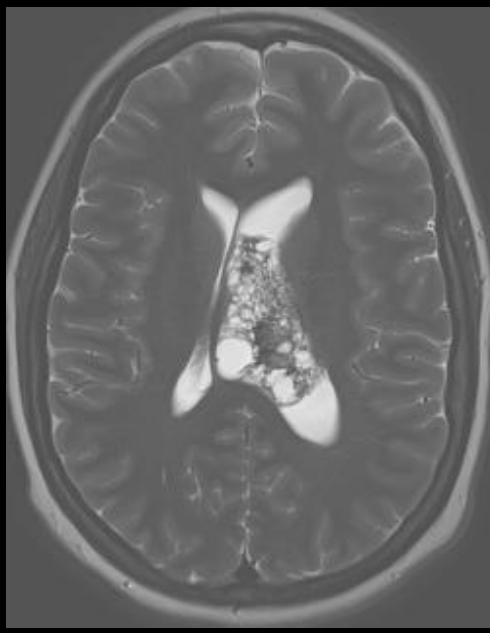




Central neurocytoma: Clinical features

- Arises from progenitor cells, WHO grade II
- 0.25 – 0.5% of intracranial neoplasms
- Occurs in lateral ventricle, arising from septum pellucidum or ventricular wall
- With or without extension into the 3rd ventricle
- Mean age 29 years, wide age range
- Presenting symptoms from increased intracranial pressure
- Gross total resection is usually curative

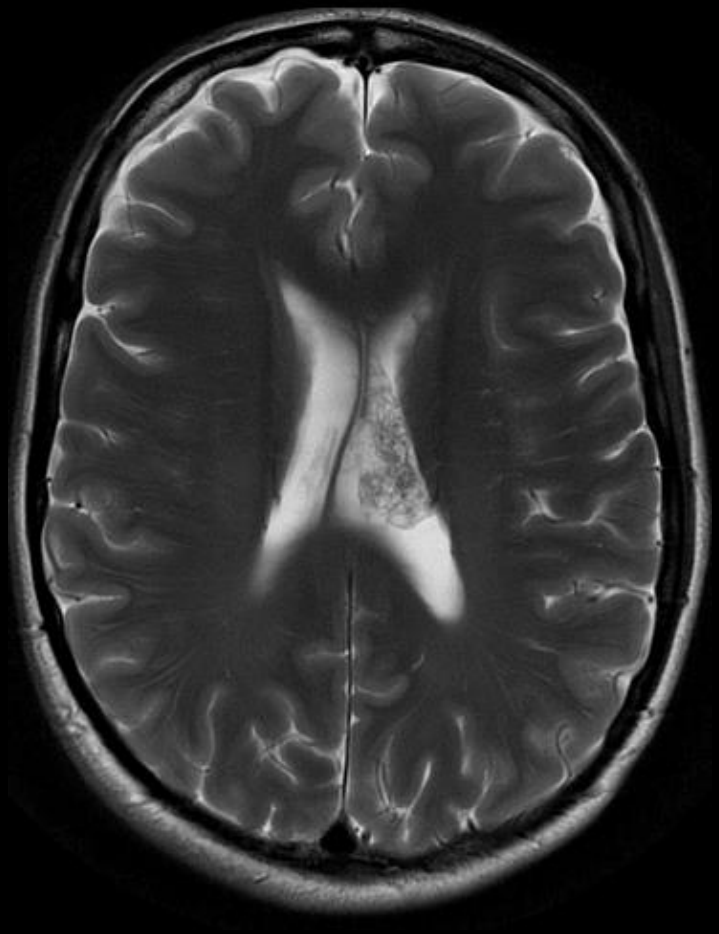
Central neurocytoma: Imaging features

- Well-circumscribed, lobulated mass that frequently have cyst-like areas— "bubbly appearance" arising from septum pellucidum or ventricular wall of the lateral ventricle
- 50% calcifications, rare hemorrhage
- MR:
  - Isointense on T1W, hyperintense on T2W
  - Variable enhancement, moderate to strong typically seen
  - Increased T2 intensity in adjacent white matter may be seen
- MR spectroscopy often reveals the presence of glycine

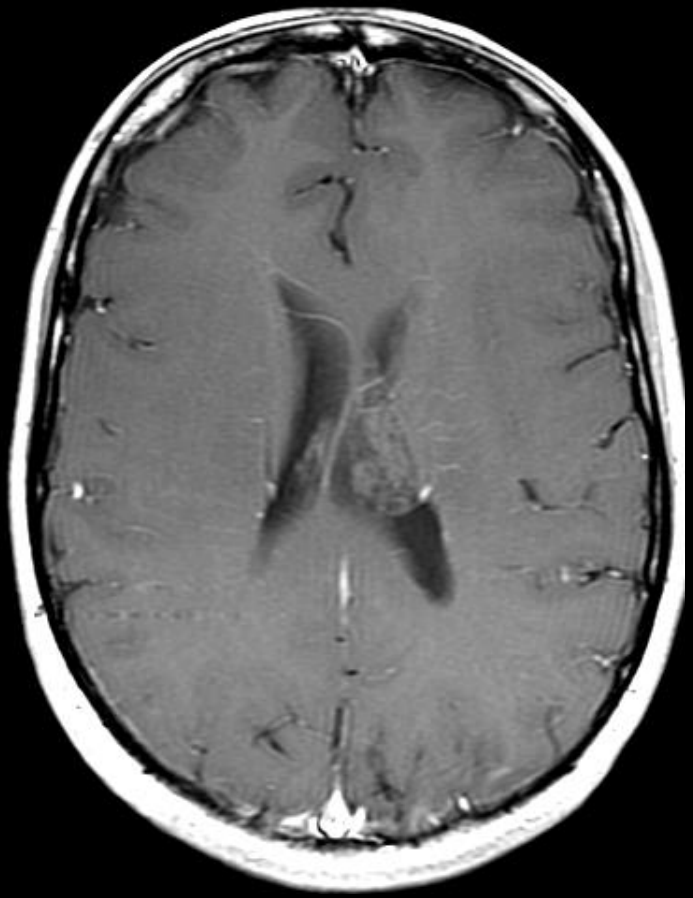


Central neurocytoma: Imaging features

Companion Case



T2W

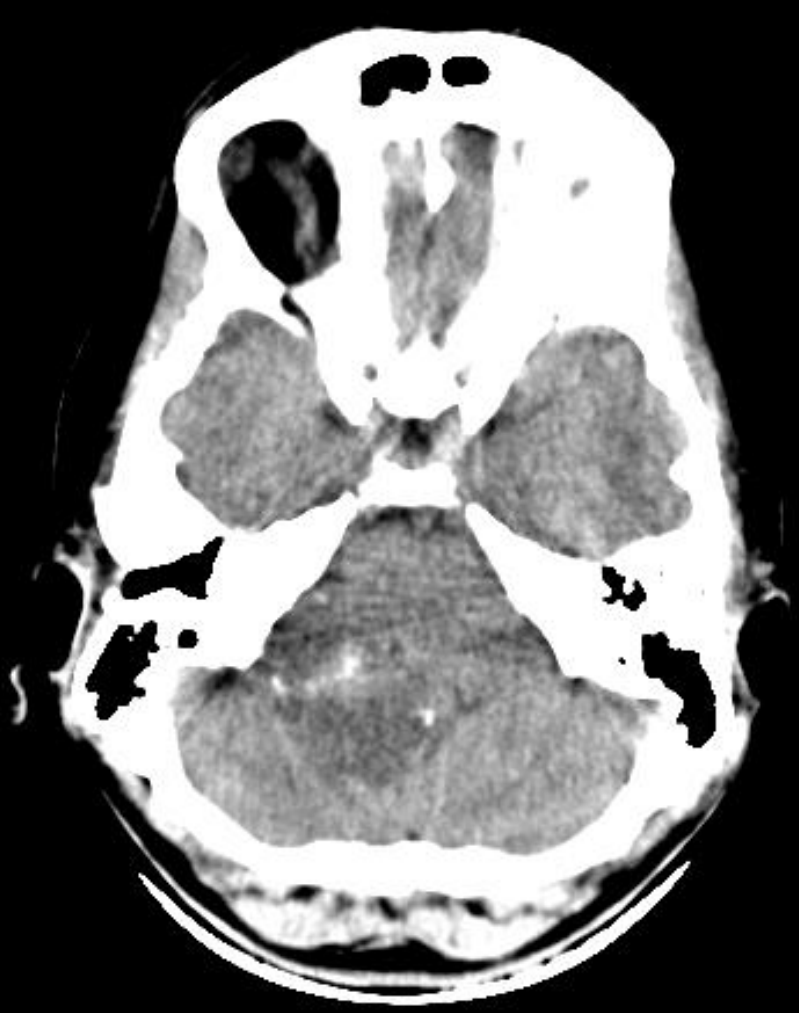


T1W post contrast

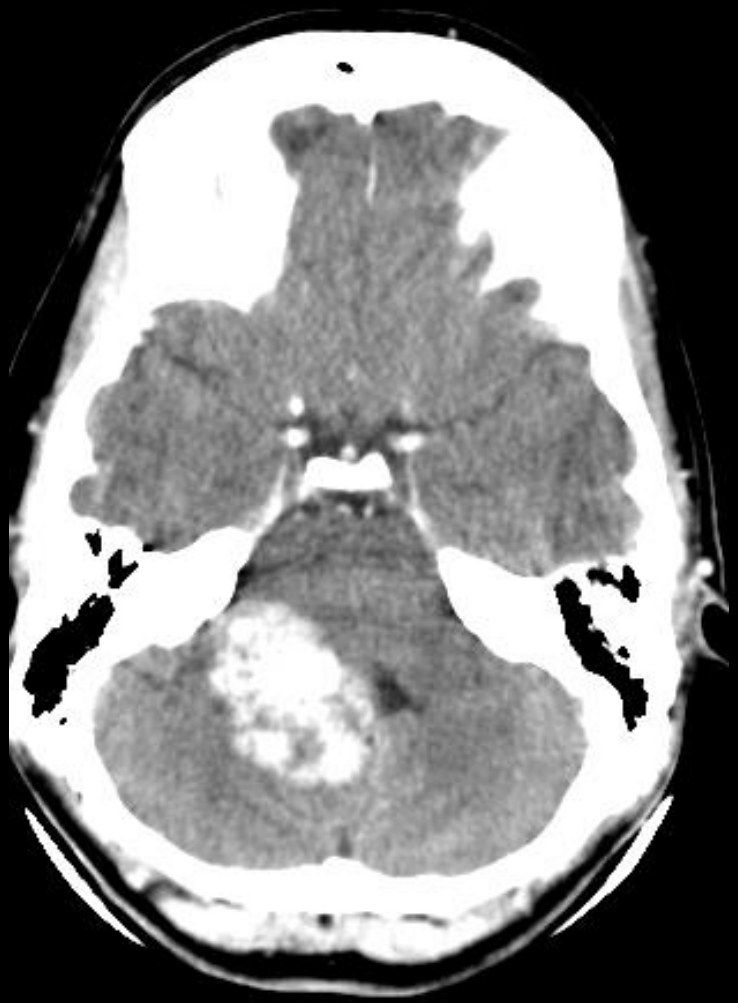
Case 4

29-year-old female referred by neurologist for MRI after 1 year headache and vertigo with worsening of the symptoms over 6 months. Headaches are base of occiput, pressure, transient, with no predilection for time of day.

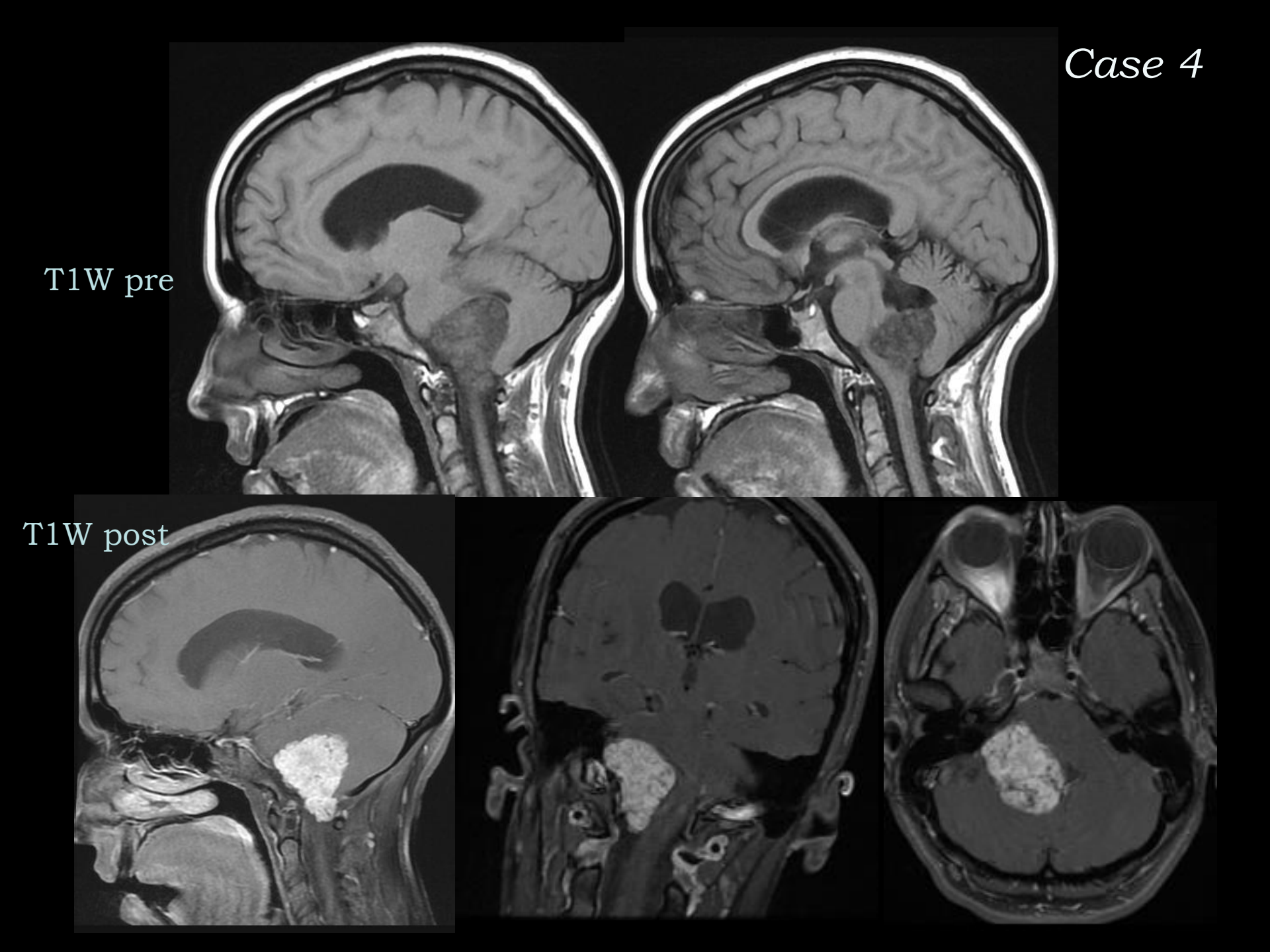
Case 4



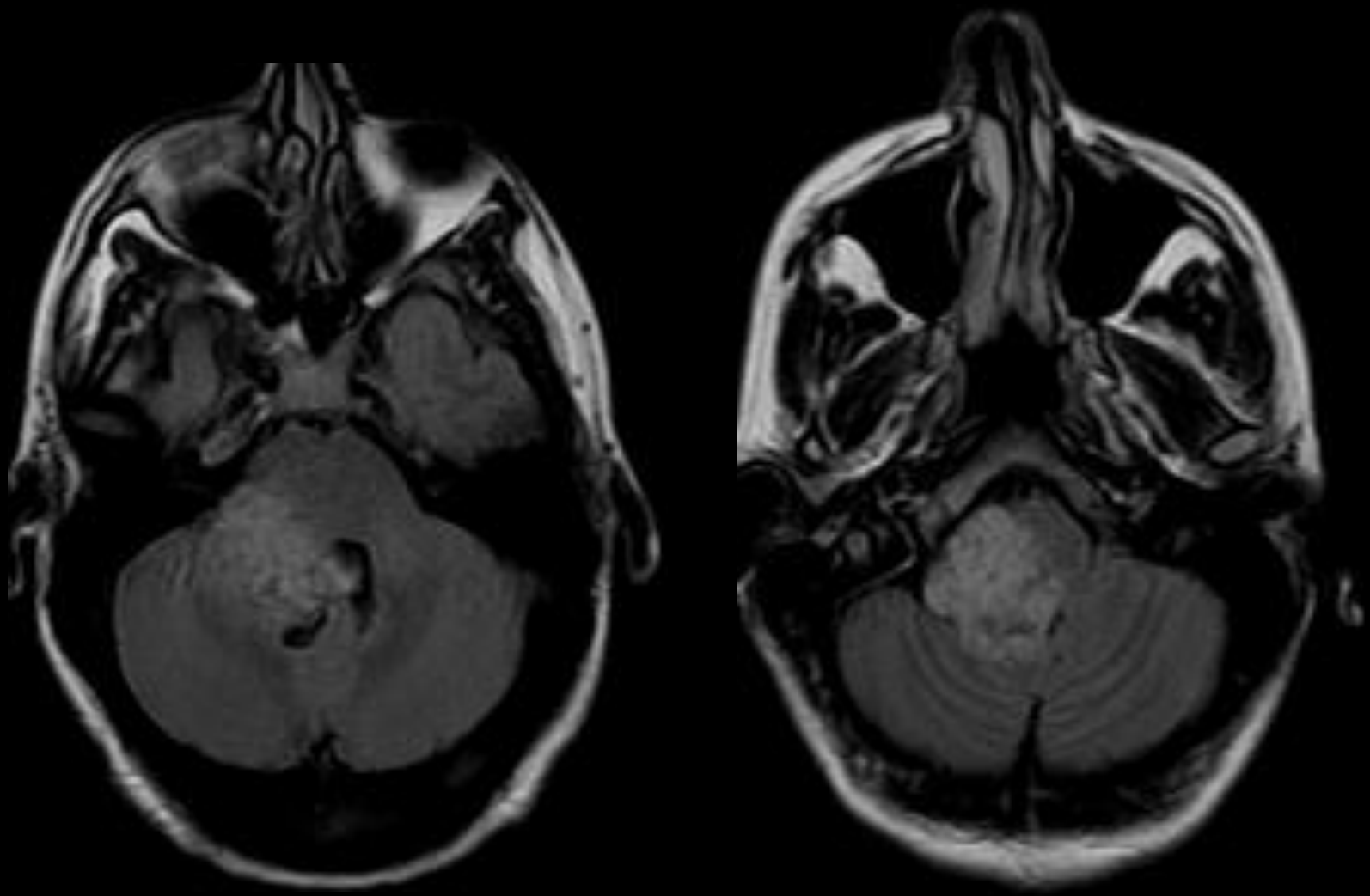
Pre contrast



Post contrast


Case 4
T1W pre
T1W post

Case 4



FLAIR

Case 4: Summary of findings

- 2.9 x 3.5 cm heterogeneous, avidly enhancing posterior fossa mass which appears to centered within the right foramen of Luschka, protruding inferiorly into the cisterna magna.
- There is resulting mass effect on the brainstem and fourth ventricle with moderate hydrocephalus.

Case 4: Differential diagnosis

Based on cell origins:

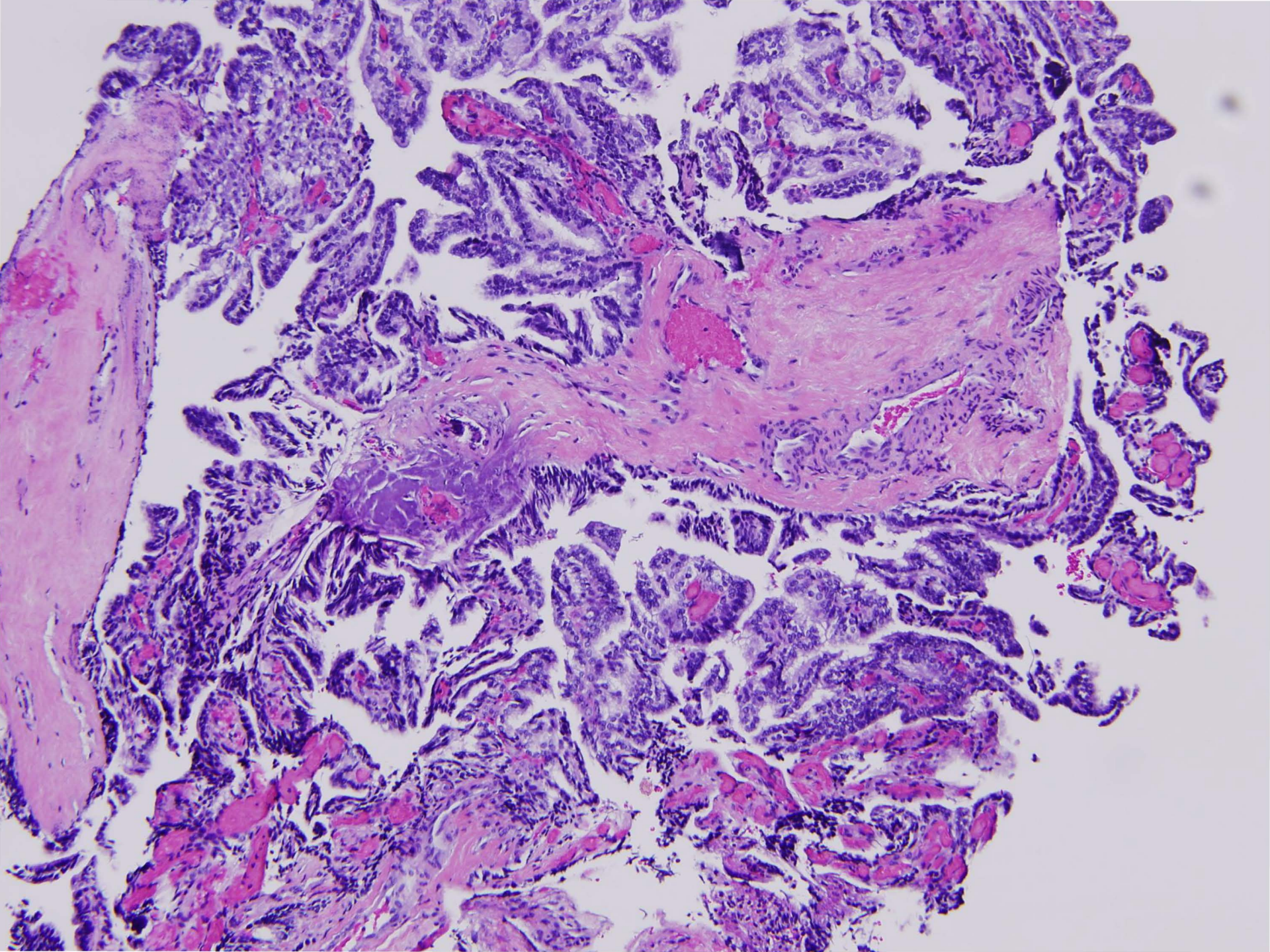
- *Ependymoma*
- *Subependymoma*
- *Choroid plexus neoplasms including choroid plexus papilloma, atypical CP papilloma and CP carcinoma*
- *Central neurocytoma*
- Meningioma
- *Metastasis*

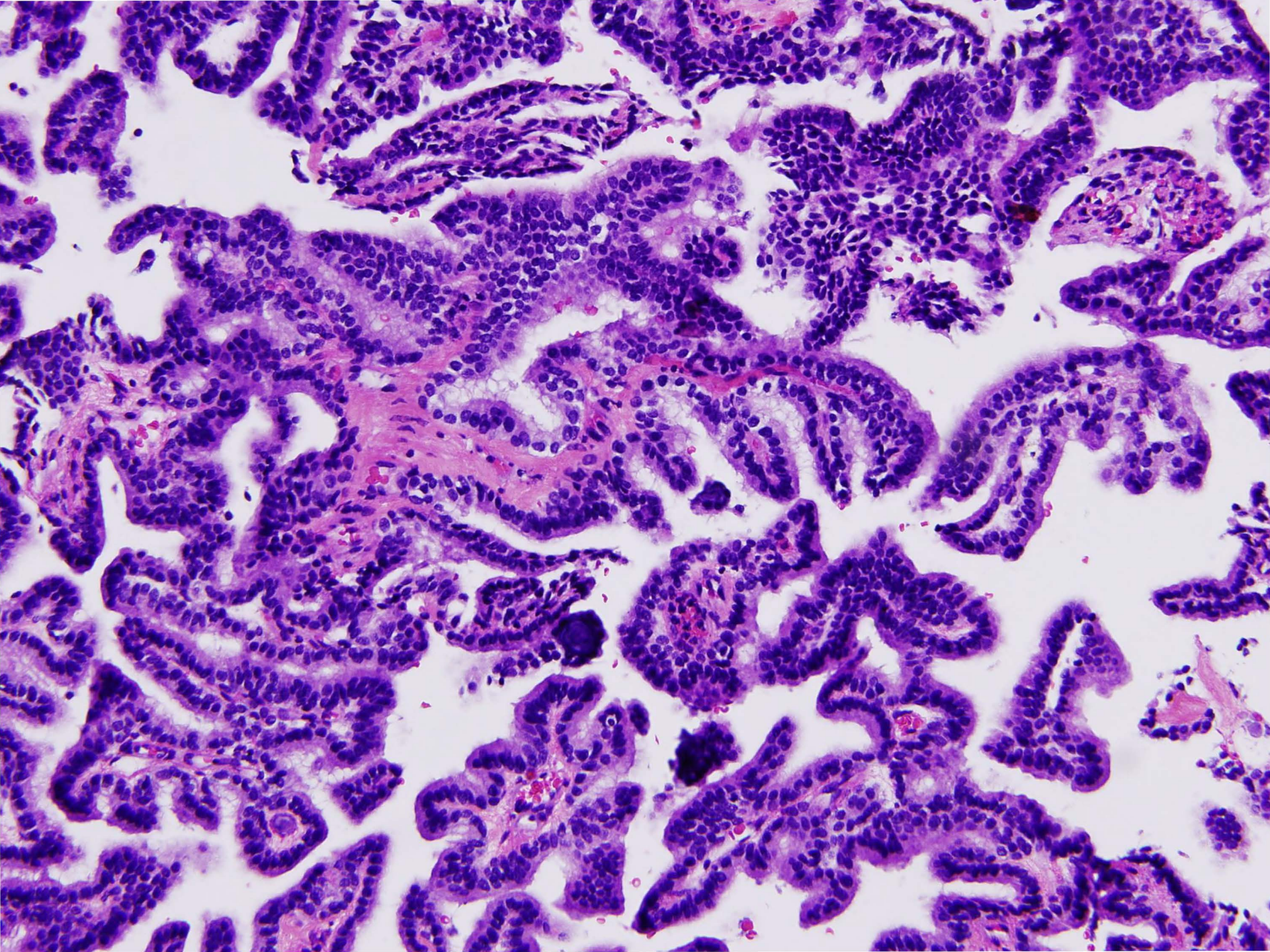
Miscellaneous:

- *Subpendymal giant cell tumor*
- *Chordoid glioma*
- *Rosette-forming glioneuronal tumor*

Case 4: Pathology

- MRN: 24211575, BS-09-43614





Choroid Plexus Papilloma

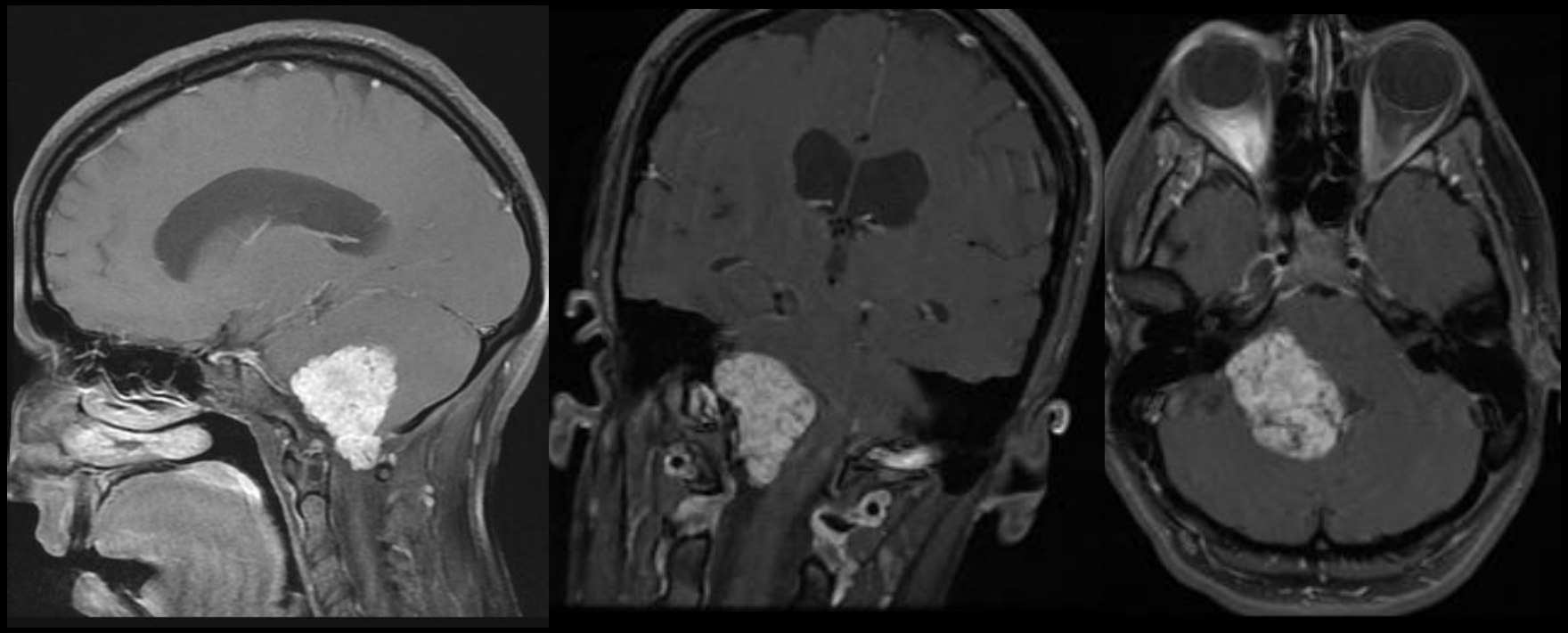
SPECIMEN LABELED " Right brain lesion" :

- - CHOROID PLEXUS PAPILLOMA, W.H.O. Grade I

- NOTE: Tumor is composed of delicate fibrovascular connective tissue fronds covered by uniform cuboidal epithelial cells with round to oval nuclei. No solid tumor areas, mitoses or necrosis are seen.

Case 4: Discussion

CHOROID PLEXUS PAPILLOMA

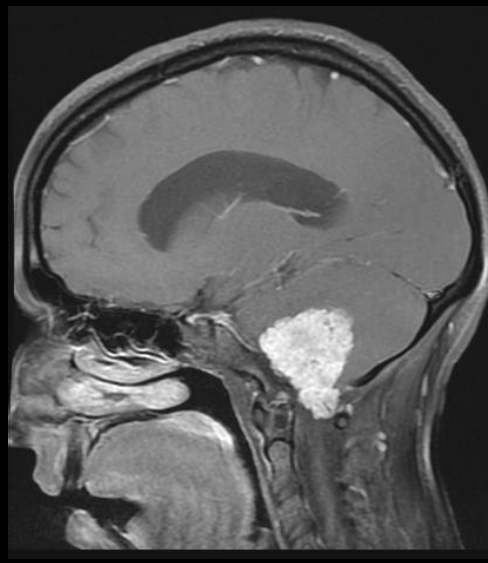


Choroid plexus neoplasms: Clinical features

- Arises from choroid plexus epithelium, subdivided into
  - Choroid plexus papilloma (WHO grade I)
  - Atypical choroid plexus papilloma (WHO grade II)
  - Choroid plexus carcinoma (WHO grade III)
- 2 – 4% of pediatric brain tumors (up to 20% occur in the 1st year of life)
- 50% lateral ventricle, 40% 4th ventricle, 10% 3rd ventricle
- Lateral ventricle lesions are more common in children, whereas 4th ventricle lesions are evenly distributed among all age groups
- CPC is found almost entirely in pediatric population
- Presenting symptoms due to hydrocephalus either from CSF overproduction, obstruction of CSF absorption or CSF flow
- 5-year survival rate 97% for CPP, 26-43% for CPC

Choroid plexus neoplasms: Imaging features

- Imaging does not allow distinction between three types of CP tumors
- Avid enhancement due to rich vascular supply
- Calcifications, hemorrhage may be seen
- Hydrocephalus common
- CPP and atypical CPP tend to have lobulated appearance, where as CPC tend to have irregular contour
- All may demonstrate CSF dissemination → recommend imaging of the entire neuroaxis
- MR:
    - Iso-hypointense on T1W, iso-hyperintense on T2W
    - Flow voids are common
    - Periventricular vasogenic edema may occur



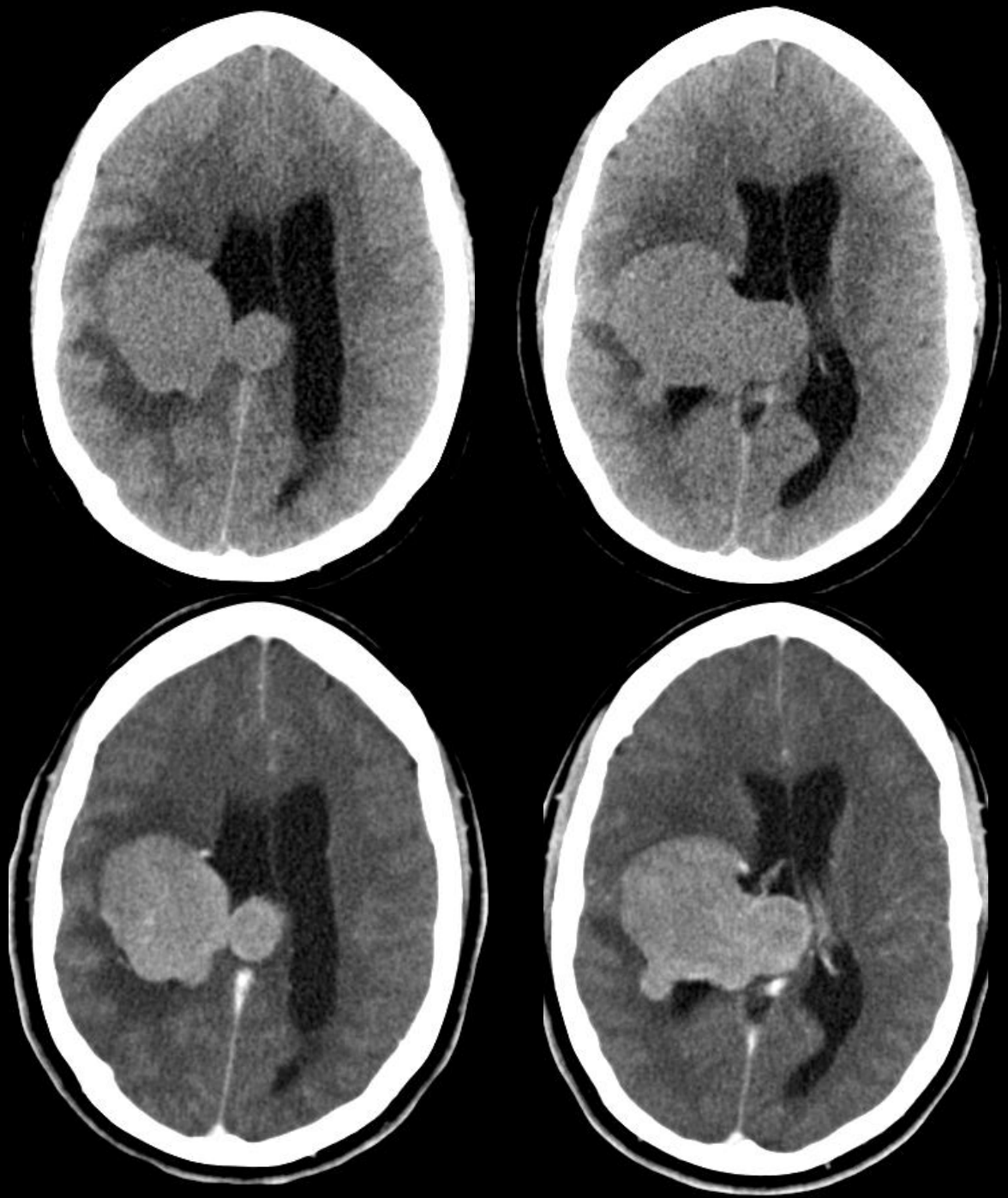
Case 5

45-year-old female with balance difficulty, headache, which are worse in the late evening, and clumsiness of the left leg and arm. These symptoms have worsened over the past year, but have been gradually progressive over the past several years.

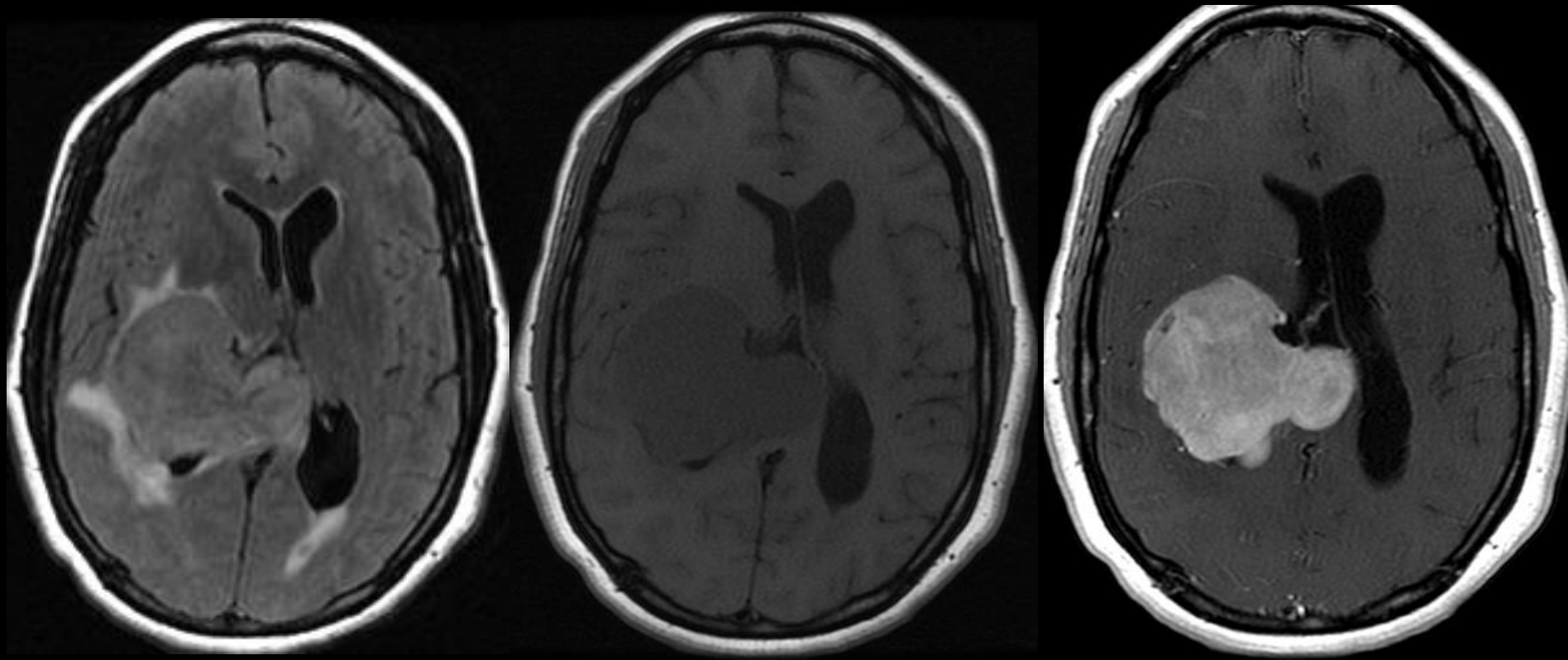
Case 5

Pre contrast

Post contrast



Case 5



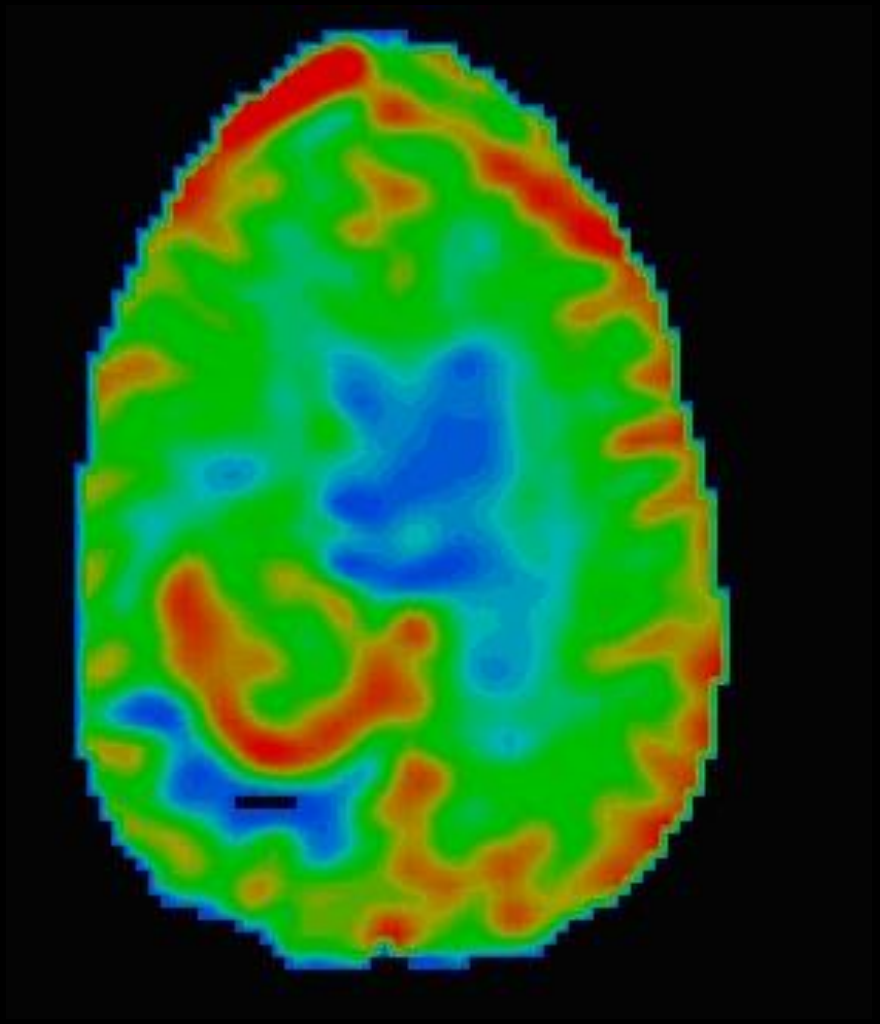
FLAIR

T1W pre contrast

T1W post contrast

Case 5

Perfusion imaging



Case 5: Summary of findings

- CTA: Smoothly marginated, bilobed 6.3 x 4.5 cm uniformly enhancing mass centered within the right atrium.
- MR brain:
  - Right frontoparietal well-circumscribed, gently lobulated mass likely arising from the atrium of the lateral ventricle, displacing the brain parenchyma laterally and causing midline shift
  - Isointense on T1W, heterogeneous signal on T2 with surrounding edema, homogeneous enhancement
  - Mild bilateral hydrocephalus
  - Increased rCBV at the rim of the lesion

Case 5: Differential diagnosis

Based on cell origins:

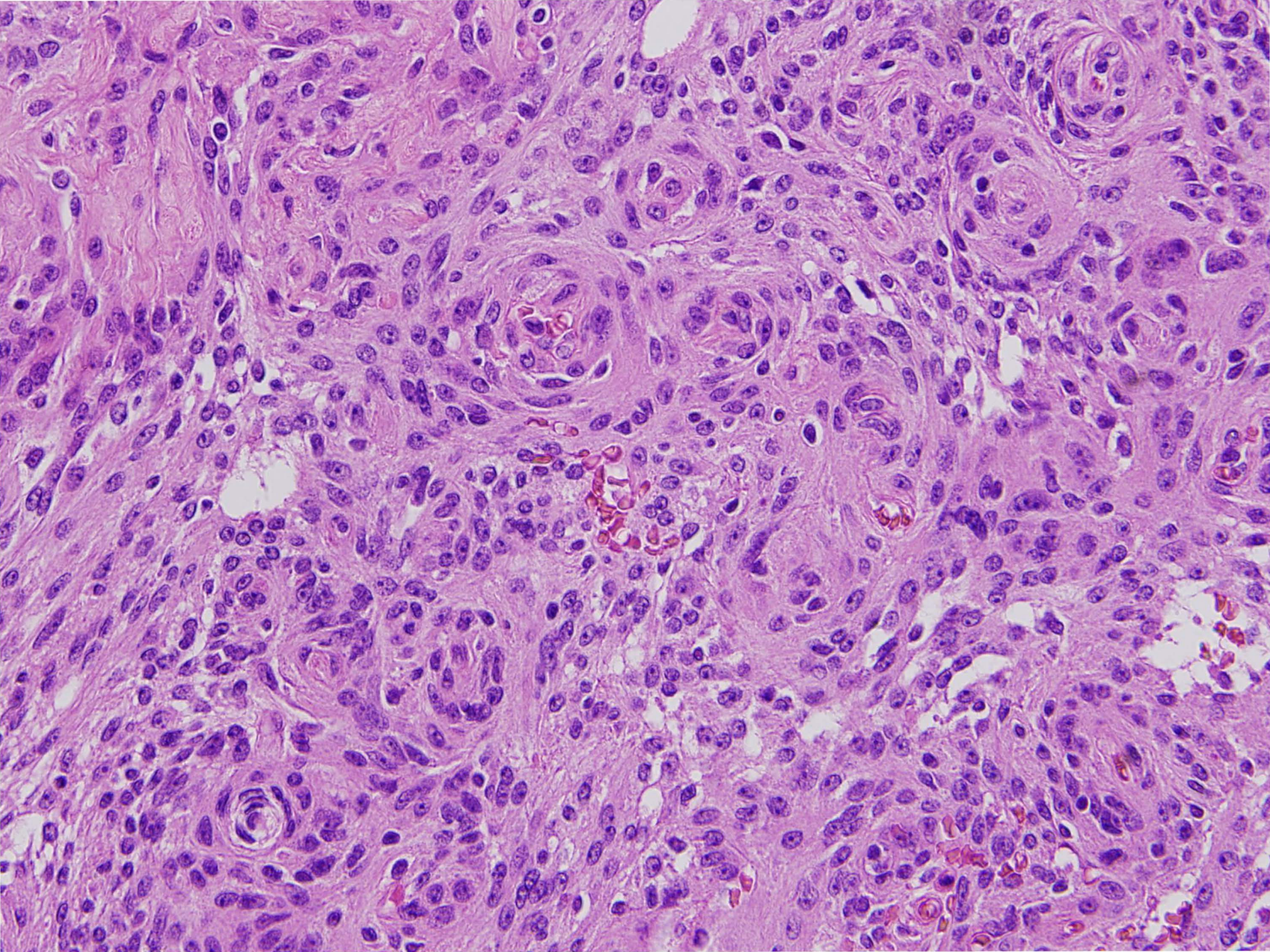
- *Ependymoma*
- *Subependymoma*
- *Choroid plexus neoplasms*
- *Central neurocytoma*
- Meningioma
- *Metastasis*

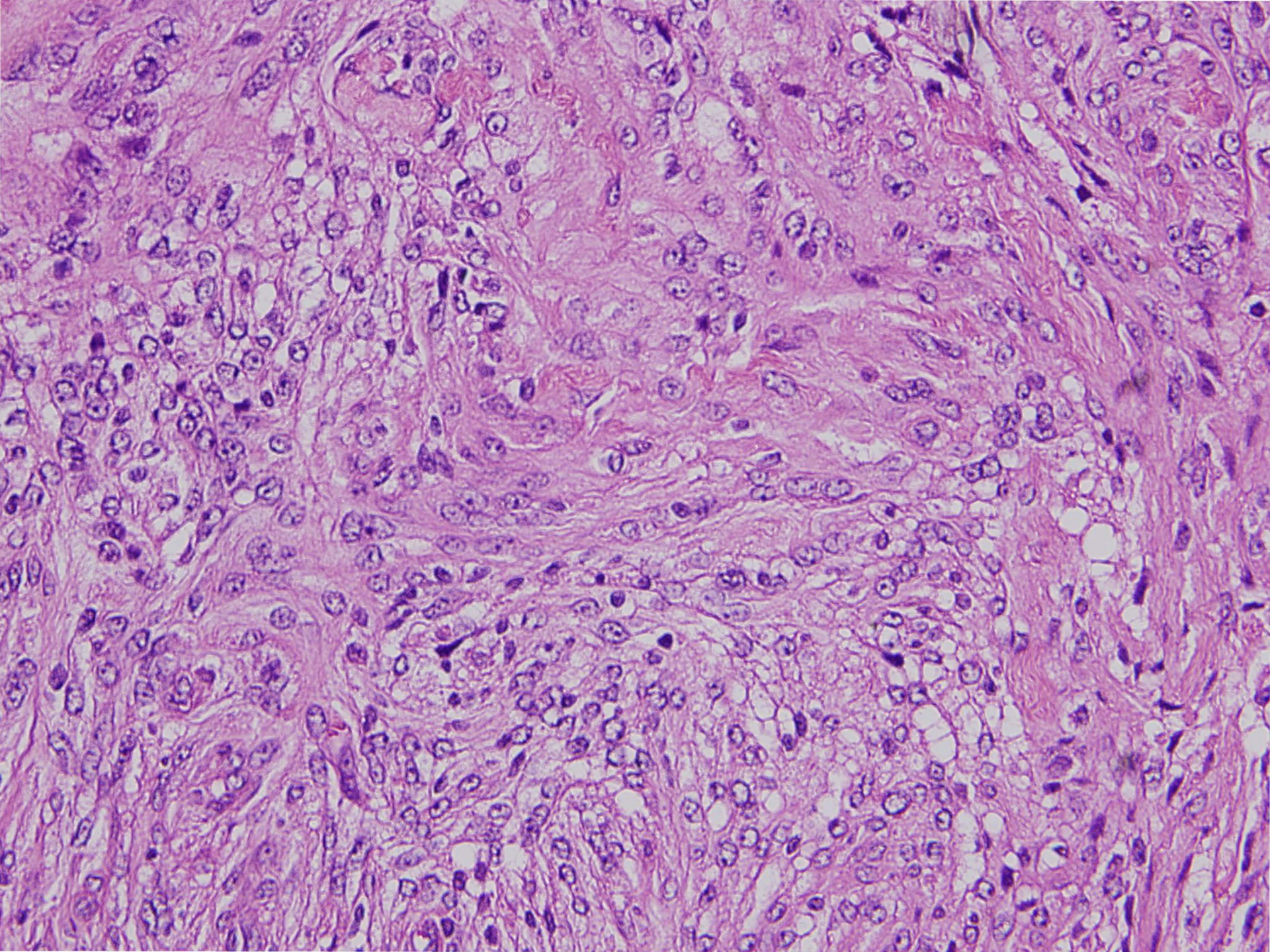
Miscellaneous:

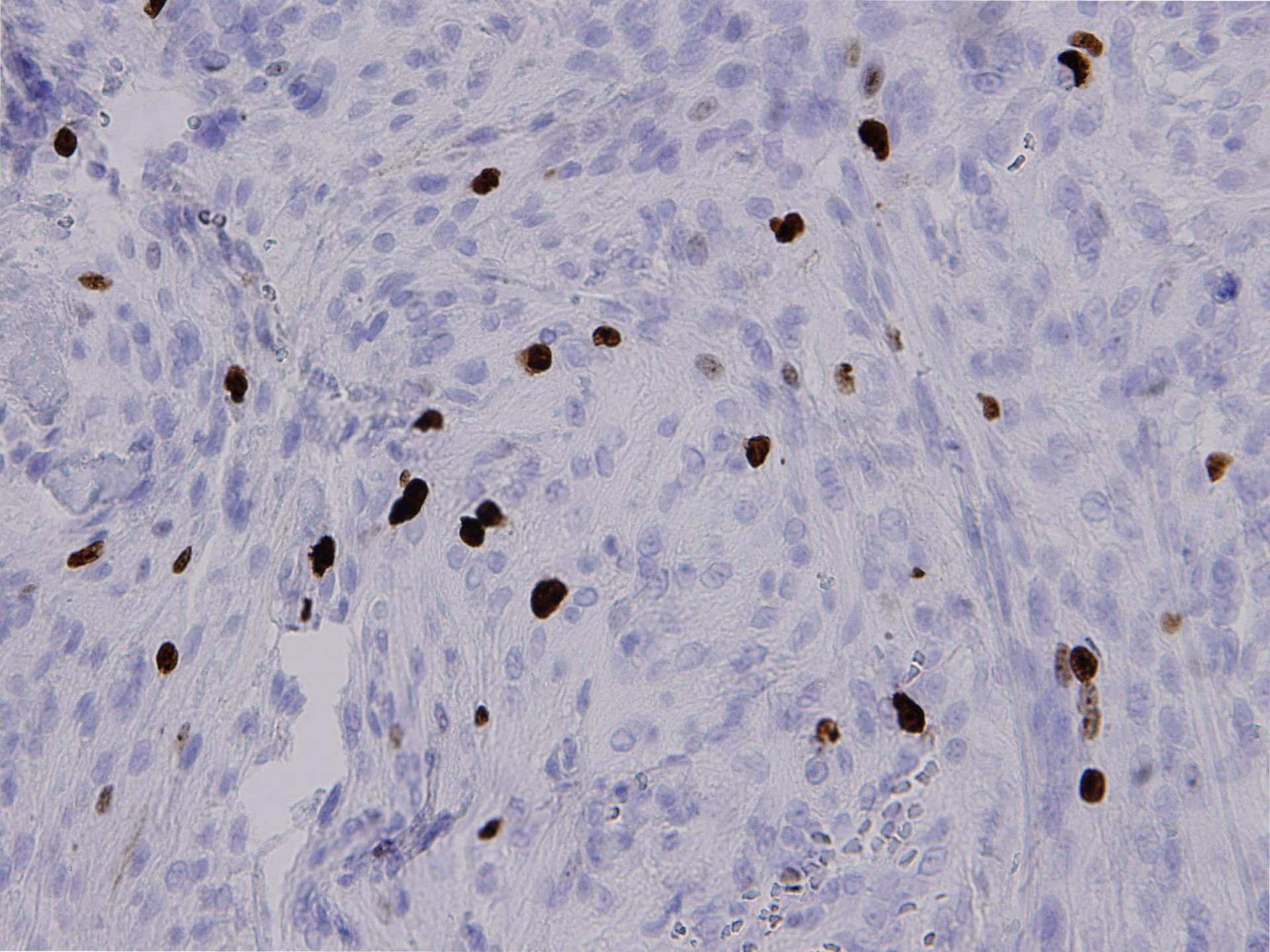
- *Subpendymal giant cell tumor*
- *Chordoid glioma*
- *Rosette-forming glioneuronal tumor*

Case 5: Pathology

- MRN: 26105452, BS-11-56884







Atypical meningioma

RIGHT INTRAVENTRICULAR MASS:

- ATYPICAL MENINGIOMA, W.H.O. Grade II, see NOTE.

W.H.O. Histologic Grading Criteria

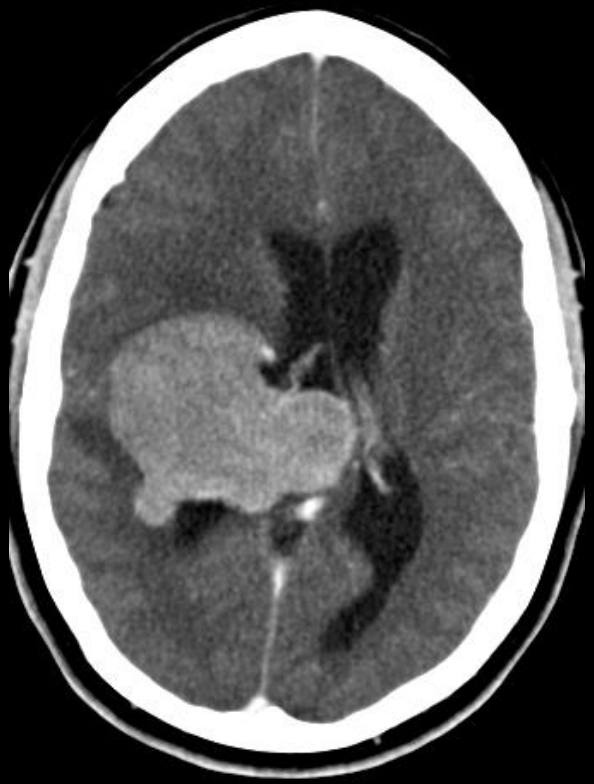
- Cellularity: dense focally
- Nuclear atypia: moderate
- Prominent nucleoli: present focally
- Sheet-like growth: present focally
- Mitoses: present (2/10 hpf)
- Necrosis: not present

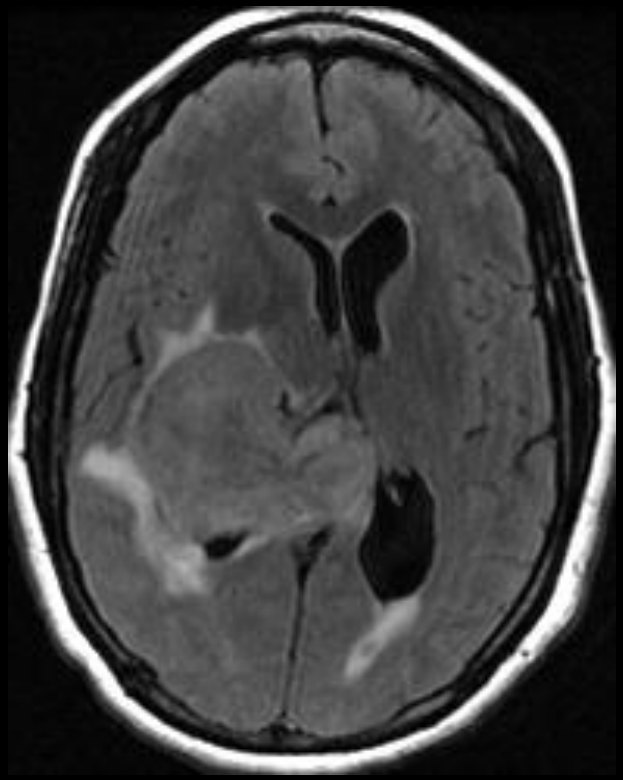
- Brain tissue: not present
- Bone: not present
- Psammomatous calcifications: not present

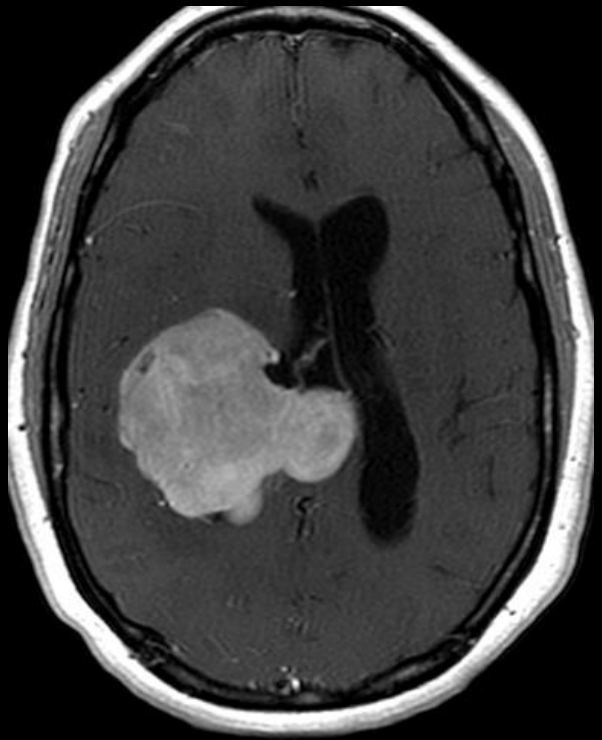
- The estimated MIB-1 proliferative index is approximately 8% (block A2, B2, not formally quantified).

Case 5: Discussion

INTRAVENTRICULAR MENINGIOMA





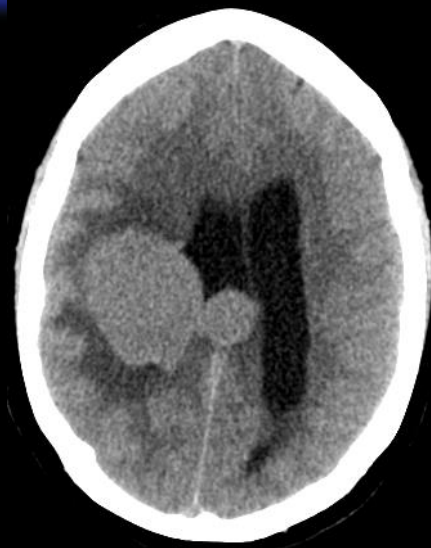


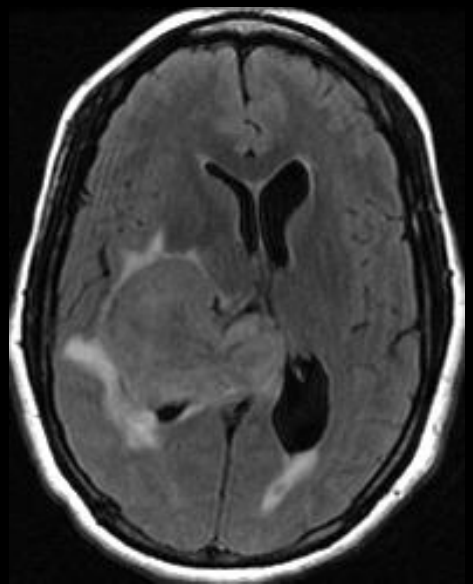
Meningioma: Clinical features

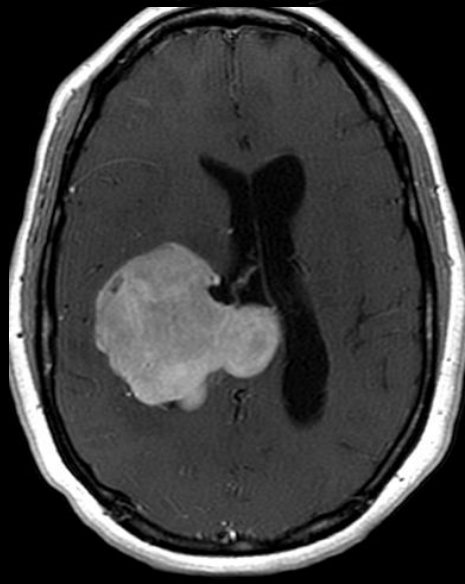
- Arises from arachnoidal cap cells trapped in choroid plexus
- 0.5-3.7% of intracranial meningiomas in adults
- Less than 3% of pediatric intracranial tumors but 17% of pediatric meningiomas are intraventricular (consider NF2)
- Atrium of the lateral ventricles most common
- Female-to-male ratio, 2:1 in adult population
- Peak age, 30-60 years
- Usually reaches a large size before becomes symptomatic
- Most demonstrate indolent behavior. Cases of atypical and anaplastic lesions are rare

Meniongioma: Imaging features

- CT: well-defined, iso- to hyperattenuating lobular mass
- MR:
  - Iso-to hypointense on T1W, iso-hyperintense on T2W
  - Reduced diffusion may be seen
  - Perfusion imaging demonstrate high blood volume
- Avid enhancement
- Calcifications in 50% of cases
- Cystic areas may be present
- Periventricular edema may be seen

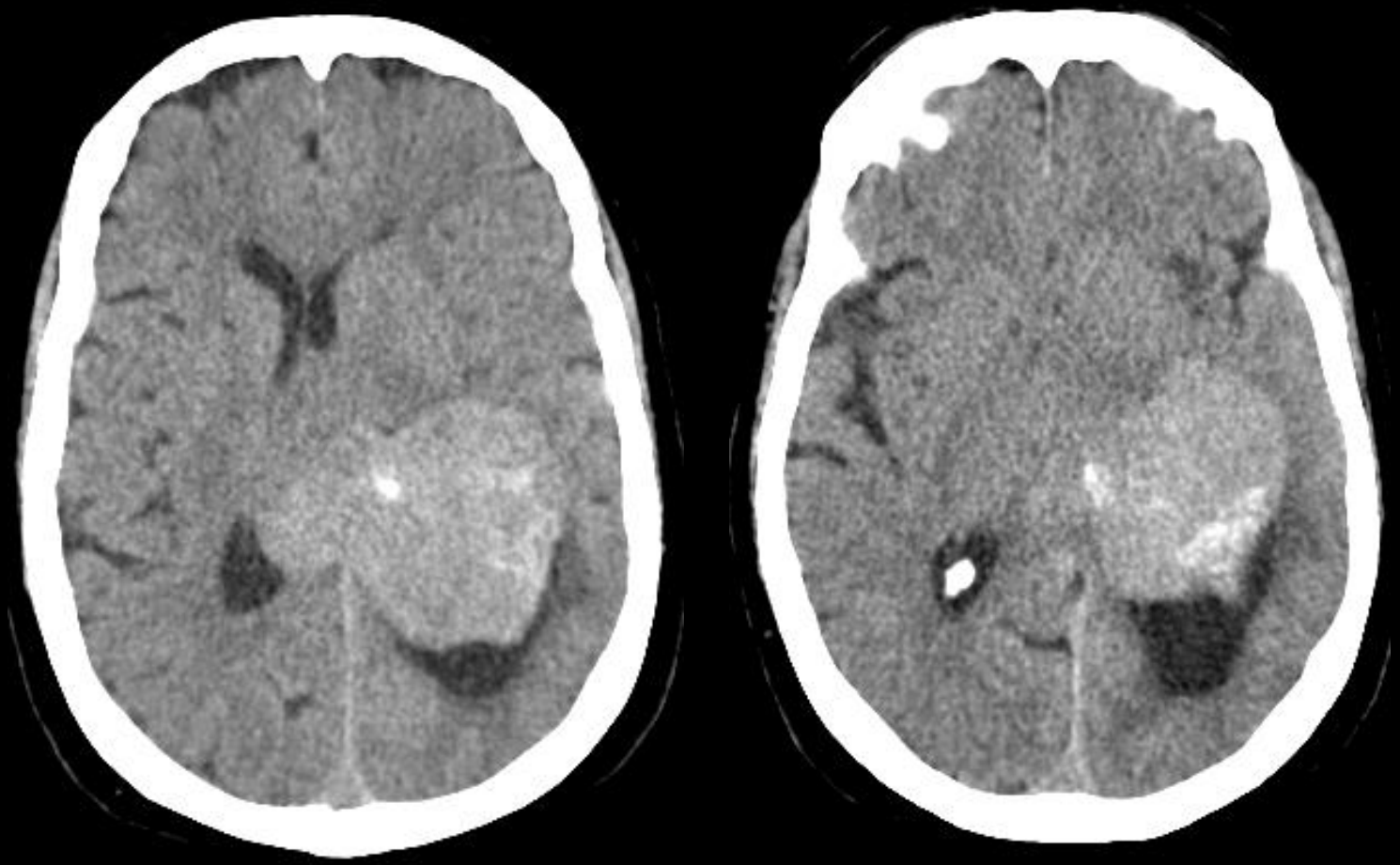






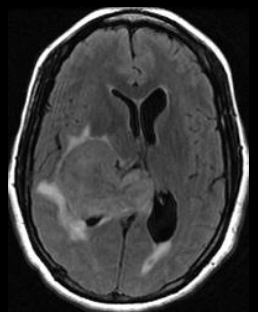
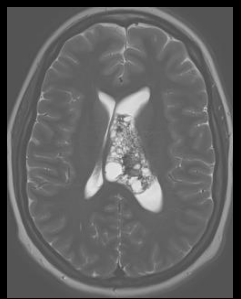
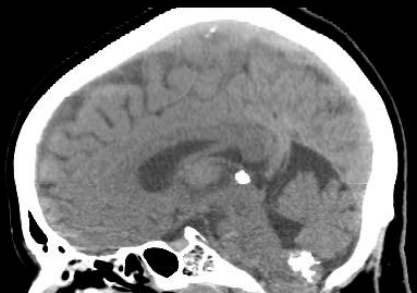
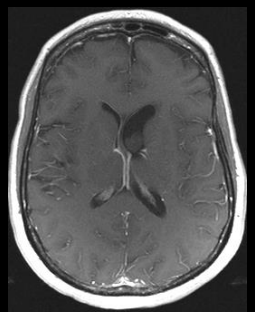
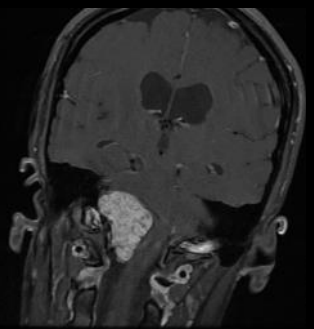
Meniongioma: Imaging features

Companion Case



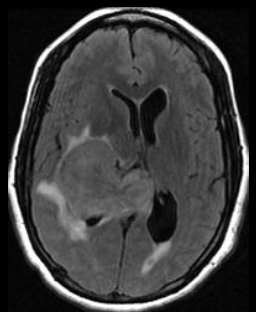
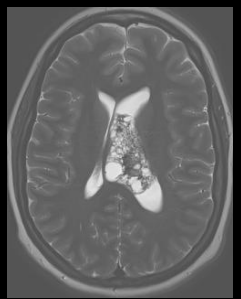
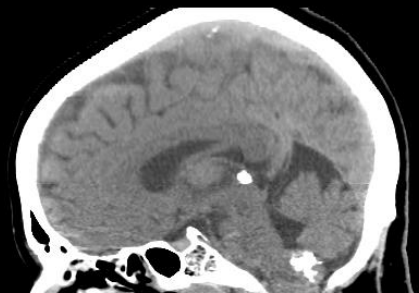
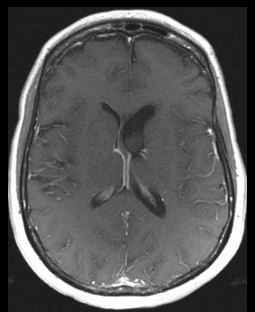
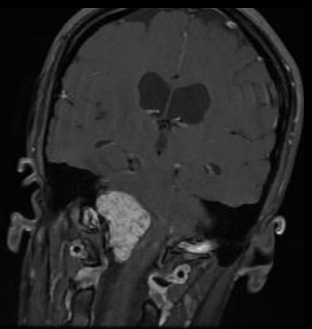
Conclusion

- To understand the different entities that can occur in an intraventricular location, it is important to understand the embryology, anatomy, and histologic structure of the ventricular system.
- Consideration of imaging features, the tumor location combined with the patient's age, gender and underlying condition helps narrowing the differential diagnosis.

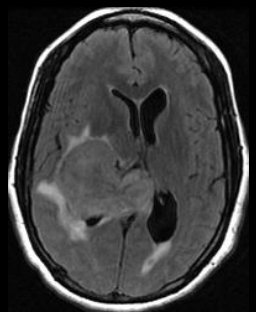
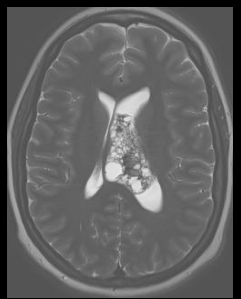
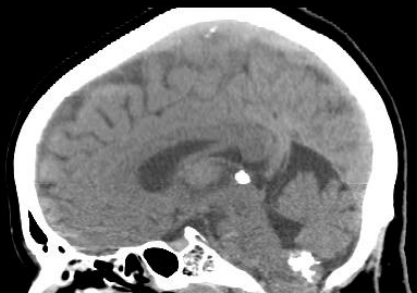
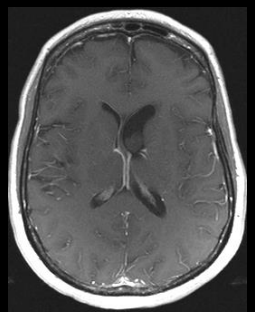
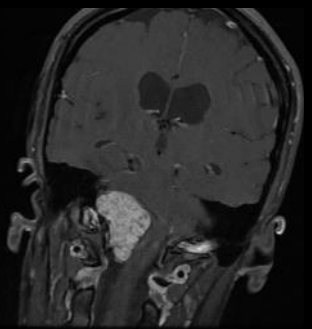
Conclusion

- Characteristic features:
  - Ependymoma: fills the ventricle—like a plaster cast or toothpaste, calcs, heterogeneous enhancement
  - Subependymoma: Non-enhancing mass in the fourth or lateral ventricle in an asymptomatic middle-aged male.
  - Central neurocytoma: Bubbly mass in the lateral ventricle, arising from septum pellucidum

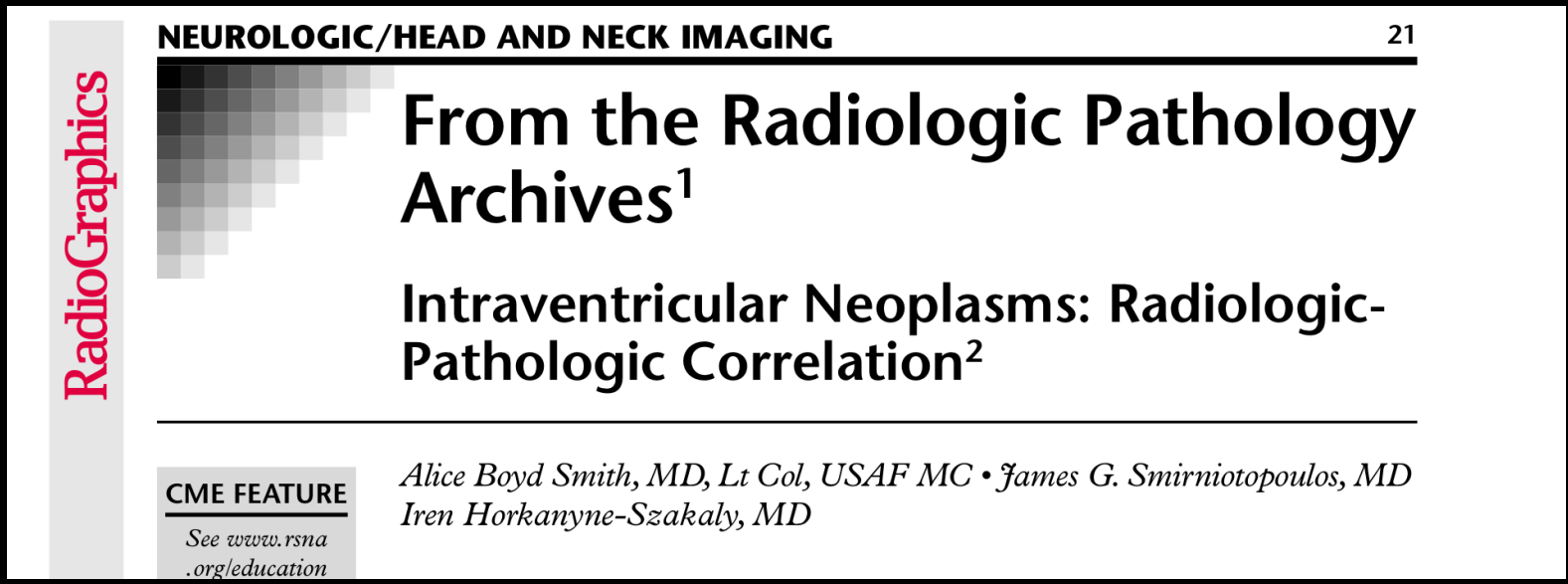






Conclusion

- Characteristic features:
  - Choroid plexus neoplasm: Lobulated strongly enhancing tumor in the lateral, 4th or 3rd ventricle
  - Intraventricular meningioma: Well-circumscribed mass centered in the atrium of the lateral ventricle with intermediate to high attenuation and vasogenic edema in the adjacent parenchyma

Reference



RadioGraphics

NEUROLOGIC/HEAD AND NECK IMAGING 21

From the Radiologic Pathology Archives[1]

Intraventricular Neoplasms: Radiologic-Pathologic Correlation[2]

CME FEATURE

See www.rsna.org/education

Alice Boyd Smith, MD, Lt Col, USAF MC • James G. Smirniotopoulos, MD
Iren Horkanyne-Szakaly, MD

Acknowledgement

- Tejus Bale, MD
- Narayan Viswanadhan, MD
- Angela Giardino, MD
- Kate Mullen, MD