

RadPath: Occipital Bone

Andrew Hill, MD
Gabriel Griffin, MD



Why the Occipital Bone?

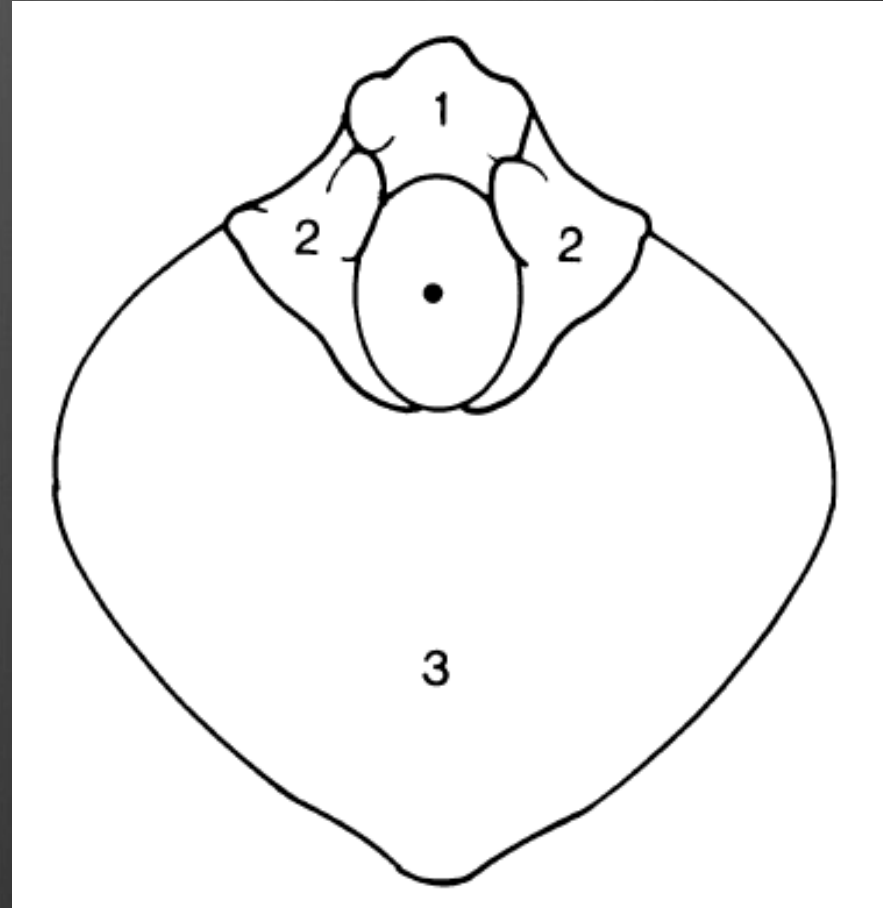
- The temporal bone gets all the love when it comes to skull base
- The occipital bone is an important part of the craniovertebral junction (CVJ)
- Occipital bone lesions are often insidious in their growth and presentation

Objectives

- Review the embryology and anatomy of the occipital bone
- Review 2 synchondroses of the posterior skull base
 - Petro-occipital
 - Spheno-occipital
- Understand classic important differential diagnoses based on 2 locations:
 - Jugular Foramen
 - Clivus

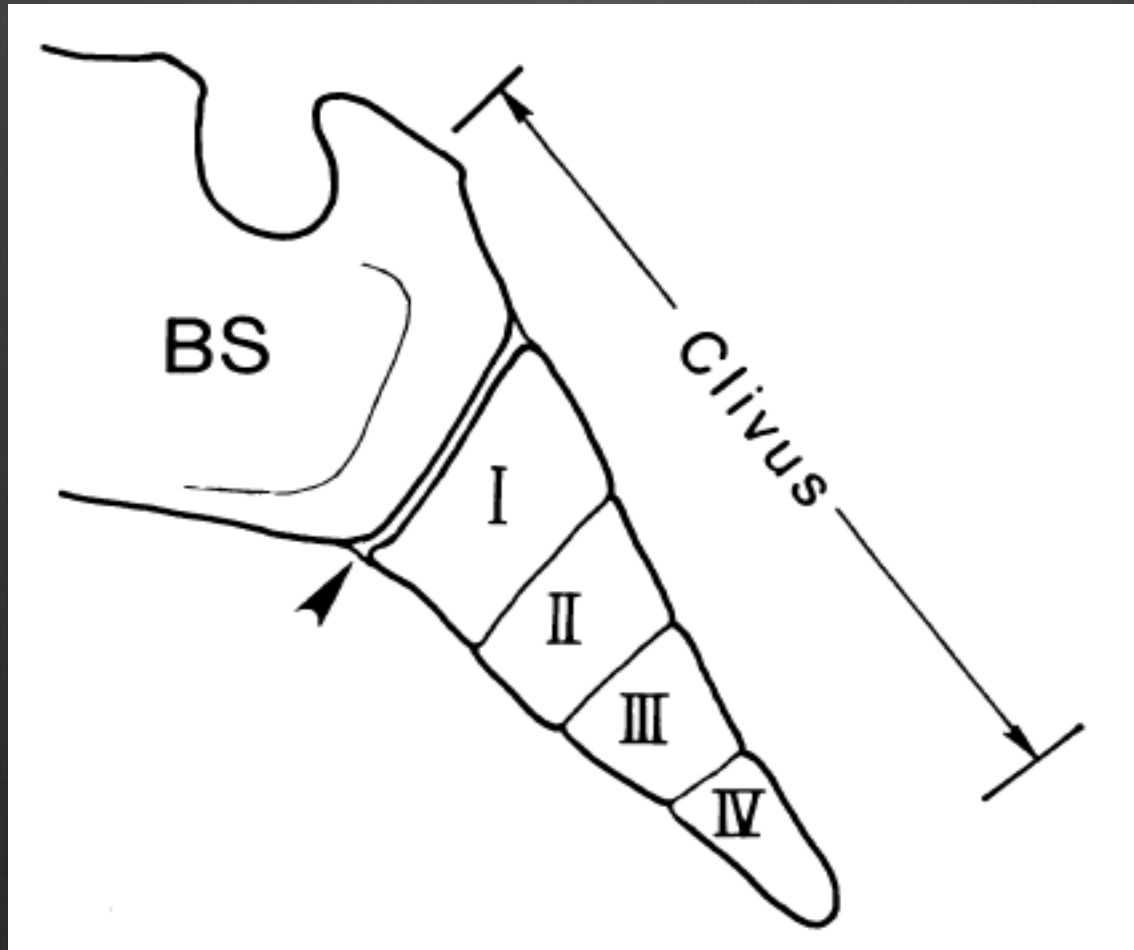
Occipital Bone Anatomy

- Composed of:
 1. Basioccipital
 2. Exoccipital
 3. Supraoccipital



Sclerotomes

Majority of the Occipital Bone is derived from 4 occipital “sclerotomes”



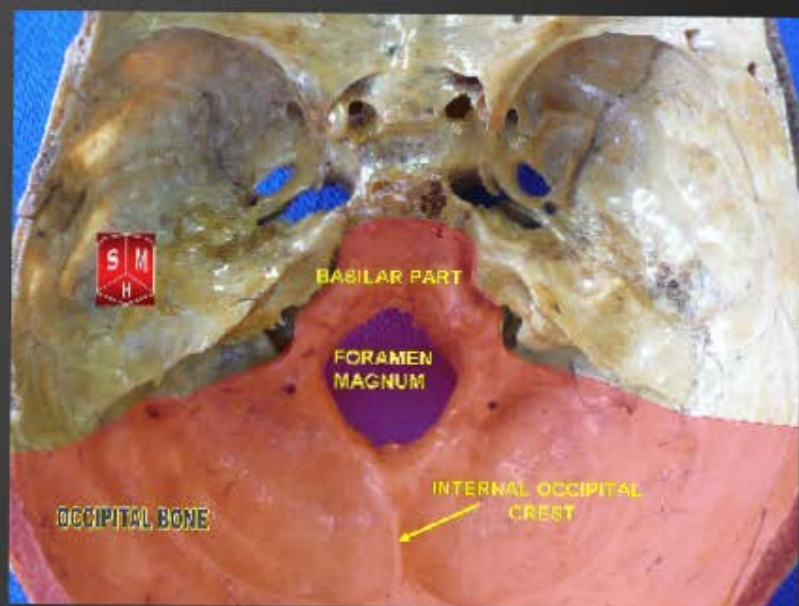
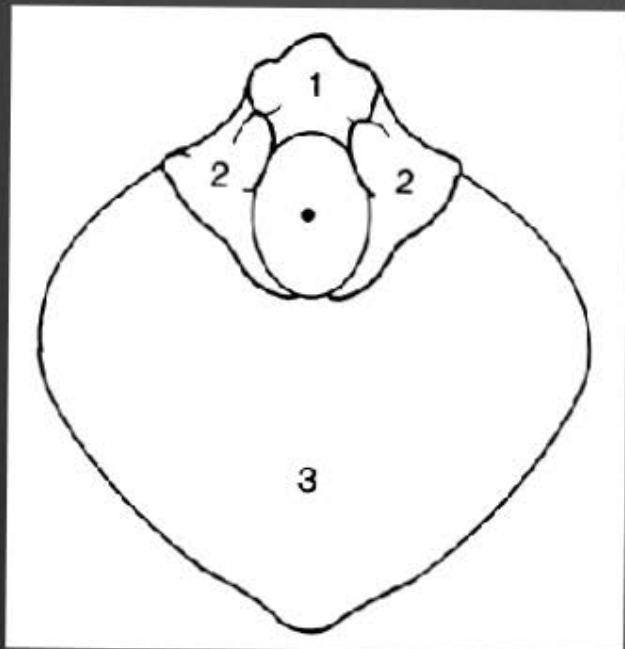
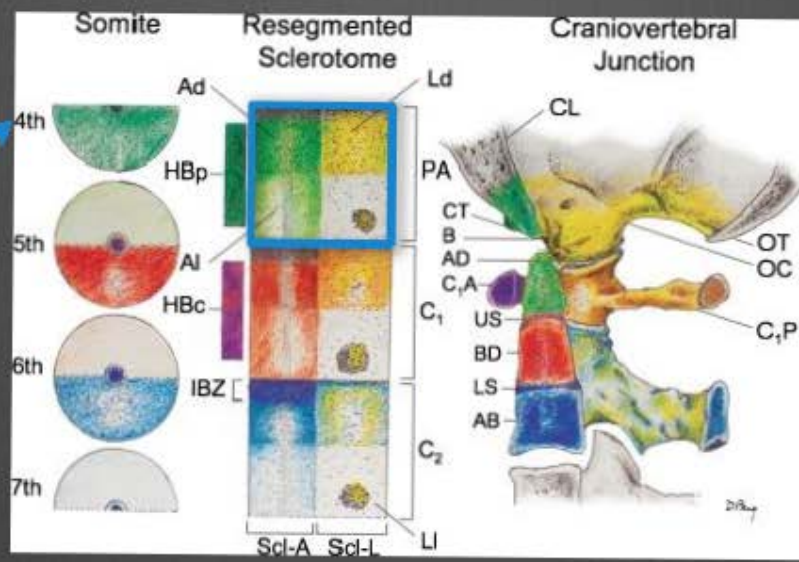
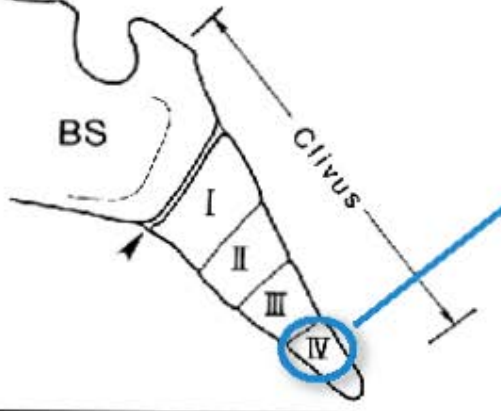
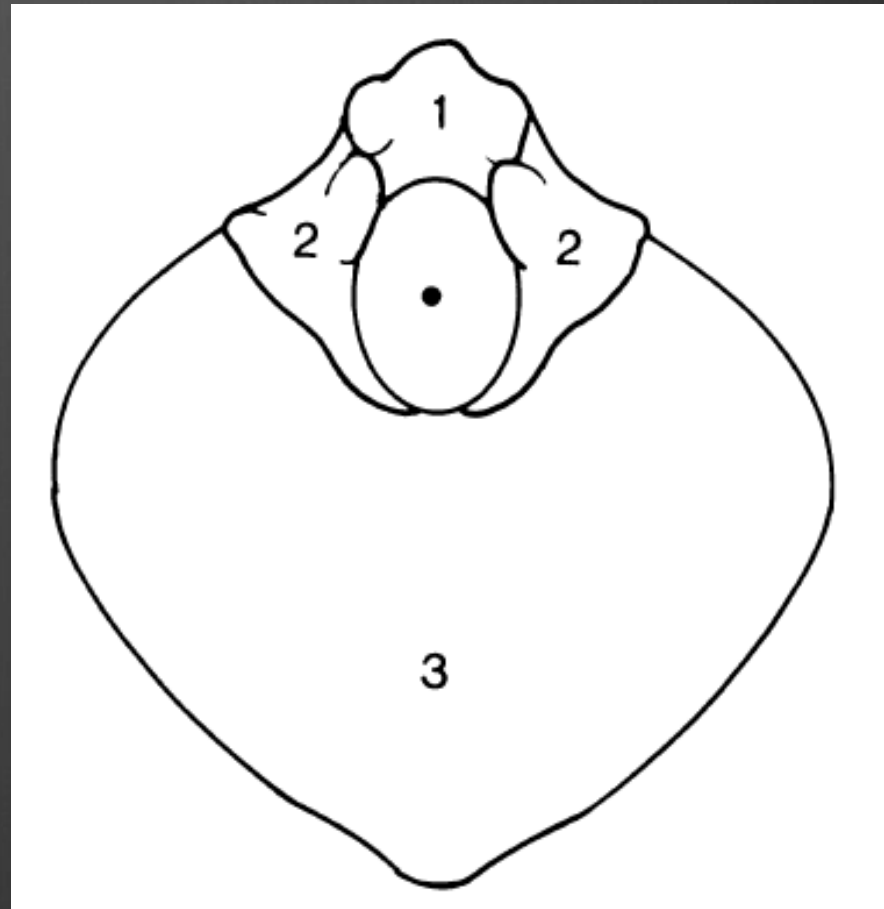
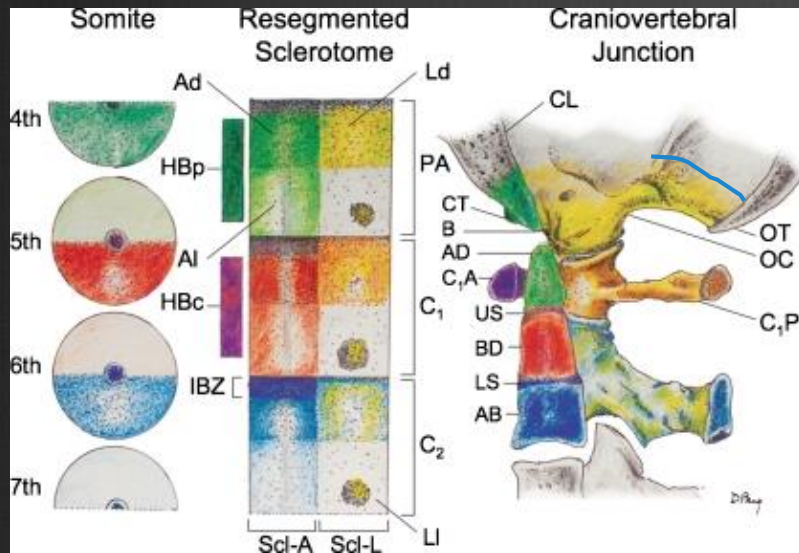




Figure 10. Condylus tertius and platybasia. Mid-sagittal T1-weighted (600/20) MR image reveals marked skull base flattening, with a Welcher basal angle of 150° (dotted line). Note the marked bow-string deformity of the cervicomedullary junction. The C-1 arch (A) lies directly above the tip of the odontoid process (O). Marrow within accessory ossification centers (condylus tertius) (black dots) is seen at the tip of the basion.

Supraoccipit

- Not purely derived from sclerotomes
- Endochondral and Membranous ossification



Differentiating a Mendosal Suture from a Skull Fracture

A 6-day-old infant was brought to her pediatrician with a right clavicle fracture. This injury could have happened at birth; however, the history provided by her mother was concerning enough that a skeletal survey and additional tests were performed and child protective services was notified. The lateral skull film seemed to show an occipital skull fracture (Figure 1), and the results of computed tomography of the head showed a single defect in the bone window that seemed to correlate with the finding on plain film. Computed tomography of the head with 0.625-mm slices and 3-dimensional reconstruction (Figures 2 and 3) was performed on day of life 14. As shown by the arrows in Figures 2 and 3,



Figure 1. Lateral skull film. Arrows show what seems to be an occipital skull fracture.

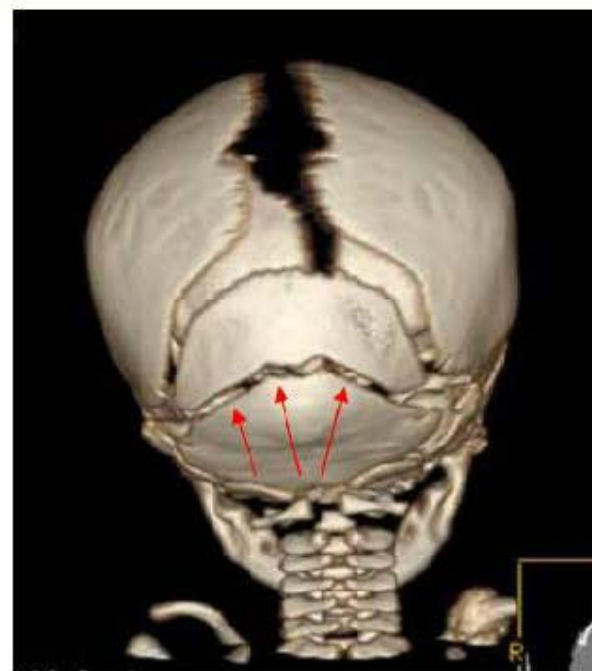
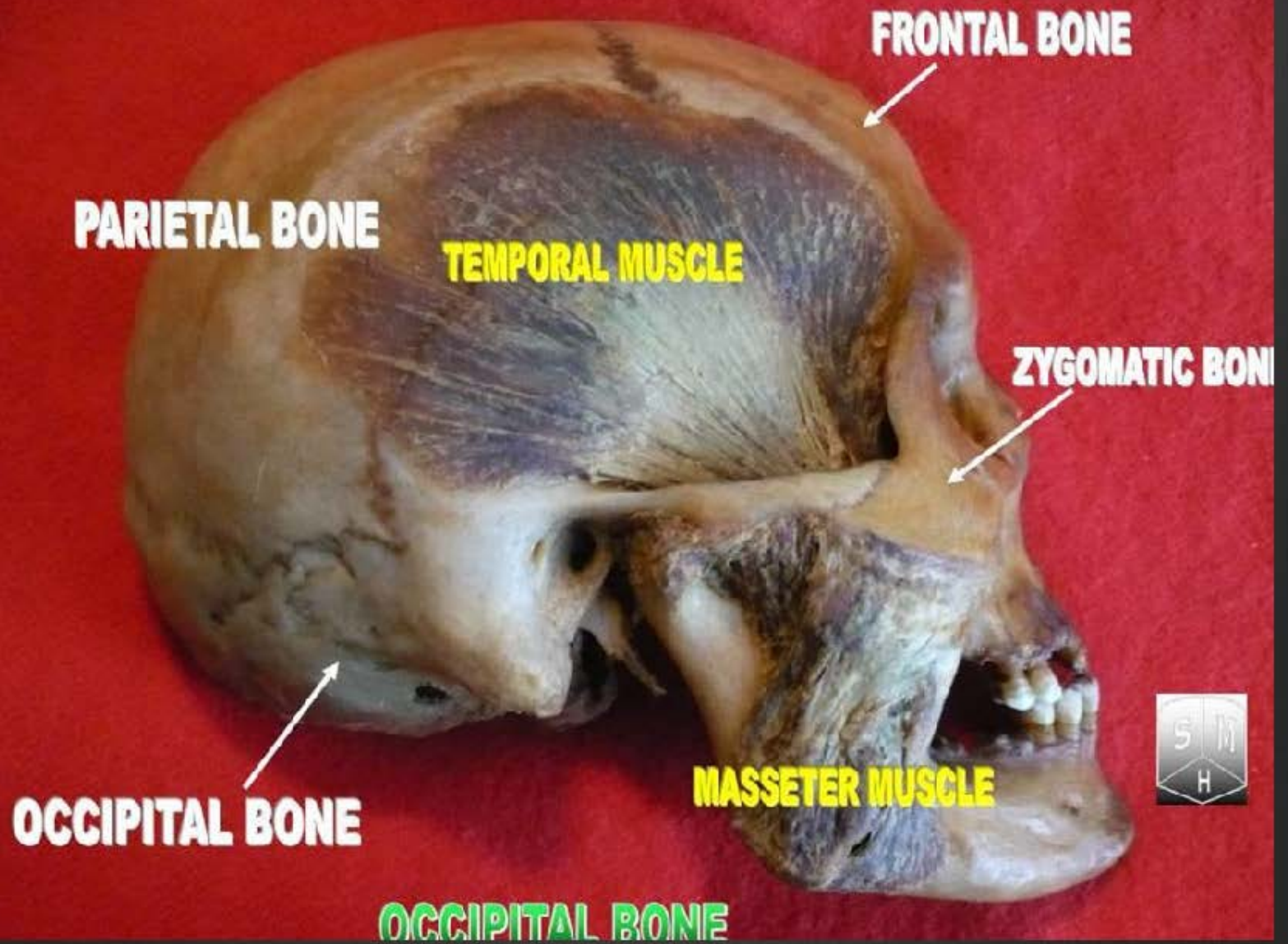
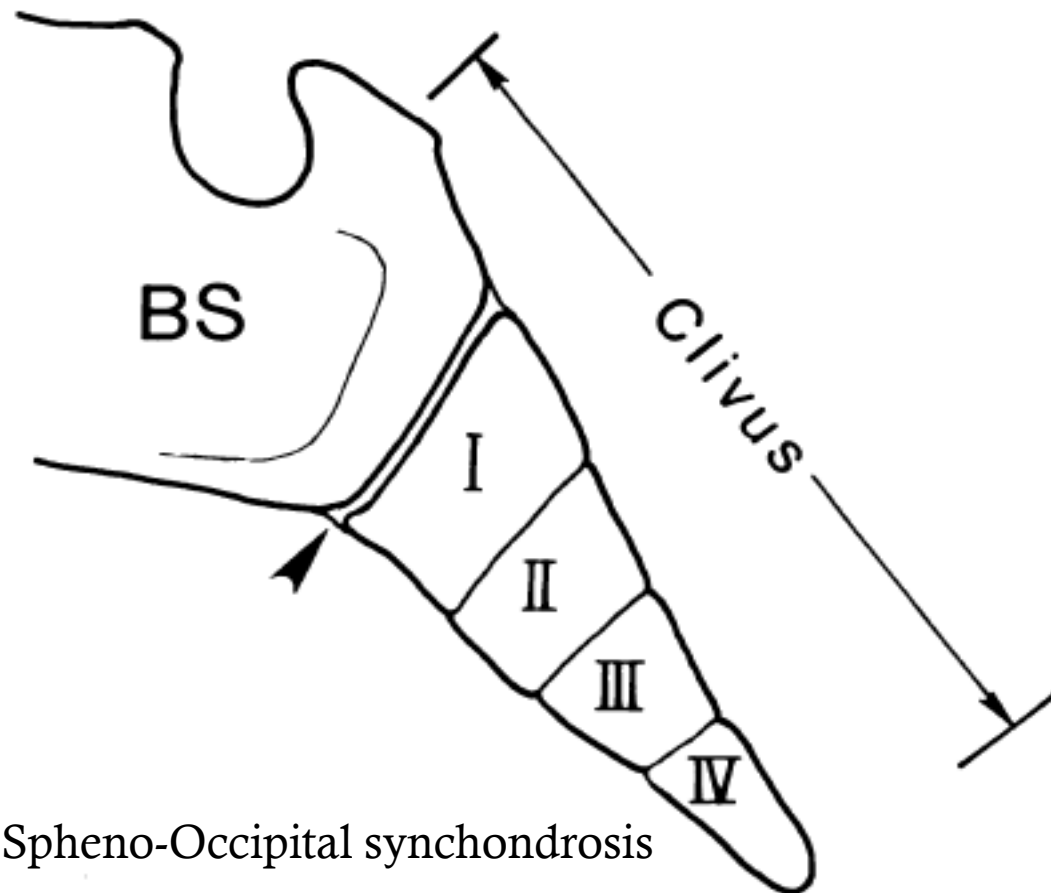


Figure 3. Three-dimensional reconstruction of computed tomography of the head, 0.625-mm slices. Arrows show persistent mendosal suture.

this patient has a rare, but classic, persistent mendosal suture. In utero, the mendosal suture separates the supraoccipital bone from the interparietal bone, and this suture usually closes in utero or in the first few days of life.¹ Performing the 3-dimensional reconstruction was key to determining that the





Spheno-Occipital synchondrosis

Roentgen Determination of the Time of Closure of the Spheno-Occipital Synchondrosis¹

GEORGE LEIGH IRWIN, M.D.²



MR Imaging of the Normal and Abnormal Clivus

and the upper cervical spine, visualized on the cranial midline T1-weighted sagittal images, also showed diffuse loss of yellow marrow in these patients.

On T2-weighted images, lesions appeared as an area of homogeneously high intensity in three patients with chordoma, two with pituitary adenoma and one with nasopharyngeal carcinoma (Fig. 12). In five of eight patients with metastasis to the clivus, lesions were hyperintense relative to the normal clivus. The signal intensity was mixed with areas of iso- and hyperintensity in two patients with lymphomatous infiltration of the clivus (Fig. 10). The signal intensity was isointense relative to the normal clivus in three patients with metastasis, one with periclavial meningioma and two with marrow reconversion. The diminished signal-to-noise ratio encountered on longer TR/TE images resulted in poorer anatomic definition of lesions.

Twelve of 21 patients with an abnormal clivus had gadopentetate-dimeglumine-enhanced scans. The low-intensity tumor, focal or diffuse, was intensely enhanced in all patients (Figs. 8 and 9). The reconverted clivus was mildly enhanced in one patient and moderately enhanced in the other (Fig. 11).

Discussion

Okada et al. [11], in their study of a group of normal children and young adults, demonstrated that signal from clival mar-

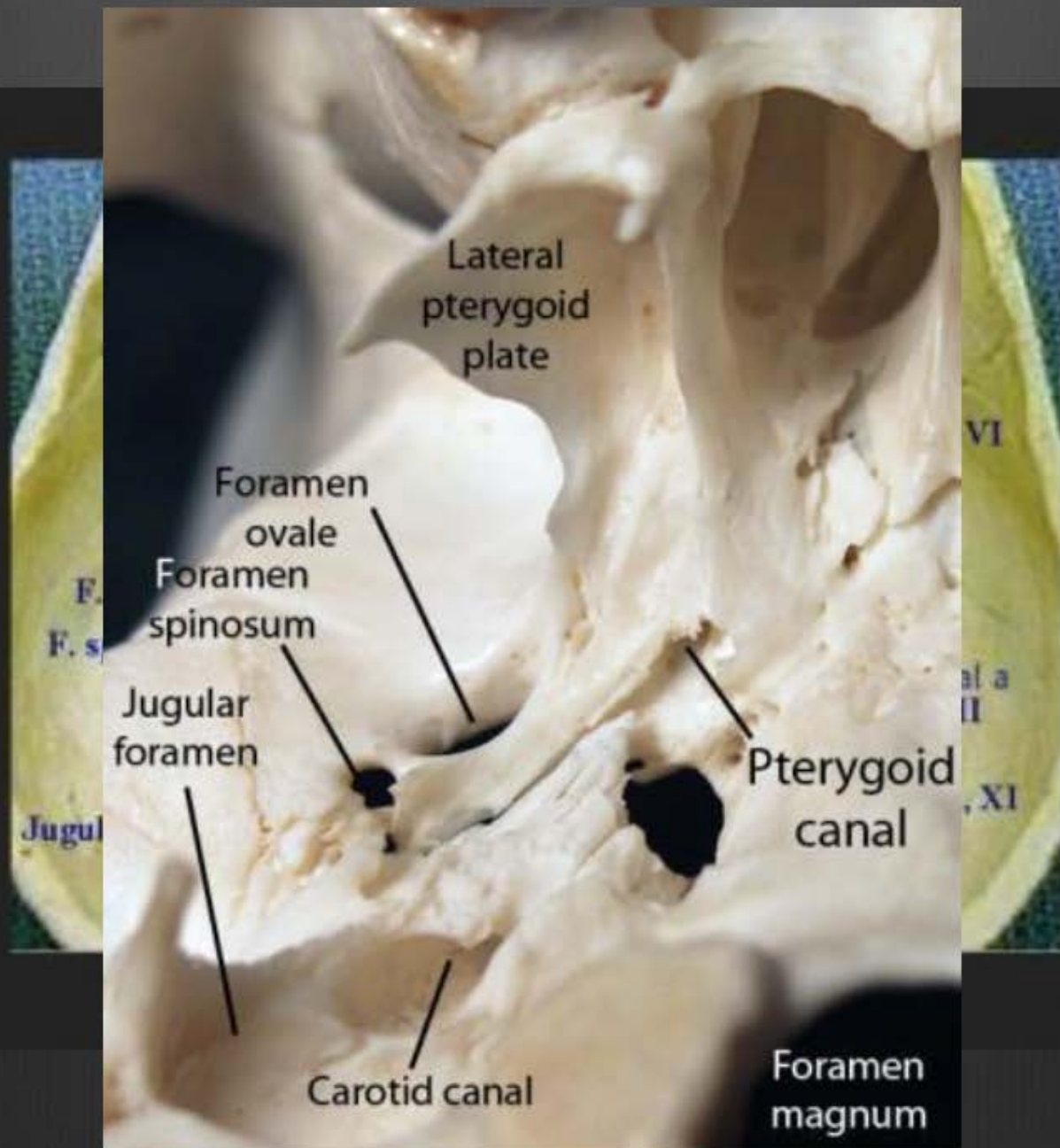
row was age-related and successively changed from uniformly low to uniformly high signal intensity on T1-weighted images. By the age of 24 years, 95% of their patients had a clivus of uniformly high signal intensity. The rate of conversion was somewhat slower in our patients. The clivus was uniformly bright in only one third of our normal patients in the third decade. The proportion of uniformly bright clivus gradually increases with advancing age. The rate increased to 80% by the eighth decade and reached 100% in the ninth decade. Our results show that interindividual variations are large in the composition of clival marrow in an adult population and that the conversion of red to yellow marrow occurs in a predictable and orderly pattern.

The hallmark of MR signal intensity caused by diseased marrow is T1 prolongation. The major factors determining the T1 changes are increased cellularity associated with tumor replacement of normal marrow and increased water content owing to bone marrow edema [10].

A clivus with uniformly low signal intensity that is hypointense relative to the pons should be considered abnormal in an adult patient. Conversely, a clivus of uniformly bright signal intensity is highly unlikely to be abnormal. The uniformly low signal intensity is attributable to diffuse tumor invasion of the clivus or marrow reconversion. In the normal adult, hematopoietic needs are met by the amount of red marrow existing

normal clivi. When contrast material is used, normal and abnormal clivi generally show different patterns of enhancement.





Jugular Foramen

Foramen lacerum

Carotid canal

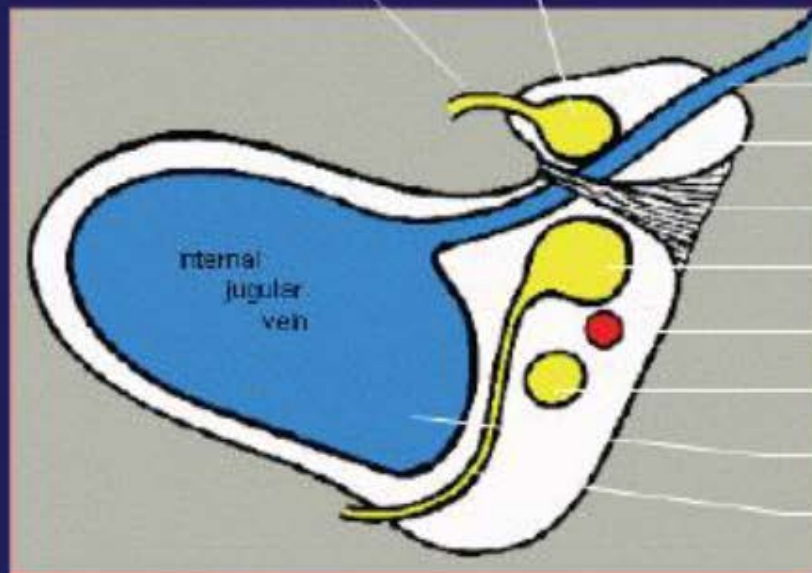
Pars Nervosa

Jugular spine

Pars vascularis



IX - Glossopharyngeal nerve
Jacobson's nerve



Inferior petrosal sinus

Pars Nervosa

Fibrous septum

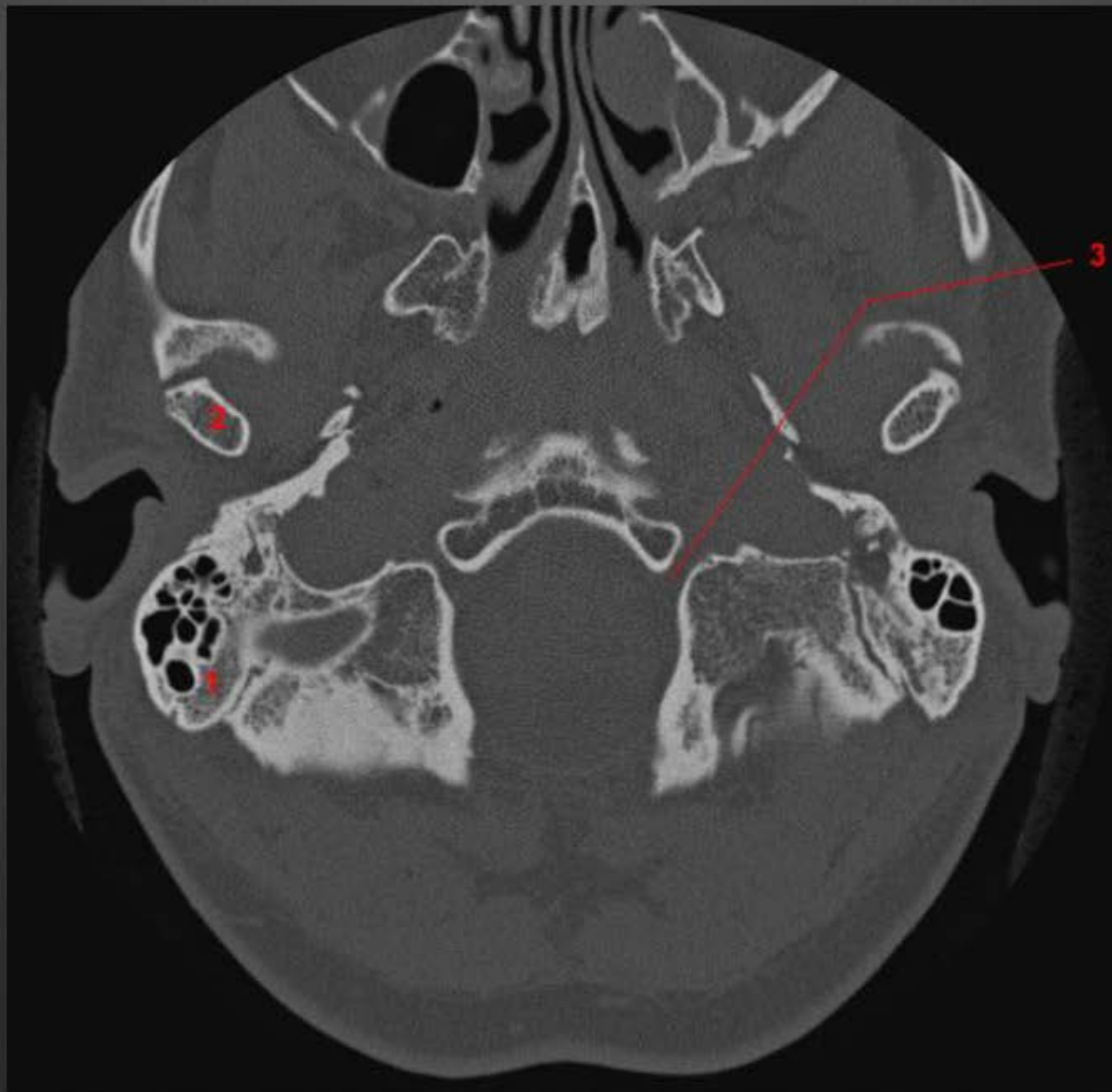
X - Vagus nerve

Posterior meningeal artery

XI - Spinal accessory nerve

Pars vascularis

Nerve of Arnold

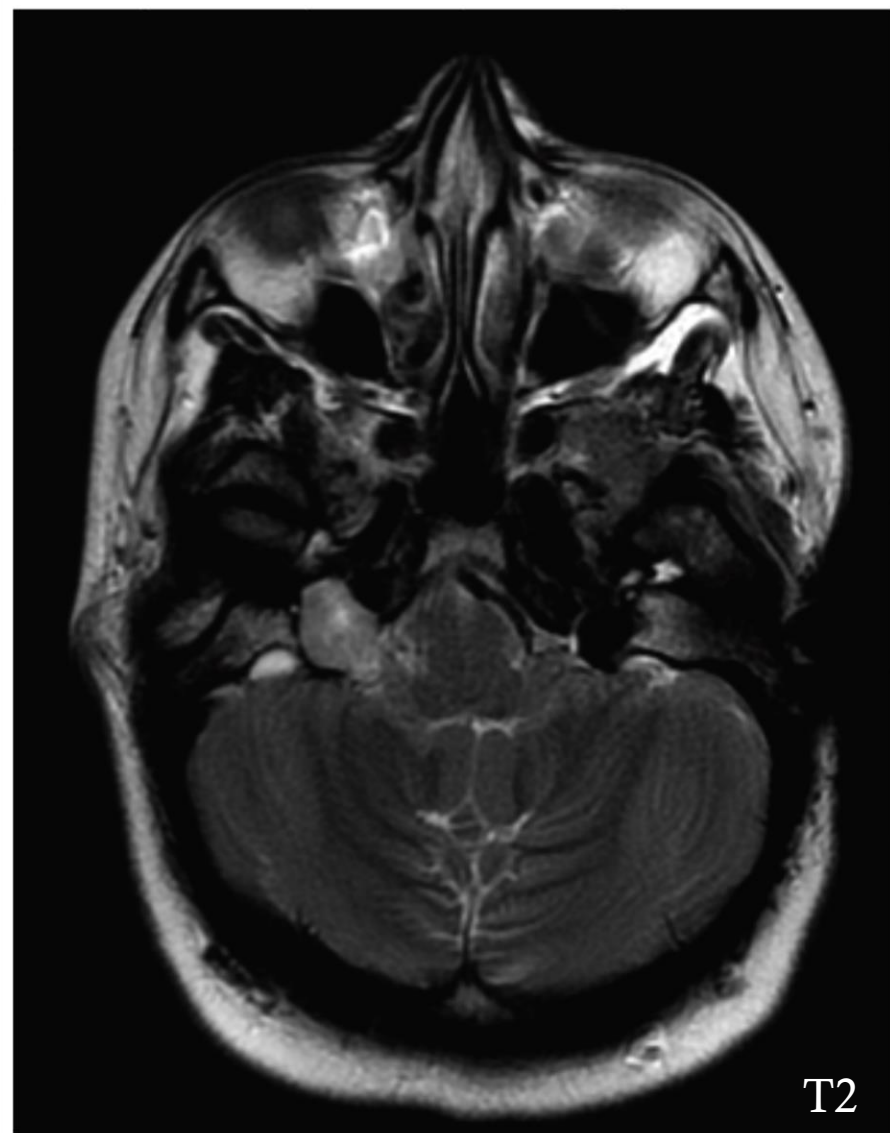
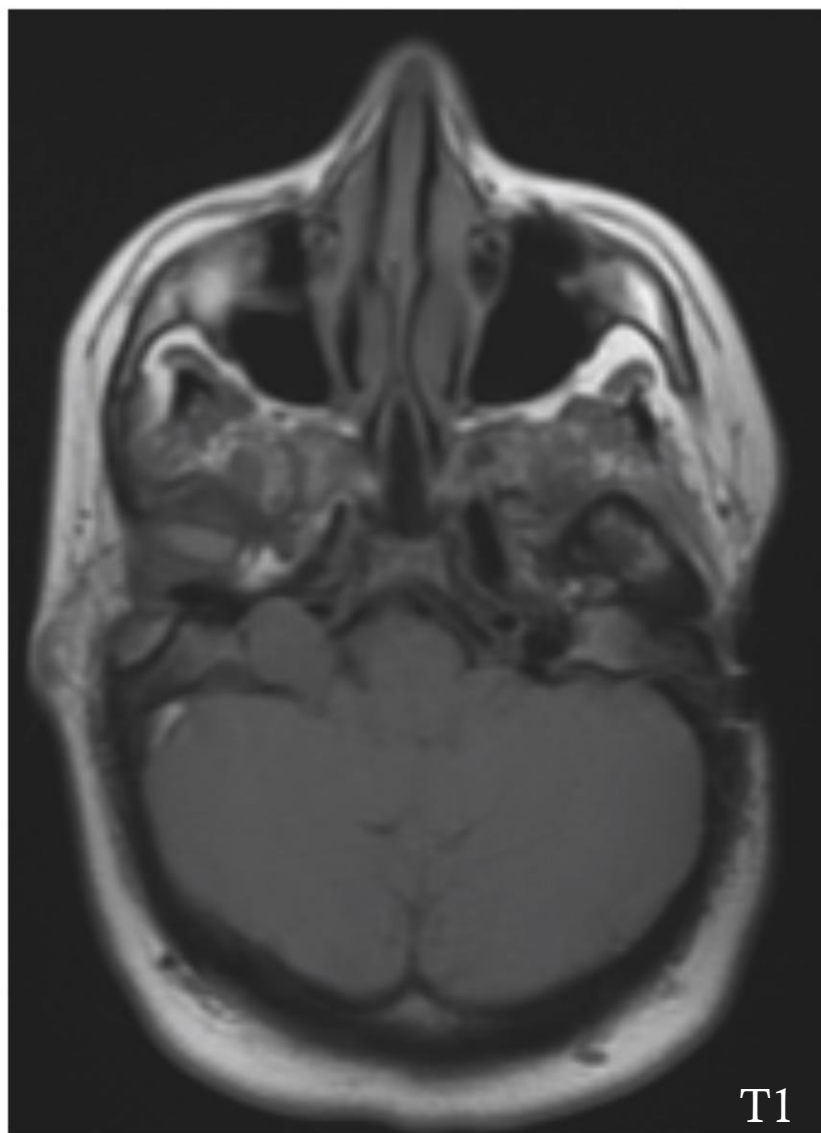


CASE 1

24-year-old woman with right side of tongue numbness, occasional hoarseness of her voice and difficulty swallowing.



IGNORE LEFT COCHLEAR IMPLANT



Lesions of the Jugular Foramen

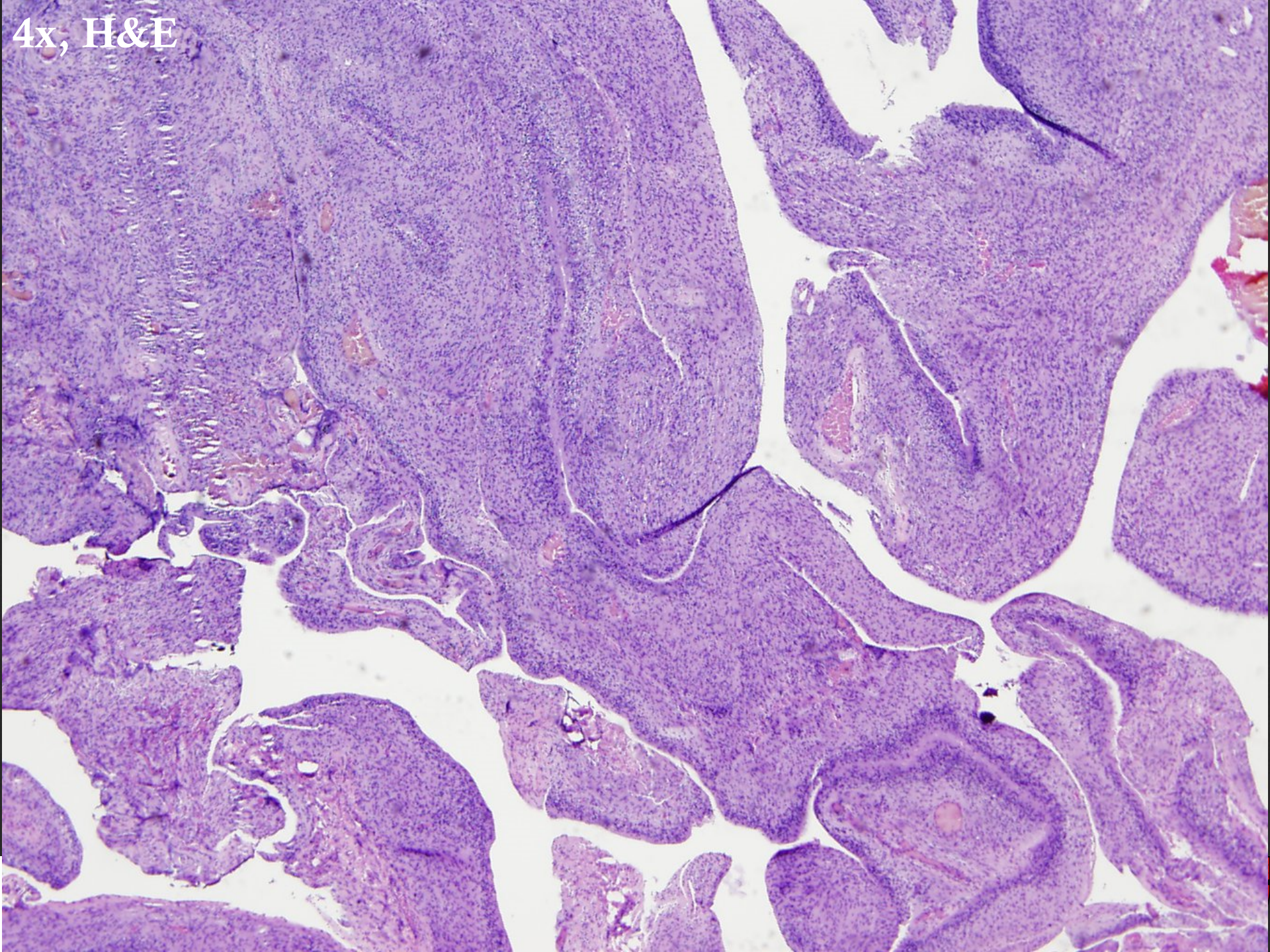
- Glomus Tumor
- Schwannoma or Neurofibroma
- Meningioma
- Nasopharyngeal Carcinoma
- Metastatic Disease
- Slow Flow
- Enlarged or high riding bulb; diverticulum

Pathology

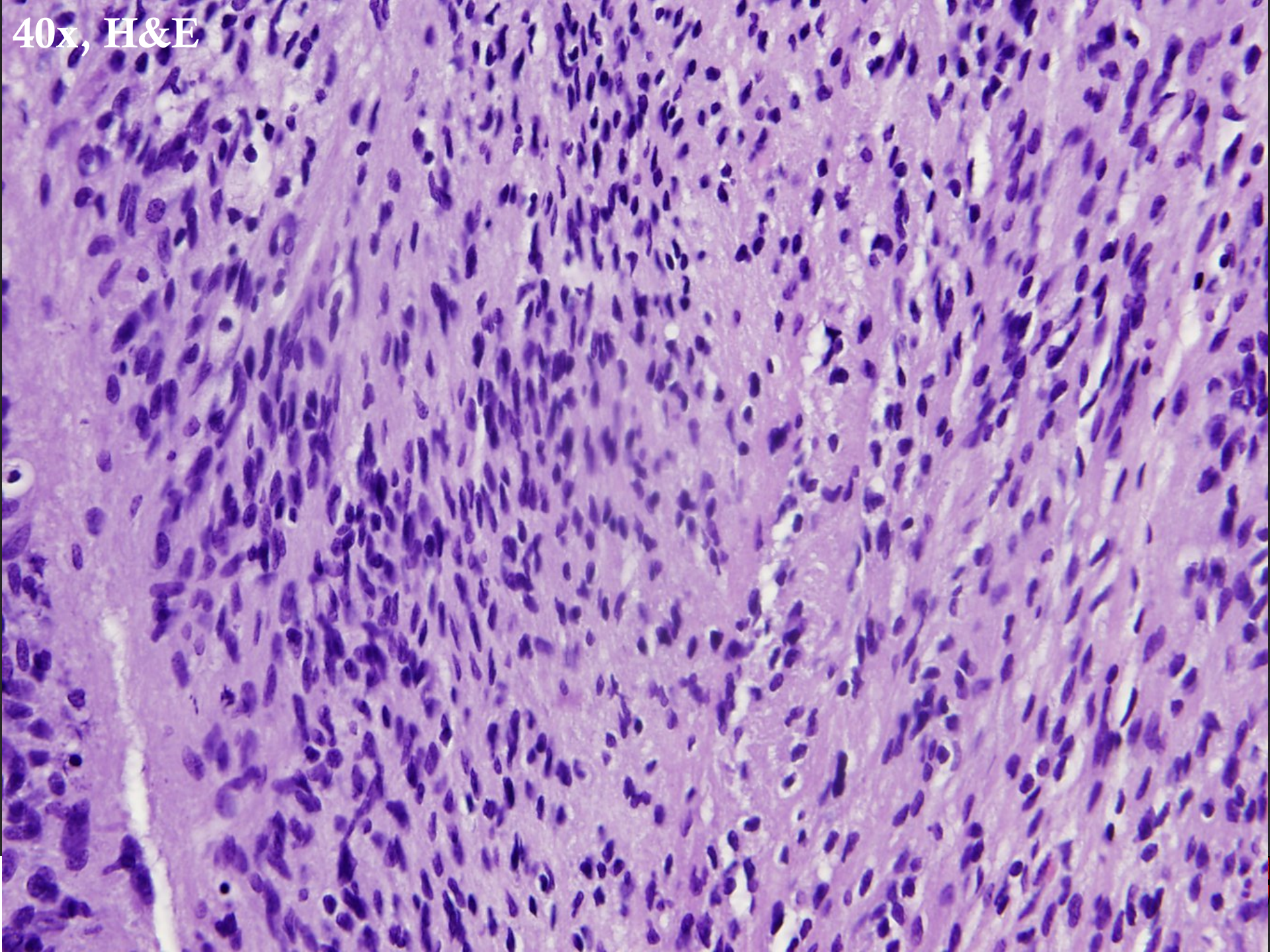


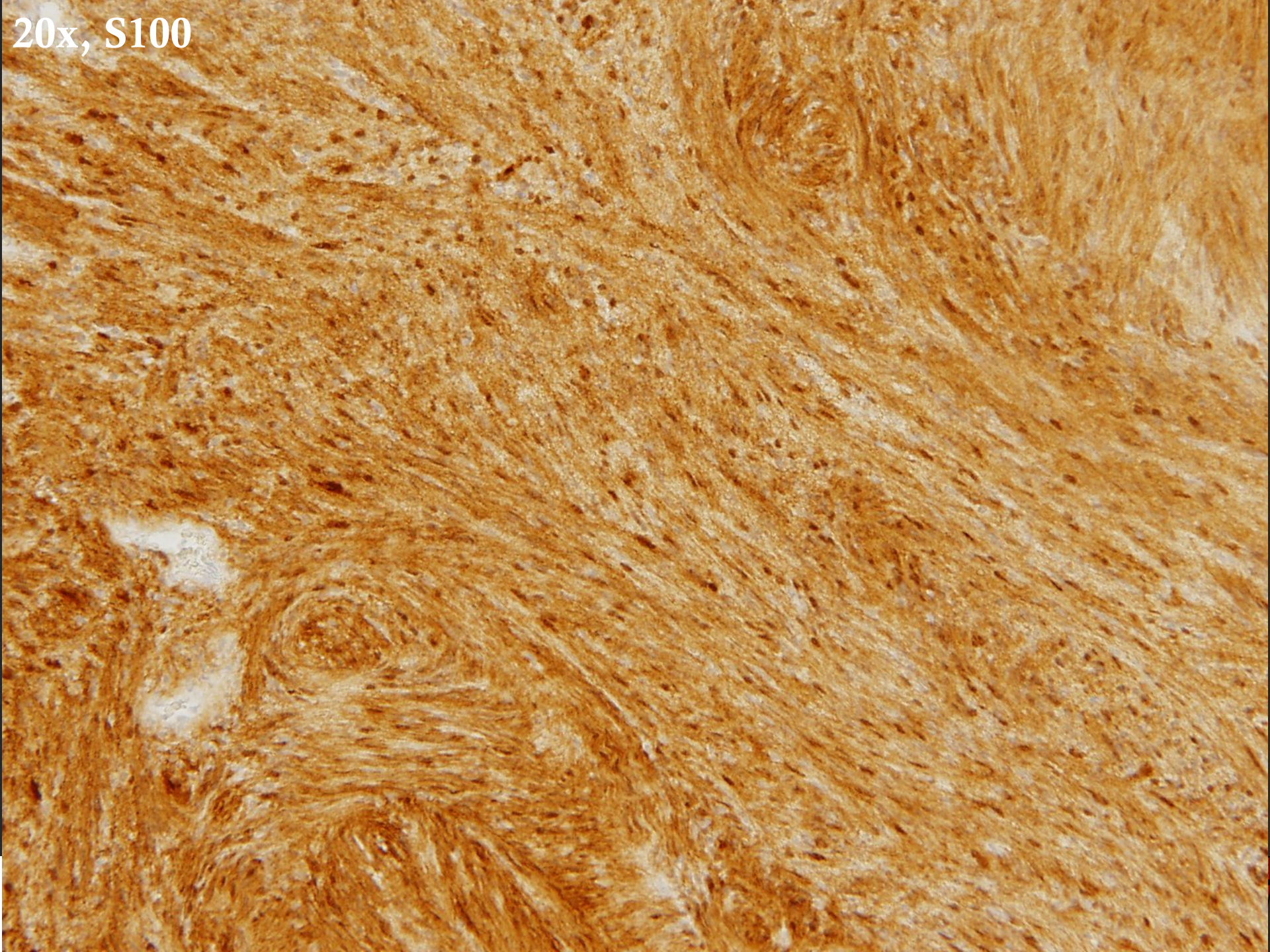


4x, H&E



40x, H&E





20x, S100

Final Pathologic Diagnosis

- **SCHWANNOMA**
 - Pos: S-100

Lesions of the Jugular Foramen

- Glomus Tumor
- Schwannoma or Neurofibroma
- Meningioma
- Nasopharyngeal Carcinoma
- Metastatic Disease
- Slow Flow
- Enlarged or high riding bulb; diverticulum

The Jugular Foramen: A Review of Anatomy, Masses, and Imaging Characteristics¹

Karen S. Caldemeyer, MD

Vincent P. Mathews, MD

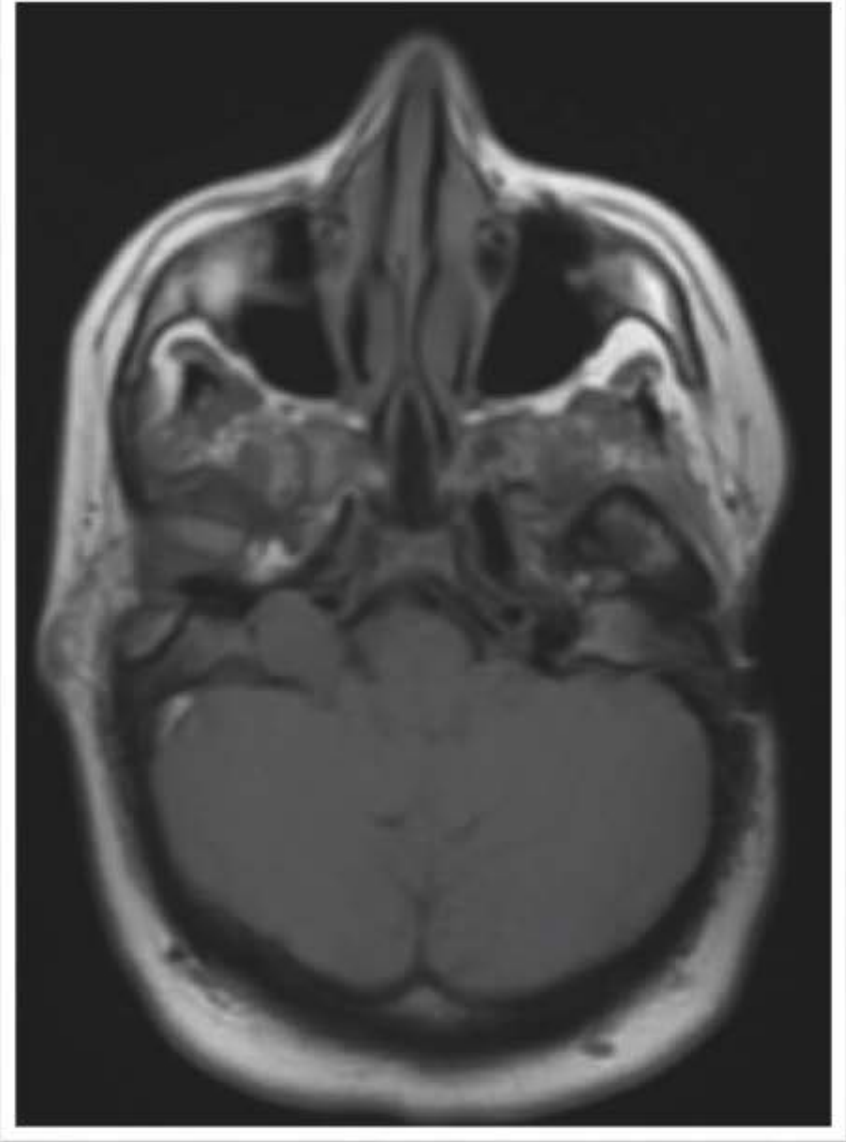
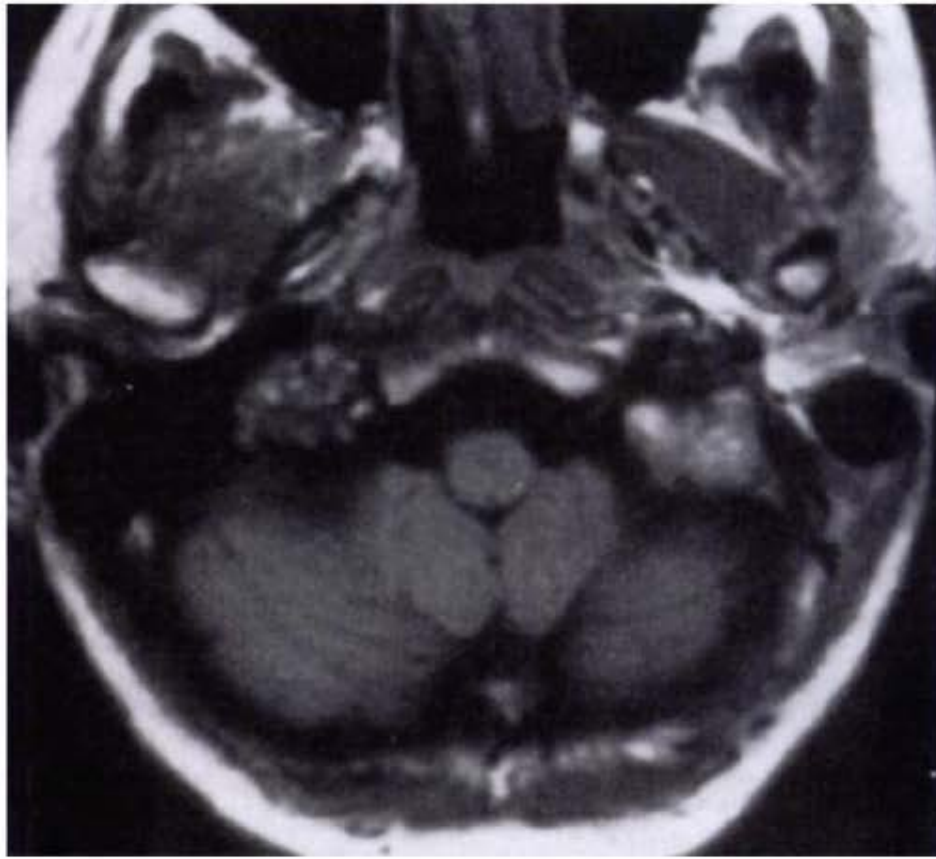
Biago Azzarelli, MD

Richard R. Smith, MD



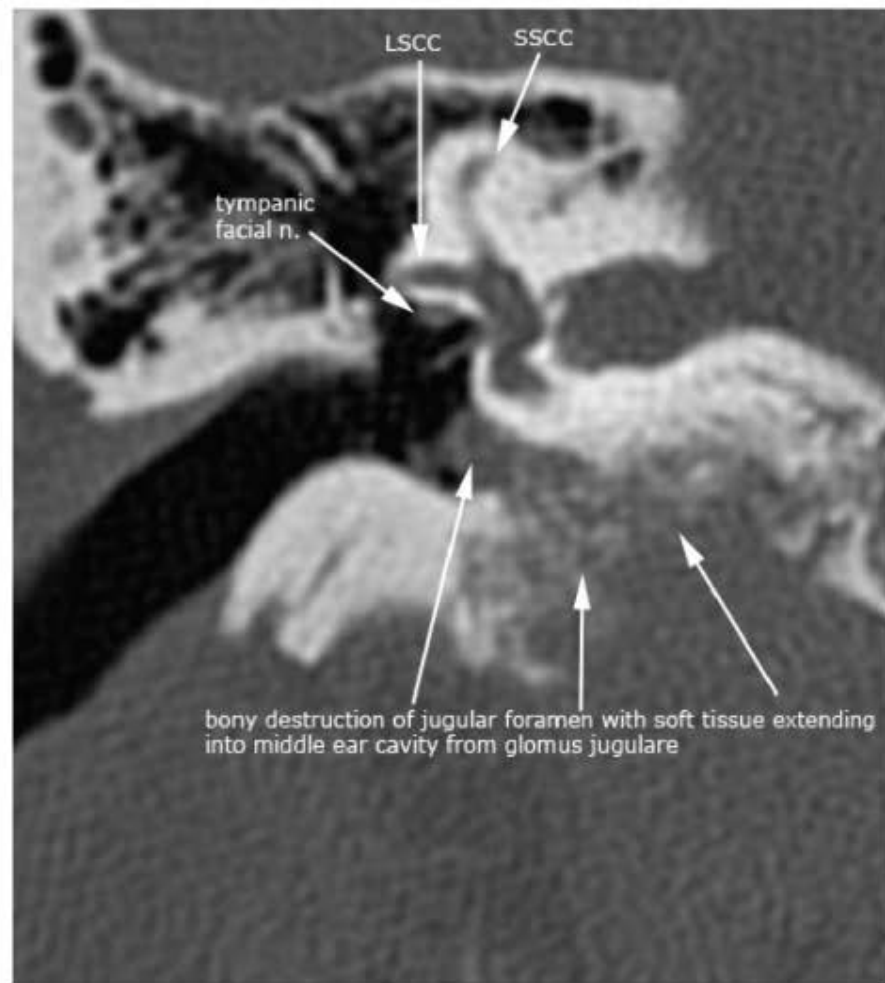
Glomus Tumor vs. Schwannoma

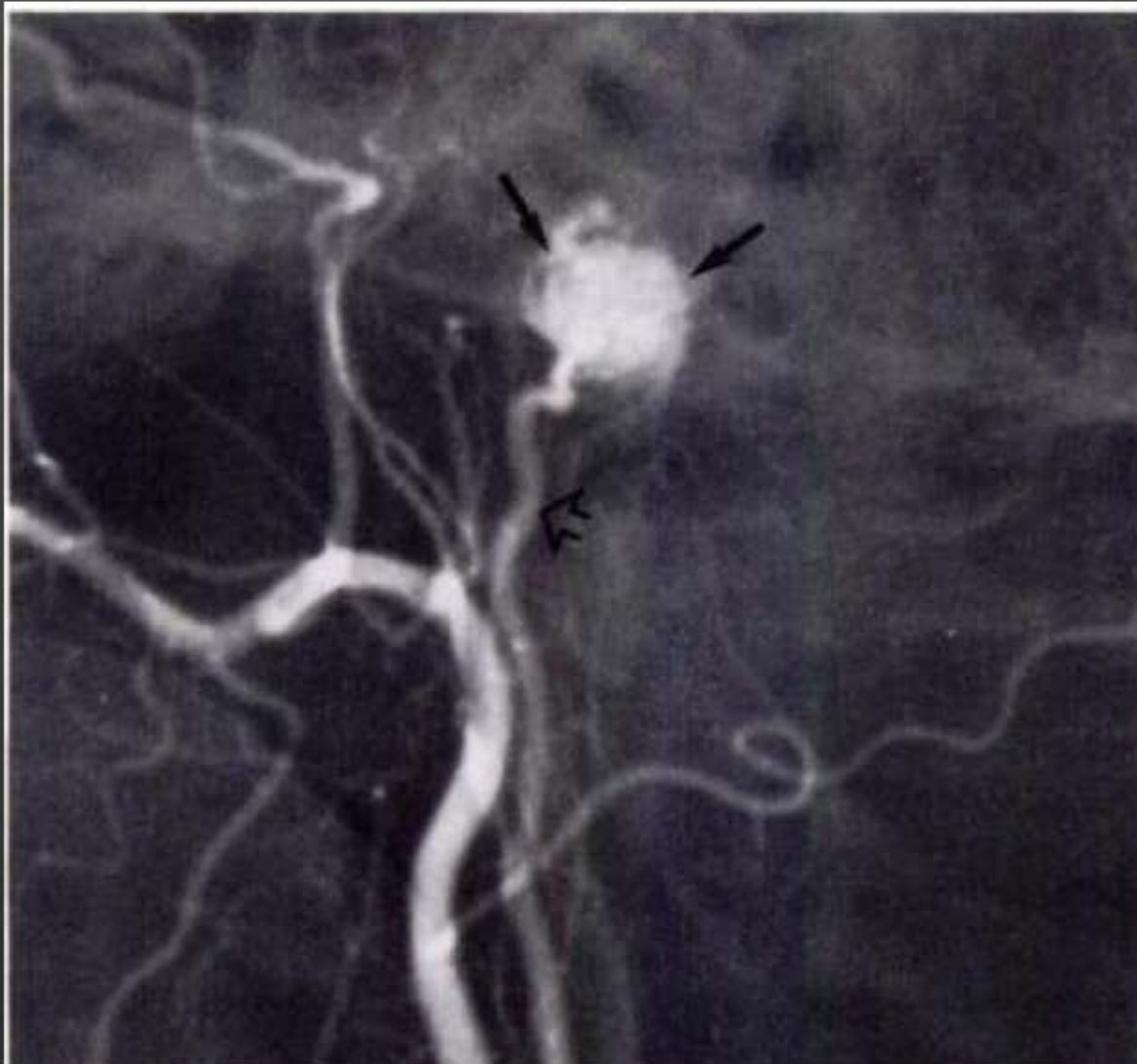
| | GLOMUS TUMOR | SCHWANNOMA |
|-----------------|--------------------------------------|--|
| CT | Irregular Bone Erosion Hyperdense | Bone Expansion Sharp Borders Sclerotic Rim Isodense |
| T1 (rel. to WM) | 'Salt and Pepper' | Hypo to Isointense |
| T2 (rel. to WM) | 'Salt and Pepper' | Hyperintense Occasionally Cystic |
| Enhancement | +++ Early Drop Out | + |
| Behavior | Invade vein | Compress vein Dumbell shape |

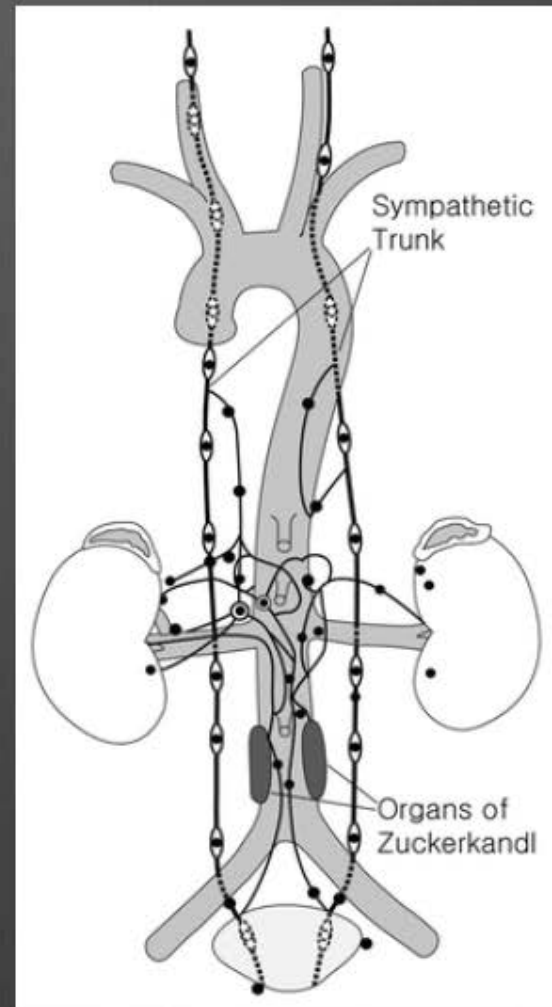
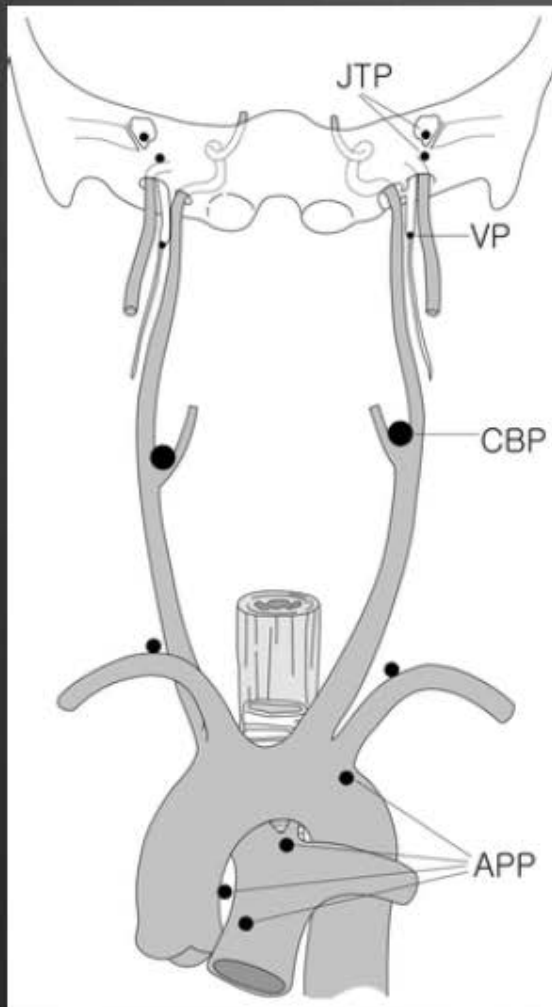


September-October 1997

Caldemeyer et al ■ *RadioGraphics*





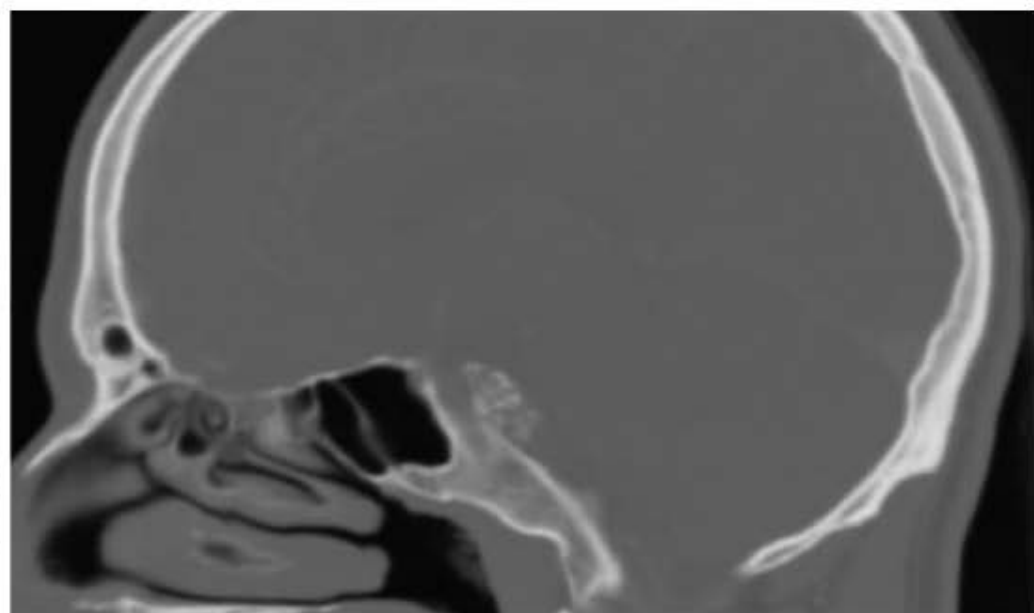


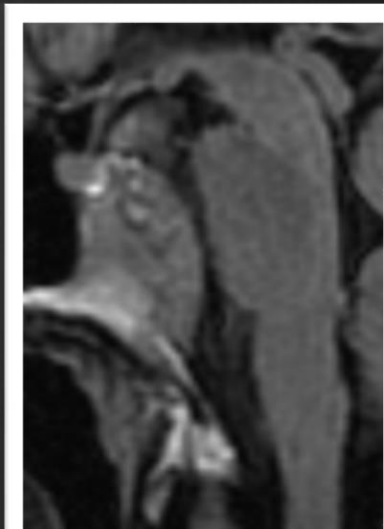
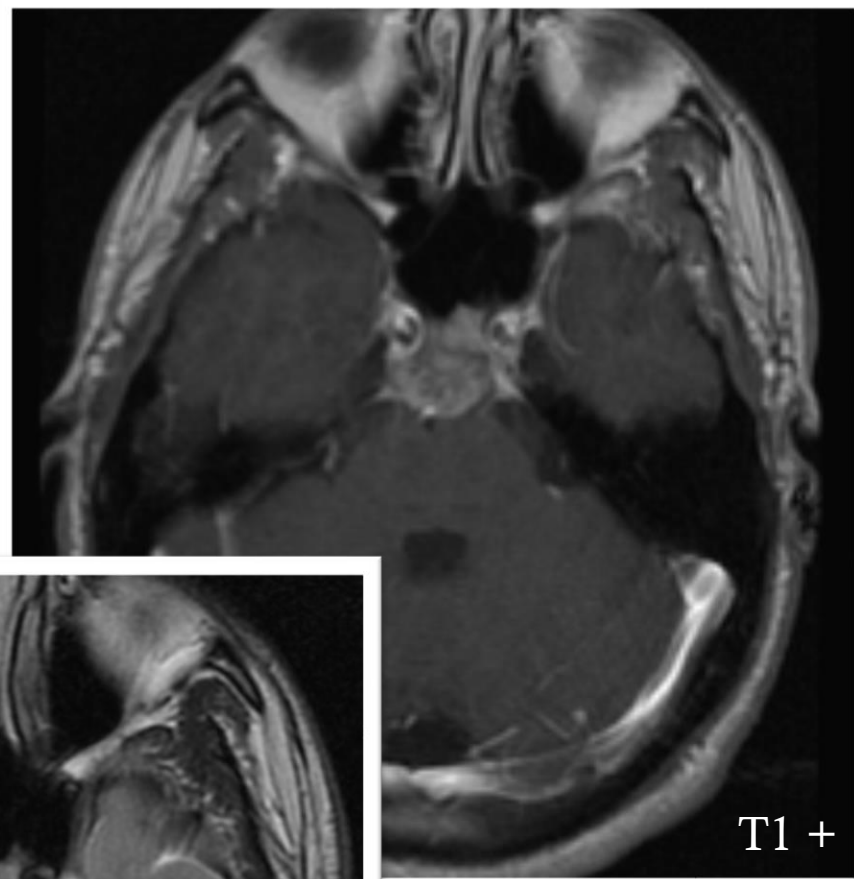
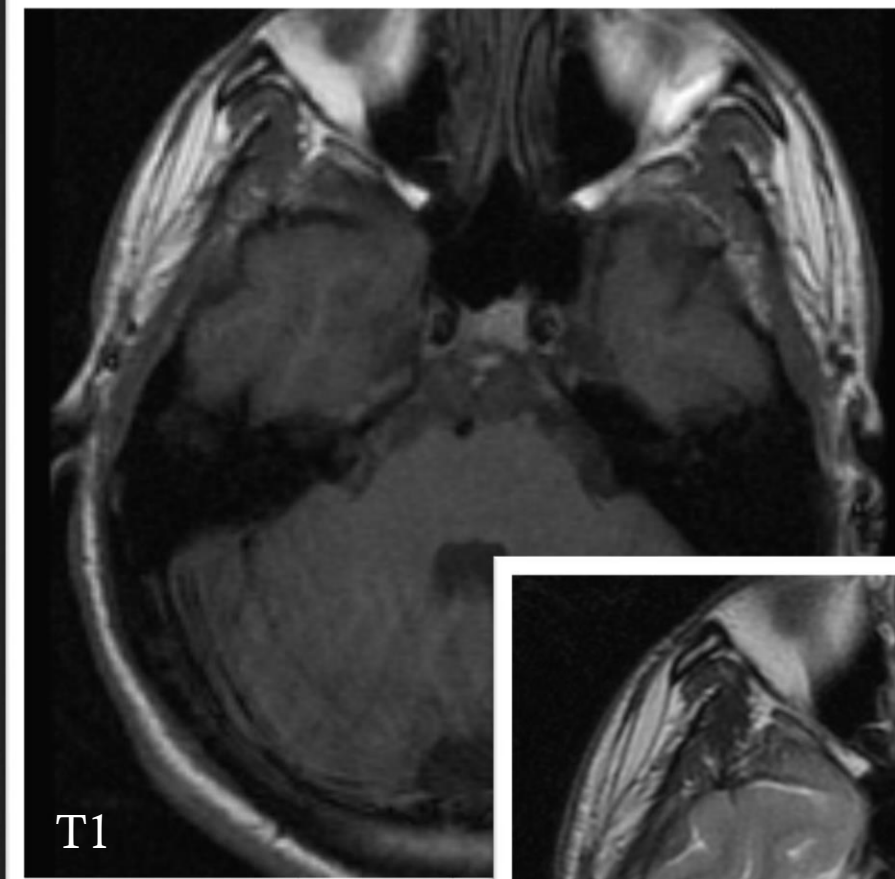
Lee et al.

AJR:187, August 2006

Case 2

22 year old male with diplopia
and mild headache





Lesions of Clivus

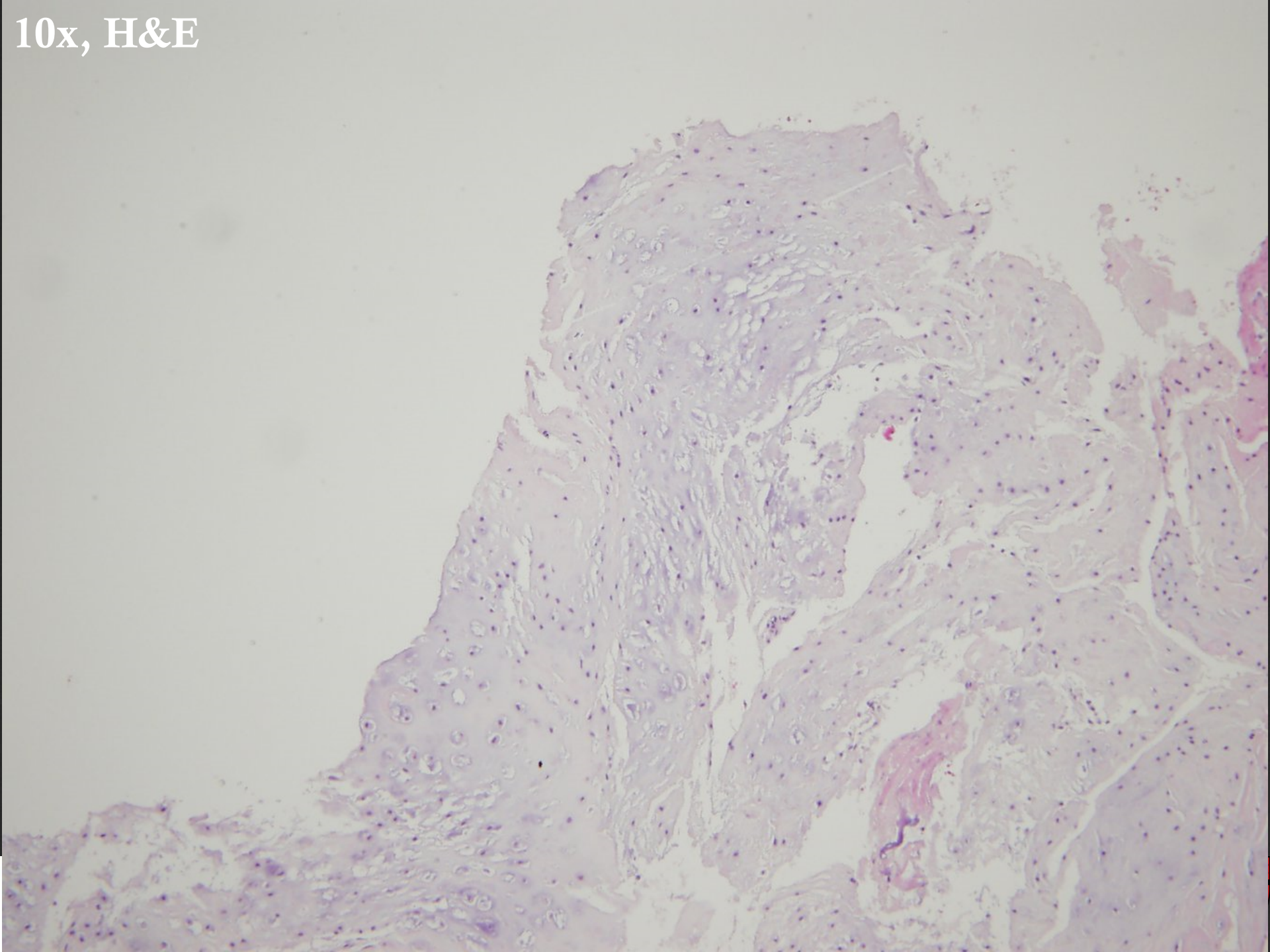
- Chondrosarcoma
- Chordoma
- Meningioma
- Plasmacytoma
- Metastasis
- Lymphoma
- Langerhans Histiocytosis
- Fibrous Dysplasia
- Infection

Pathology

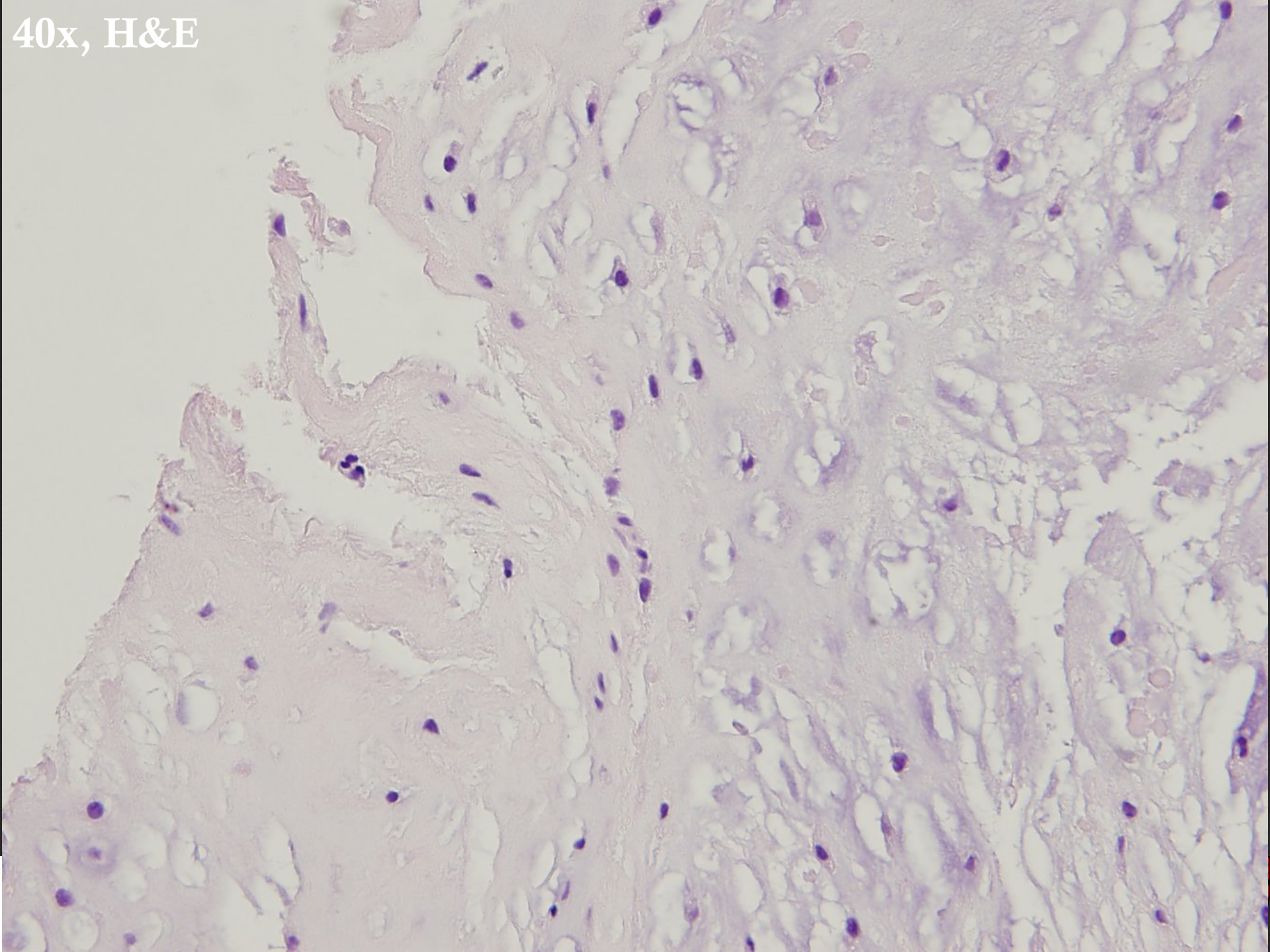


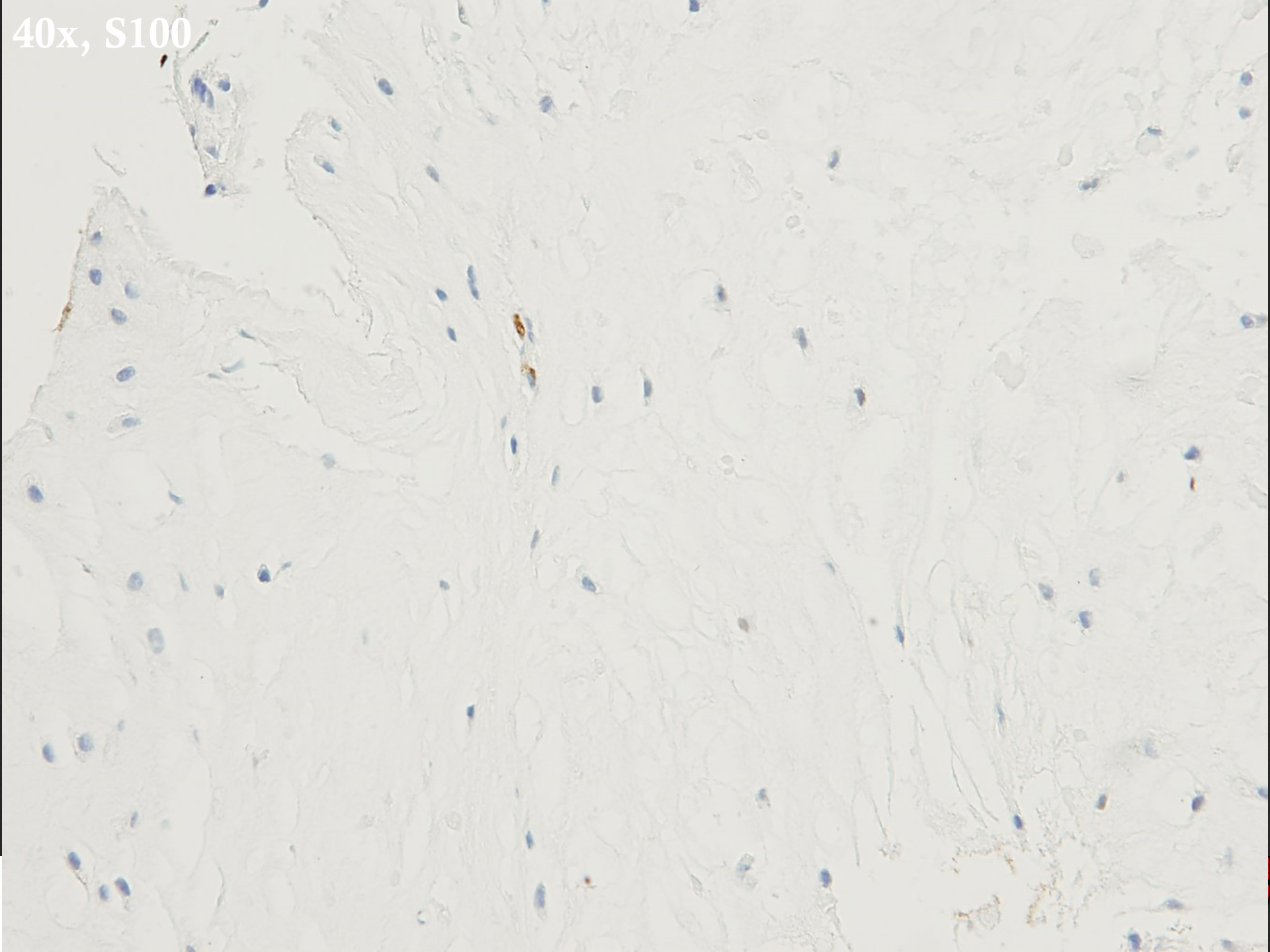


10x, H&E



40x, H&E





40x, S100

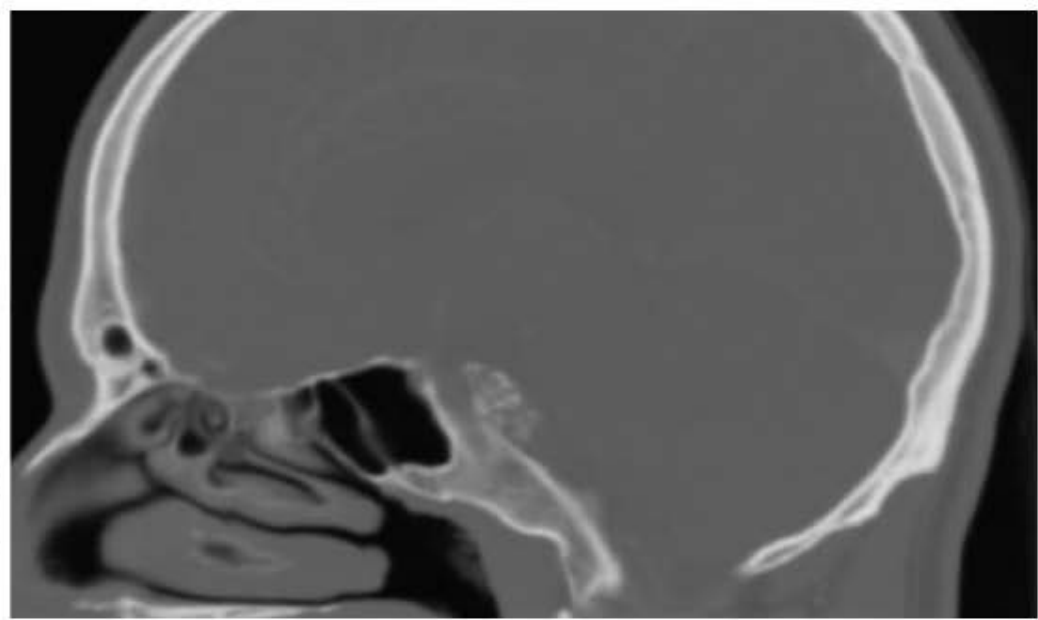
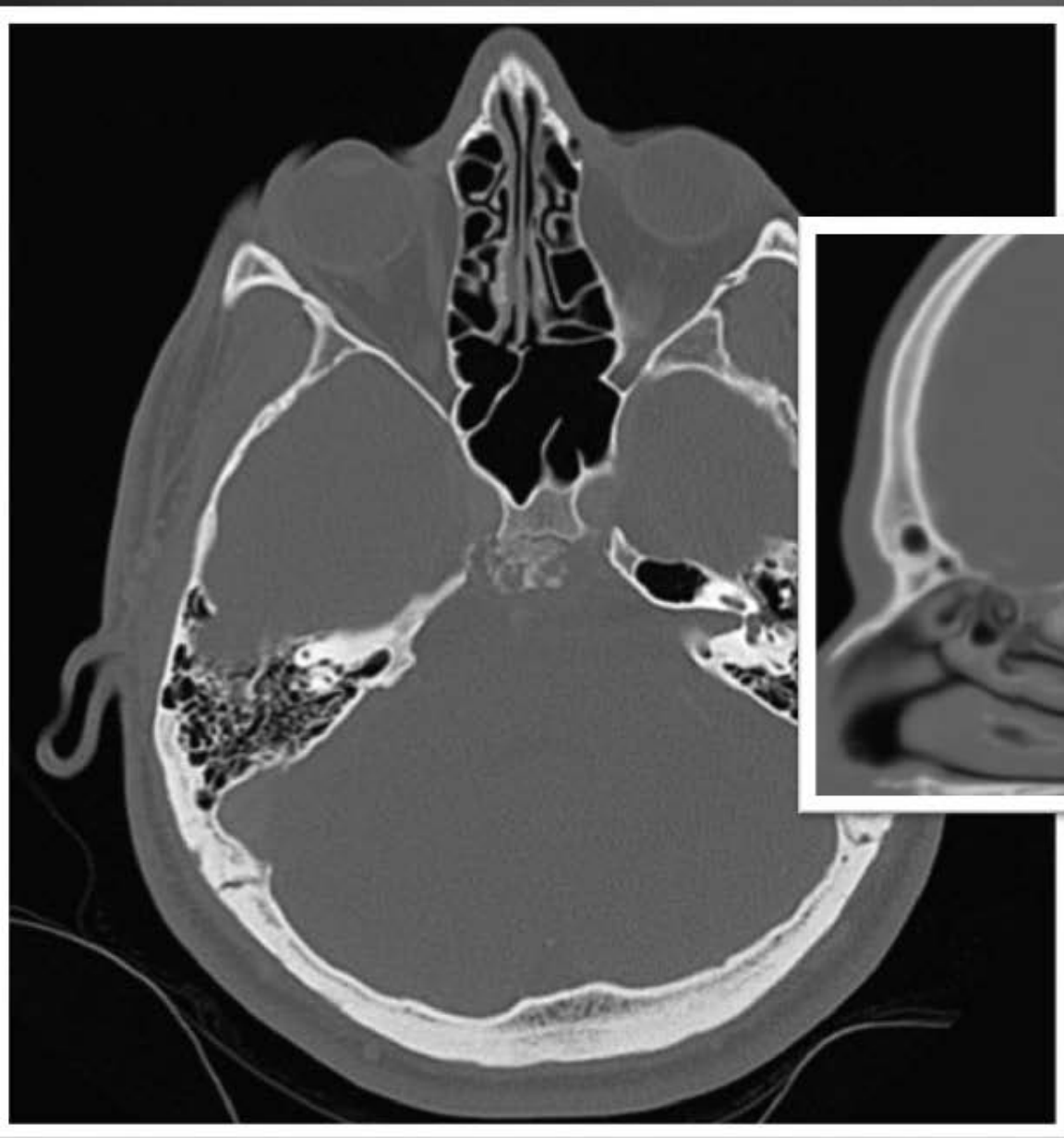
Final Pathologic Diagnosis

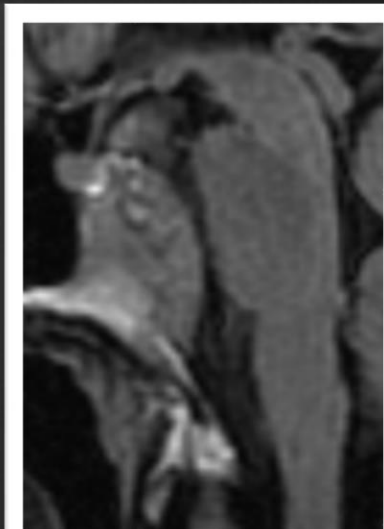
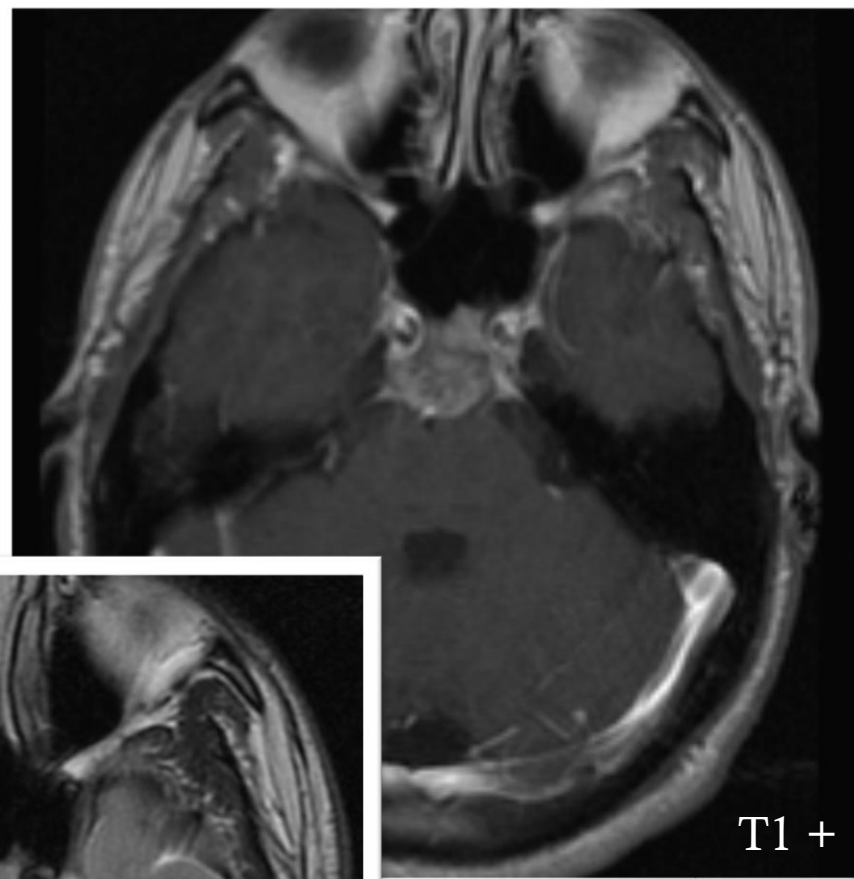
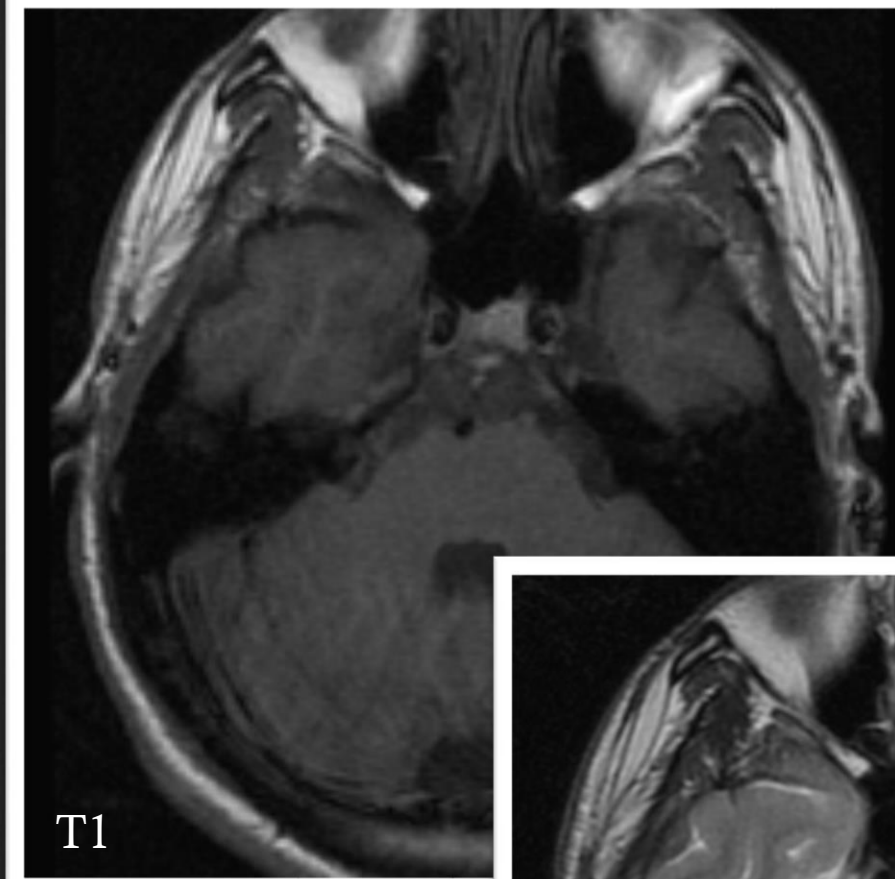
- **Well differentiated cartilaginous neoplasm, most c/w CHONDROSARCOMA**
 - Pos: S-100
 - Neg: EMA, Pan-K, CAM 5.2, AE1/AE3

Chondrosarcoma

- Mesenchymal origin malignant tumor with cells that produce cartilage matrix
 - Primary vs. Secondary
- Behavior is grade dependent

| | Chondrosarcoma | Chordoma |
|-----------------|---|--|
| CT | Rings and Arcs Linear Globular *Myxoid subtype | Well-circumscribed Variably Hyperattenuating Extensive lytic destruction *Chondriod subtype |
| T1 (rel. to WM) | Intermediate to Low | Intermediate to Low |
| T2 (rel. to WM) | Hyperintense | Hyperintense |
| Enhancement | +, heterogeneous | +, heterogeneous |
| Behavior | Less Common Majority arise from petro-occipital fissure Better prognosis | More common Midline typically Local Recurrence common Metastasis Rare |





DWI: Hot of the Press

Diffusion-Weighted MRI Distinction of Skull Base Chordoma and Chondrosarcoma

K.W. Yeom, I

Table 2: ADC values for each tumor type ($10^{-6} \text{ mm}^2/\text{s}$)

| Tumor | Mean ADC (Median) | Minimum ADC (Median) | Maximum ADC (Median) |
|--|-------------------|----------------------|----------------------|
| Chondrosarcoma (n = 9) | 2051 ± 262 (1977) | 1488 ± 360 (1352) | 2503 ± 512 (2392) |
| Classic Chordoma (n = 7) | 1474 ± 117 (1460) | 905 ± 118 (860) | 2199 ± 255 (2217) |
| Poorly Differentiated Chordoma (n = 3) | 875 ± 100 (871) | 491 ± 210 (469) | 1503 ± 127 (1557) |

3. Edwards, and N.J. Fischbein



RESULTS: Chondrosarcoma was a from classic chordoma (1474 ± 11 differentiated chordoma was char ment and/or lesion location did n

) and was significantly different $10^{-6} \text{ mm}^2/\text{s}$ ($P < .001$). Poorly IR imaging features of enhance-

Table 3: MRI features for each tumor type

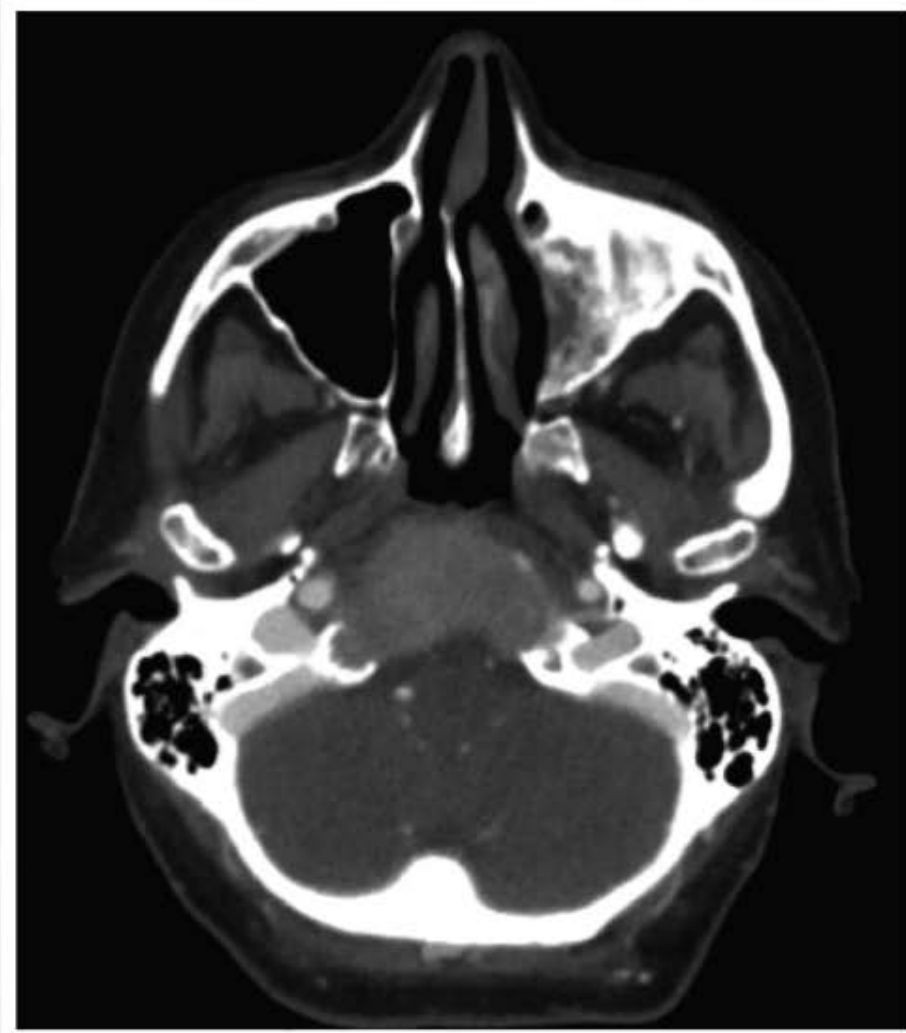
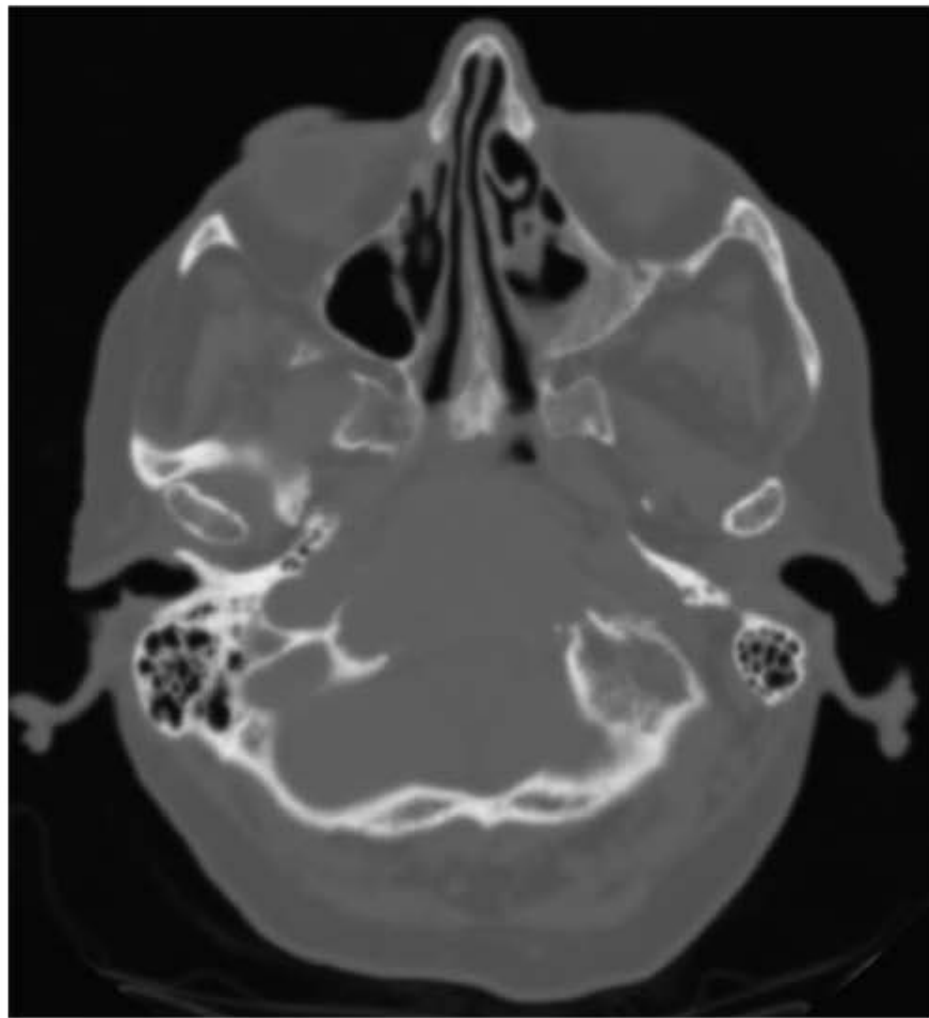
| Tumor | T2 Hypointensity | >90% Enhancement | Clival Location |
|--|------------------|------------------|-----------------|
| Chondrosarcoma (n = 9) | 0 (0%) | 5 (56%) | 4 (44%) |
| Classic chordoma (n = 7) | 0 (0%) | 1 (14%) | 4 (57%) |
| Poorly differentiated chordoma (n = 3) | 3 (100%) | 0 (0%) | 3 (100%) |

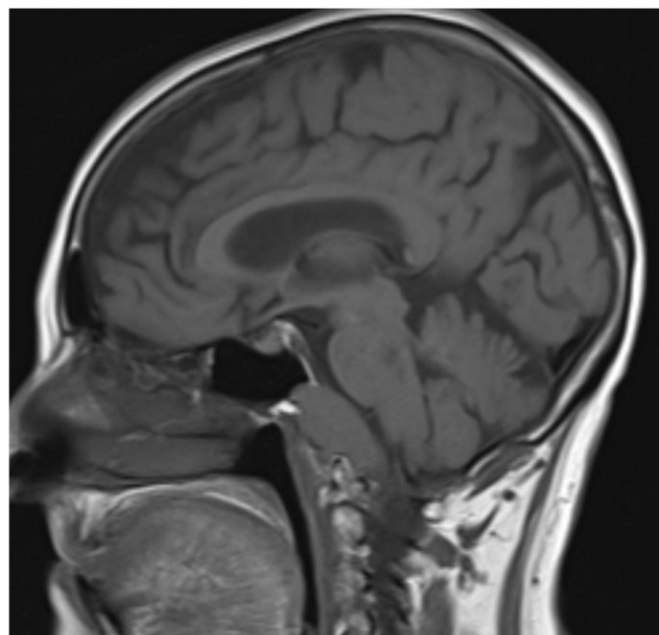
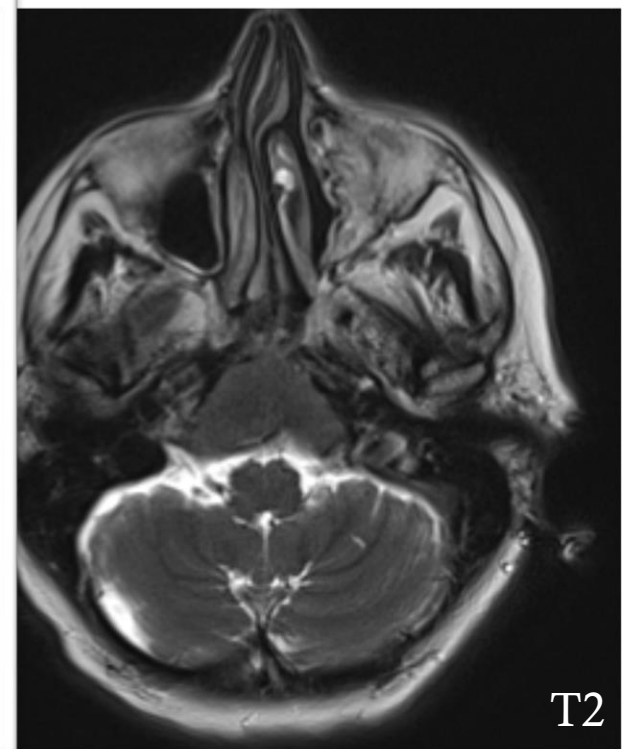
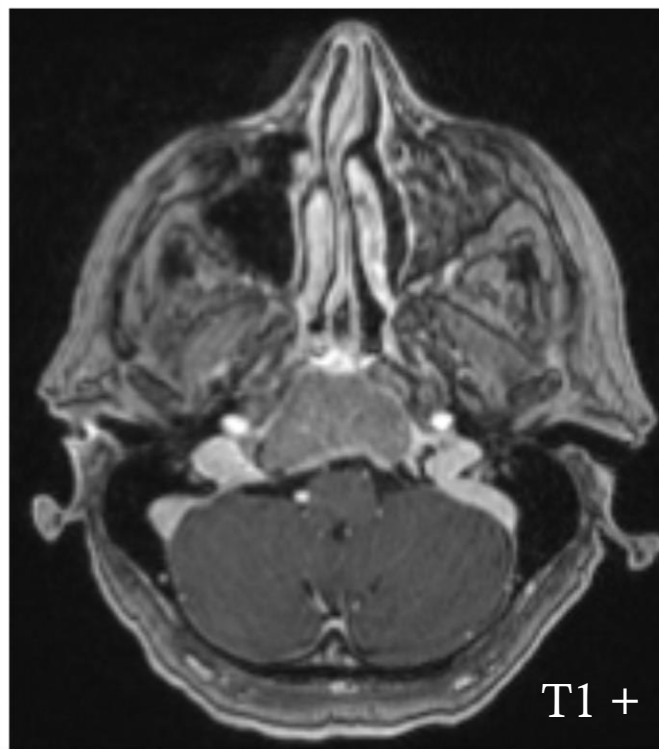
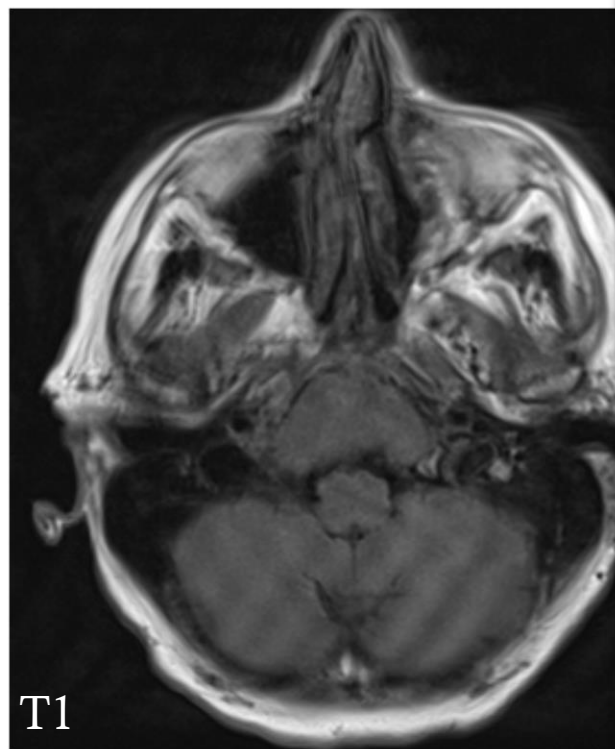
differentiating chordoma from C in predicting histopathologic

CONCLUSIONS: Diffusion-weight chondrosarcoma. A prospective s diagnosis.

Case 3

74 year old Female with fatigue
and back pain, followed by voice
changes.





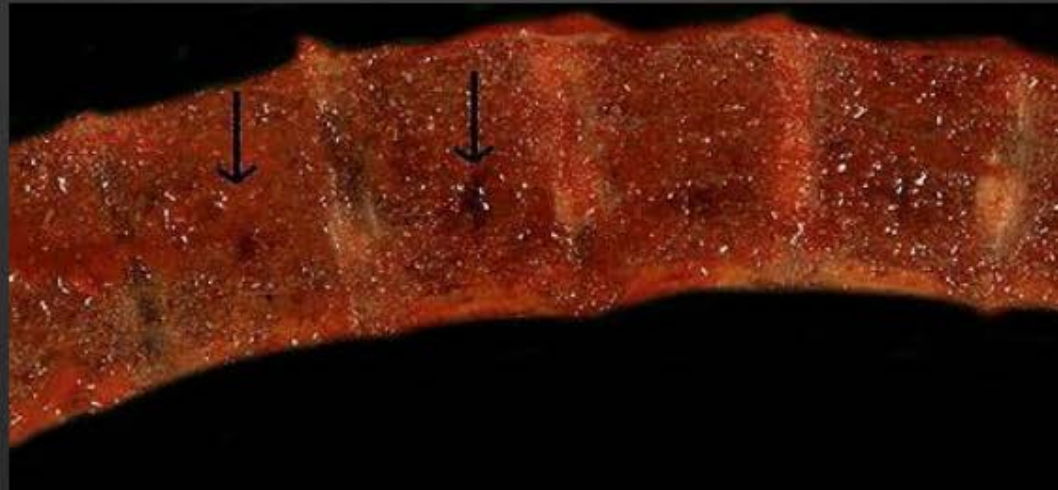


Lesions of Clivus

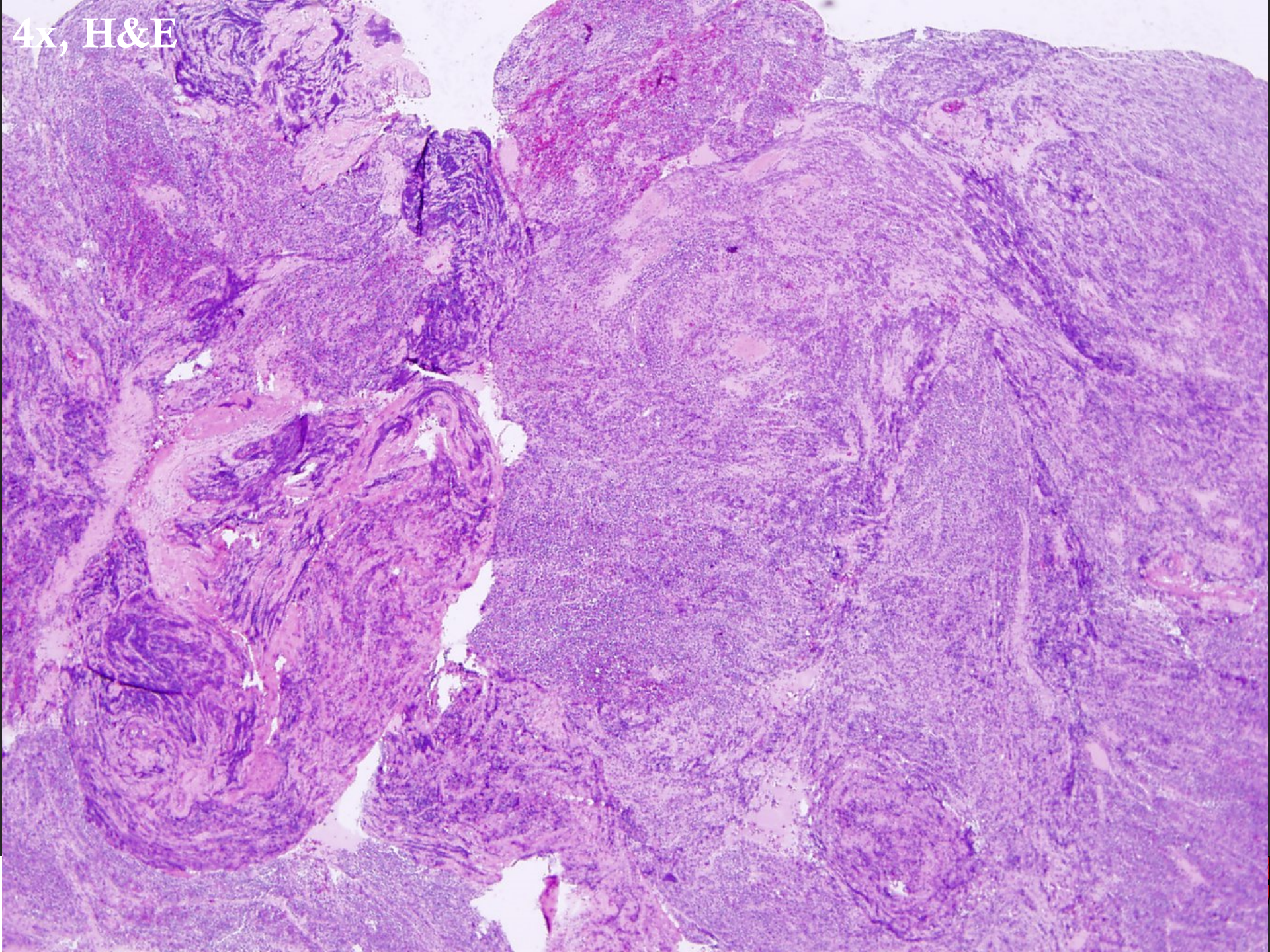
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- Chordoma
- Meningioma
- Plasmacytoma
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- Lymphoma
- Langerhans Histiocytosis
- Fibrous Dysplasia
- Infection

Pathology

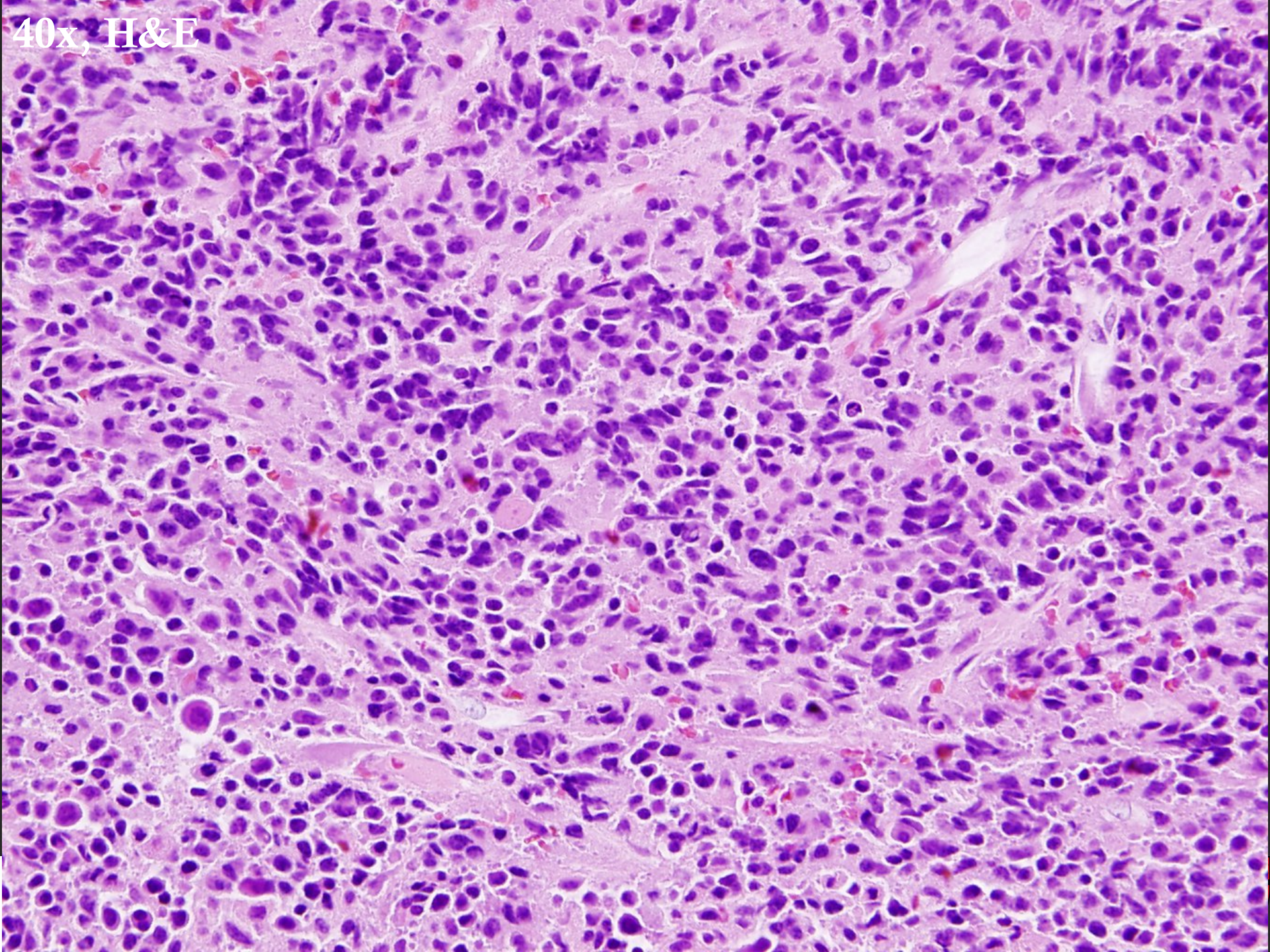




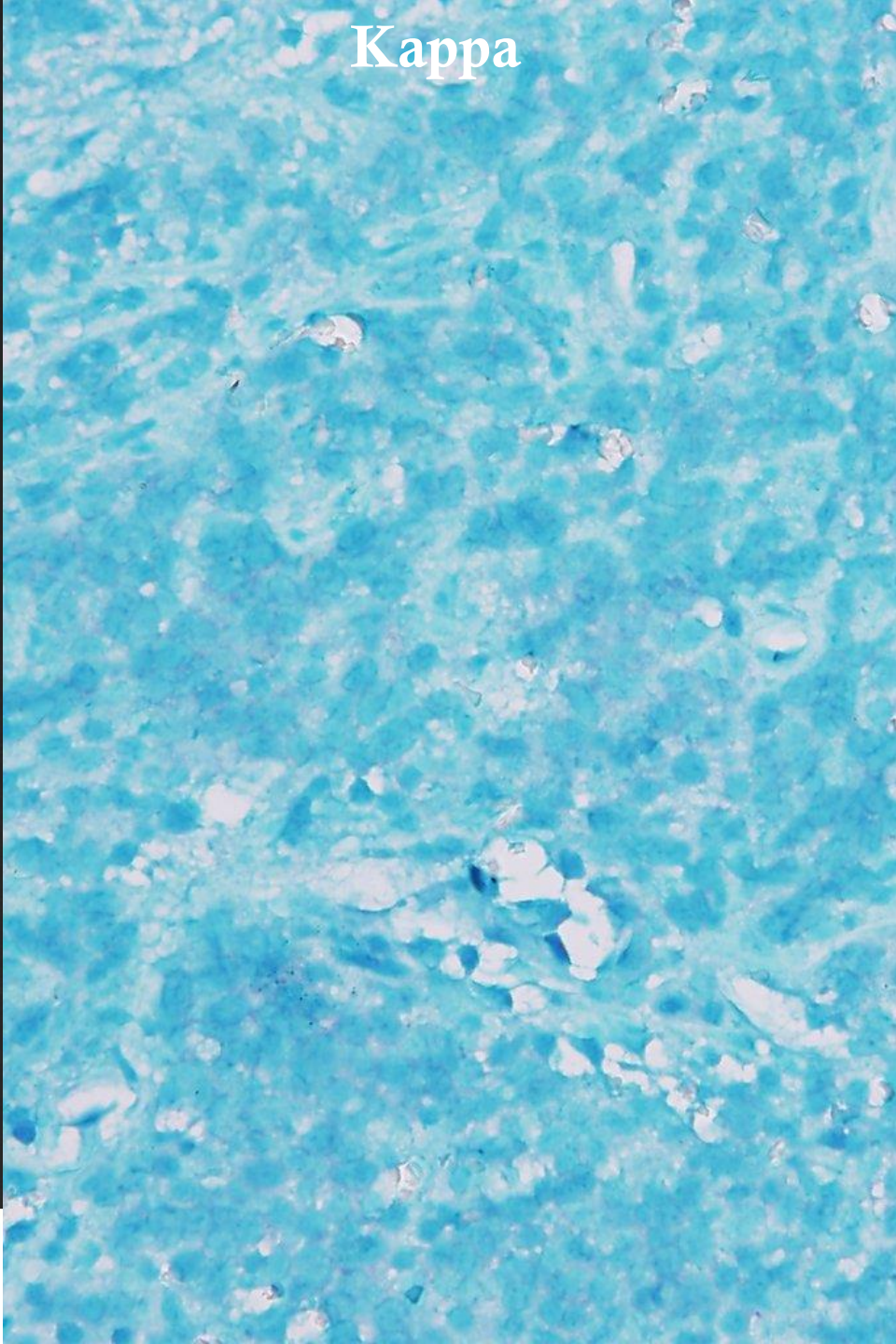
4x, H&E



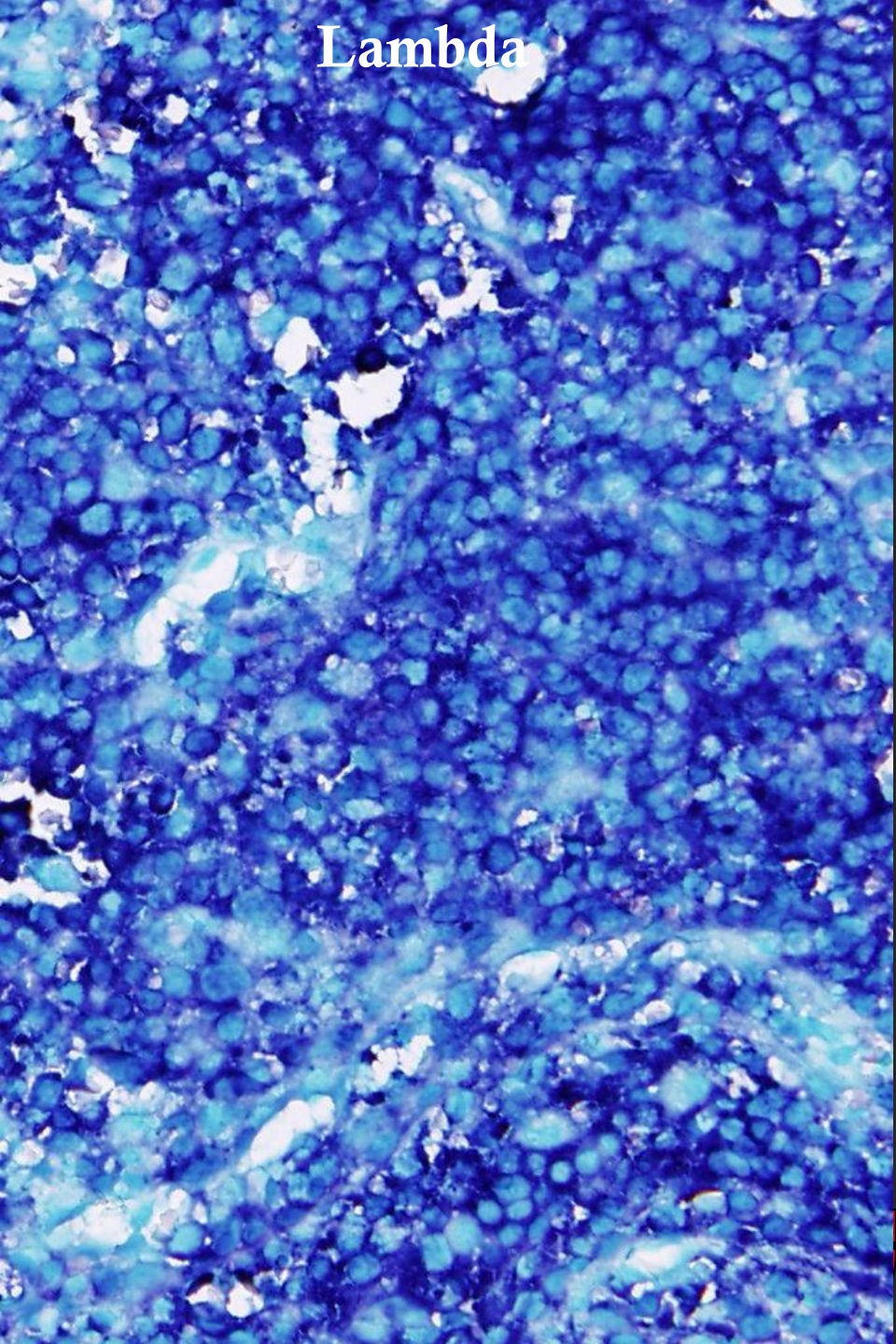
40x, H&E



Kappa



Lambda



Final Pathologic Diagnosis

- **PLASMA CELL NEOPLASM (multiple myeloma)**
 - **Cytogenetics:**
 - t(11;14) -> CyclinD1 – IgH fusion
 - Most common translocation in MM (also present in MCL)
 - **Paraproteins:**
 - No serum M spike
 - No urine free light chains

Multiple Myeloma

Review

Radiology

Edgardo J. C. Angtuaco, MD
Athanasios B. T. Fassas, MD
Ronald Walker, MD
Rajesh Sethi, MD
Bart Barlogie, MD

Index terms:
Bone marrow, diseases, 48.3452
Bones, CT, 48.12111
Bones, MR, 48.121411, 48.121412

Multiple Myeloma: Clinical Review and Diagnostic Imaging¹

Multiple myeloma (MM) is a malignant clonal neoplasm of plasma cells of B-lymphocyte origin that commonly results in overproduction of large amounts of monoclonal immunoglobulins. Important advances in the therapeutic management

Multiple Myeloma

- Estimates suggest 50% destruction must occur before radiographic demonstration
- 75% of MM patients will have positive findings
- Four forms of involvement
 - Plasmacytoma
 - Diffuse skeletal involvement (myelomatosis)
 - Diffuse skeletal osteopenia
 - Sclerosing myeloma



TABLE 1
Durle-Salmon Staging System for MM

| Stage and Criteria | Value* |
|-------------------------------|---|
| I: Low tumor burden† | |
| Hemoglobin level | >10 g/dL (100 g/L) |
| Serum calcium level | <12 mg/dL (3 mmol/L) |
| → Radiograph | No bone destruction, or solitary plasmacytoma |
| Low paraprotein level | |
| Serum IgG | <5 g/dL (0.05 g/L) |
| Serum IgA | <3 g/dL (0.03 g/L) |
| Urine light chain | <4 g/24 h |
| II: Intermediate tumor burden | |
| All criteria | Between values for stage I and values for stage III |
| III: High tumor burden‡ | |
| Hemoglobin level | <8.5 g/dL (85 g/L) |
| Serum calcium level | >12 mg/dL (3 mmol/L) |
| → Radiograph | More than two advanced lytic lesions |
| High paraprotein level | |
| Serum IgG | >7 g/dL (0.07 g/L) |
| Serum IgA | >5 g/dL (0.05 g/L) |
| Urine light chain | >12 g/24 h |
| Associated renal involvement | |
| A: serum creatinine level | <2 mg/dL (177 µmol/L) |
| B: serum creatinine level | >2 mg/dL |

* Value in parentheses is in SI unit.

† All criteria must be satisfied.

‡ Any criterion must be satisfied.

TABLE 2**Patterns of Normal Signal Intensity on T1-weighted MR Images of Spine in Different Age Groups**

| Pattern and Site | Age Distribution |
|---|------------------|
| 1: Hypointense | |
| Cervical | 92% <40 y |
| Thoracic | 70% <30 y |
| Lumbar | 99% <30 y |
| 2: Hypointense with bandlike and triangular hyperintensity at endplates | |
| Cervical | 87% >40 y |
| Thoracic | 88% >50 y |
| Lumbar | 86% >40 y |
| 3: Hyperintense* | |
| Cervical | 75% >50 y |
| Thoracic | Uniform |
| Lumbar | 76% >40 y |

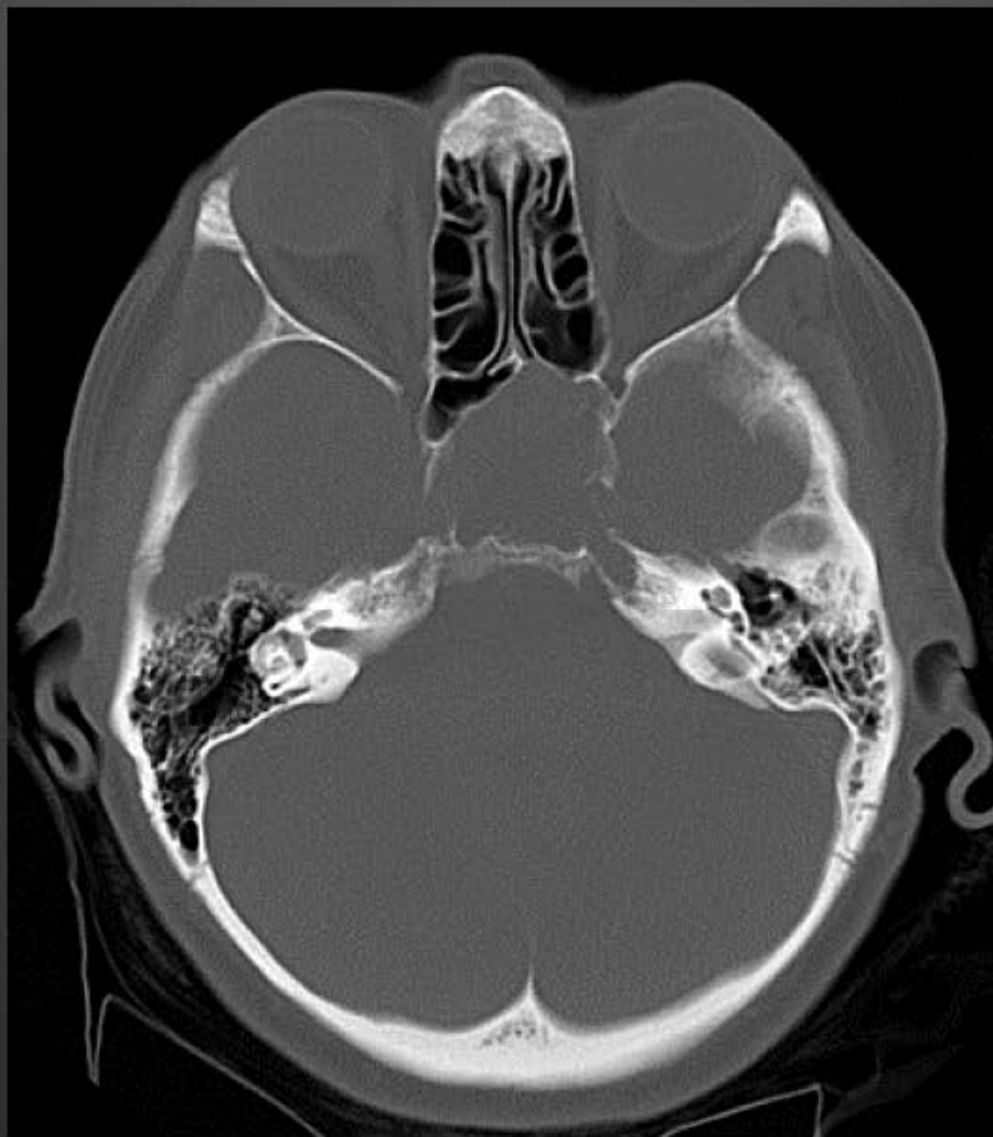
* Pattern 3a is homogeneous; pattern 3b is heterogeneous.

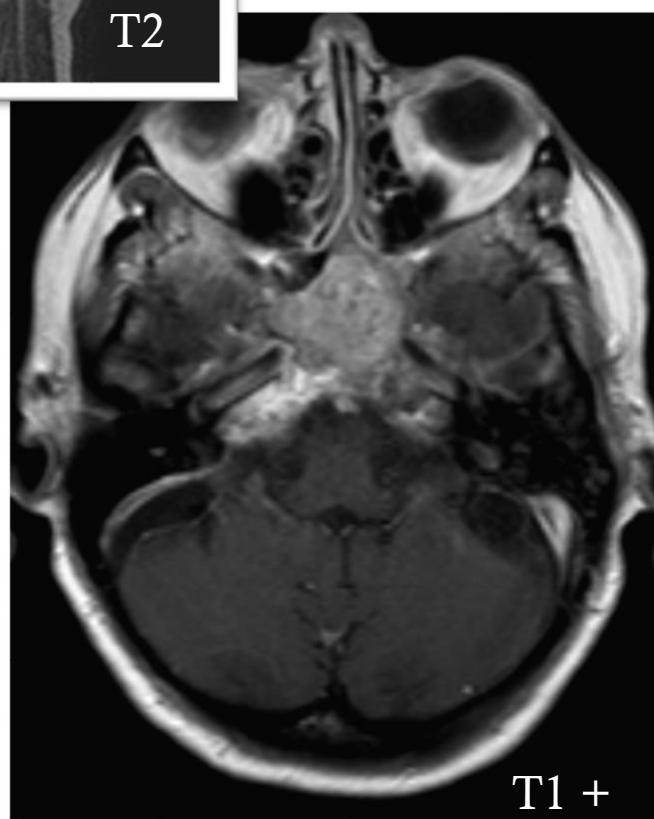
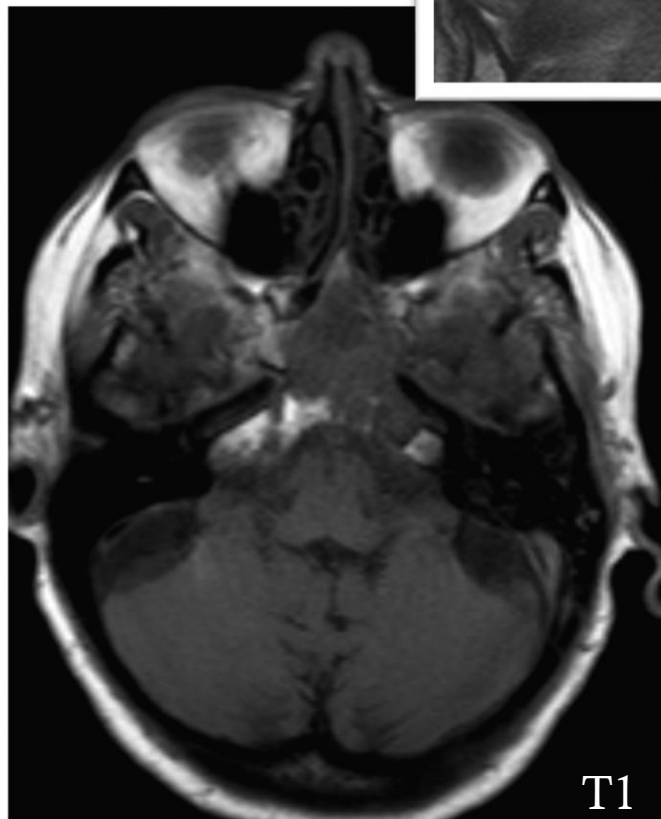
Plasmacytoma

- Lytic lesion that effects the spine, pelvis, skull, ribs, sternum, and proximal appendages
- T1 hypointense area within a generally hyperintense background
- T2 hyperintense area within a generally hypointense background
- Enhance

Case 4

82 year old Female with
diplopia





Lesions of Clivus

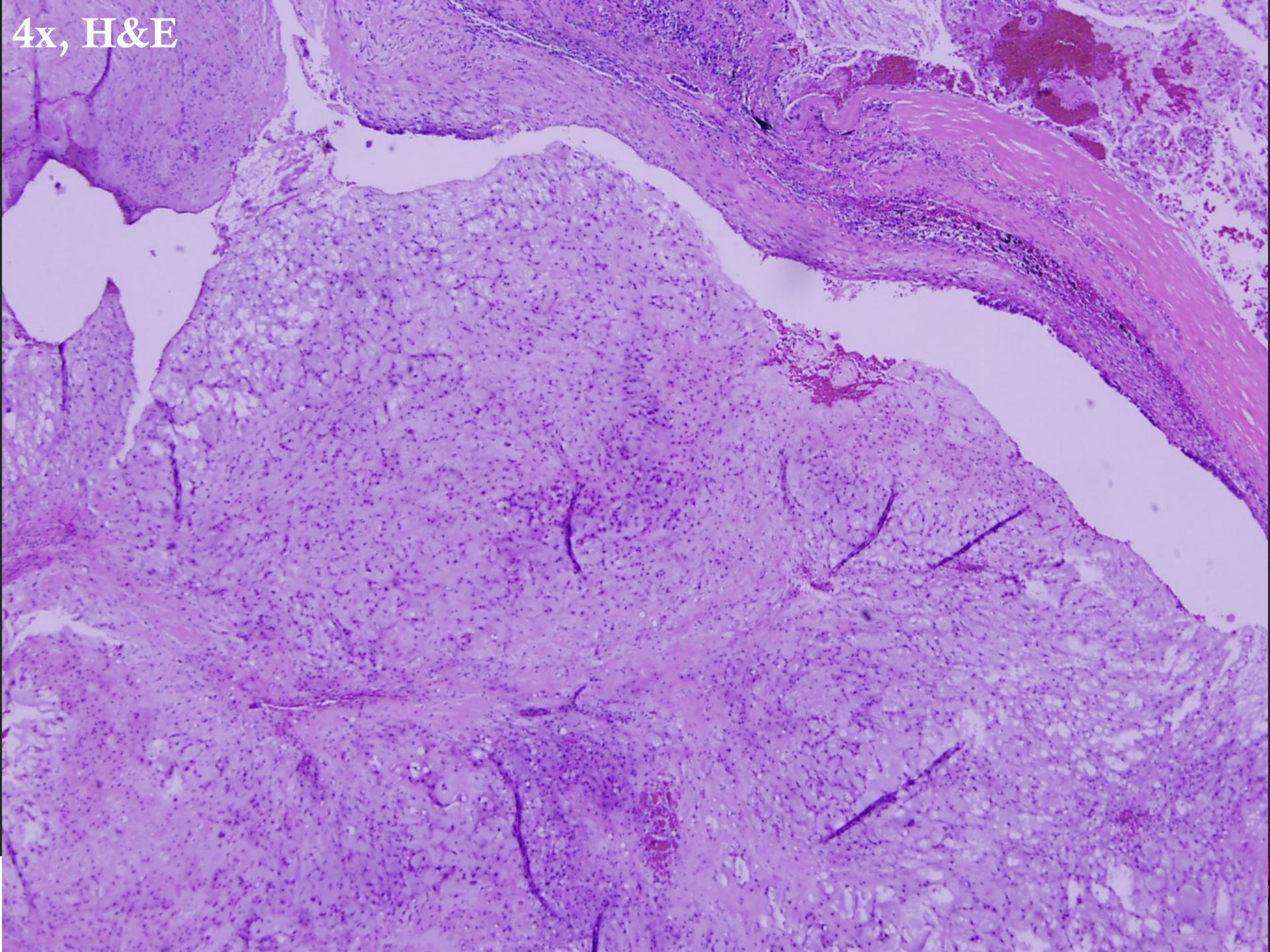
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- Metastasis
- Lymphoma
- Langerhans Histiocytosis
- Fibrous Dysplasia
- Infection

Pathology

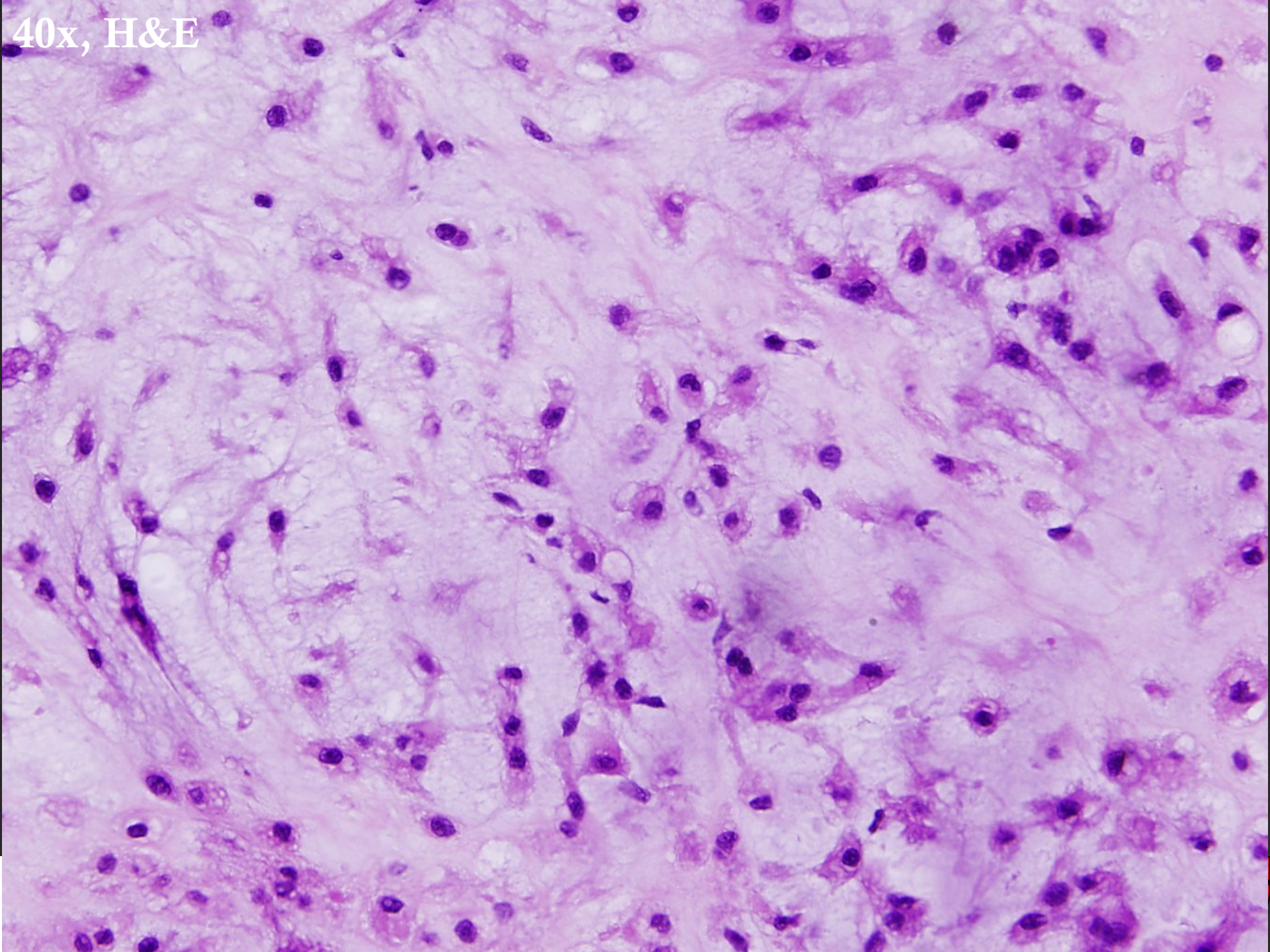




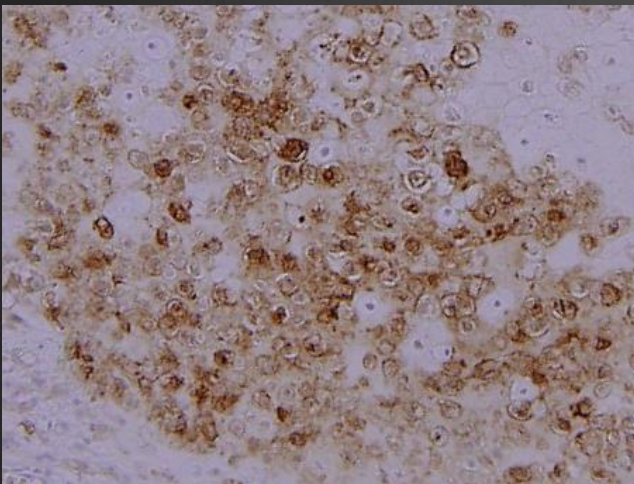
4x, H&E



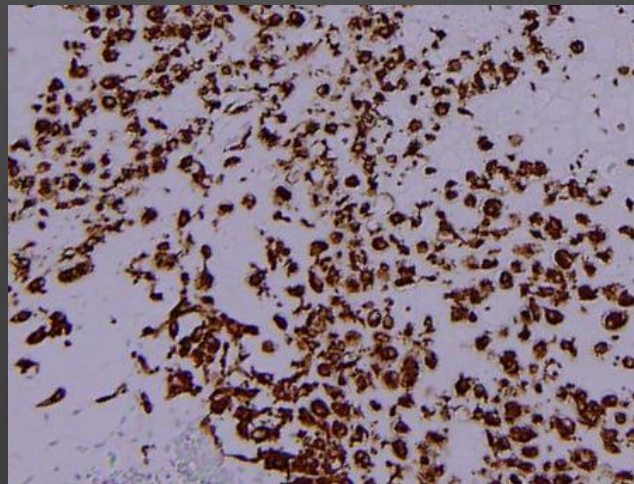
40x, H&E



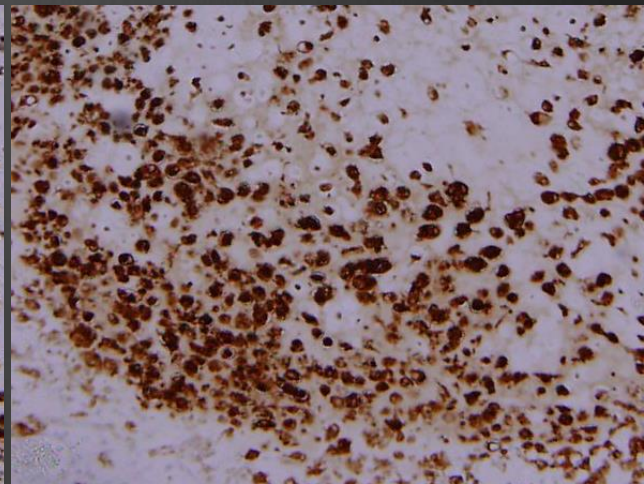
EMA



AE1/AE3



S100



Final Pathologic Diagnosis

- **CHORDOMA**
 - POS: EMA, AE1/AE3, S-100, Brachyury
 - NEG: GFAP

Chordoma

- Embryologic remnant of notochord, entrapped within bone
- Any age, peak prevalence in 4th decade, M:F 2:1, Caucasian
- Distribution: Roughly even across 3 locations

Intracranial

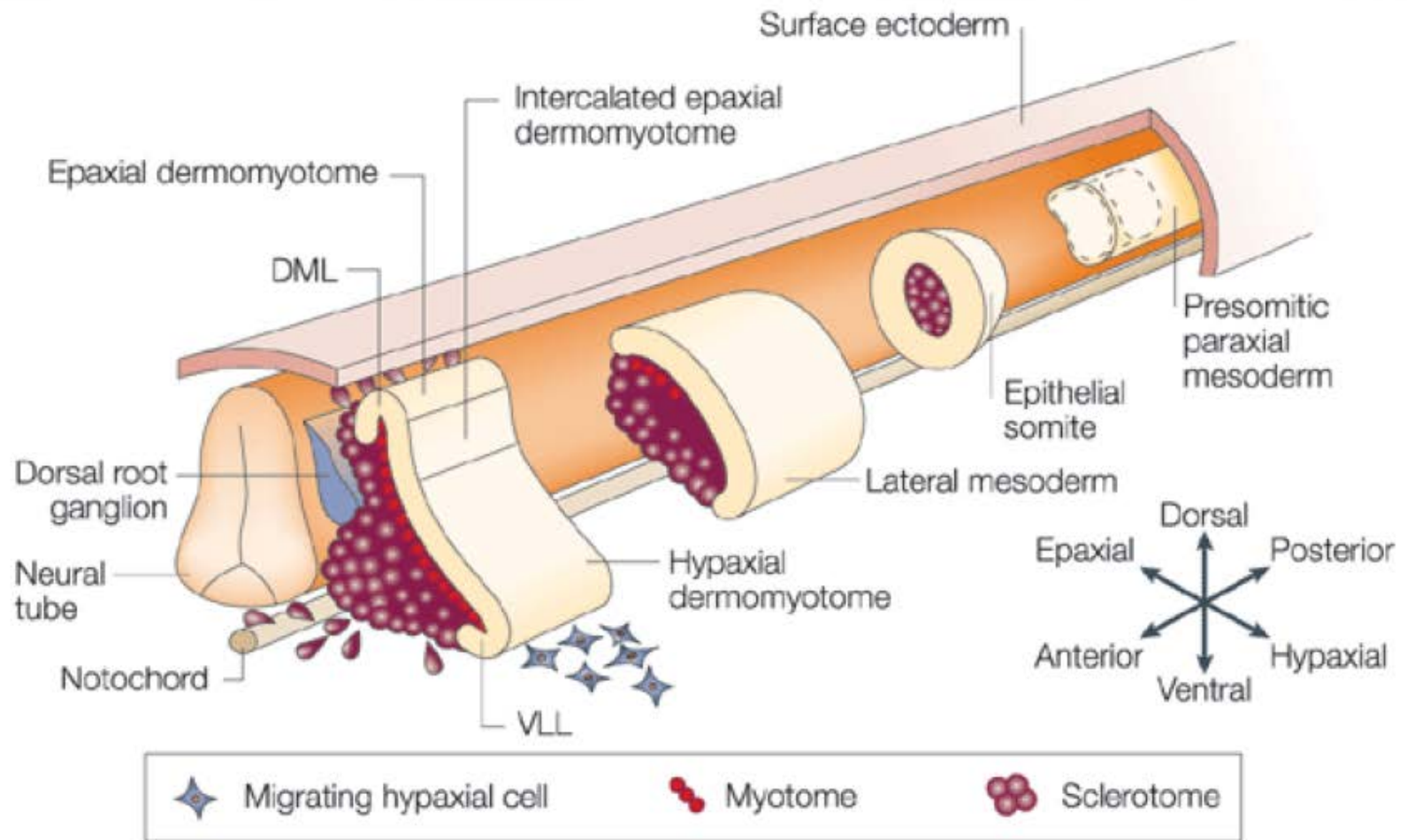


Spinal Cord



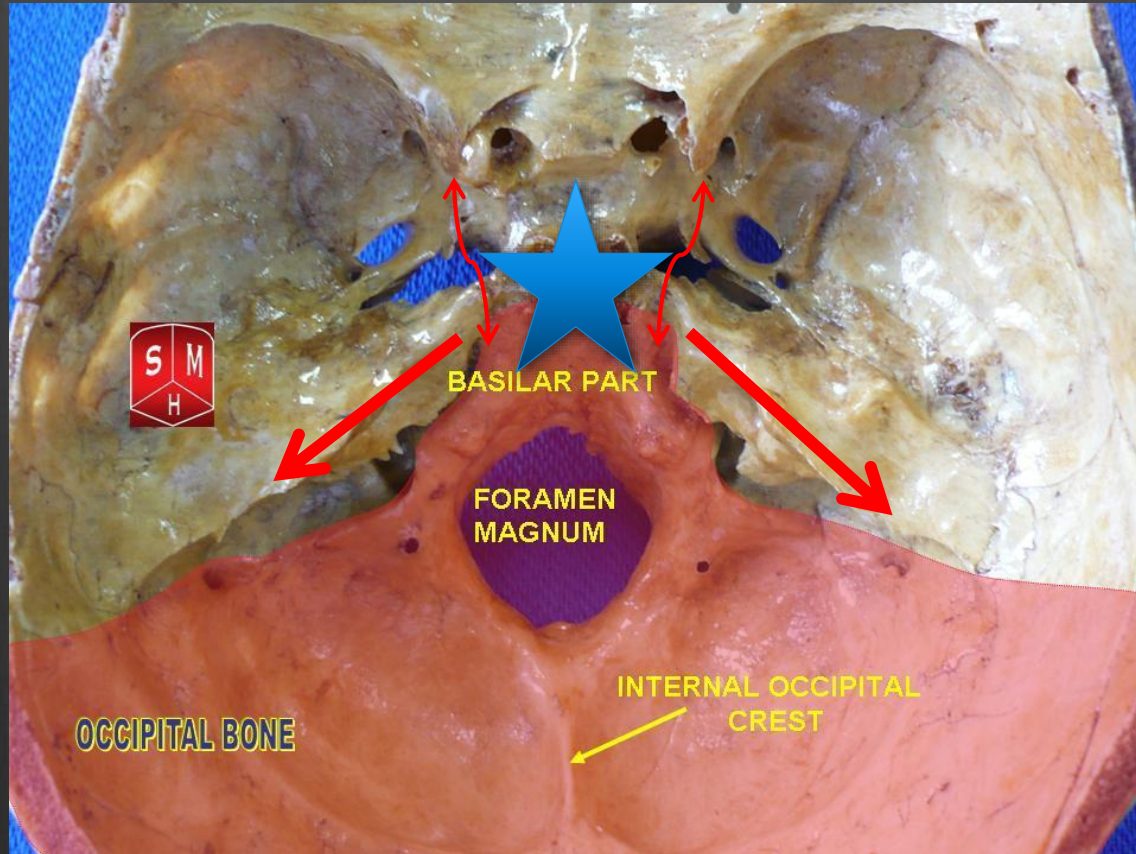
Sacral





Nature Reviews | **Genetics**

Chordoma



| | Chondrosarcoma | Chordoma |
|-----------------|---|--|
| CT | Rings and Arcs Linear Globular *Myxoid subtype | Well-circumscribed Variably Hyperattenuating Extensive lytic destruction *Chondriod subtype |
| T1 (rel. to WM) | Intermediate to Low | Intermediate to Low |
| T2 (rel. to WM) | Hyperintense | Hyperintense |
| Enhancement | +, heterogeneous | +, heterogeneous |
| Behavior | Less Common Majority arise from petro-occipital fissure Better prognosis | More common Midline typically Local Recurrence common Metastasis Rare |

DWI: Hot of the Press

Diffusion-Weighted MRI Distinction of Skull Base Chordoma and Chondrosarcoma

K.W. Yeom, I

Table 2: ADC values for each tumor type ($10^{-6} \text{ mm}^2/\text{s}$)

| Tumor | Mean ADC (Median) | Minimum ADC (Median) | Maximum ADC (Median) |
|--|-------------------|----------------------|----------------------|
| Chondrosarcoma (n = 9) | 2051 ± 262 (1977) | 1488 ± 360 (1352) | 2503 ± 512 (2392) |
| Classic Chordoma (n = 7) | 1474 ± 117 (1460) | 905 ± 118 (860) | 2199 ± 255 (2217) |
| Poorly Differentiated Chordoma (n = 3) | 875 ± 100 (871) | 491 ± 210 (469) | 1503 ± 127 (1557) |

3. Edwards, and N.J. Fischbein



RESULTS: Chondrosarcoma was a from classic chordoma (1474 ± 11 differentiated chordoma was char ment and/or lesion location did n

) and was significantly different $10^{-6} \text{ mm}^2/\text{s}$ ($P < .001$). Poorly IR imaging features of enhance-

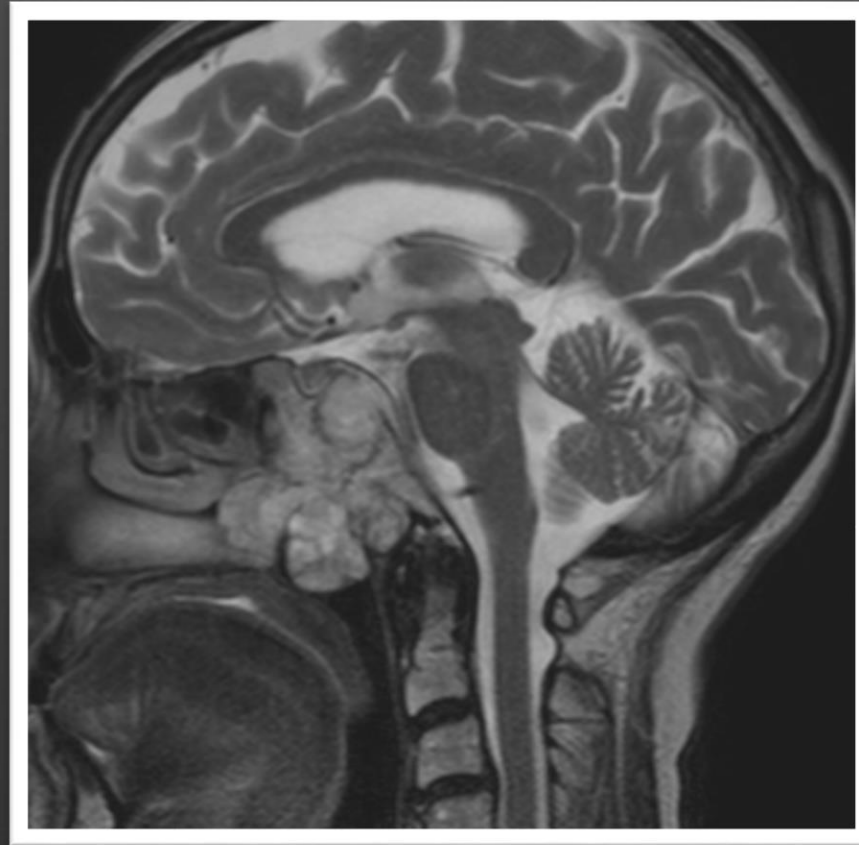
Table 3: MRI features for each tumor type

| Tumor | T2 Hypointensity | >90% Enhancement | Clival Location |
|--|------------------|------------------|-----------------|
| Chondrosarcoma (n = 9) | 0 (0%) | 5 (56%) | 4 (44%) |
| Classic chordoma (n = 7) | 0 (0%) | 1 (14%) | 4 (57%) |
| Poorly differentiated chordoma (n = 3) | 3 (100%) | 0 (0%) | 3 (100%) |

differentiating chordoma from C in predicting histopathologic

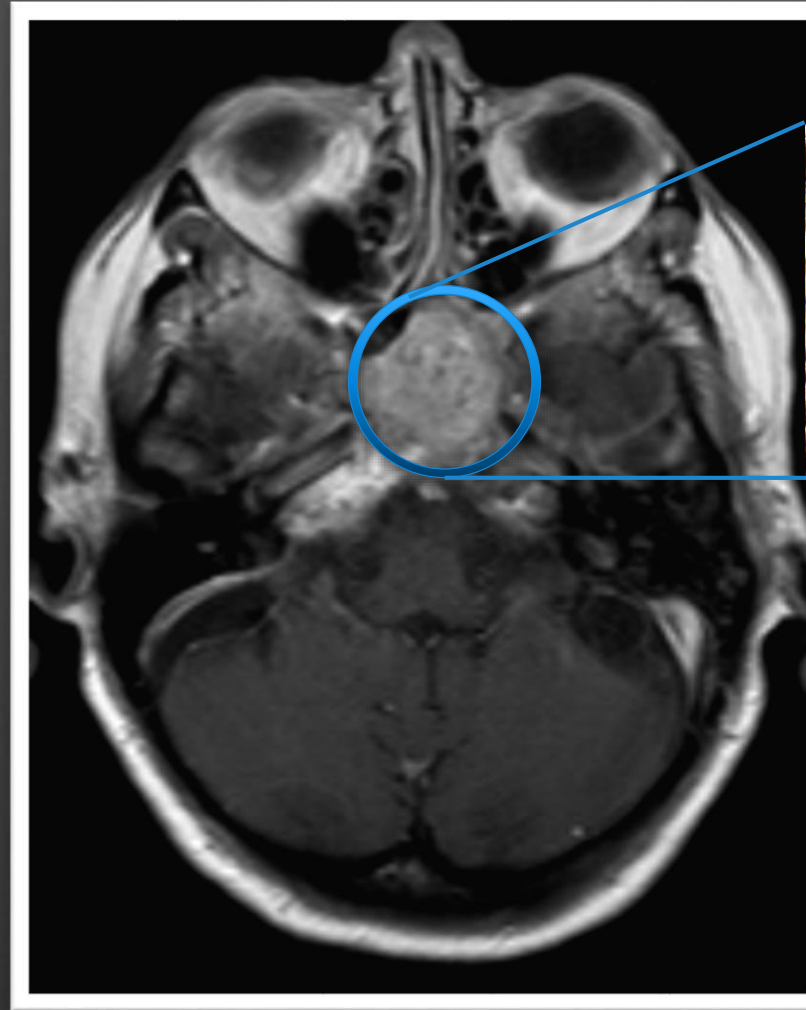
CONCLUSIONS: Diffusion-weight chondrosarcoma. A prospective s diagnosis.

Chordoma



Low signal intensity septations that separate high signal intensity lobules commonly seen

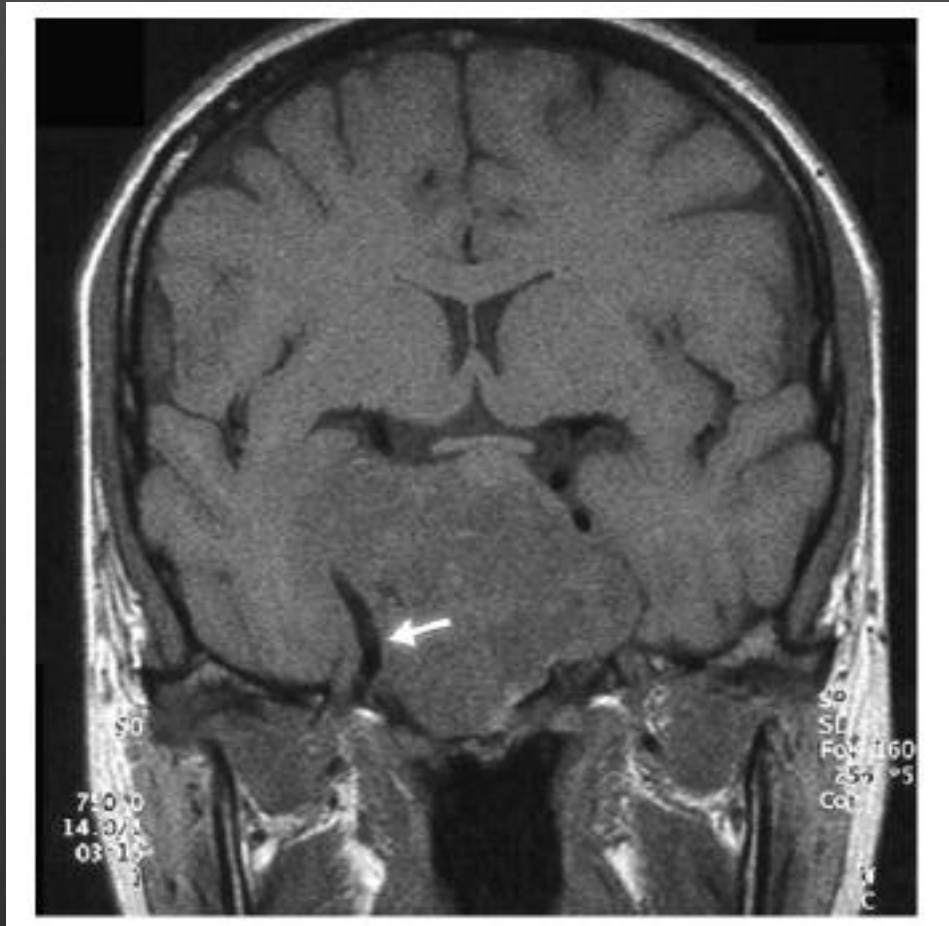
Chordoma



Honeycomb

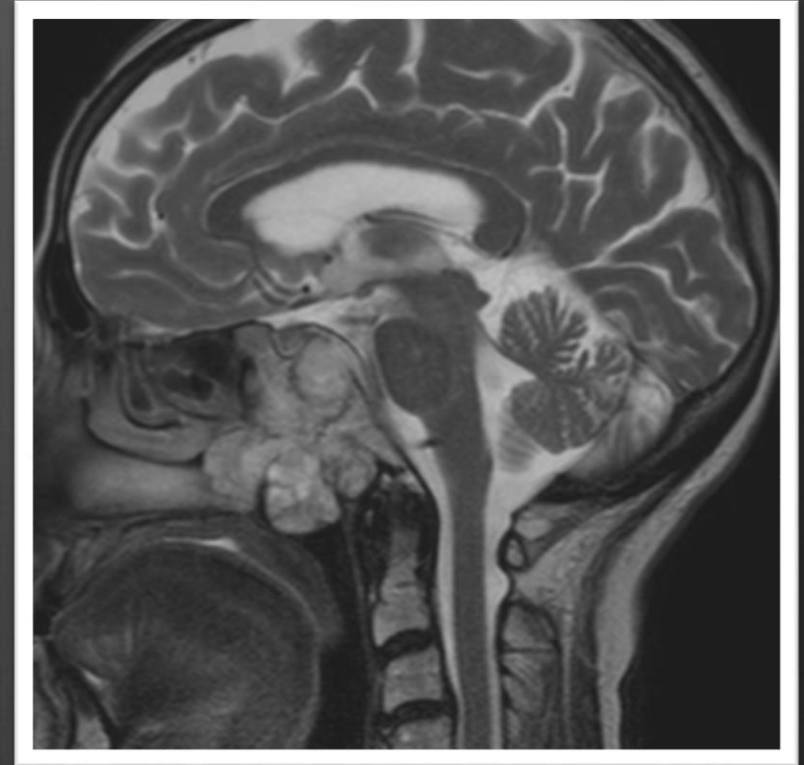
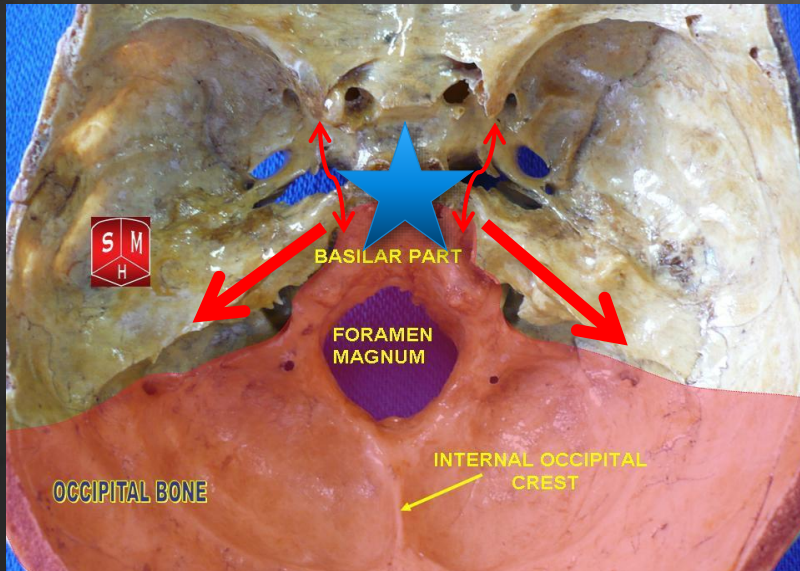
Post contrast, may see low signal areas of non-enhancement, reflecting necrosis or mucin

Chordoma



Vascular displacement common, narrowing rare

Chordoma: Behavior



Treated with resection followed by proton beam radiation
Local recurrence common; Metastasis rare

The End

