The Multiple Faces of Meningiomas: Demonstrating Atypical Appearances of Meningiomas

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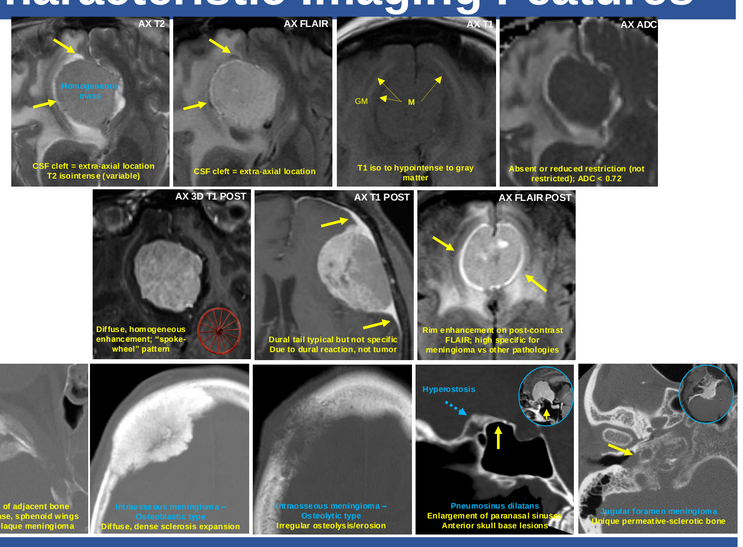
Purpose

Meningiomas are the most common primary central nervous system (CNS) neoplasm, making up more than a third of CNS tumors. While the majority of meningiomas exhibit classic imaging features in characteristic locations such as along the convexity and skull base, they can occasionally present with atypical features. Additionally, extra-axial masses can present with imaging features that mimic meningiomas; both scenarios can cause diagnostic dilemmas.

While there are no definite imaging criteria differentiating typical (WHO grade 1) meningiomas from atypical (grade 2) and anaplastic (grade 3) meningiomas, certain features may help predict higher tumor grades which may aid in pre-operative planning and management.

Recognizing certain imaging features that may alert clinicians of potentially lesions, this poster aims to provide an overview of typical and atypical imaging features of meningiomas, highlights features that can predict higher grade lesions, address common pitfalls in imaging interpretation, and offer a differential diagnosis of important alternative considerations.

Characteristic Imaging Features



Cytogenomics and Locations

HISTOLOGIC SUBTYPES

Grade 1:
- Meningoepithelial
- Fibroblastic
- Transitional
- Secretory
- Psammomatous
- Angiomatous
- Microcystic
- Lymphoplasmacyte-rich
- Metaplastic

Grade 2:
- Clear cell
- Choroid
- Atypical

Grade 3:
- Anaplastic
- Rhabdoid
- Papillary

MOLECULAR MARKERS

Less aggressive:
- KLF4
- TRAF7: Hyperostosis
- KLF4 + TRAF7 = Secretory type
- AKT1: Meningoepithelial type
- POLR2: Sellar/parasellar; only in grade 1

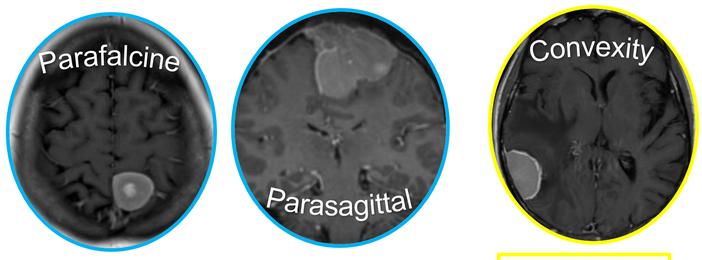
More aggressive:
- NF2: Common in sporadic and syndromic/familial and all grades
- TERT
- CDKN2A/B
- BAP1
- PBRMI

Grade 3

Hyperostosis:
- TRAF7
- Lymphoplasmacyte-rich
  * En plaque meningiomas

Peritumoral edema:
- Secretory
- Angiomatous
- Microcystic
- Lymphoplasmacyte-rich

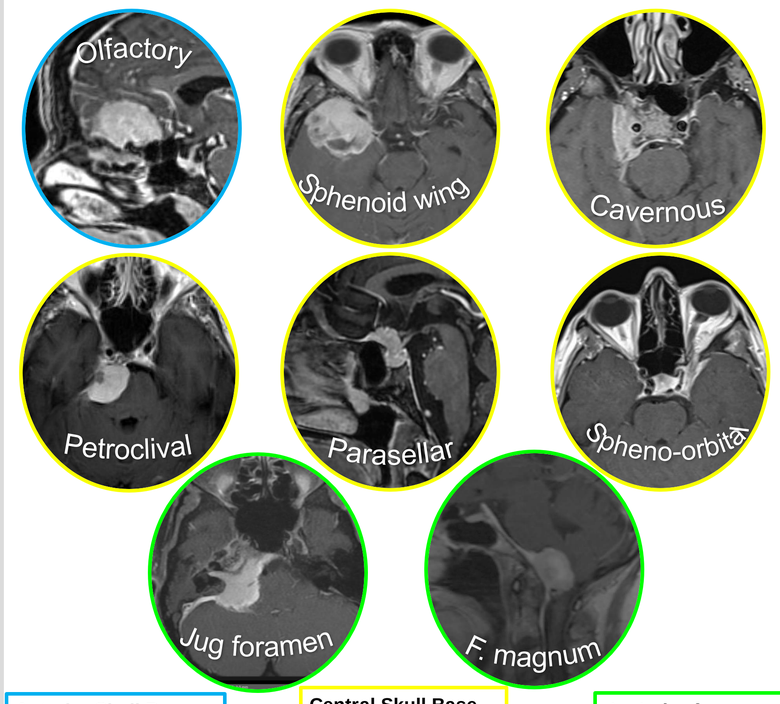
NON-SKULL BASE • More likely to be WHO grade 2/3



Parasagittal/falcine
- NF2 deletion
- SMARCB1
- TERT

Convexity
- NF2 deletion
- BAP1
- TERT
- CDKN2A/B

SKULL BASE • More likely to be WHO grade I, have hyperostosis



Anterior Skull Base
- SMO
- AKT1E17K
- TRAF7

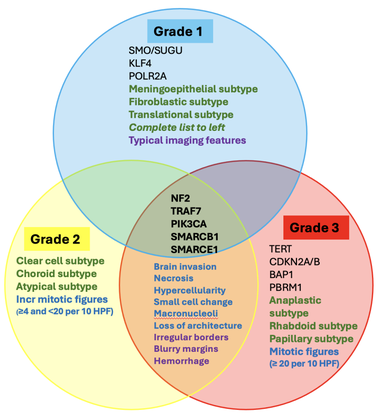
Central Skull Base
- KLF4
- TRAF7
- POLR21
- AKT1

Posterior fossa
- NF2
- AKT1E17K

Meningioma Grading

Meningiomas are divided into WHO grade 1 (benign), grade 2 (atypical), and grade 3 (anaplastic) based on histopathology criteria with the addition of molecular profiles in the 2021 WHO update. Grades 2/3 meningiomas exhibit increased parenchymal invasion and have an increased rate of recurrence with decreased five-year survival rates.

LEGEND
Molecular profile
Histologic subtype
Pathologic criteria
Imaging features



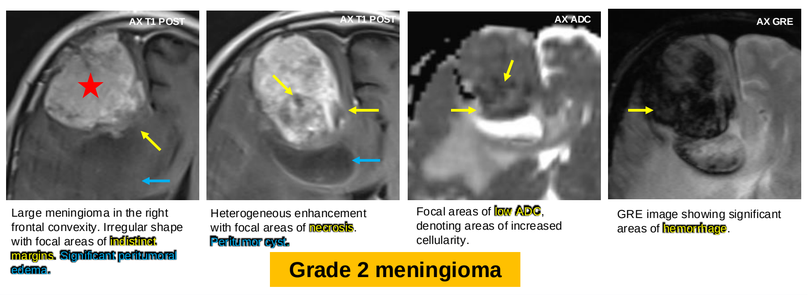
Imaging Features Predictive of Higher Grade

While there are no imaging features to accurately distinguish between grade 1 and grade 2/3 meningiomas, the following features can help predict higher grade tumors and which tumors may have increased risk of recurrence. Ultimately however, grade and recurrence risk will be based on histologic features and molecular markers.

HIGHER GRADE
- Larger tumor volumes
- Ill-defined/blurry tumor-brain interface
- Low ADC (<0.72)
- Irregular or non-spherical shape
- Heterogeneity
- Necrosis
- Non-skull base location

❖ Older age
❖ Radiation-induced

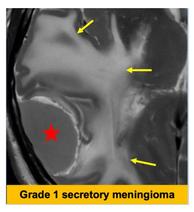
RECURRENCE RISK
- Larger tumor volume
- Significant peritumoral edema



Grade 2 meningioma

Potential Pitfalls

Peritumoral Edema



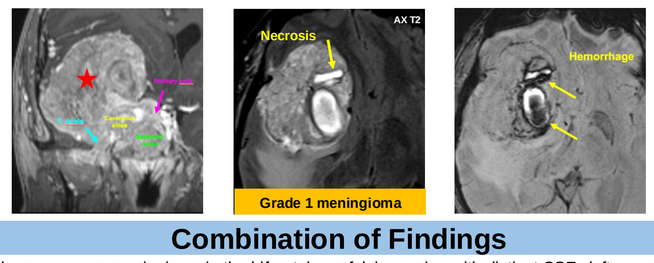
Small, well-defined, homogenous meningioma in the right temporal convexity with extensive, disproportionate peritumoral edema (PTE).

Prominent PTE is not always due to higher grade and can be related to benign causes, such as:
- Histologic subtype: Secretory, angiomatous, microcystic, lymphoplasmacyte-rich
- Highly vascular
- Expression of hormone receptors

Large Tumor Volumes

Very large right sphenoid wing meningioma with extension into the cavernous sinus, sphenoid sinuses, pituitary sella, and foramen ovale. Marked heterogeneity with central necrosis and hemorrhage. Pathology showed ≤1 mitotic figure per HPF; no hypercellularity or necrosis; no brain invasion.

Large volume masses can have more aggressive features despite their benign nature



Combination of Findings

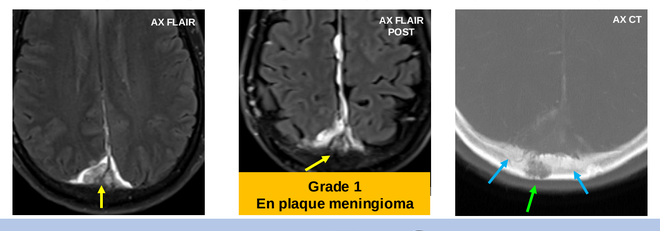
Large, homogeneous meningioma in the bifrontal parafalcine region with distinct CSF cleft, prominent PTE, typical spoke-wheel pattern of enhancement. The left margins of the mass are well-defined; however, the right margins are indistinct. Pathology showed prominent macronucleoli, 4 mitoses per 10 HPFs, rare foci of necrosis. No hypercellularity, small cell change or loss of architectural pattern. Ki-67 of 15%.

Important to evaluate all areas of the meningiomas as only certain areas may exhibit more aggressive features.

Bony erosion and Cognitive Biases

43-year-old woman with metastatic breast cancer presents for tumor staging. Thick, nodular dural enhancement along the falx bilaterally with invasion into and occlusion of the superior sagittal sinus. Associated calvarial sclerosis *plus* bony erosion seen on CT. Constellation of findings were concerning for dural metastases. Pathology showed a grade 1 meningioma without mitotic figures, hypercellularity, macronucleoli, small cell change, or architectural distortion.

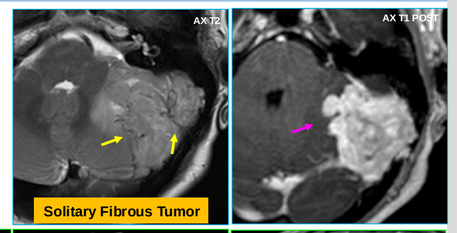
En plaque meningiomas are usually osteoblastic but may have areas of osteolysis which can appear aggressive. Additionally, anchoring bias (relying too heavily on clinical hx) can cause interpretation errors



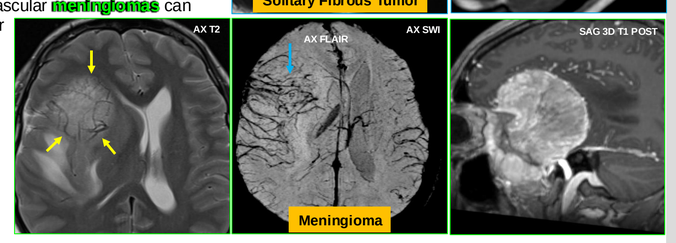
Differential Diagnoses

Benign Extra-Axial Masses

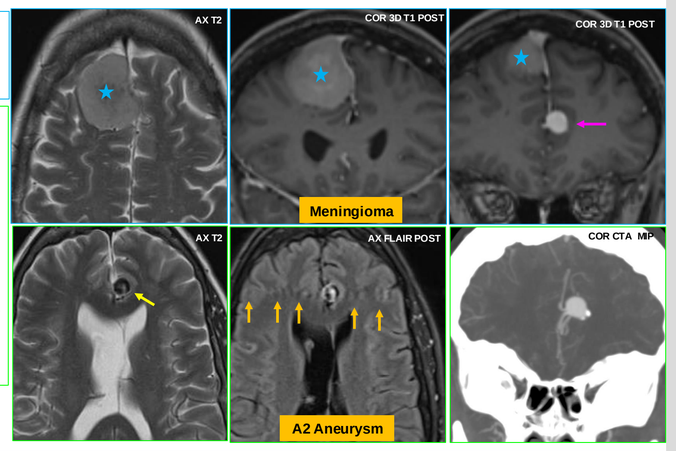
Solitary fibrous tumors are rare, dural based masses of mesenchymal origin. They have similar imaging findings as meningiomas: well-defined, solidly enhancing, dural-based masses. They may even have dural tails. SFTs, however, more commonly have flow voids, lobulated margins and usually cause bony erosion rather than hyperostosis.



However, hypervascular meningiomas can can have a similar appearance with flow voids and increased surrounding vascularity.



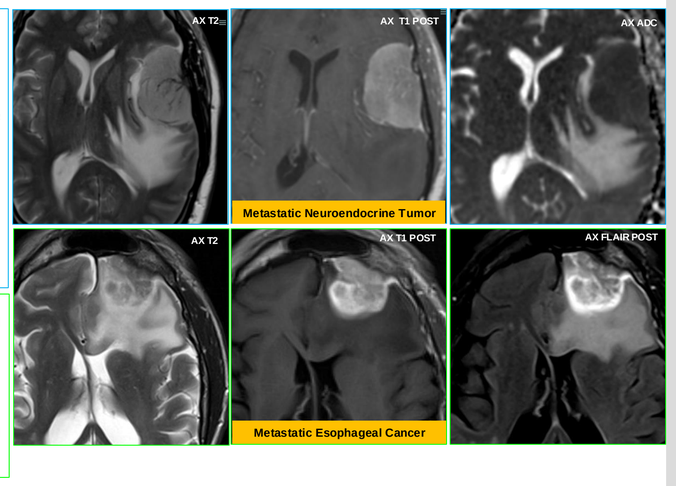
Typical right parafalcine meningioma without superior sagittal sinus invasion.



Inferior to this is another homogeneously enhancing extra-axial lesion. However, unlike typical meningiomas, there is more avid enhancement, marked T2 hypointensity, and pulsation artifact on post contrast FLAIR. A CTA was recommended, which confirmed a left A2 saccular aneurysm.

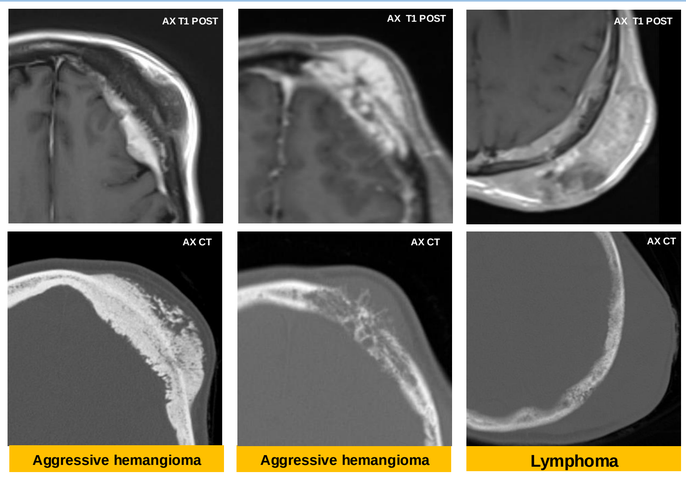
Malignant Extra-Axial Masses

This metastatic neuroendocrine tumor (NET) has characteristic imaging features of a meningioma. Meningioma should be the leading and possibly the only, pre-operative dx provided. As meningiomas have many mimickers, it is important to scrutinize assoc findings which may warrant a ddx; however, in this case, there were none.



This esophageal CA met has dural tails and even rim E+ on FLAIR post contrast, which is highly specific for meningiomas! The irregular shape, areas of T2 hypo, and clinical hx were helpful to suggest a more aggressive dx.

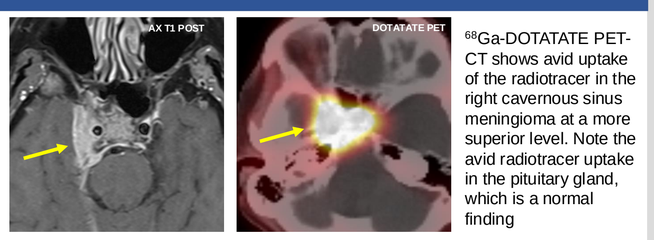
Malignant Extra-Axial Masses



Rarely, meningiomas can have extracranial extension, which present as a slow growing "bump" on the head. The associated bony changes can be either osteoblastic (more common), osteolytic, or a combination, as seen in this example. Rare lesions that can have a similar appearance with both dural and extracranial involvement are aggressive hemangiomas (much more common in the spine) and primary dural lymphomas. Primary dural lymphomas are uncommon and are usually a low-grade, B-cell marginal lymphoma.

The Use of DOTATATE PET

Homogeneously enhancing right cavernous sinus meningioma with extension along the prepontine dura and right tentorium



^{68}Ga-DOTATATE PET-CT shows avid uptake of the radiotracer in the right cavernous sinus meningioma at a more superior level. Note the avid radiotracer uptake in the pituitary gland, which is a normal finding

Meningiomas are known to express somatostatin receptor 2 (SSTR2). PET-CT using the radiotracer ^{68}Ga-DOTATATE, which is an SSTR2 analog, has a higher sensitivity and specificity for detecting meningiomas than MRI or ^{18}FDG PET-CT.

^{68}Ga-DOTATATE PET-CT is instrumental in:
- Differentiating meningiomas from other dural-based masses
  - Caveat: Cancers that also express SSTR2 (Breast, neuroendocrine tumors, etc)
- Differentiating post-surgical changes from residual tumor; particularly in guiding radiation therapy in grade 2 meningiomas
- Detect recurrence

Contact

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