

AMSER Case of the Month

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36 year old woman with intractable headaches

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Patient Presentation

HPI: A 36 year old woman presents to primary care clinic with complaints of intermittent, intractable headaches, vision changes, balance issues and confusion since delivery of her first child one year prior.

Medical History: Asthma, intrahepatic cholestasis of pregnancy, HPV infection and recurrent UTI's

Past Surgical History: None

Family History: No known history of genetic syndromes or brain cancer

Social History: 1.25 pack-year history and occasional alcohol use

Physical exam: left eyelid ptosis with no other focal neurological deficits noted

Pertinent Labs

- Myasthenia Gravis Evaluation Panel: Negative
- CMP/CBC: within normal limits
- TSH: 1.37
- ANA: Negative at 1:80

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 7:

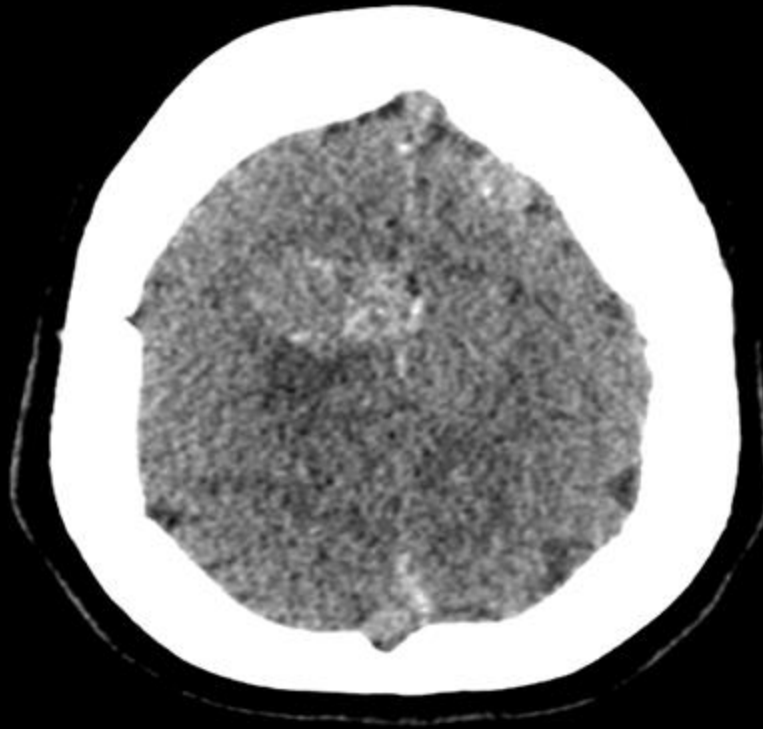
Headache with one or more of the following “red flags”: increasing frequency or severity, fever or neurologic deficit, history of cancer or immunocompromise, older age (>50 years) of onset, or posttraumatic onset. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	○
MRI head without IV contrast	Usually Appropriate	○
CT head without IV contrast	Usually Appropriate	⊗⊗⊗
Arteriography cervicocerebral	Usually Not Appropriate	⊗⊗⊗
MRA head with IV contrast	Usually Not Appropriate	○
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without and with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⊗⊗⊗
CT head without and with IV contrast	Usually Not Appropriate	⊗⊗⊗
CTA head with IV contrast	Usually Not Appropriate	⊗⊗⊗
CTV head with IV contrast	Usually Not Appropriate	⊗⊗⊗

This imaging modality was ordered by the FM physician

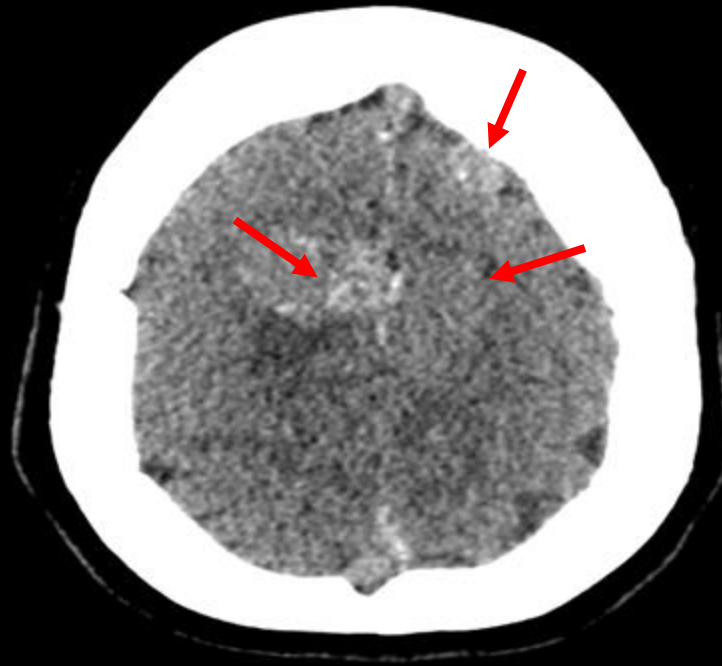


Findings (unlabeled)



CT head without contrast

Findings: (labeled)



Bilateral partially calcified masses (red)

Interval History

- The patient was referred to neurosurgery based on initial imaging results. Further history was obtained and a physical exam noted a **large birthmark**. Based on these findings, neurosurgery recommended **repeat imaging** and **genetics counseling**.

What Imaging Should We Order Next?

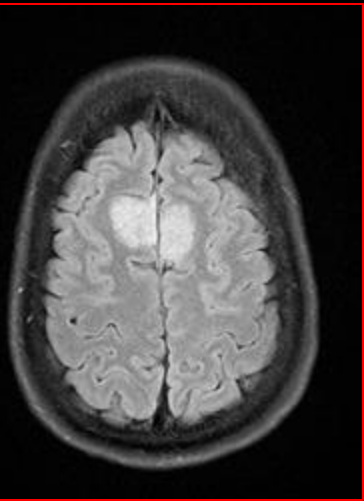
Select the applicable ACR Appropriateness Criteria

Variant 4: Adult. Suspected extraaxial brain tumor based on prior imaging. Pretreatment evaluation.

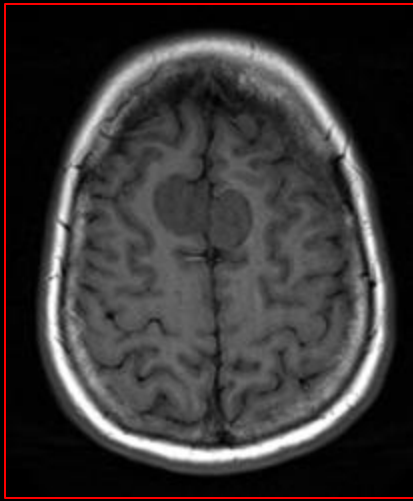
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	○
MRI complete spine without and with IV contrast	May Be Appropriate	○
MRI functional (fMRI) head without IV contrast	May Be Appropriate	○
MRI head without IV contrast	May Be Appropriate	○
DOTATATE PET/CT brain	May Be Appropriate	⊗⊗⊗
DOTATATE PET/MRI brain	May Be Appropriate	⊗⊗⊗
MR spectroscopy head without IV contrast	Usually Not Appropriate	○
MRI complete spine with IV contrast	Usually Not Appropriate	○
MRI complete spine without IV contrast	Usually Not Appropriate	○
MRI head perfusion with IV contrast	Usually Not Appropriate	○
MRI head perfusion without IV contrast	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRI head without IV contrast with DTI	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⊗⊗⊗
CT head without and with IV contrast	Usually Not Appropriate	⊗⊗⊗
CT head without IV contrast	Usually Not Appropriate	⊗⊗⊗
FDG-PET/CT brain	Usually Not Appropriate	⊗⊗⊗
FDG-PET/MRI brain	Usually Not Appropriate	⊗⊗⊗
Fluciclovine PET/MRI brain	Usually Not Appropriate	⊗⊗⊗
Fluciclovine PET/CT brain	Usually Not Appropriate	⊗⊗⊗⊗

This imaging modality was ordered by the neurosurgeon

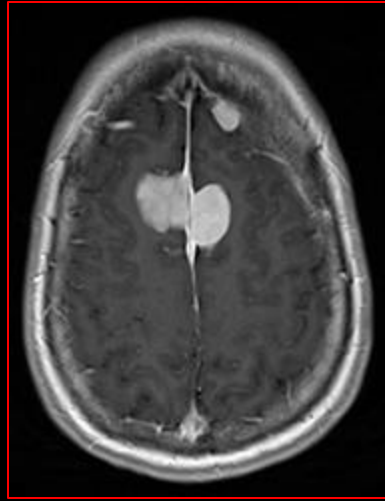
Findings (unlabeled)



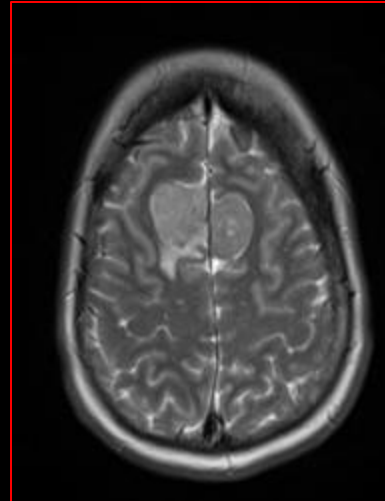
T2 Flair



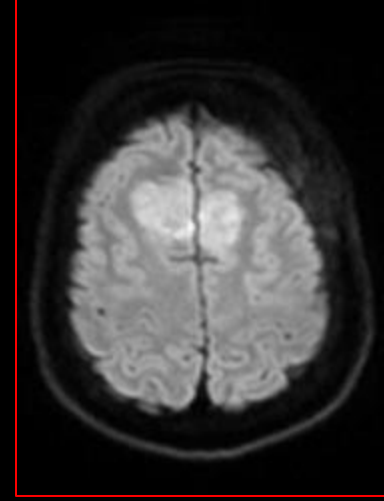
T1 precontrast



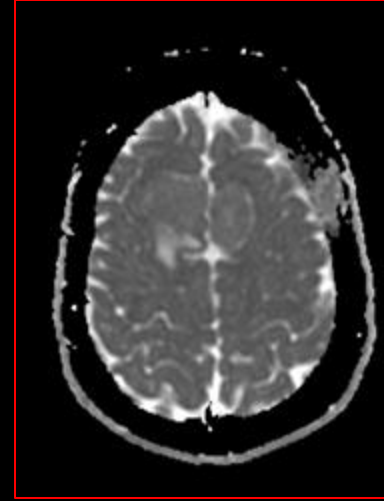
T1 post contrast



T2

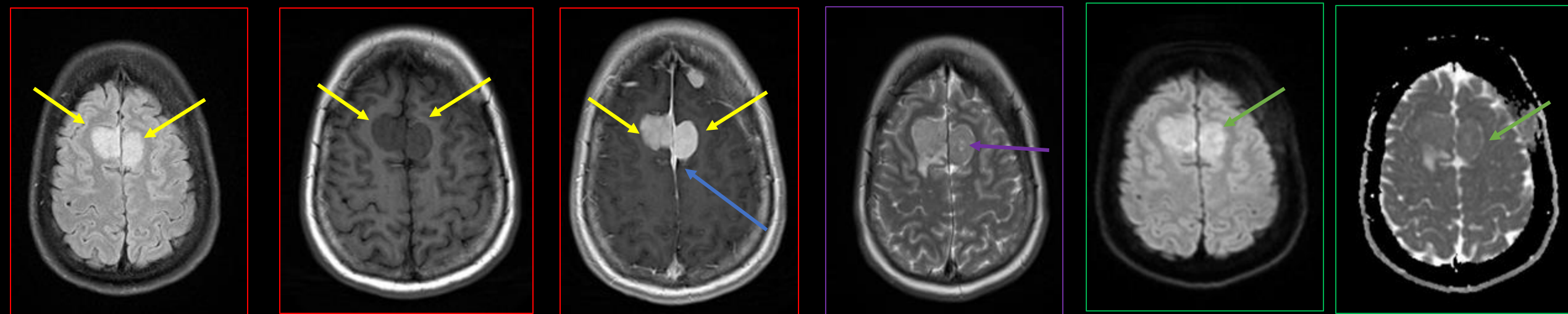


DWI



ADC

Findings (labeled)



- Multiple uniformly enhancing extra-axial masses, the largest of which are along the bilateral parafalcine region (yellow)
- Enhancing dural tails (blue)
- The extraaxial meningiomas range from isointense to mildly hyperintense as compared to the adjacent gray matter on T2 imaging (purple)
- There is demonstrated diffusion restriction of the extraaxial meningiomas. They are hyperintense on DWI images and hypointense ADC images (green)

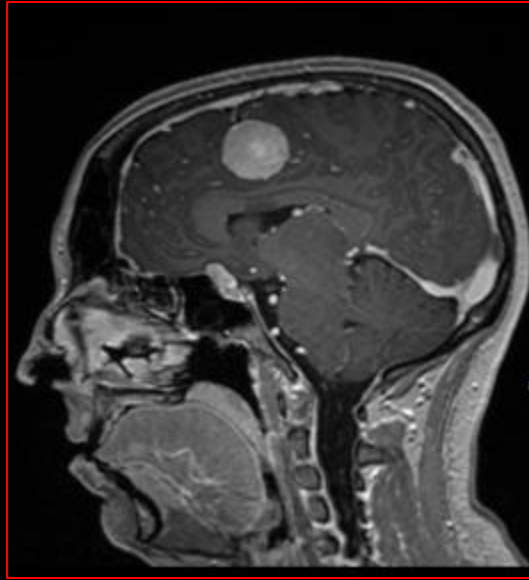
Interval History

- Given the findings of **multiple enlarging meningiomas** on interval imaging, the patient's neurosurgeon recommended surgical intervention.
- A bifrontal craniotomy was performed with resection of **six dural based lesions and one falcine mass lesion**.
- The patient recovered well post-op and returned for a follow-up visit after six months with no new symptoms or complaints and a follow-up **MRI scan** was obtained
- Of note, genetic counseling did not result in any findings

Findings (unlabeled)



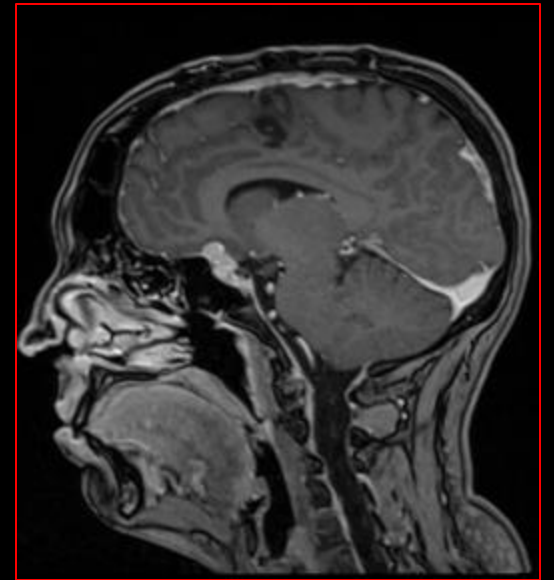
T2 Flair



T1 post contrast (pre-op)

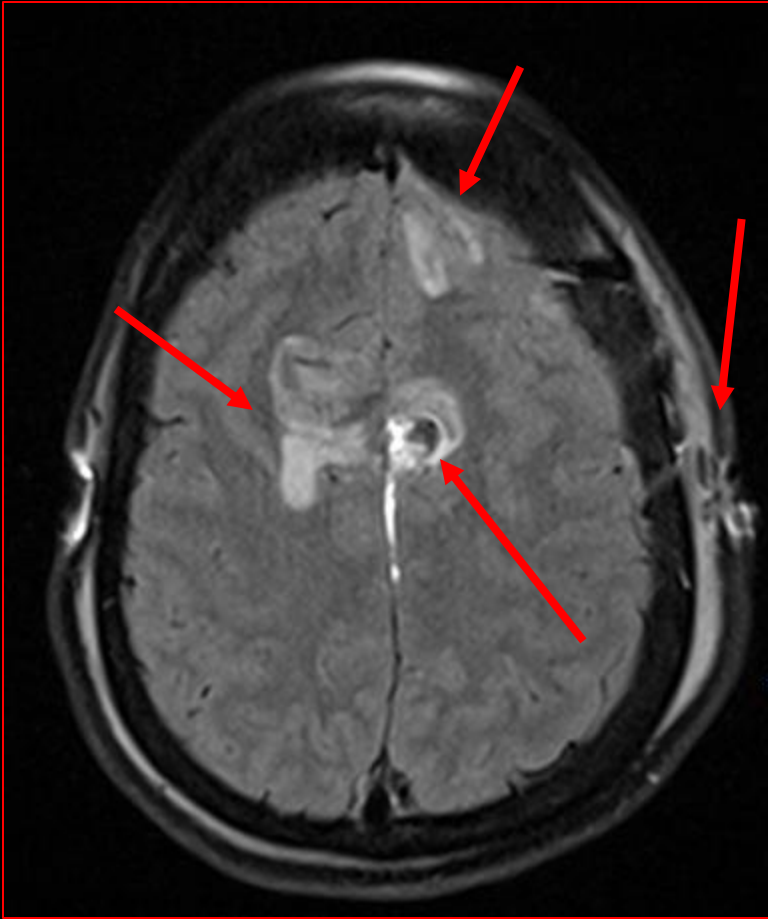


T1 post contrast (pre-op)



T1 post contrast (post-op)

Findings (labeled)



- T1 post contrast. Post op
Demonstrates a patent
superior sagittal sinus



- Status post bifrontal craniotomy and resection of multiple extra axial meningiomas
- Soft tissue edema and post surgical changes (red)
- Post-operative images demonstrate a stable tuberculum sella meningioma (red)
- No significant mass effect on suprasellar cistern (yellow)

Final Dx:

Meningioma

Case Discussion: Etiology of Meningiomas

- Meningiomas are typically benign, extra-axial tumors arising from the arachnoid cap cells of the meninges in the brain and spinal cord.
- They account for approximately 37.6% of all primary central nervous system (CNS) tumors with the highest incidence amongst female adults.
- Differential diagnoses of meningiomas include dural-based lesions that exhibit homogenous enhancement
 - CNS solitary fibrous tumors, gliosarcoma, leiomyosarcoma, intracranial Hodgkin's lymphoma, plasma cell granuloma, melanocytic tumors, neurosarcoidosis, sinus histiocytosis, plasmacytoma, dural metastatic tumors and intracranial hemangiopericytomas.

Case Discussion: Etiology of Meningiomas (cont'd)

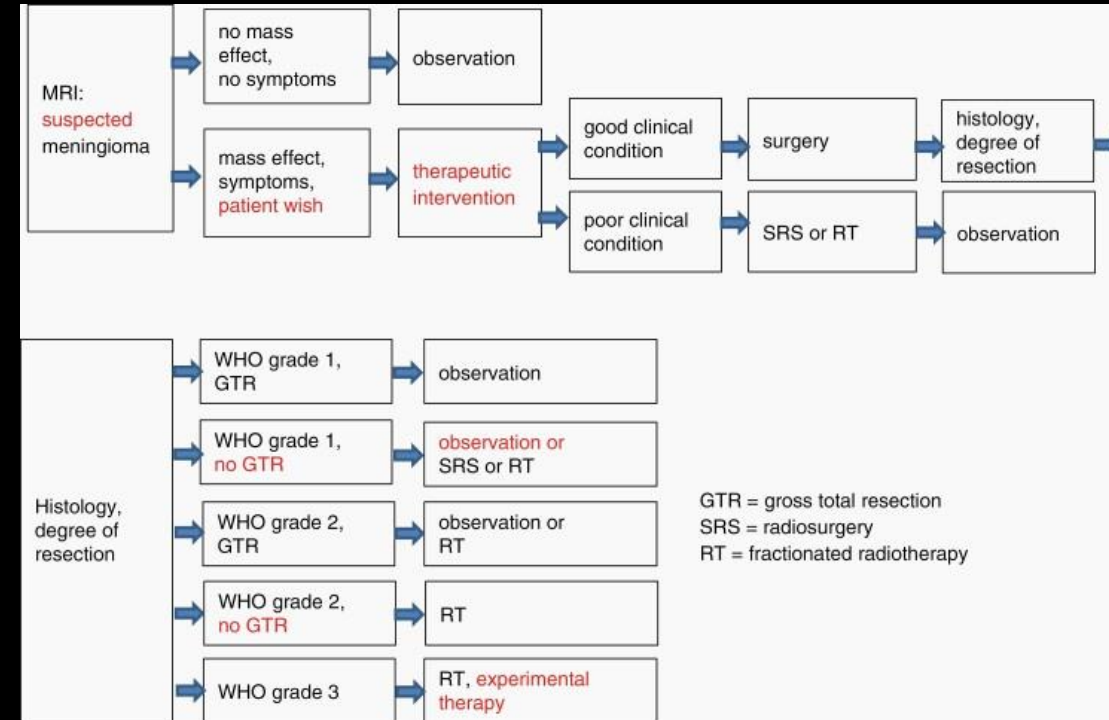
- Several genetic syndromes confer increased risk for development of **meningiomas**:
- **Neurofibromatosis Type 2 (NF2)**
 - Most associated genetic syndrome; meningioma incidence of 50%
 - Caused by mutations of the **NF2 gene** (merlin protein dysfunction)
- **Gorlin Syndrome (Basal Cell Nevus Syndrome)**
 - **PCTH-1 gene** mutation (Hedgehog pathway dysregulation)
- **Cowden Syndrome**
 - **PTEN gene** dysfunction (Tumor suppressor dysfunction)
- **Li-Fraumeni Syndrome**
 - **TP53 gene** mutation (Defective tumor suppression)
- Genetic syndromes associated with meningiomas often involve **molecular pathways** altered in sporadic cases highlighting **potential targets for therapy**.

Case Discussion: Presentation and Diagnosis of Meningioma

- **History and Physical exam**
 - Meningioma's are commonly asymptomatic and found incidentally on neuroimaging
 - Depending on the size and location, meningiomas may present with headache, seizures, visual disturbances, endocrine dysfunction and focal neurologic deficits due to compression of local structures
 - Physical exam may reveal papilledema, cranial nerve deficits or visual field deficits
- **Imaging**
 - Brain magnetic resonance imaging (MRI) with and without IV contrast is the gold standard for diagnosis of meningioma (ACR)
 - They typically appear as extra-axial, well-circumscribed lesions with homogenous contrast enhancement.
 - Presence of a dural tail (linear enhancement of contrast along the dura) is highly suggestive of a meningioma
 - Brain MRI may also help find cystic lesions within the meningioma
 - Calcifications and peritumoral edema may also occur with edema visualized as hyperintensity in T2 weighted images.
 - The white matter buckling sign denotes inward compression of the white matter which can distinguish extra-axial lesions from intra-axial lesions.

Case Discussion: Treatment and Prognosis of Meningiomas

- Management of meningiomas depends on tumor size, symptoms and histopathology.
 - Asymptomatic, small tumors: observation with follow-up imaging annually or biannually
 - Symptomatic or fast-growing lesions: gross total surgical resection with subsequent histological grading
 - Radiation therapy may be used in grade I tumors without gross total resection or grades II and III tumors



Reference 2

References:

- 1) Alruwaili AA, De Jesus O. Meningioma. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; August 23, 2023.
- 2) Goldbrunner R, Stavrinou P, Jenkinson MD, et al. EANO guideline on the diagnosis and management of meningiomas. *Neuro Oncol*. 2021;23(11):1821-1834. doi:10.1093/neuonc/noab150
- 3) Kerr K, Qualmann K, Esquenazi Y, Hagan J, Kim DH. Familial Syndromes Involving Meningiomas Provide Mechanistic Insight Into Sporadic Disease. *Neurosurgery*. 2018;83(6):1107-1118. doi:10.1093/neuros/nyy121
- 4) Watts J, Box G, Galvin A, Brotchie P, Trost N, Sutherland T. Magnetic resonance imaging of meningiomas: a pictorial review. *Insights Imaging*. 2014;5(1):113-122. doi:10.1007/s13244-013-0302-4