Olfactory Groove Meningiomas

Neurosurgery I Elective
Carle Illinois College of Medicine
Carle Foundation Hospital

Joseph Policarpio, M3
joep3@illinois.edu

Elective Preceptors
Dr. Kevin Teal MD, Dr. Paul Arnold MD, Dr. Wael Hassaneen Mostafa MD PhD

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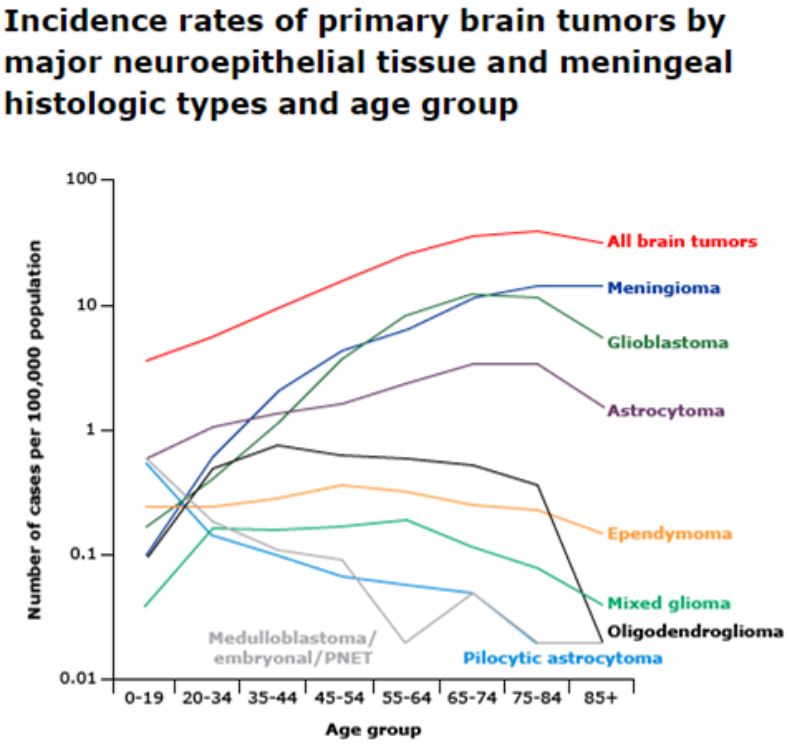
Epidemiology of Meningioma

- Meningioma is the most common *primary* intracranial neoplasm in adults (metastases are most common)
  - 20-36% of intracranial tumors
  - incidence of 1.8-13 per 100,000
  - Approximately 80% of these are benign

- Olfactory groove represent:
  - 2% of all intracranial tumors
  - 5%–18% of all intracranial meningiomas



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Risk Factors for Meningioma

- **Ionizing radiation (most important acquired risk factor)**
  - radiation treatment for malignancy
  - incidental radiation (radiographs, CT, atomic bombs)
- Genetic predisposition - 22q alterations
  - NF2 (75% lifetime risk)
  - MEN1
  - Schwannomatosis (SMARCB1 tumor suppressor)
- Reproductive age female
  - Hormonal factors: progesterone, androgen, estrogen)
- Breast cancer
- High-dose cyproterone (anti-androgen, not approved in the US)
- Ever-use estrogen or estrogen-progestin therapy (RR 1.35)
- Obesity (OR 1.4 to 2.1)
- Age > 65 years
- Higher in African American than other ethnic groups in the US

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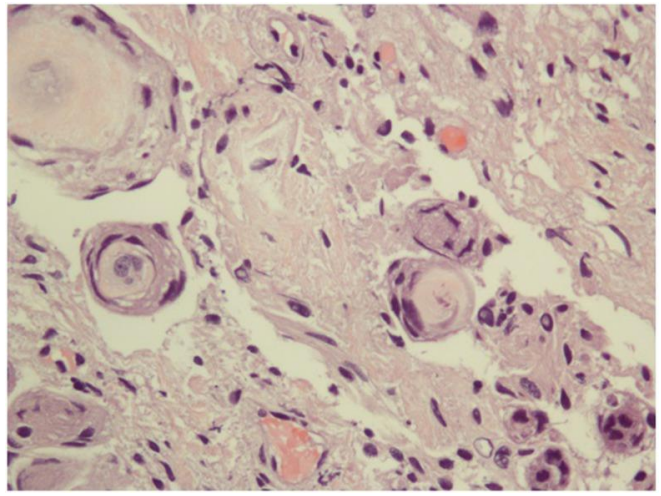
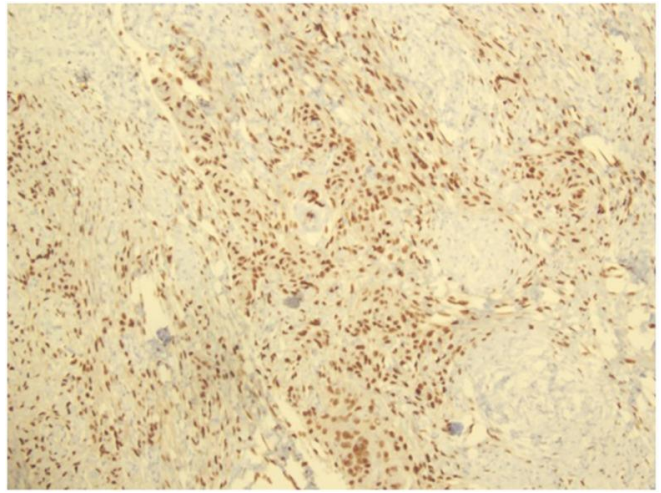
Typical Clinical Presentation

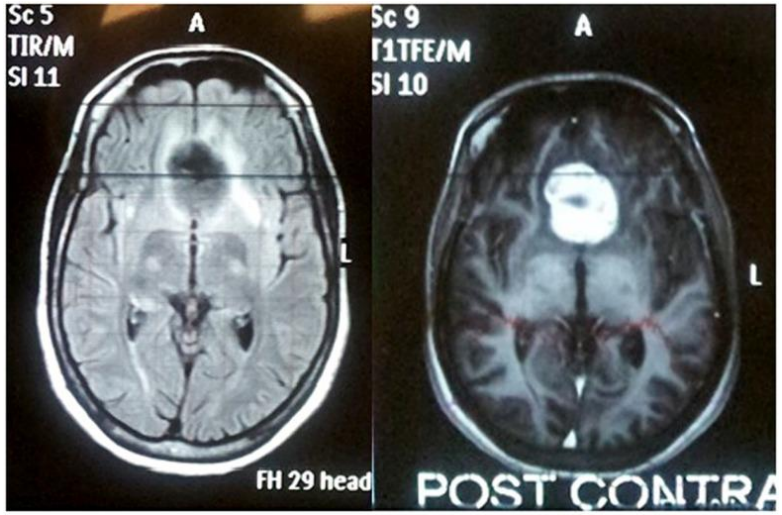
- Asymptomatic
- Seizure
- Focal neurological deficits

Typical Clinical Presentation





- Asymptomatic
  - Discovered incidentally
- Seizure
  - Found in approx. 30%
- Focal neurological deficits
  - Visual changes
    - Foster Kennedy syndrome
      - Unilateral optic atrophy
      - Contralateral papilledema
      - Anosmia
    - Progressive unilateral visual loss
    - Weakness of extraocular movements
  - Loss of smell (olfactory groove or sphenoid ridge)
  - Mental status changes
    - Apathy
    - Inattention



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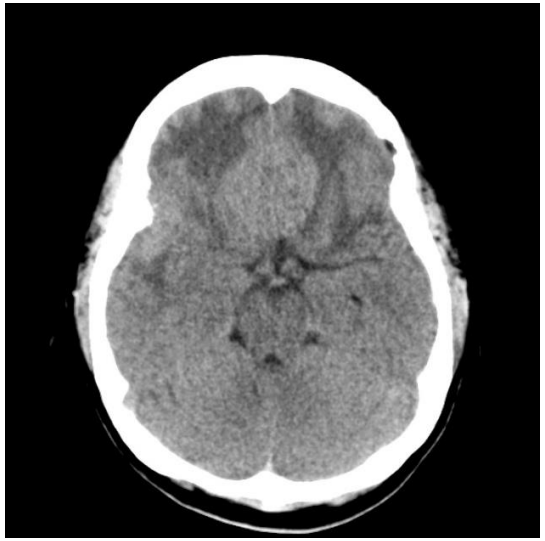
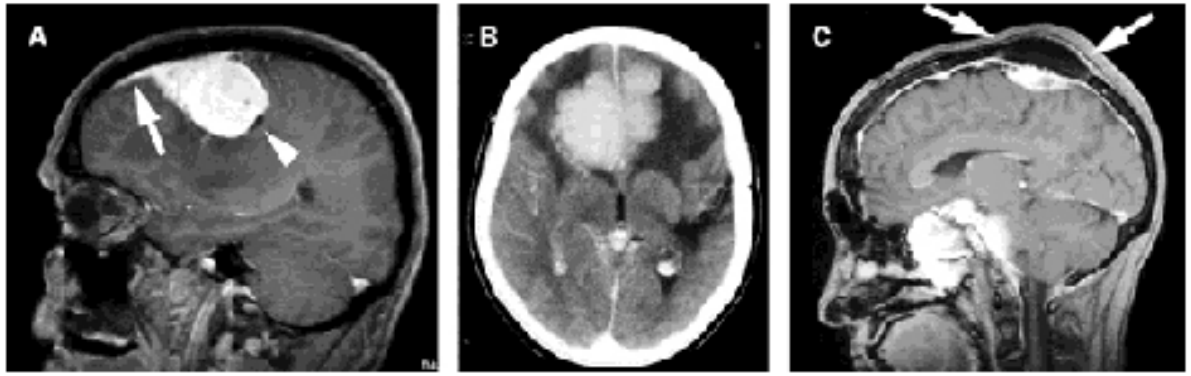
Typical Workup



- H&P – consider risk factors
  - especially prior therapeutic radiation
  - Genetic syndromes (NF2)
- Physical Exam is usually normal
- Labs:
  - CBC, CMP – if hypercalcemia or anemia, may consider further testing for multiple myeloma or hematologic malignancy
  - Lumbar puncture / CSF labs not usually necessary unless atypical features on imaging such as leptomeningeal enhancement
- Imaging
  - CT with contrast (72% of meningiomas contrast enhance vs. 60% that appear hyperdense without contrast)
  - MRI with contrast

Neuroimaging Features



Typical	Atypical
▪ Smooth contour ▪ Homogeneous enhancement ▪ Dural tail ▪ Calcification	▪ Large or disproportionate amount of associated edema ▪ Intratumoral cystic change ▪ Extensive bone involvement ▪ Brain or leptomeningeal invasion ▪ Low apparent diffusion coefficient ▪ Elevated cerebral blood volume

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- A: Tail sign: tapering dural thickening (arrow) due to direct tumor involvement or reactive change in the dura, is a highly characteristic sign of meningioma; note also the small reactive arachnoid cyst, reflecting the extra-axial site of this lesion (arrowhead).
- B: Reactive cerebral white matter changes.
- C: Bony reactive changes: reactive bone sclerosis, reflected in the increased vault thickness (arrows) is seen most commonly in patients with multiple meningiomas associated with neurofibromatosis type 2; invasive tumors such as these can cause problematic extracranial facial masses.

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Differential Diagnosis

- Lymphoma
- Plasmacytoma
- Metastatic carcinoma
- Melanocytic neoplasms
- Solitary fibrous tumor
- Gliosarcoma
- Inflammatory lesions such as
  - Sarcoidosis
  - Granulomatosis with polyangiitis
- Infections such as tuberculosis

WHO Classificiation

- Distinguished by histological subtype

World Health Organization (WHO) Meningioma Classifications

WHO Grade I Benign	WHO Grade II Atypical	WHO Grade III Malignant
Meningiothelial	Chordoid	Papillary
Fibrous (fibroblastic)	Clear Cell	Rhabdoid
Transitional (mixed)	Atypical	Anaplastic
Psammomatous		
Angiomatous		
Microcystic		
Secretory		
Lymphoplasmacyte-rich		
Metaplastic		

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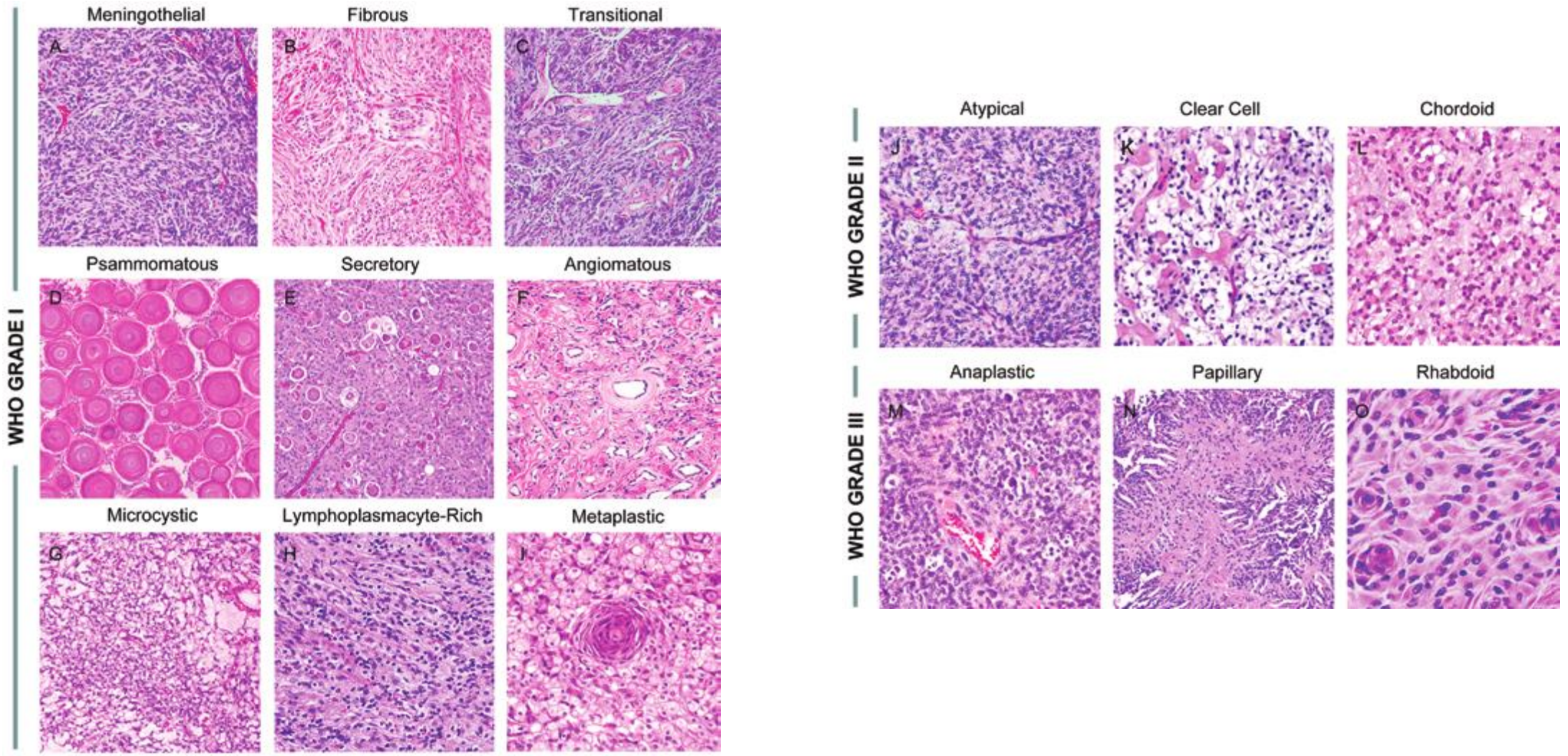
Histopathology of Meningioma



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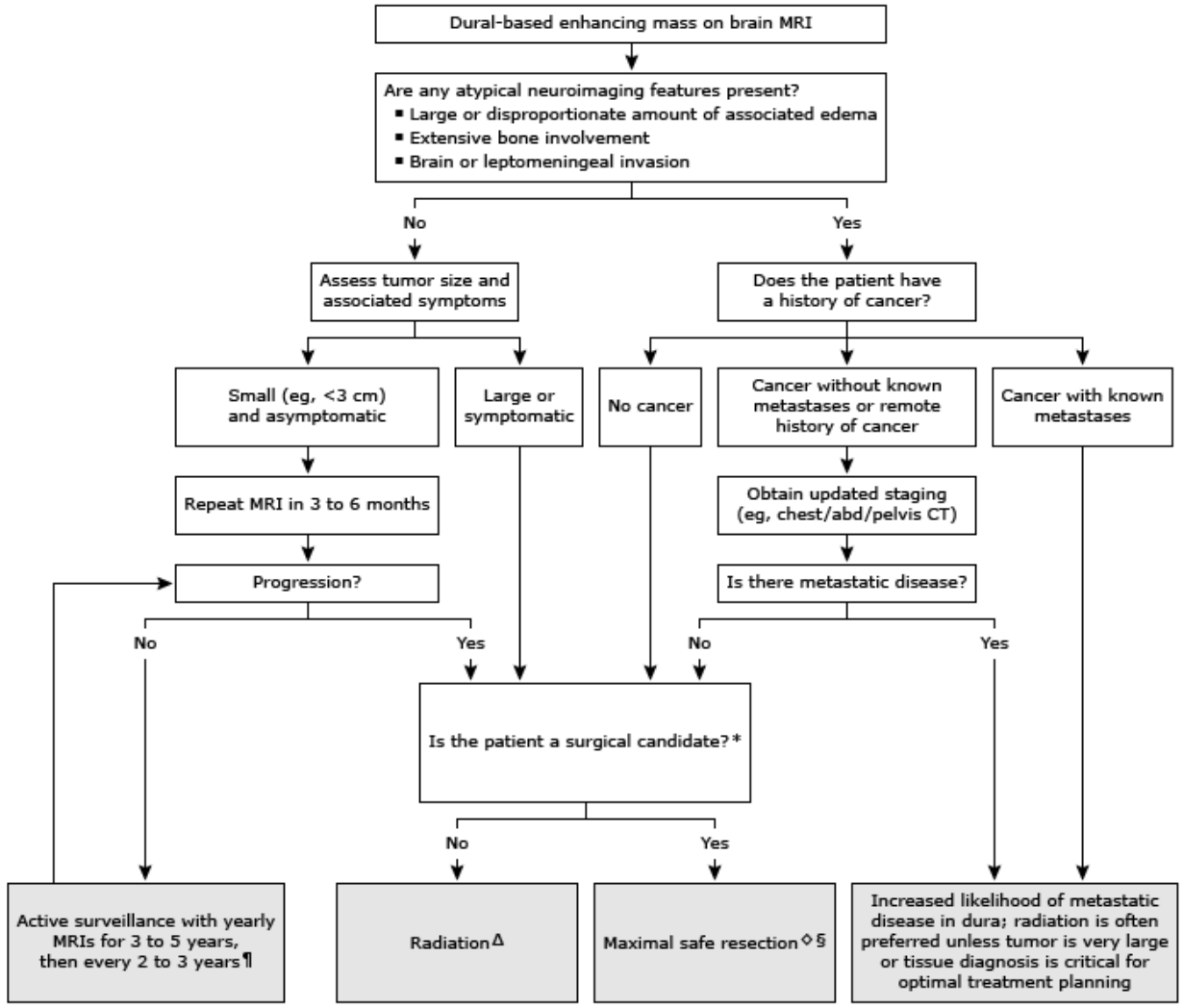
Treatment

- Some may not require treatment:
  1. Incidental discovery
  2. Elderly patients, consider expected survival
  3. Patients with significant comorbidities
  4. Risks outweigh benefits
  5. Meningiomas that have been stable for a long period (likely highly calcified)
- Consider reducing hormone exposure if possible
- Surgical candidacy varies based on tumor location (eg, deep vs superficial, proximity to critical structures), imaging evidence of invasion, and presurgical status of the patient
- Radiation



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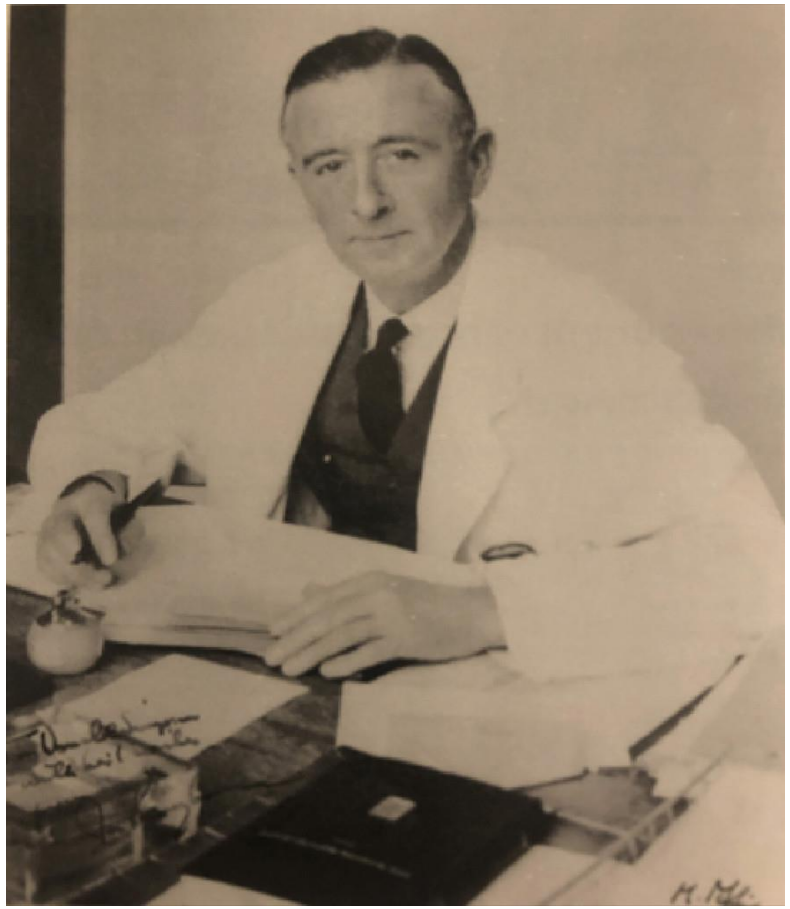
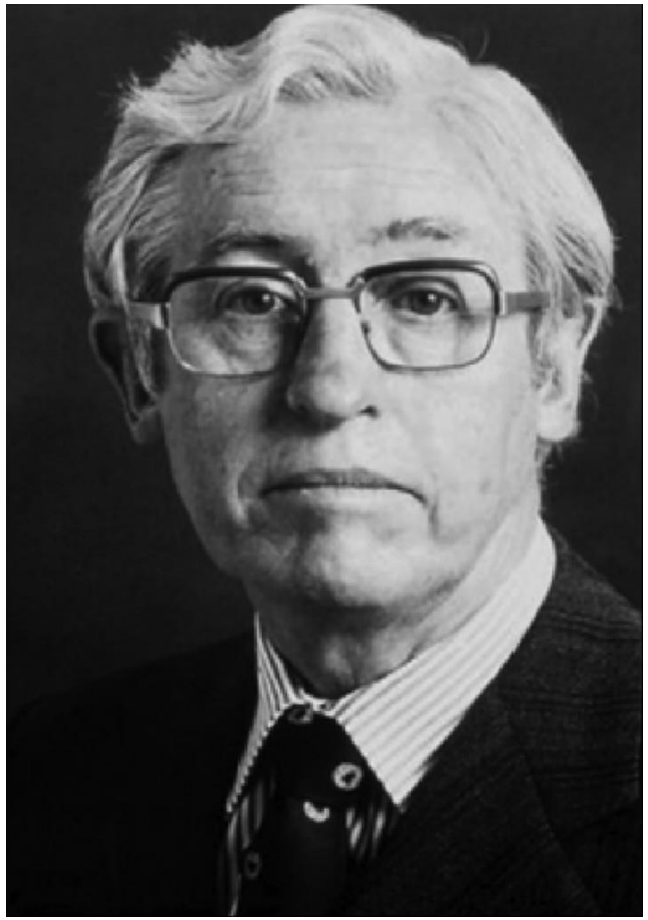
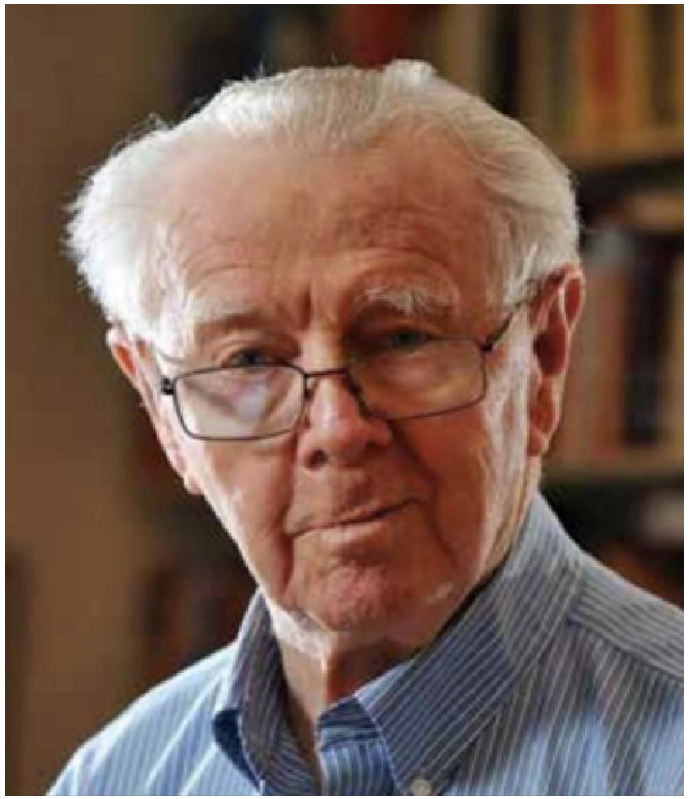
Donald Simpson (1927-2018)







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Simpson Grading of Meningioma Resection

- **grade I**
  - complete removal including resection of underlying bone and associated dura
  - **9%** symptomatic recurrence at 10 years
- **grade II**
  - complete removal and coagulation of dural attachment
  - **19%** symptomatic recurrence at 10 years
- **grade III**
  - complete removal w/o resection of dura or coagulation
  - **29%** symptomatic recurrence at 10 years
- **grade IV**
  - subtotal resection
  - **44%** symptomatic recurrence at 10 years
- **grade V**
  - simple decompression with or without biopsy
  - **100%** symptomatic recurrence at 10 years (small sample in original paper)

Original 1957 paper:

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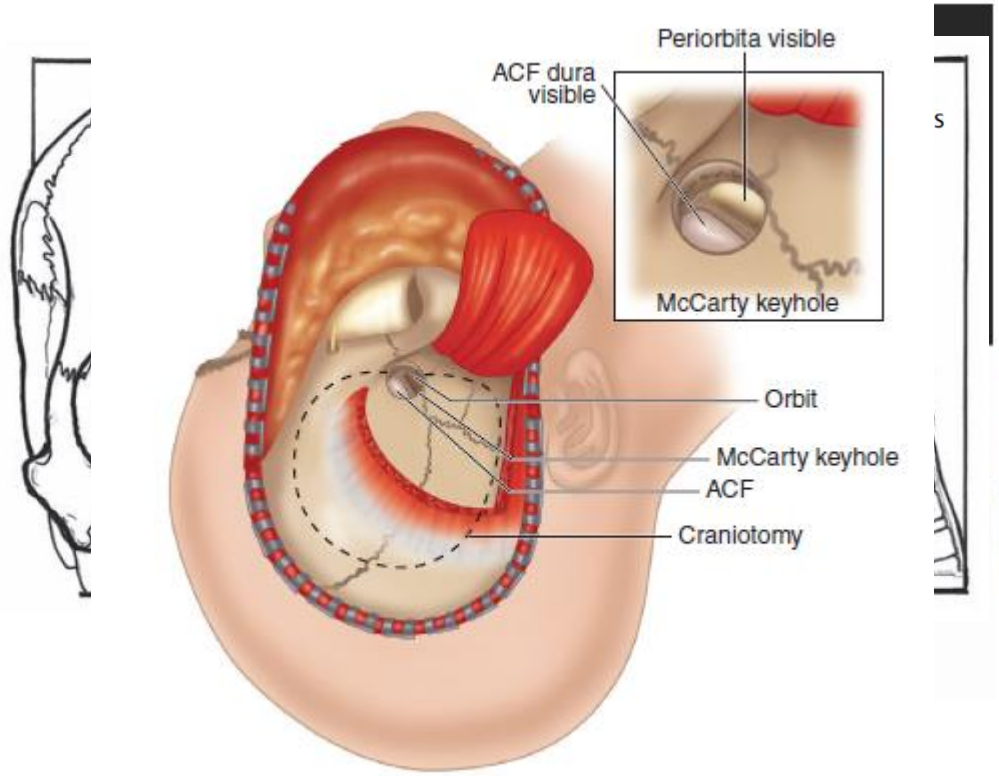
Surgical Approaches

- Endoscopic endonasal transcribiform
- Bifrontal transbasal
- Pterional
- in a series of 25 patients, 19 patients underwent bifrontal approach, and pterional approach was used in the remaining six



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Pathology

Immunohistochemistry

- 80% positive for Epithelial Membrane Antigen (EMA)
- 39-80% positive for Progesterone Receptor (PR)
- 33% positive for S-100
- 15% positive for CD34
- pHH3 – can be used for prognostication
- Ki-67 – can be used for prognostication
- Usually GFAP negative

Molecular risk stratification

- TERT promotor region mutation

OR

- Homozygous CDKN2A/B deletion

are independent WHO classification criteria for Grade 3 meningioma

- loss of trimethylation of lysine 27 of histone 3 (H3K27me3) associated with increased rate of recurrence

Case 1 presentation

- 46yo right-handed African-American female
- h/o iron deficiency anemia, HTN, tobacco use
- Presents with new onset, witnessed generalized tonic-clonic seizure, 4 min long
  - Systolic BP in 190s
- Personality change over the past several years
  - irritability, confusion
- Menorrhagia for some time
- Twin sister had seizures at age 25, controlled medically then weaned, now seizure free
- Familial history of leukemia
- No focal neurological deficits

- admitted to ICU

Case 1 Workup

- CBC, CMP
- LTM EEG showed no epileptiform activity
- CTA brain and neck
- MRI brain
- CT brain without contrast

WBC	4.00 - 11.00 10^3/uL	8.94
RBC	3.50 - 5.20 10^6/uL	4.12
HGB	11.0 - 16.0 g/dL	8.6 ↓
HCT	34.0 - 47.0 %	29.6 ↓
MCV	80.0 - 100.0 fL	71.8 ↓
MCH	26.0 - 33.0 pg	20.9 ↓
MCHC	31.0 - 35.0 g/dL	29.1 ↓
RDW	12.0 - 15.0 %	25.8 ↑
RDW-SD	38.0 - 52.0 fL	64.3 ↑
PLATELET	140 - 400 10^3/uL	303
MPV	9.0 - 12.0 fL	10.4
PLATELET EST		AGREE
GIANT/ENLARGED PLT		4.30
MICRO	%	<25
MACRO	%	<25
HYPO	%	<25
POLY	%	1-5
SCHISTOCYTE	%	1-3
TARGET	%	5-10
TEAR DROP	%	1-3
CRENATED	%	10-25
SEG	%	93.9
LYMPHOCYTE	%	4.4
MONOCYTE	%	0.0
EOSINOPHIL	%	0.0
BASOPHIL	%	1.7
ABSOLUTE NEUTR	1.60 - 7.70 10^3/uL	8.39 ↑
ABSOLUTE LYMPH	1.00 - 4.90 10^3/uL	0.39 ↓
ABSOLUTE MONO	0.00 - 1.10 10^3/uL	0.00
ABSOLUTE EOS	0.00 - 0.50 10^3/uL	0.00
ABSOLUTE BASO	0.01 - 0.20 10^3/uL	0.18

CALCIUM	8.9 - 10.6 mg/dL	9.0
GLUCOSE	74 - 100 mg/dL	120 ↑
BUN	7 - 19 mg/dL	5 ↓
CREATININE	0.55 - 1.02 mg/dL	0.66
TOTAL PROTEIN	6.0 - 8.0 g/dL	7.6
ALBUMIN	3.5 - 5.0 g/dL	3.9
BILIRUBIN, TOTAL	0.2 - 1.2 mg/dL	1.5 ↑
AST	5 - 34 U/L	37 ↑
ALT	0 - 55 U/L	28
ALKALINE PHOSPHATASE	40 - 150 U/L	67
SODIUM	136 - 145 mmol/L	139
POTASSIUM	3.5 - 5.1 mmol/L	3.1 ↓
CHLORIDE	98 - 107 mmol/L	111 ↑
CO2	22.0 - 29.0 mmol/L	19.0 ↓
GFR:NON-AFRICAN AMERICAN	arbitrary unit	>60
GFR:AFRICAN AMERICAN	arbitrary unit	>60

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Case 1 Surgical Resection

Surgeon – Dr. Mostafa
Resident – Dr. Johnson
CSA – Justin Maxey

- Right fronto-temporal / pterional craniotomy, subfrontal approach
- Steal neuro navigation
- Devascularized from skull base and debulked with ultrasonic aspirator
- Intraoperative frozen section favored meningioma
- Small piece of tumor adherent to right ACA A1 segment was left (Simpson grade IV?)
- Enormous amount of irrigation prior to closure

Case 1 Pathology

- FINAL DIAGNOSIS :

  A. Skull base tumor:
  Meningioma, WHO grade 1.

  B. Skull base tumor:
  Meningioma, WHO grade 1.

  Comment: Sections show fragments of meningothelial cells, fibrin, red cells, and cell debris.
  Well-controlled immunohistochemical stains as follows:
  Positive for epithelial membrane antigen (EMA), strongly positive for progesterone receptor, with 2 to 3% positivity for Ki-67 proliferation marker.

Meningothelial meningiomas (also known as syncytial or endothelial meningiomas) are the most common histological subtype of meningioma, found in ~60% of all meningiomas, most frequently combined with fibrous meningioma (40%) or in isolation (17%)

Case 1 Post-Op

- post-operatively sent to ICU, intubated
- Extubated on POD#1
- POD#2
  - no neurological deficits
  - Some right periorbital edema
- POD#3
  - Advanced to regular diet
- Discharged on POD#5

Case 2 Presentation

- 44yo right-handed female
- h/o vit D deficiency, ADD, tobacco use
- Presented to clinic with gradual worsening of vision L>R, anosmia for 7 years
- MR brain ordered after ophthalmological assessment showed 7cm presumed meningioma in olfactory groove

- Admitted for elective surgical resection preceded by endovascular embolization

Case 2 Workup

- CBC, CMP
- MRI brain with and without contrast
- MRI brain stealth
- Cerebral four vessel angiogram
- CT brain without contrast

WBC	4.00 - 11.00 10^3/uL	9.24
RBC	3.50 - 5.20 10^6/uL	4.64
HGB	11.0 - 16.0 g/dL	14.5
HCT	34.0 - 47.0 %	43.4
MCV	80.0 - 100.0 fL	93.5
MCH	26.0 - 33.0 pg	31.3
MCHC	31.0 - 35.0 g/dL	33.4
RDW	12.0 - 15.0 %	12.6
RDW-SD	38.0 - 52.0 fL	43.1
PLATELET	140 - 400 10^3/uL	296
MPV	9.0 - 12.0 fL	8.9 ↓
SEG	%	61.9
LYMPHOCYTE	%	31.5
MONOCYTE	%	4.7
EOSINOPHIL	%	1.5
BASOPHIL	%	0.3
IMMATURE GRANULOCYTE	%	0.1
ABSOLUTE NEUTR	1.60 - 7.70 10^3/uL	5.72
ABSOLUTE LYMPH	1.00 - 4.90 10^3/uL	2.91
ABSOLUTE MONO	0.00 - 1.10 10^3/uL	0.43
ABSOLUTE EOS	0.00 - 0.50 10^3/uL	0.14
ABSOLUTE BASO	0.00 - 0.20 10^3/uL	0.03
ABSOLUTE IMMATURE GRANULOCYTE	0.00 - 0.09 10^3/uL	0.01

CALCIUM	8.4 - 10.2 mg/dL	8.8
GLUCOSE	74 - 106 mg/dL	116 ↑
BUN	7 - 17 mg/dL	5 ↓
CREATININE	0.52 - 1.04 mg/dL	0.62
TOTAL PROTEIN	6.3 - 8.2 g/dL	6.7
ALBUMIN	3.5 - 5.0 g/dL	3.8
BILIRUBIN, TOTAL	0.2 - 1.3 mg/dL	0.4
AST	14 - 36 U/L	22
ALT	0 - 34 U/L	25
ALKALINE PHOSPHATASE	38 - 126 U/L	72
SODIUM	137 - 145 mmol/L	144
POTASSIUM	3.5 - 5.1 mmol/L	3.2 ↓
CHLORIDE	98 - 107 mmol/L	112 ↑
CO2	22.0 - 30.0 mmol/L	26.0

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Case 2 Surgical Resection

Preceded by left meningeal and left sphenopalatine branch embolization with Onyx

Surgeon – Dr. Mostafa
CSA – Tammi Holmes

- Left pterional subfrontal approach
- Devascularization and debulking with bipolar electrocautery and elliquence device
- Gross total resection was achieved
- Both optic nerves and optic chiasm were decompressed
- Enormous amount of irrigation prior to closure

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Case 2 Pathology

FINAL DIAGNOSIS:

A and B: Skull base tumor:
Meningioma, fibrous type, WHO Grade 1.

Comment: Well-controlled immunostains show the lesional cells to be positive for EMA, diffusely positive for PR; negative for S100 and GFAP. Ki67 proliferative index is approximately 10%.

Fibrous meningiomas (also known as fibroblastic meningiomas) are the second most common histological subtype of meningioma, found in ~50% of all meningiomas, usually along with meningothelial histology (40%) or in isolation (7%). They are, for some reason, the most common intraventricular meningioma histological subtype.

Case 2 Post-Op

- Post-operatively sent to ICU, intubated
- POD#1
  - Extubated on room air
  - hypernatremic to 154, concern for DI
- POD#2
  - Agitated, drowsy, impulsive, not following commands
- POD#5
  - Hypernatremia resolved
- POD#7
  - Admitted to inpatient rehab for deficits in mobility, ADLs, IADLs, speech, language, cognition
- Discharged on POD#17

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Images:

1. Case courtesy of Dr Victor Yang, Radiopaedia.org, rID: 36459
2. Rodríguez-Porcel F, Hughes I, Anderson D, Lee J, Biller J. Foster Kennedy Syndrome Due to Meningioma Growth during Pregnancy. Front Neurol. 2013;4:183. Published 2013 Nov 11. doi:10.3389/fneur.2013.00183
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