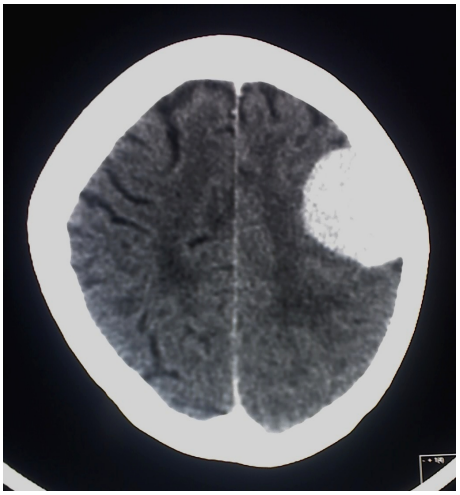
WORLD SKULL BASE E-LEARNING MATERIAL

Meningioma

Meningioma

Meningioma	
Classification and external resources	
 A contrast enhanced CT scan of the brain, demonstrating the appearance of a meningioma	
ICD-10	C70 [1], D32 [2]
ICD-9	225.2 [3]
ICD-O:	M9530/0 [4]
OMIM	607174 [5]
DiseasesDB	8008 [6]
eMedicine	neuro/209 [7] radio/439 [8]
MeSH	D008579 [9]

Meningiomas are a diverse set of tumors arising from the meninges, the membranous layers surrounding the central nervous system.[] They arise from the arachnoid "cap" cells of the arachnoid villi in the meninges.[] These tumors are usually benign in nature; however, a small percentage are malignant.[] Many meningiomas are asymptomatic, producing no symptoms throughout a person's life, and require no treatment other than periodic observation. Symptomatic meningiomas are typically treated with either radiosurgery or conventional surgery. Historical evidence of meningiomas has been found going back hundreds of years, with some successful surgeries for their removal beginning in the 1800s.

History and nomenclature

The neoplasms currently referred to as meningiomas were referred to with a wide range of names in older medical literature, depending on the source. Various descriptors included "fungoid tumors", "fungus of the dura mater", "epithelioma", "psammoma", "dural sarcoma", "dural endothelioma", "fibrosarcoma", "angioendothelioma", "arachnoidal fibroboastoma", "endotheliosis of the meninges", "meningeal fibroblastoma", "meningoblastoma", "mestothelioma of the meninges", "sarcoma of the dura", and others. The modern term of "meningioma" was first used by Harvey Cushing (1869–1939) in 1922, to describe a set of tumors that occur throughout the neuraxis (brain

and spinal cord), but have various commonalities.[10][11] Charles Oberling then separated these into subtypes based on cell structure, and several other researchers over the years have defined dozens of different subtypes as well. In 1979, the World Health Organization (WHO) classified seven subtypes, upgraded in 2000 to a classification system with nine low-grade variants (grade I tumors) and three variants each of grade II and grade III meningiomas.[11] The most common subtypes are Meningotheliomatous (63%), transitional or mixed-type (19%), fibrous (13%), and psammomatous (2%).[12]

The earliest evidence of a probable meningioma is from a skull approximately 365,000 years old, which was found in Germany. Other probable examples have been discovered in other continents around the world, including North and South America, and Africa. The earliest written record of what was probably a meningioma is from the 1600s, when Felix Plater (1536–1614) of the University of Basel performed an autopsy on Sir Caspar Bonecurtius.[13] Surgery for removal of meningiomas was first attempted in the 18th century, with the first successful surgery for removal of a meningioma performed in 1835 by Zanobi Pecchioli, Professor of Surgery at the University of Siena.[14] Other notable meningioma researchers have been William Macewen (1848–1924), and William W. Keen (1837–1932).[13] Medical science has continued to make dramatic improvements in meningioma research and treatment over the last century, both in terms of the surgical techniques for resection (removal) of the tumor, and related improvements in anesthesia, antiseptic methods, techniques to control blood loss, and better ability to determine which tumors are and are not operable.[15]

Causes

The causes of meningiomas are not well understood.[16] Most cases are sporadic, appearing randomly, while some are familial. Persons who have undergone radiation, especially to the scalp, are more at risk for developing meningiomas, as are those who have suffered brain injury at some time.[17] Atomic bomb survivors from Hiroshima were more likely to develop meningiomas, with the incidence increasing the closer that they were to the site of the explosion. Dental x-rays increase the risk of meningioma, in particular for patients who had frequent dental x-rays in the past, when the x-ray dose of a dental x-ray was higher than in the present.[18]

Patients with neurofibromatosis type 2 (NF-2) have a 50% chance of developing one or more meningiomas. Studies of cell phones have found no association between cell phone use and incidence of meningiomas.[12][]

Many individuals have meningiomas but remain asymptomatic (no symptoms) for their entire life, so the meningiomas are not discovered until after an autopsy. One to two percent of all autopsies reveal meningiomas that were unknown to the individuals during their lifetime, since there were never any symptoms. In the 1970s, tumors causing symptoms were discovered in 2 out of 100,000 people, while tumors discovered without causing symptoms occurred in 5.7 out of 100,000, for a total incidence of 7.7/100,000. With the advent of modern sophisticated imaging systems such as CT scans, the discovery of asymptomatic meningiomas has tripled. Meningiomas are more likely to appear in women than men, though when they appear in men, are more likely to be malignant. Meningiomas can appear at any age, but most commonly are noticed in men and women age 50 or older, with meningiomas becoming more likely with age. They have been observed in all cultures, Western and Eastern, in roughly the same proportion as other possible brain tumors.[12]

Ninety-two percent of meningiomas are benign, with 8% being either atypical or malignant.[12]

Genetic causes

The most frequent genetic mutations (~50%) involved in meningiomas are inactivation mutations in the neurofibromatosis 2 gene (merlin) on chromosome 22q.

Other possible genes/loci include:

- AKT1
- MN1[]
- PTEN[]
- SMO
- an unknown gene at 1p13[]

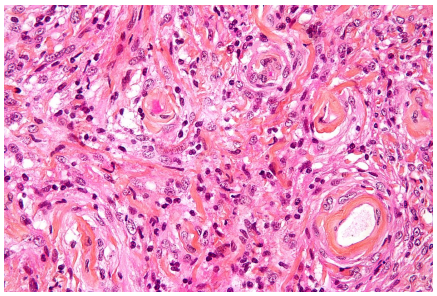
Signs and symptoms

Small tumors (e.g., < 2.0 cm) are usually incidental findings at autopsy without having caused symptoms. Larger tumors can cause symptoms depending on the size and location.

- Focal seizures may be caused by meningiomas that overlie the cerebrum.
- Progressive spastic weakness in legs and incontinence may be caused by tumors that overlie the parasagittal frontoparietal region.[*citation needed*]
- Sylvian tumors may cause myriad motor, sensory, aphasic, and seizure symptoms, depending on the location.
- Increased intracranial pressure eventually occurs, but is less frequent than in gliomas.
- Diplopia (Double vision) or uneven pupil size can be symptoms if related pressure causes a 3rd and/or 6th nerve palsy.

Mechanism

Meningiomas arise from arachnoidal cells,[] most of which are near the vicinity of the venous sinuses, and this is the site of greatest prevalence for meningioma formation. They are most frequently attached to the dura over the superior parasagittal surface of frontal and parietal lobes, along the sphenoid ridge [19], in the olfactory grooves, the sylvian region, superior cerebellum along the *falx cerebri*, cerebellopontine angle, and the spinal cord. The tumor is usually gray, well-circumscribed, and takes on the form of space it occupies. They are usually dome-shaped, with the base lying on the dura.



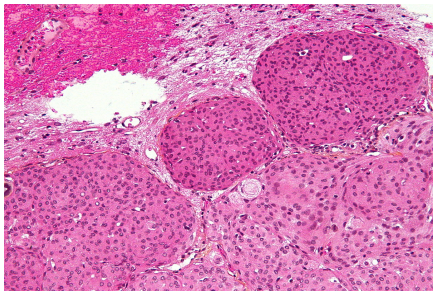
Micrograph of a meningioma showing the characteristic whorling. HPS stain

Locations

- Parasagittal/falcine (25%)
- Convexity (surface of the brain) (19%)
- Sphenoid ridge (17%)
- Suprasellar (9%)
- Posterior fossa (8%)
- Olfactory groove (8%)
- Middle fossa/Meckel's cave (4%)
- Tentorial (3%)
- Peri-torcular (3%)

Other uncommon locations are the Lateral ventricle, Foramen magnum, and the orbit/optic nerve sheath.[12] Meningiomas can also occur as a spinal tumor, more often in women than in men, which occurs more often in Western countries than Asian.

Histologically, meningioma cells are relatively uniform, with a tendency to encircle one another, forming whorls and psammoma bodies (laminated calcific concretions).[] They have a tendency to calcify and are highly vascularized.



Micrograph of a meningioma with brain invasion (WHO Grade II); the tumour (bottom/right of image) has the typical "pushing border" invasion into the cerebral cortex (top/left of image). HPS stain

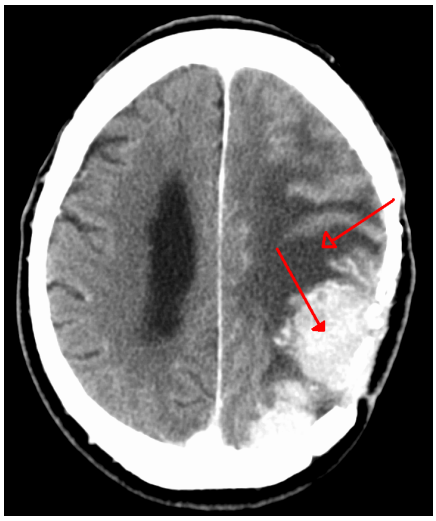
Meningiomas are often considered benign tumors curable by surgery, but most recurrent meningiomas correspond to histologic benign tumors. The metabolic phenotype of these benign recurrent meningiomas indicated an aggressive metabolism resembling that observed for atypical meningioma.[]

Diagnosis

Meningiomas are readily visualized with contrast CT, MRI with gadolinium,[20] and arteriography, all attributed to the fact that meningiomas are extra-axial and vascularized. CSF protein is usually elevated if lumbar puncture is attempted.

Though the majority of meningiomas are benign, they can have malignant presentations. Classification of meningiomas are based upon the WHO classification system.[]

- Benign (Grade I) – (90%) – meningothelial, fibrous, transitional, psammomatous, angioblastic (most aggressive)
- Atypical (Grade II) – (7%) – chordoid, clear cell, atypical (includes brain invasion)
- Anaplastic/malignant (Grade III) – (2%) – papillary, rhabdoid, anaplastic



A meningioma that had previously been operated on with surrounding edema,

In a 2008 review of the latter two categories, atypical and anaplastic-meningioma cases, the mean overall survival for atypical meningiomas was found to be 11.9 years vs. 3.3 years for anaplastic meningiomas. Mean relapse-free survival for atypical meningiomas was 11.5 years vs. 2.7 years for anaplastic meningiomas.[21]

Malignant anaplastic meningioma is an especially malignant tumor with aggressive behavior. Even if – by general rule – neoplasms of the nervous system (brain tumors) cannot metastasize into the body because of the blood–brain barrier, anaplastic meningioma can. Although they are inside the cerebral cavity, they are located on the bloodside of the BBB, because meningiomas tend to connect themselves to blood vessels to feed. Cancerized cells can thus escape into the bloodstream, which is why meningiomas, when they metastasize, often turn up around the lungs. Anaplastic meningioma and hemangiopericytoma are difficult to distinguish even by pathological means as they look similar, especially if the first occurrence is a meningeal tumor, and both tumors occur in the same places (same types of tissue).[*citation needed*]

Treatment

Observation

Observation with close imaging follow-up can be used in select cases if a meningioma is small and asymptomatic. In a retrospective study on 43 patients, 63% of patients were found to have no growth on follow-up, and the 37% found to have growth grew at an average of 4 mm / year.[22] In this study, younger patients were found to have tumors that were more likely to grow on repeat imaging; thus are poorer candidates for observation. In another study, clinical outcomes were compared for 213 patients undergoing surgery vs. 351 patients under watchful observation.[23] Only 6% of the conservatively treated patients later developed symptoms, while among the surgically treated patients, 5.6% developed persistent morbid condition, and 9.4% developed surgery-related morbid condition.

Observation is not recommended in tumors already causing symptoms. Furthermore, close follow-up with imaging is required with an observation strategy to rule out an enlarging tumor.[24]

Surgical resection

Meningiomas can usually be surgically resected (partially removed) and result in a permanent cure if the tumor is superficial on the dural surface and easily accessible. Transarterial embolization has become a standard preoperative procedure in the preoperative management.[25] If invasion of the adjacent bone occurs, total removal is nearly impossible. It is rare for benign meningiomas to become malignant.

The probability of a tumor recurring or growing after surgery can be estimated by comparing the tumor's WHO (World Health Organization) grade and by the extent of surgery by the Simpson Criteria.[26]

Simpson Grade	Completeness of Resection	10-year Recurrence
Grade I	complete removal including resection of underlying bone and associated dura	9%
Grade II	complete removal + coagulation of dural attachment	19%
Grade III	complete removal w/o resection of dura or coagulation	29%
Grade IV	subtotal resection	40%

Radiation therapy

Radiation therapy may include photon-beam or proton-beam treatment, or fractionated external beam radiation. Radiosurgery can be used in lieu of surgery in small tumors located away from critical structures.[27] Fractionated external-beam radiation can also be used as primary treatment for tumors that are surgically unresectable, or for patients who are inoperable for medical reasons.

Radiation therapy is often considered for WHO grade I meningiomas after subtotal (incomplete) tumor resections. The clinical decision to irradiate after a subtotal resection is somewhat controversial, as no class I randomized, controlled trials exist on the subject.[28] Numerous retrospective studies, however, have strongly suggested the addition of postoperative radiation to incomplete resections improves both progression-free survival (i.e. prevents tumor recurrence) and improves overall survival.[29]

In the case of a grade III meningioma, the current standard of care involves postoperative radiation treatment regardless of the degree of surgical resection.[30] This is due to the proportionally higher rate of local recurrence for these higher-grade tumors. Grade II tumors can behave variably and there is no standard of whether to give radiotherapy following a gross total resection. Subtotally resected grade II tumors should be radiated.

Conventional chemotherapy

Current chemotherapies are likely not effective. Antiprogestin agents have been used, but with variable results.[31] A 2007 study that hydroxyurea has the capacity to shrink unresectable or recurrent meningiomas is being further evaluated.[32]

Notable cases

- Leonard Wood (1860–1927) underwent successful surgery by Dr. Harvey Cushing for a meningioma around 1910, a major advance in neurosurgery at the time[33]
- Crystal Lee Sutton (1940–2009), American union organizer and inspiration for the film *Norma Rae*, died of a malignant meningioma[][34]
- Kathi Goertzen (1958–2012),[35] TV news anchor in Seattle who underwent a very public battle with recurring tumors. She died on August 13, 2012 of complications related to her treatment. [36] [37][38][39]
- Hillary Howard, American TV news reporter, had a successful surgery to remove her meningioma in 2011[40][41]
- Mary Tyler Moore (b. 1936), American actress, underwent surgery in May 2011 to remove a benign meningioma[42][43]
- Sheryl Crow (b. 1962), American singer-songwriter, revealed to an audience in 2012 that she was diagnosed with meningioma, but her representative has stated that it is quite common. Crow is "happy and healthy."[44]

Notes

[1] http://apps.who.int/classifications/icd10/browse/2010/en#/C70
[2] http://apps.who.int/classifications/icd10/browse/2010/en#/D32
[3] http://www.icd9data.com/getICD9Code.ashx?icd9=225.2
[4] http://www.progenetix.net/progenetix/I95300/
[5] http://omim.org/entry/607174
[6] http://www.diseasesdatabase.com/ddb8008.htm
[7] http://www.emedicine.com/neuro/topic209.htm
[8] http://www.emedicine.com/radio/topic439.htm#
[9] http://www.nlm.nih.gov/cgi/mesh/2013/MB_cgi?field=uid&term=D008579
[10] Okonkwo et al. p. 5.
[11] Prayson, Richard A. "Pathology of Meningiomas". *Meningiomas*. pp. 31–41.
[12] Park, Bong Jin; Kim, Han Kyu; Sade, Burak; Lee, Joung H. "Epidemiology", *Meningiomas*, pp. 11–13.
[13] Okonkwo et al. pp. 7–8.
[14] Mirimanoff. p. 18.
[15] DeMonte. *Al-Mefty's Meningiomas*.
[19] http://radiology.casereports.net/index.php/rcr/article/viewArticle/357/718
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[30] Goyal LK et al. "Local control and overall survival in atypical meningioma: a retrospective study," Int J Radiat Oncol Biol Phys. 2000 Jan 1;46(1):57–61.
[31] Wahab M et al.: Meningioma and hormonal influences. Climacteric. 2003 Dec;6(4):285–92.
[32] Newton HB. Hydroxyurea chemotherapy in the treatment of meningiomas. Neurosurg Focus. 2007;23(4):E11.
[33] Okonkwo, David O.; Laws, Edward R. Jr. "Meningiomas: Historical perspective". *Meningiomas*. p. 8.

[35] Kathi Goertzen dies after long battle with brain tumors (http://www.komonews.com/kathi-goertzen/Kathi-Goertzen-dies-after-long-battle-with-brain-tumors-166022836.html) KOMO News, August 13, 2012. Retrieved on August 13, 2012.
[36] >
[37] KOMO's Goertzen cuts back anchor duties (http://www.seattlepi.com/tv/150349_goertzen29.html) Seattle Post-Intelligencer, November 29, 2003. Retrieved on February 25, 2012.
[38] Kathi Goertzen goes back to late nights on KOMO/4 (http://www.seattlepi.com/tv/295264_tf209.html) Seattle Post-Intelligencer, December 9, 2006. Retrieved on February 25, 2012.

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- DeMonte, Franco; Ossama Al-Mefty, Michael McDermott (2011). *Al-Mefty's Meningiomas* (2nd ed.). Thieme. ISBN 978-1-60406-053-9.

Further reading

- Cushing, Harvey; Eisenhardt, Louise (1938). *Meningiomas: their classification, regional behavior, life history, and surgical end results*. Springfield, Illinois: Charles C. Thomas.

External links

- (https://sites.google.com/site/meningiomainformation/) Epidemiology and more information about meningioma.
- MR/CT scans of meningioma (http://rad.usuhs.edu/medpix/medpix.html?mode=image_finder&srchstr=meningioma&srch_type=all&action=search#top) from MedPix
- CancerBackup (http://www.cancerbackup.org.uk/Cancertype/Brain/Typesofbraintumour/Meningioma)
- MR/CT scans of pneumosinus dilatans (http://rad.usuhs.edu/medpix/raw_image.html?mode=cow_viewer&pt_id=13349&imid=51843) from MedPix
- Meningioma Mommas (http://meningiomamommas.org), online support group for men and women with meningiomas, and their caregivers
- Meningioma Awareness Day (http://www.brainsciencefoundation.org/bAttendbEvents/UpcomingEvents/MeningiomaAwarenessDay/tabid/318/Default.aspx), annual conference hosted by Harvard University for patients, doctors, caregivers, and researchers
- Cancer.Net: Meningioma (http://www.cancer.net/patient/Cancer+Types/Meningioma)

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