Meningiomas
basic level

Overview
A meningioma is a type of tumor that grows from the protective membranes, called meninges, which surround the brain and spinal cord. Most meningiomas are benign (not cancerous) and slow growing; however, some can be malignant. Symptoms typically appear gradually and vary depending on the location and brain area affected. Because these are slow growing tumors, not all meningiomas need to be treated immediately. Treatment options focus on removing the tumor and relieving the compression on the brain.

What is a meningioma?
Three layers of membranes, called meninges, lying just under the skull, protect the brain and spinal cord. From the outermost layer inward they are: the dura mater, arachnoid mater, and pia mater. A meningioma grows from the arachnoid cells that form the middle layer, and are firmly attached to the dura. Some meningiomas contain cysts or calcified mineral deposits, and others contain hundreds of tiny blood vessels. Because meningiomas tend to grow inward, they commonly cause pressure on the brain or spinal cord (Fig. 1). They can also grow outward causing the skull to thicken (hyperostosis).

The World Health Organization (WHO) developed a classification system for all known tumor types, including meningiomas. Tumors are classified by their cell type and grade by viewing the cells taken during a biopsy under a microscope. Treatment varies depending on the grade of your meningioma.

- **WHO, Grade I** meningiomas are the slowest growing. If the tumor is not causing symptoms, it may be best to observe its growth over time with periodic MRI scans. If there is a chance the tumor will grow enough in your lifetime to cause symptoms, then surgical removal may be recommended. Most Grade I meningiomas are treated with surgery and continued observation.
- **WHO, Grade II** meningiomas are called atypical. They are slightly more aggressive in growth than Grade I and have a slightly higher risk of recurrence. Surgery is the first line treatment for these types of meningiomas. Some Grade II meningiomas require radiation after surgery.

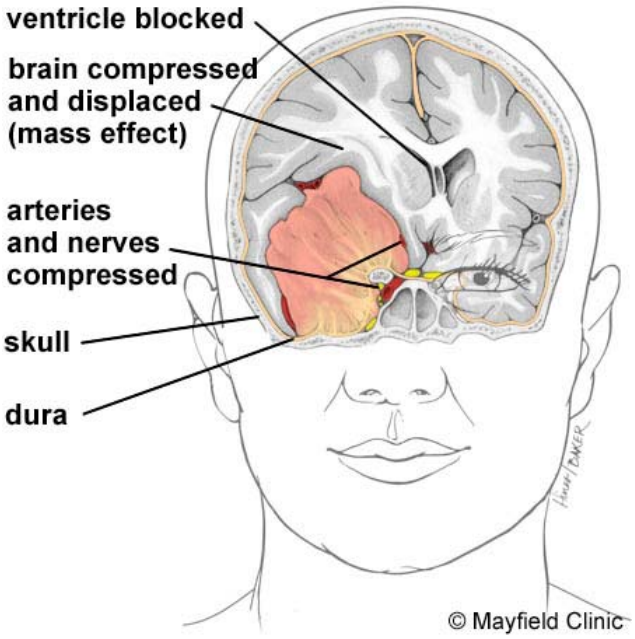

Figure 1. Meningiomas grow from the dura covering of the brain. As the tumor grows, it compresses and displaces normal brain tissue. Increasing size, pressure, and swelling cause neurologic symptoms.

- **WHO, Grade III** meningiomas are the most aggressive and are called malignant or anaplastic. Malignant meningiomas account for less than 1% of all meningiomas. Surgery is the first line treatment for Grade III meningiomas followed by radiation. If the tumor recurs, chemotherapy is used.

What are the symptoms?
Meningiomas grow slowly; it may take years before they cause symptoms. Some people with meningiomas have no symptoms. The tumor may be found incidentally on a diagnostic scan performed for another reason such as a trauma. Symptoms of a meningioma vary by location and size of the tumor. They often first appear as headaches and seizures, primarily due to increased pressure of the growing tumor. Weakness in the arms or legs, or loss of sensation, may occur with spinal cord meningiomas.

Meningiomas are often named according to their location and symptoms:

- **Convexity meningiomas:** grow on the surface of the brain. They may not produce symptoms until they reach a large size. Symptoms include seizures, neurological deficits, or headaches.
- **Falx and parasagittal meningiomas:** grow from the dural fold, called the falx, which runs between the left and right sides of the brain. The falx contains two large blood vessels (superior and inferior sagittal sinuses). Because of the danger of injuring the sinuses, removing a tumor in the falx or parasagittal region can be difficult. Symptoms may include personality changes, headache, vision problems, and arm or leg weakness.
- **Olfactory groove meningiomas:** grow along the olfactory nerves that run between the brain and the nose. These tumors often cause a loss of smell. They can compress the frontal lobes causing personality changes that may be mistaken for depression. They can also compress the optic nerves to the eyes, causing visual problems such as loss of specific areas within your field of vision, or even blindness.
- **Sphenoid meningiomas:** grow along the sphenoid ridge, which lies behind the eyes. These tumors can cause visual problems, loss of sensation in the face, or facial numbness. Tumors in this location can sometimes encase the major blood vessels of the brain (e.g. cavernous sinus, or carotid arteries) as well as the cranial nerves in the area making them difficult to completely remove.
- **Posterior fossa meningiomas:** grow along the underside of the brain near the brainstem and cerebellum. These tumors can compress the cranial nerves causing facial symptoms or loss of hearing. Petroclival tumors can compress the trigeminal nerve, resulting in facial pain (trigeminal neuralgia) or spasms of the facial muscles. Foramen magnum meningiomas grow near the area where the spinal cord connects to the brain and can cause headaches, or other signs of brainstem compression such as difficulty walking.
- **Intraventricular meningiomas:** grow inside the fluid-filled ventricles deep inside the brain. They can block the flow of cerebrospinal fluid (CSF) causing hydrocephalus, which can produce headaches and dizziness.
- **Intraorbital meningiomas:** grow around the eye sockets of the skull and can cause pressure in the eyes to build up, giving a bulging appearance. They can also cause loss of vision.
- **Spinal meningiomas:** grow predominantly in the thoracic spine. They can cause back pain (typically at night) or loss of sensation and paralysis of the legs from compression of the spinal nerves.

What are the causes?

Scientists are not certain what causes meningioma tumors, although several theories are being investigated. Most agree that a malformed chromosome is the most common abnormality in meningiomas, but the cause of this abnormality is unknown. People with a genetic disorder known as neurofibromatosis type 2 (NF2) are more likely to develop meningiomas. Of people with malignant meningiomas, a higher percent have mutations in NF2. Studies have also found that patients who received radiation treatment to the head for medulloblastomas, ependymomas, or other tumors are at higher risk for developing meningiomas later in life secondary to the radiation.

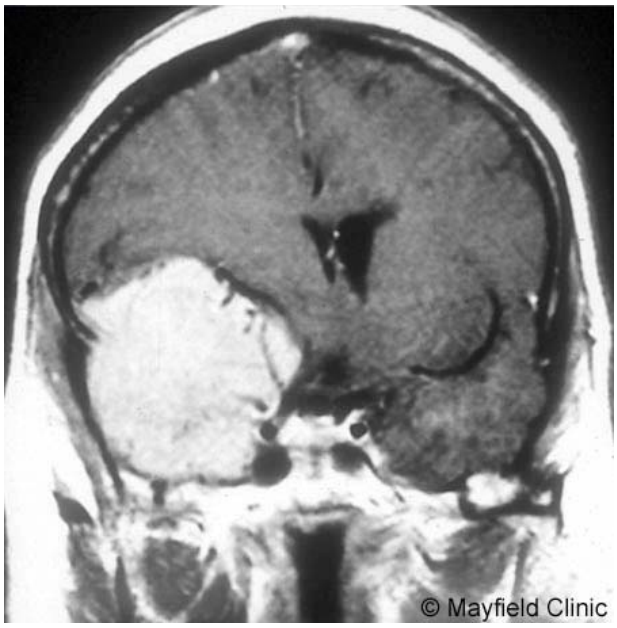

Figure 2. MRI of a sphenoid wing meningioma. The tumor has filled the area where the temporal lobe normally lies and has compressed the arteries and nerves causing vision problems.

Who is affected?

Meningiomas represent about 20% of all primary brain tumors and 12% of all spinal cord tumors. They can occur in children, but most often occur in adults between the ages of 40 and 60 years. Most meningiomas are benign (not cancerous), as less than 10% of meningiomas are malignant. While malignant meningiomas occur in both women and men, benign meningiomas occur most often in women.

How is a diagnosis made?

First, the doctor will ask about your personal and family medical history and perform a complete physical examination. In addition to checking your general health, the doctor performs a neurological exam. This includes checks for mental status and memory, cranial nerve function (sight, hearing, smell, tongue and facial movement), muscle strength, coordination, reflexes, and response to pain. If a problem is found, the doctor may order diagnostic imaging tests such as computerized tomography (CT) or magnetic resonance imaging

(MRI) scans to help determine the size, location, and type of tumor, if one exists (Fig. 2). Skull x-rays may be obtained if the tumor is believed to involve the bone. For spinal cord tumors, a myelogram may be done, and in some cases, angiograms, or x-rays of the blood vessels, are necessary. The diagnosis can be confirmed by a biopsy.

What treatments are available?

There are a variety of treatment options for meningiomas. The treatment that is right for you will depend on your age, general health status, and the location, size, and grade of the meningioma. Each treatment has benefits, risks and side effects that should be discussed and understood.

Observation (growth monitoring)
Because of the slow growth of meningiomas, patients with no or few symptoms may be monitored instead of undergoing surgical removal of the tumor. The doctor will monitor the growth of the tumor with periodic MRI scans. Patients should promptly report any symptom change immediately.

Surgery
Surgical removal is the most common treatment for meningiomas causing symptoms. A neurosurgeon performs a craniotomy to open the skull and remove the tumor (Fig. 3). A biopsy of tissue is examined by a pathologist to determine the tumor grade. Although total removal can provide a cure for meningiomas, total resection is not always possible. The tumor location determines how much can be safely removed. If the tumor cannot be completely removed, the remainder of the tumor can be treated with radiation.

Improvements in surgical techniques, particularly image-guided stereotaxy, intraoperative MRI/CT, and functional brain mapping have improved the surgeon's ability to precisely locate the tumor, define the tumor's borders, avoid injury to vital brain areas, and confirm the amount of tumor removal while in the operating room.

- **Image-guided surgery** (IGS) is a technology that helps the surgeon pinpoint the exact location of a tumor - similar to a GPS for the brain. Before surgery, a special MRI scan is performed with fiducial markers placed on the skin around the patient's head. During surgery, the fiducial markers correlate the "real patient" lying on the table to a 3D computer model of the patient created from their MRI or CT scans. Using a hand-held probe, the surgeon can track the probe's position in real time on the computer model of the patient's anatomy. IGS allows very precise planning of the approach by pinpointing the tumor location and guiding the skin and bone openings.

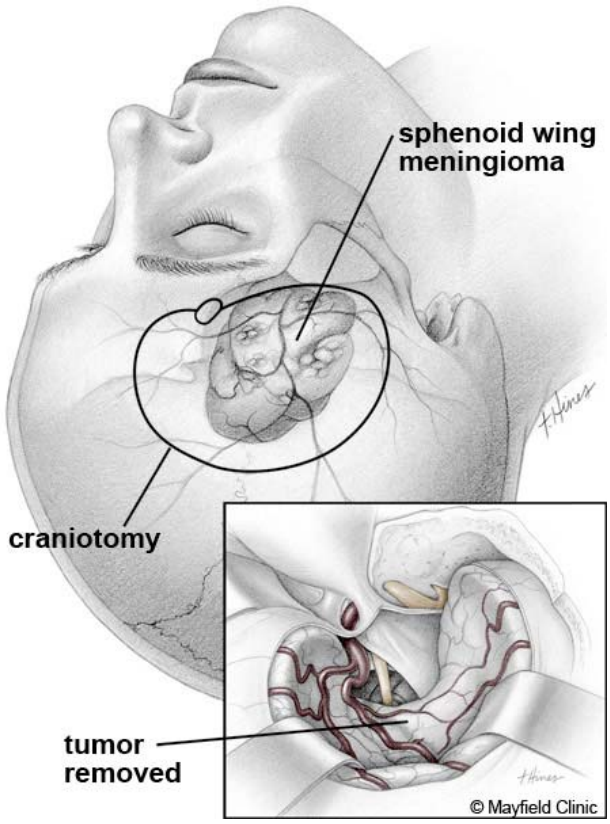

Figure 3. A craniotomy is cut in the skull to expose the tumor. After the tumor is removed, brain tissue can re-expand in the cavity.

- **Interventional MRI or CT** is a specially designed operating room in which the patient can undergo an MRI or CT scan before, during, and after surgery while still under anesthesia. This enables the surgeon to have real-time images of the patient's brain and to know exactly how much tumor has been removed prior to ending the procedure. This technology improves the ability for total tumor removal and reduces the need for a second operation.

Radiation
Some tumors may be considered inoperable because of their location near areas of the brain that control vital functions like breathing or intellect. Some malignant meningiomas grow back after surgical removal. In these cases, radiation may be used to damage the DNA inside the cells making them unable to divide and reproduce. The goal of radiation treatment is to maximize the dose to abnormal tumor cells and minimize exposure to normal healthy cells. The benefits of radiation are not immediate but occur over time. Gradually, the tumor will stop growing, shrink, and in some cases, completely disappear. There are two ways to deliver radiation: multiple low doses (radiotherapy) or a single high dose (radiosurgery).

- **Fractionated Stereotactic Radiotherapy (FSR)** delivers a low dose of radiation over many visits. A facemask is used to precisely locate the tumor and accurately reposition the patient for each treatment session. Patients return daily over 5 to 7 weeks to receive the complete radiation dose.

- **Stereotactic Radiosurgery (SRS)** delivers a high dose of radiation during a single session. Although it is called surgery, no incision is made. Because a single radiosurgery dose is more damaging than multiple fractionated doses, the target area must be precisely located and completely immobilized with a stereotactic head frame or mask. Patients spend most of the day at the center while the tumor is precisely located, a treatment plan is developed, and a radiation dose is delivered.

Clinical trials

Clinical trials are research studies in which new treatments - drugs, diagnostics, procedures, vaccines, and other therapies - are tested in people to see if they are safe and effective. Research is always being conducted to improve the standard of medical care and explore new drug and surgical treatments. Information about current clinical trials, including eligibility, protocol, and locations, are found on the Web. Studies can be sponsored by the National Institutes of Health (clinicaltrials.gov) as well as private industry and pharmaceutical companies (www.centerwatch.com).

Recovery

The location of the tumor is the most important factor in determining the outcome. Convexity, parasagittal and lateral sphenoid wing meningiomas usually are completely removable and surgery can yield excellent results. Optic, cavernous sinus, and skull base meningiomas have a higher rate of complication and are more difficult to completely remove. The patient's age and overall health prior to surgery may also affect the results. Meningiomas do sometimes recur after surgery or radiation. Regular follow-up MRI or CT scans (every one to three years) are an important part of long-term care for anyone diagnosed with a meningioma.

Sources & links

If you have more questions, please contact the Mayfield Clinic at 800-325-7787 or 513-221-1100. For information about the University of Cincinnati Neuroscience Institute Brain Tumor Center, call 866-941-8264.

Support

Support groups provides an opportunity for patients and their families to share experiences, receive support, and learn about advances in treatments and medications. Local support groups in the Cincinnati area include:

- Wellness Community of Greater Cincinnati, 513-791-4060
- Wellness Community of Northern Kentucky, 859-331-5568
- Pediatric Brain Tumor Support Group at Cincinnati Children's Hospital, 513-636-6369

Links

American Brain Tumor Association
(www.ABTA.org) 800-886-2282

National Brain Tumor Society
(www.braintumor.org) 800-934-2873

Glossary

anaplastic: when cells divide rapidly and bear little or no resemblance to normal cells in appearance or function.
benign: does not invade nearby tissues or spread; not cancerous.
biopsy: a sample of tissue cells for examination under a microscope to determine the existence or cause of a disease.
edema: tissue swelling caused by the accumulation of fluid.
hyperostosis: an excessive growth of bone.
meninges: three membranes (pia mater, arachnoid mater, and dura mater) that surround the brain and spinal cord.
hemangiopericytoma: very uncommon type of meningioma, highly vascular and usually fast growing.
mass effect: damage to the brain due to the bulk of a tumor, the blockage of fluid, and/or excess accumulation of fluid within the skull.
malignant: having the properties of invasive growth and ability to spread to other areas; cancerous.
meningioma: a tumor that grows from the meninges, the membrane that surrounds the brain and spinal cord.

updated > 10.2009
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