

Meningioma tumors

Overview

A meningioma is a tumor that grows from the meninges — the protective membranes that cover the brain and spinal cord. Most meningiomas are benign (not cancer) and slow growing; however, some can be malignant. Symptoms typically appear gradually and vary depending on the tumor location. Because of their slow growth, 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, arachnoid, and pia. A meningioma grows from the arachnoid cells that form the middle layer, and are firmly attached to the dura (Fig. 1). 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). They can also grow outward, causing the skull to thicken (hyperostosis).

A pathologist classifies meningiomas by their cell type and grade by viewing the cells taken during a biopsy under a microscope. Treatment varies, depending on the grade of the meningioma.

- **Grade I, benign** 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. Patients who undergo complete removal of a grade I meningioma usually do not require additional treatment. Patients who undergo incomplete removal may need radiation after surgery.
- **Grade II, atypical** meningiomas are slightly more aggressive in growth than grade I and have a slightly higher risk of recurrence. Surgery is the first line of treatment. Some will require radiation after surgery.
- **Grade III, malignant** meningiomas are the most aggressive and are called anaplastic.

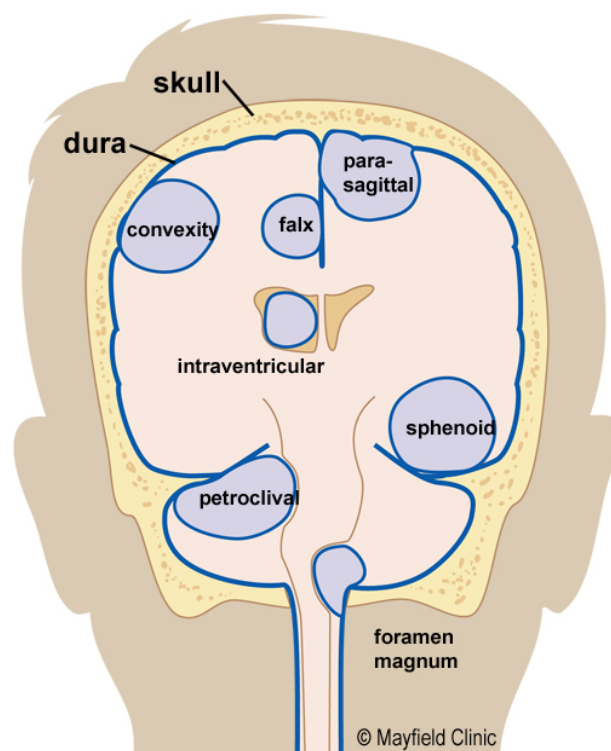


Figure 1. A meningioma is a tumor that grows from the dura membrane (blue) covering the brain and spinal cord. Meningiomas are named according to their location.

Malignant meningiomas account for less than 1% of all meningiomas. Surgery is the first line of treatment, followed by radiation. If the tumor recurs, chemotherapy is used.

What are the symptoms?

Meningiomas grow slowly and may not cause symptoms for years. Some people with meningiomas have no symptoms. The tumor may be found incidentally on an MRI or CT scan performed for another reason. Symptoms of a meningioma vary by the location and size of the tumor. They often first appear as headaches and seizures caused by increased pressure of the growing tumor. Weakness in the arms or legs, or loss of sensation, may occur with spinal cord meningiomas.

Meningiomas are named according to their location (Fig. 1) and cause various symptoms:

- **Convexity meningiomas:** grow on the surface of the brain. They may not produce symptoms until they reach a large size. Symptoms may include seizures, neurological deficits, or headaches.
- **Falx and parasagittal meningiomas:** grow from the dura fold that runs between the left and right sides of the brain. The falx contains two large blood vessels (sinuses) that can make surgical removal more difficult. Symptoms may include personality changes, headache, vision problems, and arm or leg weakness.
- **Olfactory groove meningiomas:** grow along the nerves that run between the brain and the nose and 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, causing visual problems or even blindness.
- **Tuberculum sella meningiomas:** grow near the pituitary gland and optic nerves. They often cause visual field problems.
- **Sphenoid meningiomas:** grow along the bony ridge behind the eyes. These tumors can cause visual problems, loss of sensation in the face, or facial numbness. They often encase major blood vessels (e.g., cavernous sinus, or carotid arteries) as well as the cranial nerves, 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 or spasms of the facial muscles.
- **Foramen magnum meningiomas:** grow where the spinal cord connects to the brain; they can cause headaches, brainstem compression, and difficulty walking.
- **Intraventricular meningiomas:** grow inside the fluid-filled ventricles of the brain. They block the flow of cerebrospinal fluid, causing hydrocephalus, headaches and dizziness.

- **Intraorbital meningiomas:** grow around the eye sockets of the skull and can cause a buildup of pressure in the eyes, giving a bulging appearance. They can also cause loss of vision.
- **Spinal meningiomas:** grow mainly 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 brain tumors. Most agree that an alteration in chromosome 22 (involved in tumor suppression) is the most common abnormality in meningiomas. People with the genetic disorder neurofibromatosis type 2 (NF2) are more likely to develop meningiomas. Studies show that patients who received radiation to the head for other conditions are at higher risk for developing meningiomas later in life. There is a correlation between hormones and meningiomas.

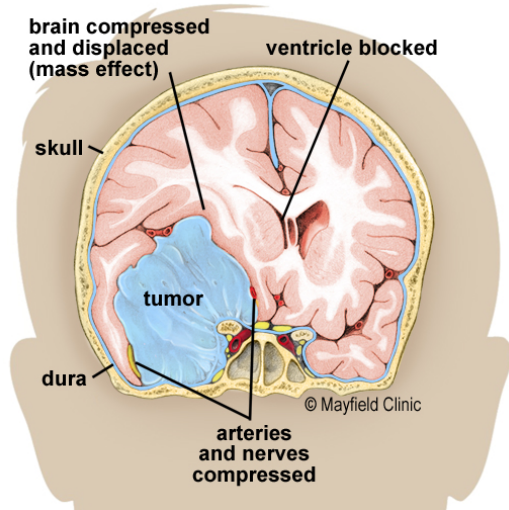
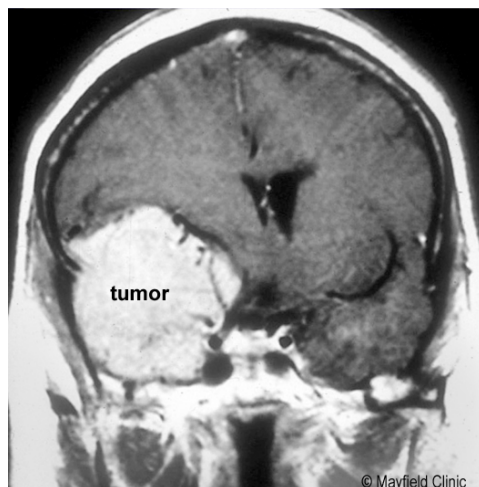
Who is affected?

Meningiomas account for 37% of all primary brain tumors and 12% of all spinal cord tumors. They are most often found in adults between the ages of 40 and 60 years and rarely occur in children. Most are benign (not cancer); less than 10% are malignant. While malignant meningiomas occur in both women and men, benign meningiomas are three times more common in women.

How is a diagnosis made?

The doctor will ask about your personal and family medical history and perform a physical exam. In addition to checking your general health, the doctor performs a neurological exam to check mental status and memory, cranial nerve function, muscle strength, coordination, reflexes, and response to pain. The doctor may order imaging scans such as CT or MRI to help determine the size, location, and type of tumor (Fig. 2). In some cases, angiograms of the blood vessels are necessary.

Figure 2. MRI scan of a sphenoid meningioma. The tumor has filled the area where the temporal lobe normally lies. The illustration shows that as the tumor grows, it compresses and displaces normal brain tissue and can encase the arteries and nerves.



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 ("watch and wait")

Because meningiomas grow slowly, 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 immediately report any change in their symptoms.

Medications

Medications may be used to relieve some of the side effects of meningiomas. These include steroids to reduce swelling and edema around the tumor, and anticonvulsants to prevent or control seizures.

Surgery

If a meningioma is causing symptoms or is growing in size, surgical removal is often recommended. A neurosurgeon performs a craniotomy to open the skull and remove the tumor (Fig. 3). During surgery, samples of tumor cells are taken (biopsy) and examined by a pathologist under a microscope to confirm the tumor type and grade. Although total removal can provide a cure for meningioma, it is not always possible. The tumor location determines how much can be safely removed. If some tumor is left attached to arteries or nerves, radiation can treat the remainder. The risks of surgery depend on where the tumor is located.

Technologies 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. These include:

- **Image-guided surgery (IGS)**, which works like the GPS in your car, helps the surgeon pinpoint the exact location of a tumor. Before surgery, a special MRI scan is performed, with white 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 his or her MRI or CT scans. Using a hand-held pointer, the surgeon can track the instrument's position in real time on the computer model of the patient's anatomy. IGS allows precise pinpointing of the tumor edges, guides the skin and bone openings, and tracks tumor removal (Fig. 4).
- **Brain mapping functional MRI (fMRI)** creates a series of images of the brain in action. The images capture blood oxygen levels in parts of the brain that are responsible for movement,

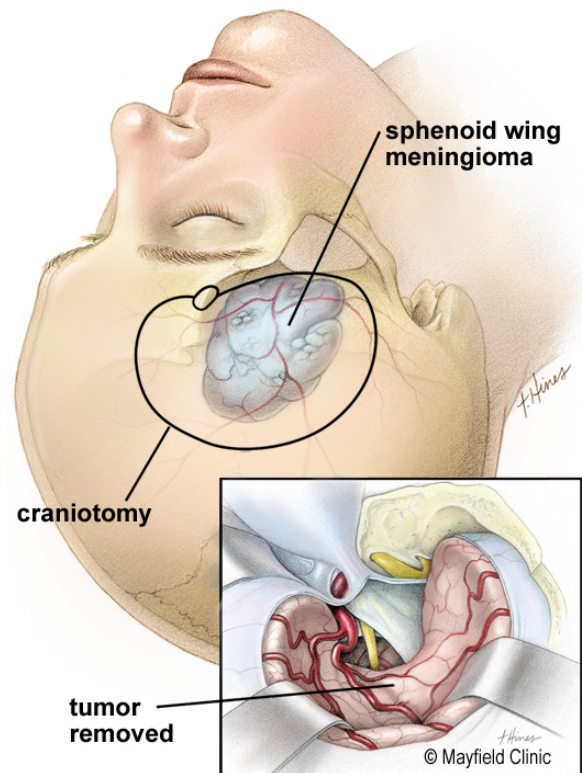


Figure 3. A craniotomy is cut in the skull to expose the tumor. After the tumor is removed, brain tissue can re-expand into the cavity. The bone flap is replaced and secured with plates and screws.

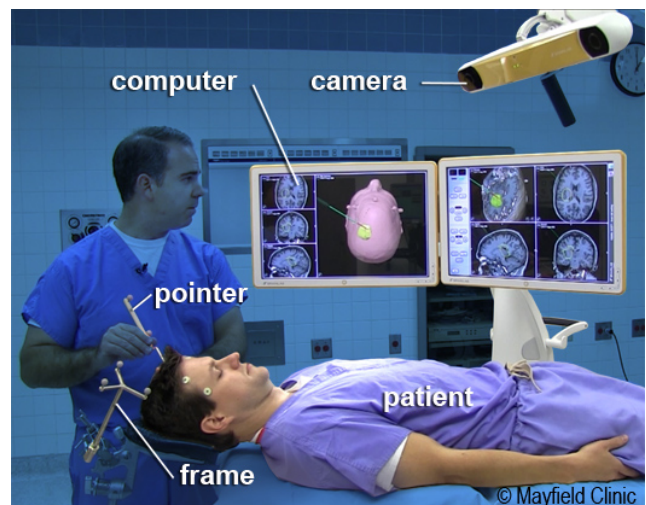


Figure 4. Image-guided surgery uses fiducials, frames, and infrared cameras to correlate the patient's anatomy to the 3D computer model. Similar to GPS in a car, it serves as a roadmap to your brain so the surgeon can locate the lesion. Instruments are detected by the cameras and displayed on the computer model. The surgeon watches instrument position on the computer screen in real-time as the tumor is removed.

perception, language, and thinking. By identifying and “mapping” these eloquent areas, surgeons can remove tumors to the greatest extent possible without harming areas that are critical to the patient’s quality of life.

- **Keyhole & endoscopic surgery** are minimally invasive techniques that enable surgeons to remove tumors at the skull base through small keyhole openings, such as through the nose, eyelid, or eyebrow. Tumors that formerly required a large craniotomy and brain retraction can be accessed with endoscopes through natural pathways such as the nose and sinuses.
- **Intraoperative 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. It improves the potential for total tumor removal and reduces the need for a second operation.

Radiation

Indications for radiation include: (1) incomplete tumor removal, (2) inoperable tumors, (3) malignant meningioma, and (4) patient choice. Radiation damages 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 (Fig 5). The benefits of radiation are not immediate but occur over time. Gradually, the tumor stops growing, shrinks, and in some cases disappears. Benign tumors, whose cells divide slowly, may take several months to a year to show an effect. Radiation therapy options for meningiomas include:

- **Stereotactic radiosurgery** delivers a high dose of radiation during a single session or 5 daily sessions. 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 completely immobilized with a stereotactic head frame or facemask. 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.
- **Fractionated radiotherapy** delivers lower doses of radiation over many visits. A facemask is used to accurately reposition the patient for each treatment session. Delivering a fraction of the total radiation dose allows normal cells time to repair themselves between treatments. It also reduces side effects. Patients return 5 days a week for 5 to 6 weeks to receive the complete radiation dose.

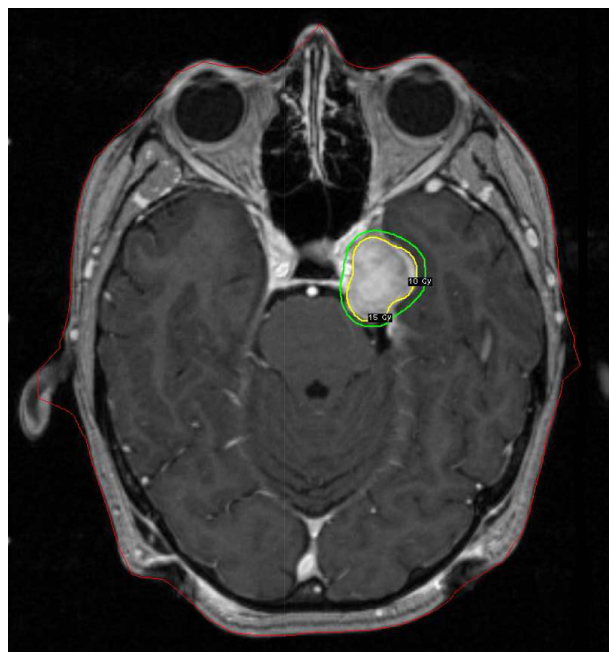


Figure 5. A radiosurgery treatment plan for a meningioma in the cavernous sinus. The rings represent the radiation dose delivered to the tumor.

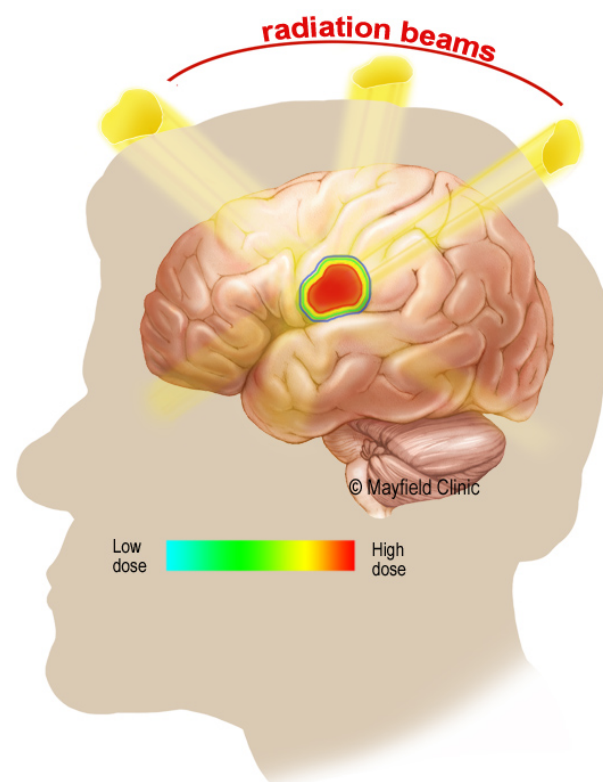


Figure 6. Radiation is delivered by a machine that rotates around the patient, aiming beams that are shaped to match the tumor and minimize exposure to normal brain tissue.

- **Proton beam therapy** delivers accelerated proton energy to the tumor at a specific depth. The radiation beam does not go beyond the tumor.

Chemotherapy and biologics

Chemotherapy is rarely used except in cases of malignant or recurrent meningiomas that don't respond to surgery and/or radiation. Medical therapies may include:

- Hydroxyurea - a chemotherapy drug taken as a pill that inhibits tumor cell DNA replication.
- Alpha interferon - a biologic therapy that inhibits tumor blood vessel growth. The drug is given by injection.
- Sandostatin - a hormone therapy that suppresses tumor growth. The drug is given by injection. A nuclear imaging study is first performed to determine if the tumor has somatostatin receptors.

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 to 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 (www.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 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 may recur after surgery or radiation. Regular follow-up MRI or CT scans (1 to 2 years) are an important part of long-term care for anyone diagnosed with a meningioma.

Sources & links

If you have more questions, please contact New England Neurological Associates at 978-687-2321.

Support

Support groups provide an opportunity for patients and their families to share experiences, receive support, and learn about advances in treatments and medications.

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: a very uncommon type of meningioma, highly vascular and usually fast growing.

mass effect: damage to the brain caused by the bulk of a tumor, 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.



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