

Meningiomas

basic level

Overview

A meningioma is a tumor that grows from the protective membranes (meninges) surrounding the brain and spinal cord. Most meningiomas are benign (not cancerous) and are usually slow growing. Because these are slow growing tumors, not all meningiomas need to be treated immediately. Meningiomas vary in their symptoms and appropriate treatment options depending on where they are located.

What is a meningioma?

Meninges are the three layers of tissue that cover and 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 gap cells that form the middle layer, the arachnoid mater, and are usually 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. Although less common, meningiomas can grow outward, causing the skull to thicken. Meningiomas grow very slowly, and it is oftentimes many years before they cause symptoms.

What are the symptoms?

Symptoms 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 meningioma are seizures, focal 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

World Health Organization Classification of Meningiomas

Grade 1

Meningiothelial
Fibrous (fibroblastic)
Transitional (mixed)
Psammomatous
Angiomatous
Microcystic
Secretory
Lymphoplasmacyte-rich
Metaplastic

Grade 2

Chordoid
Clear Cell

Grade 3

Papillary
Rhabdoid
Anaplastic

Of these subtypes, meningiothelial, fibrous and transitional are the most common. Most of these subtypes behave similarly, however anaplastics are the most aggressive.

difficult. Large parasagittal meningiomas may result in bilateral leg weakness.

- **Olfactory groove meningiomas:** grow along the olfactory nerves that run between the brain and the nose and allow you to smell. Often tumors growing here cause loss of smell. If they grow large enough, they can also compress the nerves to the eyes, causing visual symptoms. Similarly, meningiomas growing on the optic nerve can cause visual problems, including loss of patches 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 involve the blood sources of the brain (e.g. cavernous sinus, or carotid arteries), making them difficult or impossible 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 sharp pain in the face (trigeminal neuralgia) or spasms of the facial muscles. Foramen magnum meningiomas grow near the area where your spinal cord connects to your brain and can cause headaches, or other signs of brain stem compression like trouble walking.
- **Intraventricular meningiomas:** grow inside the fluid-filled ventricles deep inside the brain. They can block the flow of cerebrospinal fluid (CSF) causing pressure to build up, 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 an increasing loss of vision.
- **Spinal meningiomas:** grow intradural (within or enclosed within the dura mater), extramedullary (outside or unrelated to any medulla) tumors occurring predominantly in the thoracic spine. They can cause back pain, or pain in the limbs from compression of the nerves where they run into the spinal cord.

What are the causes?

Neuroscientists are not certain of the cause of meningioma, 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.

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 be obtained if the tumor, if one exists. 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 people with meningiomas. The treatment that is right for you will depend on your age, general health status, and the location and size of the tumor. Each treatment has benefits, risks and side effects that should be discussed and understood before a decision is made.

Observation (growth monitoring)

Because of the slow growth of meningiomas, elderly patients with this tumor may be monitored instead of undergoing surgical removal of the tumor. The doctor will monitor the growth of the tumor with annual MRI scans. Patients should promptly report any symptom change immediately.

Surgery

Surgery is the most common treatment for meningiomas. A neurosurgeon performs a craniotomy to open the skull and remove the tumor. Although "total" removal theoretically can provide a "cure" for benign meningiomas, total resection is not always possible. For meningiomas, the tumor location determines how much of the tumor can be safely removed. If the tumor cannot be completely removed, treatments can be combined, and the remainder of the tumor may be treated with radiotherapy.

Advances in laser microsurgical techniques as well as pioneering skull base surgical approaches, have now made previously inoperable tumors reachable. A skull base surgery team comprises experts in all sub-specialties of neuroscience: neurosurgeons, head-and-neck surgeons, otologists, neuro-radiologists, oculoplastic surgeons and reconstructive surgeons. Working together, they have charted new pathways into complex areas at the base of the brain where the vital nerves and blood vessels exit the skull.

Technology has improved the surgeon's ability to precisely locate the tumor, define its borders, and confirm its removal while in the operating room.

Image-guided surgery (IGS) uses stereotactic techniques to help the neurosurgeon pinpoint the exact location of a tumor within normal tissue. Stereotactic means to locate a structure by use of three-dimensional coordinates. During surgery, skin markers and infrared cameras correlate the "real patient" to a 3D computer model of the

patient created from their MRI or CT scans. Image-guided surgery allows very precise planning of the approach by pinpointing the tumor location and allowing precise skin and bone openings. This leads to shorter operating time, improved surgical visibility, and shorter length of stay.

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. 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 possibility of a second operation.

Radiation

Some tumors, because of their location near areas of the brain that control vital bodily functions like breathing or intellect, may be considered inoperable. Others grow back after surgical removal. In these instances, radiation therapy may be used, either by conventional means or through stereotactic radiosurgery.

Radiotherapy

Radiation therapy uses controlled high-energy rays to treat tumors. The radiation dose is delivered over many visits and uses a mask to precisely locate the tumor and accurately reposition the patient for each treatment session. With this therapy, radiation beams are manipulated by the computer to precisely conform to the shape of the tumor. Three-dimensional conformal radiation treatments significantly spare normal brain tissue and allow much higher doses to be delivered to the tumor. For patients with tumors that are usually successfully treated with radiation (e.g., pituitary tumors, meningiomas, craniopharyngiomas) three-dimensional conformal radiation should result in fewer early and late side effects.

Stereotactic Radiosurgery

Stereotactic radiosurgery uses a highly concentrated and precisely directed beam of radiation to treat a pituitary tumor during a single session. 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. Although it is called surgery, no incision is made. 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. The effects of treatment occur over a period of time so the tumor will not show immediate results. Gradually, the lesion will stop growing, shrink, and in some cases, completely disappear. Treatment

with stereotactic radiosurgery has proved to be very successful for patients with meningiomas. It is often effective in treating tumors that have not responded to surgery.

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. You can find information about current clinical investigations, including their eligibility requirements, protocol, and participating locations, on the web at: the National Institutes of Health (NIH) at clinicaltrials.gov, sponsors many trials; private industry and pharmaceutical companies also sponsor trials www.centerwatch.com

Current studies

Go to www.mayfieldclinic.com/ClinicalTrials.htm for information about clinical trials conducted by our doctors at local Cincinnati hospitals or call 1-800-325-7787 ext. 5260.

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 yield a higher rate of complication and are more difficult to completely resect. The patient's age and overall health prior to surgery may also affect the results. Meningiomas do sometimes reoccur after surgery or radiation. Regular follow-up MRI or CT scans (once every one to three years) are an important part of long-term health 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. Additional information is available on the web.

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) 1.800.886.2282

National Brain Tumor Foundation
(www.braintumor.org) at 1-800-934-CURE

Glossary

benign: not cancerous.

meninges: thin layer of tissue covering the brain and spinal cord.

syncytial meningioma: most common type of meningioma, with plump cells.

fibroblastic meningioma: tumor spindle-shaped cells.

transitional meningioma: tumor with both plump and spindle-shaped cells.

angioblastic meningioma: less common type of tumor of the meninges.

hemangiopericytoma: very uncommon type of meningioma, but usually fast growing.

malignant: cancerous.

meningioma: a tumor that grows from the meninges, the membrane that surrounds the brain and spinal cord.

neuroscientist: a physician or scientist dedicated to research, education, and clinical care of neurological diseases and disorders.

tumor: an abnormal growth of tissue resulting from uncontrolled multiplication of cells and serving no physiological function. A tumor can be benign or malignant.

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