

AMERICAN BRAIN TUMOR ASSOCIATION

# Meningioma



American  
Brain Tumor  
Association®

*Providing and pursuing answers®*

## **ABOUT THE AMERICAN BRAIN TUMOR ASSOCIATION**

Founded in 1973, the American Brain Tumor Association (ABTA) was the first national nonprofit organization dedicated solely to brain tumor research. The ABTA has since expanded our mission and now provides comprehensive resources to support the complex needs of brain tumor patients and caregivers, across all ages and tumor types, as well as the critical funding of research in the pursuit of breakthroughs in brain tumor diagnoses, treatments and care.

To learn more, visit **[abta.org](http://abta.org)**.

*We gratefully acknowledge Santosh Kesari, MD, PhD, FANA, FAAN chair of department of translational neuro-oncology and neurotherapeutics, and Marlon Saria, MSN, RN, AOCNS®, FAAN clinical nurse specialist, John Wayne Cancer Institute at Providence Saint John's Health Center, Santa Monica, CA; and Albert Lai, MD, PhD, assistant clinical professor, Adult Brain Tumors, UCLA Neuro-Oncology Program, for their review of this edition of this publication.*

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# Meningioma

## INTRODUCTION

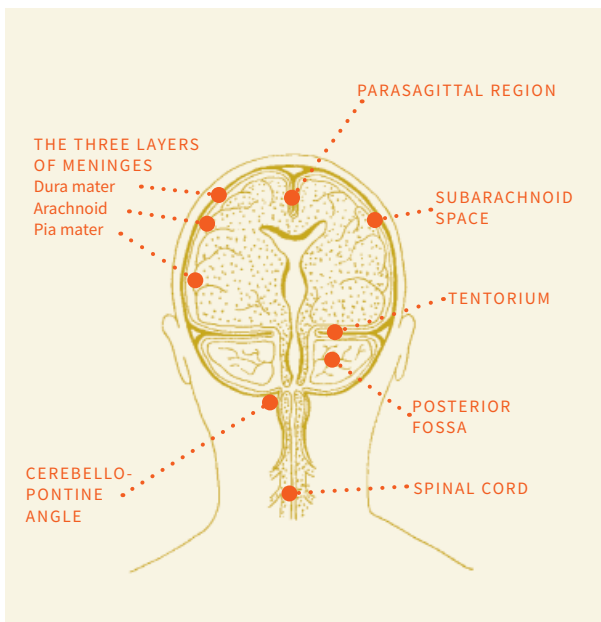
Although meningiomas are considered a type of primary brain tumor, they do not grow from brain tissue itself, but instead arise from the meninges, three thin layers of tissue covering the brain and spinal cord. These tumors most commonly grow inward causing pressure on the brain or spinal cord, but they may also grow outward toward the skull, causing it to thicken. Most meningiomas are benign, slow-growing tumors. Some contain cysts (sacs of fluid), calcifications (mineral deposits), or tightly packed bunches of blood vessels.

There are several systems used to name, or group, these tumors. One system names meningiomas by the type of cells in the tumor. Syncytial (or meningothelial) meningiomas are the most common and feature unusually plump cells. Fibroblastic meningiomas feature long, thin shaped cells. Transitional meningiomas contain both types of cells.

Another system uses the terms benign, atypical and malignant (or anaplastic) to describe the overall grade of meningiomas. In this system, benign meningiomas contain easily recognized, well-differentiated (resembling normal) cell types which

tend to grow slowly. Atypical tumors represent 10–15% of meningiomas. They contain proliferating cells that may be faster growing and more likely to grow back after treatment, even after seemingly complete resection (surgical removal). Therefore, these tumors must be followed carefully for early signs of recurrence. Malignant or “anaplastic” tumors are poorly differentiated forms that often recur rapidly. Although they are quite rare (1–3%), malignant meningiomas can be highly aggressive and difficult to treat.

Another common practice is to attach the location of the tumor to its name. For example, a parasagittal meningioma is located near the sagittal sinus, a major blood vessel at the top of the cerebral hemispheres. A sphenoid ridge meningioma is found along the ridge of bone behind the eyes and nose. Some meningiomas can cause problems despite their benign nature, because they are difficult to remove when they are located in functionally sensitive



Meninges

or hard to reach areas. Depending on the situation, stereotactic radiotherapy or radiosurgery may be particularly helpful in some of these cases.

## INCIDENCE

Meningiomas account for about one-third of all primary brain tumors. They are most likely to be diagnosed in adults older than 60 years of age, and the incidence appears to increase with age. Meningiomas are rarely found in children. They occur about twice as often in women as in men.

## CAUSE

Although the exact cause of meningioma is not known, it has been associated with radiation exposure. The most described genetic alteration is the loss of chromosome 22, normally involved in suppressing tumor growth. It is seen in approximately 50% of patients with mutations in the neurofibromatosis type 2 (NF-2) gene. Multiple meningiomas occur in 5–15% of patients, particularly those with NF-2. Meningiomas also frequently have extra copies of the platelet-derived growth factor (PDGFR) and epidermal growth factor receptors (EGFR), which may contribute to the growth of these tumors. Other genes linked to meningiomas include DAL1, SMO, AKT1, TRAF7, and mTORC1.

Obesity (high body mass index), a history of breast cancer, head trauma, or cell phone use may be risk factors for developing meningioma, but the evidence is inconclusive.

Some meningiomas have receptors that interact with the sex hormones such as progesterone, androgen and less commonly, estrogen. The expression of progesterone receptor is seen most often in benign meningiomas, both in men and women. The function of these receptors is not fully understood, and thus, it is often challenging for doctors to advise their female

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patients about the use of hormones if they have a history of a meningioma. Although the exact role of hormones in the growth of meningiomas has not been determined, researchers have observed that occasionally meningiomas may grow faster during pregnancy.

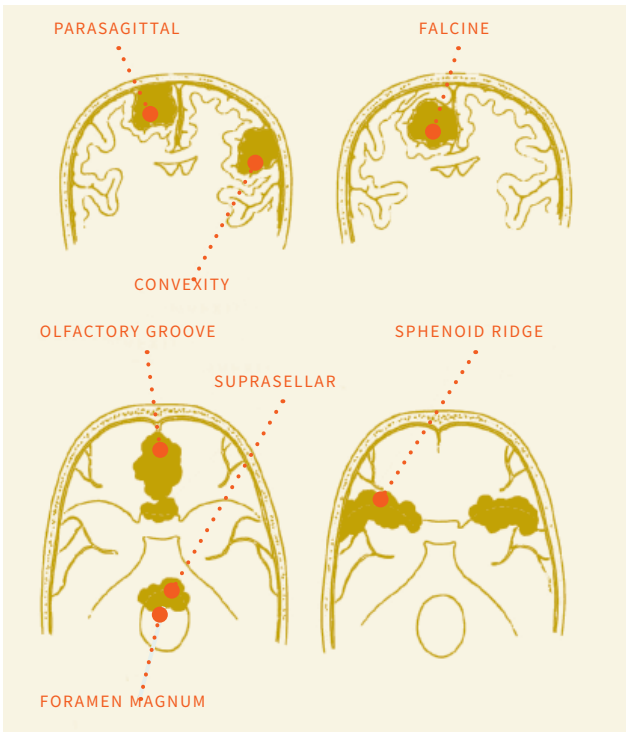
## **SYMPTOMS**

Meningiomas are usually slow growing and, may not cause any symptoms until it is big enough to compress adjacent structures. These tumors are most often found in the coverings of the parasagittal/falcine region (near the top of the brain) and the convexity (the outer curve) of the brain. Other common sites include the sphenoid ridge at the bottom of the brain, called the skull base.

As the tumor grows, it may interfere with the normal functions of the brain. The symptoms will depend on the location of the tumor. Headache and weakness in an arm or leg are the most common, although seizures, personality change or visual problems may also occur. Pain and loss of sensation or weakness in the arms or legs are the most common symptoms of spinal cord meningioma.

## **DIAGNOSIS**

Your doctor will begin with a neurological examination, followed by an MRI and/or a CT scan. MR angiography (a MRI scan of the blood vessels) or an arteriogram (a blood vessel X-ray) may be performed to help the doctors plan an embolization, a procedure to block the blood vessels in the tumor. An octreotide scan may be helpful in distinguishing meningiomas from other tumors. Used for tumors that have an extensive blood supply, embolization may help reduce bleeding during surgery.



### Common locations of meningiomas

*Burger, Scheithauer, and Vogel, Surgical Pathology of the Nervous System and Its Coverings. Fourth edition. Churchill Livingstone, New York, 2002. Diagram produced with permission.*

*If you have questions about using hormone replacement therapy (HRT) during menopause, please discuss your concerns with your doctors. Together, you can weigh the benefits and risks in light of your individual health situation.*

If you have a tumor, these tests help your doctor determine the location, size and probable type of tumor. However, only an examination of a sample of tumor tissue under a microscope confirms the exact diagnosis. Such a tissue sample can only be obtained through a surgical biopsy or excision.

## TREATMENT

### SURGERY

Surgery is the primary treatment for meningiomas located in an accessible area of the brain or spinal

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cord, although some tumors may be inoperable. Another factor that neurosurgeons consider is whether your vital organs (heart, lungs, kidneys and liver) are strong enough to withstand anesthesia and surgery.

The goals of surgery are to obtain tumor tissue for diagnosis and to remove as much tumor as possible. If the tumor cannot be removed, a biopsy to obtain a sample of tumor tissue may be performed.

A computer program that combines different MR images taken before surgery may be used to make a three dimensional, or stereotactic, map of your brain. This map helps the neurosurgeon plan the surgery to remove as much of the tumor as possible while avoiding parts of the brain that control vital functions.

During the operation, the surgeon may use stereotactic imaging and instrument guiding technologies to navigate through the brain. Occasionally, surgery is performed within a specialized MRI (intraoperative MRI), which allows the surgeon to view the tumor during the operation and determine the extent of tumor that is removed. High powered microscopes may be used to help the surgeon to better see the tumor. Ultrasonic aspirators are used to break up and suction out parts of the tumor.

In cases where the tumor cannot be removed completely, partial removal can help decrease symptoms. Radiation may then be used to treat the remaining tumor.

## **RADIATION**

Radiation therapy (external beam) may be used for inoperable tumors, tumors that are not completely removed in surgery, atypical and malignant tumors,



or recurrent tumors. There are different types of radiation, which use various doses and schedules. Most forms of radiation, however, are aimed at the tumor and a small area around the tumor.

Conventional external beam radiation is “standard” radiation given five days a week for five or six weeks. Stereotactic radiation aims converged beams of radiation at the tumor. Intensity modulated radiation therapy, also called IMRT, conforms radiation beams to the shape of the tumor. Additional information about these forms of radiation therapy is available from our office.

Stereotactic radiosurgery (SRS) utilizes numerous finely focused beams of radiation to accurately administer a single high-dose treatment to the tumor, while minimizing the effects to adjacent normal tissue. Therefore, despite the name, this is a noninvasive procedure and there is no real “surgery” involved. This may be particularly advantageous for patients that are poor surgical candidates, have tumors in high-risk regions of the brain, or have recurrences that are no longer amenable to conventional forms of surgical and radiation therapies. The disadvantages are that if no surgery or biopsy is done, no tissue is obtained for examination under the microscope; the technique may only inhibit further growth, stabilizing – rather than killing or removing – the tumor, and the technique is limited to relatively small tumors, usually those that are less than three centimeters in size.

For large tumors, or tumors located close to critical structures, conventional or stereotactic radiotherapy is often used instead. While stereotactic radiosurgery (SRS) involves the use of a single large dose of focused radiation, stereotactic radiotherapy, (SRT), involves the administration of smaller doses of focused radiation over a longer period of time (up to several

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weeks). This reduces the potential for swelling or injury to surrounding structures.

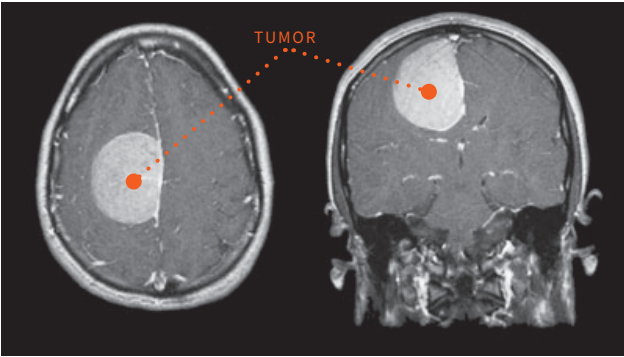
## **OTHER TREATMENTS**

Systemic therapy may be indicated for tumors that are not surgically accessible or for patients in whom further radiation is not possible. Some of these treatments are offered in organized research studies called clinical trials. Your doctor can determine if you are a candidate for treatment in one of these trials.

Several other treatment approaches have or are being explored:

- Hydroxyurea (used as a radiosensitizing drug in the treatment of other types of tumors)
- Progesterone receptor inhibitors (eg. mifepristone)
- Somatostatin analogs (hormones that prevent the release of growth hormones) (eg. octreotide)
- Targeted molecular agents (eg. everolimus)
- Epidermal growth factor receptor (EGFR) inhibitors (eg. erlotinb)
- Platelet-derived growth factor receptor (PDGFR) inhibitors (eg. imatinib)
- Vascular endothelial growth factors (VEGF) inhibitors (eg. bevacizumab)
- Immunotherapy or the use of biological agents to stimulate the immune system (eg. interferon alfa, nivolumab)

There are also several drugs used to treat the symptoms of a brain tumor. Steroids are used to decrease swelling, or edema, around the tumor. Anti-seizure drugs control seizures. Anti-nausea drugs prevent vomiting and help control nausea. Additional



MRI showing two views of a meningioma arising from the right side of the falx

*MRI scans courtesy of Patrick Wen, MD*

suggestions for managing side effects are offered on the ABTA website at [www.abta.org](http://www.abta.org).

### **WATCHFUL WAITING**

Depending on the location of the tumor, symptoms caused by the tumor and sometimes patient preference, some meningiomas may be carefully watched. Scans will be recommended during the time of observation, and it is very important to be sure those scans are done. If your doctor suggests a course of observation, remember that any new or changed symptoms should be promptly reported to your doctor.

### **RECURRENCE**

Most meningiomas are benign and treatable with surgery. However, brain tumors recur when all of the tumor cells cannot be removed with surgery or killed with other treatments. Over time, those cells multiply and result in tumor regrowth. Your doctor can talk with you about the chances of your tumor recurring. In general, at five years following surgery, about 5% of completely resected benign meningiomas, 30% of partially resected benign meningiomas and 40% of atypical meningiomas have recurred. Although rare,

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it is also possible that the meningioma may recur as a more aggressive, or higher grade, tumor.

Depending on your general health and the growth characteristics of the tumor, repeat surgery and possibly radiation therapy can be considered if the tumor recurs. Focused forms of radiation therapy, such as stereotactic radiotherapy or radiosurgery, may be repeated or used following a history of conventional radiation therapy. Treatments offered in clinical trials may also be used for recurrent tumors.

## RECOVERY

As with any brain tumor treatment, the length of recovery time varies. The age and general health of the patient, the location and size of the tumor, and the type of treatment all affect the recovery time. Prior to your surgery, ask your doctor what side effects you might expect.

Muscle coordination or speech problems may occur following surgery depending on the location of the tumor; they are often temporary. During this healing time, many brain tumor patients discover the benefits of rehabilitative services. The goal of rehabilitative medicine is to restore physical, vocational and psychological functions. Services may include physical, occupational and/or speech therapy to help reduce some of the symptoms that may accompany a tumor or treatment. Cognitive retraining – a memory training method – is used to teach another part of the brain to take over the tasks of the impaired portion. Visual aids may be required for those with tumors near the optic nerves. Support services that help patients and their families live with a brain tumor diagnosis are just as important. Call the ABTA's CareLine at 800-886-ABTA (2282) for help locating both rehabilitative

and support services in your area. For additional information, access to ABTA's online support community and to view webinars, please visit [www.abta.org](http://www.abta.org).

## PROGNOSIS

People diagnosed with a meningioma often have very specific questions regarding their future. They may want to know the risks involved in their surgery, the need for follow-up care or additional treatments, if or how the tumor might affect their life, and what the chances are for their tumor recurring. Although the medical term “prognosis” is usually associated with malignant tumors, a “predication of outcome” may be more applicable to a person with a meningioma.

We encourage you to ask your doctor these outcome questions. They can respond to your concerns based on your individual tumor. Your doctor can also explain your treatment plan, the benefits and risks of the treatment plan suggested for you, and what you can expect in the future.

## THE ABTA IS HERE FOR YOU

You don't have to go through this journey alone. The American Brain Tumor Association is here to help.

Visit us at [www.abta.org](http://www.abta.org) to find additional brochures, view free, educational webinars on demand, read about research and treatment updates, connect with a support community, join a local event and more.

We can help connect patients and caregivers with information and resources that can help support them in the brain tumor journey. Our team of caring professionals are available via email at [abtacares@abta.org](mailto:abtacares@abta.org) or via our toll-free CareLine at 800-886-ABTA (2282).

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## AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

### BROCHURES

Educational brochures are available on our website or can be requested in hard copy format for free by calling the ABTA. Most brochures are available in Spanish, with exceptions marked with an asterisk.

### GENERAL INFORMATION

About Brain Tumors: A Primer for Patients and Caregivers  
Brain Tumor Dictionary\*  
Brain Tumors Handbook for the Newly Diagnosed\*  
Caregiver Handbook\*

### TUMOR TYPES

Ependymoma  
Glioblastoma and Anaplastic Astrocytoma  
Medulloblastoma  
Meningioma  
Metastatic Brain Tumors  
Oligodendroglioma and Oligoastrocytoma  
Pituitary Tumors

### TREATMENT

Chemotherapy  
Clinical Trials  
Conventional Radiation Therapy  
Proton Therapy  
Stereotactic Radiosurgery\*  
Steroids  
Surgery

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## AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

### INFORMATION

ABTA WEBSITE | [ABTA.ORG](http://ABTA.ORG)

Offers more than 200 pages of information, programs, support services and resources, including: brain tumor treatment center and support group locators, caregiver resources, research updates and tumor type and treatment information across all ages and tumor types.

### EDUCATION & SUPPORT

- **ABTA Educational Meetings & Webinars**  
In-person and virtual educational meetings led by nationally-recognized medical professionals.
- **ABTA Peer-to-Peer Mentor Program**  
Connect with a trained patient or caregiver mentor to help navigate a brain tumor diagnosis.
- **ABTA Connections Community**  
An online support and discussion community of more than 25,000 members.
- **ABTA CareLine**  
For personalized information and resources, call 800-886-ABTA (2282) or email [abta cares@abta.org](mailto:abta cares@abta.org) to connect with a CareLine staff member.

### GET INVOLVED

- Join an ABTA fundraising event.
- Donate by visiting [abta.org/donate](http://abta.org/donate).

### CONTACT THE ABTA

CareLine: 800-886-ABTA (2282)

Email: [abta cares@abta.org](mailto:abta cares@abta.org)

Website: [abta.org](http://abta.org)

AMERICAN BRAIN TUMOR ASSOCIATION

8550 W. Bryn Mawr Avenue, Suite 550

Chicago, IL 60631

**For more information contact:**

Website: [abta.org](http://abta.org)

CareLine: 800-886-ABTA (2282)

Email: [abta cares@abta.org](mailto:abta cares@abta.org)



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