#### AMERICAN BRAIN TUMOR ASSOCIATION

# Meningioma







## 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 about the ABTA, visit abta.org.

We gratefully acknowledge Elizabeth B. Claus, MD, PhD, Professor and Director of Medical Research, Yale University School of Public Health, Attending Neurosurgeon and Director of Stereotactic Radiosurgery, Department of Neurosurgery, Brigham and Women's Hospital, and Gelareh Zadeh, MD, PhD, FRCS(C), Medical Director, Krembil Brain Institute, University Health Network, Professor, Department of Surgery, University of Toronto, Senior Scientist, MacFeeters-Hamilton Neuro-oncology Program, Princess Margaret, Chair, Wilkins Family in Brain Tumor Research for their review of this edition of this publication.

This publication is not intended as a substitute for professional medical advice and does not provide advice on treatments or conditions for individual patients. All health and treatment decisions must be made in consultation with your physician(s), utilizing your specific medical information. Inclusion in this publication is not a recommendation of any product, treatment, physician or hospital.

COPYRIGHT© 2019 ABTA

REPRODUCTION WITHOUT PRIOR WRITTEN PERMISSION IS PROHIBITED ABTA1223

## AMERICAN BRAIN TUMOR ASSOCIATION

## Meningioma

## INTRODUCTION

Meningiomas are the most common type of primary (arising from the brain and not from other parts of the body) brain tumor in adults. Most meningiomas contain easily recognized, well-differentiated (resembling normal) cells which tend to grow slowly (Grade I). Atypical (Grade II) meningioma tumors represent 10-15% of all meningiomas. They contain cells that may be faster-growing and more likely to grow back after treatment, even after seemingly complete surgical removal. Therefore, these tumors must be followed carefully for early signs of recurrence. Malignant or "anaplastic" (Grade III) meningioma tumors do not have clearly defined borders and often grow rapidly. Although they are quite rare (1-3%), malignant meningiomas can be highly aggressive and difficult to treat.

Although meningiomas are considered a type of primary brain tumor, they do not grow from brain tissue itself. They arise from the lining (meninges) of the brain and spinal cord. These tumors most commonly grow inward, causing pressure on the brain or spinal cord. They may also grow outward toward the skull, causing the bone to thicken and raising a bump on the skin surface. Some meningiomas contain cysts (sacs of fluid), calcifications (mineral deposits), or tightly packed bunches of blood vessels.

Meningioma tumors can be described based on their location. 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.



Meninges

## INCIDENCE

It is estimated that approximately 1% of the adult population has a meningioma tumor, but that many are asymptomatic and go undetected.<sup>1</sup> Meningiomas account for approximately 37% of all primary adult brain tumors. In the United States it is estimated that 31,990 people will be diagnosed with meningioma in 2019.2 Risk for developing a meningioma increases with age, and patients have a median age of 66 years at time of diagnosis.2 Females are 2-3 times more likely than males to be diagnosed with meningioma. Rates also vary by race with African Americans at greater risk than Caucasions and Hispanics.<sup>2</sup>

## ETIOLOGY

Researchers have examined a number of genetic and environmental risk factors for meningioma. A small number of meningiomas are associated with rare inherited genes (NF2, NF1, VHL, PTEN, PTCH, CREBBP). People with a family history of meningioma have up to four times the risk of being diagnosed with a meningioma compared to those without a family history.<sup>3</sup> Studies have found evidence for two genes associated with meningioma (located on chromosomes 10 and 11).<sup>4</sup>

If you have questions about your family history of meningioma or other tumors, please discuss your concerns with you health care providers.

Exposure to high-dose ionizing radiation is a primary environmental risk factor, increasing likelihood of meningioma by 6-10 fold. Other risk factors for meningioma include the following<sup>5</sup>:

Being female, most notably before menopause

- The presence of hormone receptors on meningiomas
- Having uterine fibroids or endometriosis
- Increased body mass index (BMI)

Some factors which may lead to greater risk include the following<sup>5</sup>:

- Having breast cancer
- Hormone replacement therapy (HRT) or oral contraceptive use

People with allergies and women who breast feed may be less likely to develop meningiomas. There are case reports where meningioma size can change based on the phase of the menstrual cycle and pregnancy. Estrogens may also affect the growth of meningioma-cell lines.<sup>5</sup>

No definitive associations have been identified between meningioma risk and cell phone use or head trauma.

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

## SYMPTOMS

Meningiomas are usually slow-growing and may not cause any symptoms until big enough to press into healthy brain tissue. 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 (skull base).

As a meningioma 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 changes 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. Other common symptoms may be a result of mass effect, which can cause changes in memory, personality, ability to multi-task, and other functions.

## TREATMENT Observation

When a meningioma is small or is not an immediate threat to a patient's health, health care providers might not recommend immediate treatment. They will continue to monitor the meningioma using imaging, generally an MRI. The frequency of such imaging may vary, but is typically done on a yearly basis.



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.

## Surgery

Surgery is the primary treatment for meningiomas that are large, causing symptoms, or are located in an accessible area of the brain or spinal cord. The goals of surgery are to obtain tumor tissue for diagnosis and to remove as much of the 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 MRI images taken before surgery may be used to make a three dimensional, or stereotactic, map of the brain. This map helps the neurosurgeon 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 determine how much of the tumor that was 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. Some non-malignant (benign) meningiomas are located in functionally sensitive or hard to reach areas and can be difficult to remove. Depending on the situation, radiation may be particularly helpful in some of these cases.

## Radiation

Radiation therapy may be used for tumors that are inoperable (hard to remove), not completely removed in surgery, atypical and malignant, or recurrent. Additionally, some patients prefer radiation rather than surgical treatment due to age and other medical conditions and considerations. There are different types of radiation which use various doses and schedules. Most forms of radiation 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. Intensity modulated radiation therapy, also called IMRT, conforms radiation beams to the shape of the tumor. Stereotactic radiosurgery (SRS) utilizes numerous finely focused beams of radiation to deliver a single high-dose treatment to the tumor. The accuracy of this form of radiation helps limit effects to nearby healthy tissue. This may be particularly effective for patients that are not able to undergo surgery, have tumors in high risk regions of the brain, or have recurrences that are no longer easily treated by conventional forms of surgical and radiation therapies. SRS is used to treat tumors that are less than three centimeters in size. Despite the name, SRS is a noninvasive procedure and there is no real "surgery" involved

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

Additional information about these forms of radiation therapy is available from both the ABTA website and in other ABTA educational pamphlets. The advantages and disadvantages of radiation and surgery can be discussed with a health care provider in order to make an informed decision together.

## Other Treatments

Systemic therapy, or drugs that work throughout the body, may be prescribed when tumors cannot be removed surgically or when further radiation is not possible. For meningioma, such treatments are generally offered in organized research studies called clinical trials. A number of recent studies6,7 have shown that some meningioma tumors may have changes or mutations that may respond to systemic therapy. A health care provider can determine if someone is a candidate for treatment in one of these trials.

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

## RECURRENCE

Recurrence is a regrowth of a tumor after treatment. The great majority of meningiomas are nonmalignant (benign) and do not recur. However, treated meningiomas can recur when all of the tumor cells cannot be removed with surgery or killed with other treatments. In addition, higher grade tumors are more likely to recur. Although rare, it is also possible for a meningioma to recur as a more aggressive, or higher grade, tumor. After treatment, the tumor will continue to be monitored with imaging so that any recurrence can be identified at an early stage.

Repeat surgery and radiation therapy can be considered if the tumor recurs. This decision is often based on the patient's general health and the growth characteristics of the tumor. Treatments offered through clinical trials



MRI showing two views of a meningioma arising from the right side of the falx MRI scans courtesy of Patrick Wen, MD.

may also be used for recurrent tumors.

## MANAGEMENT OF SYMPTOMS AND RECOVERY

There are several medications used to treat the symptoms of a meningioma. Steroids are used to decrease swelling, or edema, around the tumor. Anti-seizure drugs control seizures. Anti-nausea drugs prevent nausea and vomiting. In many instances, patients are able to be weaned off these medications over time. For patients who have had a seizure, it is important to remain on anti-seizure medicine and refrain from driving until/if a medical provider informs them that it is safe to resume driving.<sup>8</sup>

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 recovery time. Prior to surgery, medical providers can be asked what side effects might be expected. 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 and cognitive functions. Services may include physical, occupational and/or speech therapy. 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. For additional information, access to ABTA's online support community and to view educational webinars, please visit abta.org.

## PROGNOSIS

In general, the prognosis for patients with meningioma is good. The majority of patients are surviving at ten years from surgery.<sup>2</sup> People diagnosed with a meningioma often have 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.

Speaking with a health care provider can be helpful in answering questions and addressing concerns. It can also help patients better understand their treatment plan and what to expect in the future.

#### MENINGIOMA

## REFERENCES

- 1. https://www.nejm.org/doi/full/10.1056/nejmoa070972
- <sup>2</sup> https://academic.oup.com/neuro-oncology/article/20/suppl\_4/iv1/5090960
- 3. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3241000/pdf/ nihms-319212.pdf
- 4 https://www.ncbi.nlm.nih.gov/pubmed/29762745
- 5. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3756881/pdf/ nihms496632.pdf
- 6 https://www.ncbi.nlm.nih.gov/pubmed/23334667
- 7. https://www.ncbi.nlm.nih.gov/pubmed/23348505
- 8. https://www.epilepsy.com/driving-laws/2008871

## AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

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

## AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

## Information

#### ABTA WEBSITE | 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 Patient & Caregiver Mentor Support 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
   Ear percenalized inform

For personalized information and resources, call 800-886-ABTA (2282) or email info@abta.org to connect with a CareLine staff member.

## **Get Involved**

- Join an ABTA fundraising event.
- Donate by visiting abta.org/donate.

## **Contact The ABTA**

CareLine: 800-886-ABTA (2282) Email: info@abta.org Website: abta.org

#### AMERICAN BRAIN TUMOR ASSOCIATION

8550 W. Bryn Mawr Avenue, Suite 550 Chicago IL 60631

## For more information:

Website: abta.org CareLine: 800-886-ABTA (2282) Email: info@abta.org

