## AMERICAN BRAIN TUMOR ASSOCIATION

# Oligodendroglioma and Oligoastrocytoma







## 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. For over 40 years, the Chicago-based ABTA has been providing comprehensive resources that support the complex needs of brain tumor patients and caregivers, 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 www.abta.org.

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## Oligodendroglioma and Oligoastrocytoma

## INTRODUCTION

Oligodendroglioma and oligoastrocytoma belong to a group of brain tumors called gliomas. Gliomas are tumors that arise from the glial, or supportive cells of the brain. There are several different types of gliomas. This publication addresses two common gliomas: oligodendroglioma and oligoastrocytoma.





Astrocyte

Oligodendrocyte

- Oligodendrogliomas arise from oligodendrocytes fried egg-shaped cells within the brain. The role of normal oligodendrocytes is to form a covering layer for the nerve fibers in the brain.
- Astrocytomas are gliomas that arise from astrocytes star-shaped cells within the brain. The normal role of astrocytes is to store information and nutrients for the nerve cells in the brain.
- Oligoastrocytomas are mixed glioma tumors, containing both abnormal oligodendroglioma and astrocytoma cells.

Oligodendrogliomas are soft, greyish-pink tumors. They often contain solid mineral deposits which are mostly calcium – called calcifications. Oligodendrogliomas may also contain small areas of blood and/or cysts.

#### INCIDENCE

Primary brain tumors are tumors that arise in the brain and tend to stay in the brain. About 40% of primary brain tumors are gliomas. About 10% of those gliomas are oligodendrogliomas. Mixed gliomas, primarily oligoastrocytomas, account for about 5–10% of all gliomas. Biologic markers now help pathologists separate oligodendrogliomas from other types of gliomas.

Oligodendrogliomas are most common in adults and have a peak incidence in people ages 35–44. Anaplastic oligodendrogliomas tend to occur in slightly older adults, ages 45–74. Although these tumors are found in both men and women, they tend to occur more often in men.

Oligoastrocytomas are also most common in adults and have peak incidence in people ages 35–50. These tumors are found in both men and women.

Relatively few children are diagnosed with oligoastrocytomas; these tumors comprise only three percent of primary brain tumors in children ages 0-14. Oligodendrogliomas are more common in older children, comprising about five percent of brain tumors for those aged 15-19.

#### CAUSE

The exact cause of these tumors, as well as other types of brain tumors, is unknown. We do know that tumors develop when a normal cell, for some unknown reason, becomes abnormal. That abnormal cell may produce the wrong number of proteins or enzymes, or it may be missing genetic material containing the cell's basic

#### instructions.

When that abnormal cell reproduces itself, it creates two abnormal cells. Those two cells reproduce to create four cells, four cells create eight and so on. This reproduction continues, resulting in a "lump" of abnormal cells. That lump is called a tumor.

Scientists now know that the cells of some oligodendrogliomas contain abnormal genetic material. Deletions or absence of chromosomes 1p and 19q are frequently seen in oligodendroglioma and oligoastrocytoma tumors. Combined deletion of 1p and 19q is a predictor of prognosis and may predict response to treatment. In addition, anaplastic (malignant) tumors appear to have abnormalities on chromosomes 9 or 10, along with unusual amounts of growth factors and gene proteins.

Those substances are thought to regulate the growth of blood vessels around a tumor. The greater the blood supply, the more nutrients brought to the tumor.

Researchers also believe both oligodendrogliomas and astrocytomas originate from one mother cell whose "offspring" may follow two slightly different developmental pathways. This research helps explain the biologic relationship between these two types of gliomas. However, the initial steps that change these cells from normal brain cells to abnormal tumor cells are still uncertain. Tracing these pathways is of interest to many researchers as our understanding of the biology of brain tumors continues to advance.

If your doctor initially diagnoses you with a glioma and later tells you the tumor is an oligodendroglioma or an oligoastrocytoma, the diagnosis did not "change." These are both very specific types of glioma tumors.

#### **SYMPTOMS**

Some oligodendrogliomas grow slowly and may be present for years before diagnosis, while oligoastrocytomas can grow more aggressively. When the tumor makes its presence known, the most common symptoms are seizures, headaches and personality changes. Other symptoms vary by location and size of the tumor and can include weakness, numbness, or visual symptoms.

The frontal and temporal lobes are the most common locations for these tumors, although they can be found anywhere within the cerebral hemispheres of the brain. The frontal lobe controls the movement of your arms and legs, houses personality and behavior characteristics, controls language and maintains your ability to reason. Tumors of the frontal lobe may cause weakness on one side of the body, difficulty walking, or seizures. Difficulty remembering very recent occurrences, comments that do not match the conversation, or sudden changes in a person's usual behavior may be some of the symptoms of a tumor in the frontal lobe.

The temporal lobe of the brain generally controls memory, the ability to understand language and interpret sensations, comprehending what your eyes see and the significance of what you see and some emotions. Temporal lobe tumors generally cause few "visible" symptoms other than partial seizures and subtle language problems. Sometimes the seizures will start with unusual smells or tastes.

#### DIAGNOSIS

After a neurological examination, done in the office by your doctor, MRI and/or CT scans may be suggested. The calcifications sometimes present in an oligodendroglioma may be seen on a scan and suggest the diagnosis of oligodendroglioma. Sometimes both

an MRI and a CT scan will be ordered; MRI visualizes the softer tissues and blood vessels, while CT can better see structures such as the skull, calcifications within the tumor and blood.

Although scans may give your doctors an educated idea of the tumor type, only examination of a sample of tumor tissue by a pathologist confirms the exact diagnosis and leads to appropriate treatment. This is why surgery or a biopsy will be done to obtain tumor tissue.

Following the surgery, a pathologist will microscopically examine the removed tumor tissue. A pathology report will be sent back to your neurosurgeon. If the pathologist is part of the hospital system, the report will take about three to five days to generate. If tissue is also sent out of the hospital to another institution, the report may take a few weeks. The pathology report states the type of tumor and the "grade" of the tumor. An additional part of the report may contain the study to determine whether the tumor has deletions of 1p and 19q. This additional study may take longer to complete.

Grading tells you how close to normal, or how abnormal, the tumor cells looked when viewed under a microscope. The higher the grade, the more abnormal the cells and the more aggressive the tumor. Using the World Health Organization's grading systems of I through IV, cells appearing to be "almost normal" are assigned a grade I. The cells of a grade II tumor appear slightly abnormal. Grade III tumor cells are definitely abnormal in appearance. The cells of a grade IV tumor are very abnormal.

In this system, oligodendrogliomas and oligoastrocytomas are usually grade II or grade III tumors. Grade II tumors are considered low-grade tumors, which generally grow at a slower rate than grade III tumors. Grade II tumors may evolve over time into grade III tumors. Grade III tumors are anaplastic, or malignant tumors. Sometimes, anaplastic oglioastrocytomas contain grade IV astrocytoma cells, which are glioblastoma, or fastgrowing, aggressive tumor cells. If your tumor is an oligodendroglioma or anaplastic oligodendroglioma, additional testing may be done to determine if your tumor shows a loss of chromosomes 1p and 19q. This laboratory test looks for the presence – or absence – of bits of genetic material called chromosomes. Recent research found that oligodendrogliomas can be further subdivided based on the status of these two chromosomes. If this test is ordered for your tumor, it will take about two to three weeks for the results to be returned to your neurosurgeon.

As you learn more about brain tumors, you will often see the word "genetic." Genetic means "pertaining to the genes" – the tiny parcels that carry cell instructions. Genetic is not the same as hereditary (the ability to pass disease from one generation to another). Less than five percent of brain tumors are thought to be hereditary tumors. Those tend to be part of hereditary syndromes, such as neurofibromatosis or Li Fraumeni syndrome, which cause tumors in other parts of the body as well as the brain.

## TREATMENT

#### SURGERY

For both oligodendrogliomas and oligoastrocytomas, surgery remains the first step in treatment for most brain tumors located in an accessible area of the brain. An "accessible" tumor is one that can be removed without causing severe neurological damage. Numerous tools are available to assist the neurosurgeon in tumor removal. Computer-guided stereotactic navigational systems, along with sophisticated imaging equipment, can help define the exact tumor location. A special functional MRI may help to identify whether the tumor extends into vital areas of brain function.

Using that information, brain-mapping techniques may help outline vital parts of the brain to be avoided during surgery. Lasers and tiny microscopic instruments may be used to further remove tumor tissue. MRI scanners in or near the operating room can provide up-to-the moment images of the tumor site.

Even with the use of all of these tools, however, some tumors can be only partially removed because of their location. If the tumor is considered "inoperable," the neurosurgeon may be able to perform a biopsy to obtain a tissue sample and confirm the exact diagnosis.

#### **CHEMOTHERAPY**

If your tumor is an anaplastic tumor, or a mixed tumor such as an oligoastrocytoma, or if the tissue shows a loss (also called a "deletion") of chromosomes 1p or 19q, your doctor may talk with you about chemotherapy as part of your treatment plan. Temozolomide (Temodar) is an oral chemotherapy drug that may be suggested. PCV is an acronym for the combination of the drugs procarbazine, lomustine (CCNU), and vincristine. Bevacizumab (Avastin) is a targeted therapy that is sometimes used. It is an antibody against vascular endothelial growth factor (VEGF).

There are also several new drugs being tested for oligodendroglioma and oligoastrocytoma. The ABTA's clinical trial matching service, TrialConnect®, can help you find a clinical trial. Visit www.abtatrialconnect.org or call 877-769-4833.

Chemotherapy may also be used in infants and very young children to delay radiation therapy until the child is older. Clinical trials are underway to evaluate the most effective ways of treating these tumors in infants and children.

There are a few other drugs that may be suggested for someone with a brain tumor. It is not unusual for a tumor to cause swelling, or edema, around the tumor. Steroids are drugs used to decrease that edema. Antiepileptic drugs, also called AEDs, or anticonvulsant drugs, are used to control seizures. Antiemetic drugs prevent vomiting and help control nausea.

#### RADIATION

Radiation therapy may be suggested as treatment. The timing of radiation is determined by many factors. It may be recommended as a part of initial treatment particularly for anaplastic tumors, oligoastrocytomas or tumors that show no loss of chromosomes 1p or 19q. If the tumor is a low-grade oligodendroglioma, your doctor will determine if radiation therapy is recommended at this time. If your tumor is a highgrade tumor, radiation may be given at the time of diagnosis or deferred depending on other factors.

There are different types of radiation, using various doses and schedules. Most forms of radiation, however, are aimed at the tumor and a small area around the tumor. Radiation is usually given five days a week for five or six weeks. Intensity modulated radiation (IMRT) shapes radiation beams to the shape of the tumor. Stereotactic radiosurgery aims converged beams of radiation at small areas of the brain.

Just as in treating any disease, treatment for a brain tumor may have side effects. Ask your doctor to talk with you and your family about these potential effects. He or she can also help you balance the risks of treatment against the potential benefits.

#### RECURRENCE

Tumors recur or progress when all the tumor cells cannot be removed by surgery or killed by other treatments. Over time, those cells multiply and result in tumor regrowth. A tumor may recur as a highergrade tumor. It may contain a greater percentage of

anaplastic cells, more astrocytoma cells, or the tumor may spread into the spinal canal. Oligoastrocytoma growth generally depends on the the percentage of astrocytoma in the tumor, as astrocytomas tend to grow more rapidly than oligodendrogliomas. Because many oligodendrogliomas are generally slow-growing tumors, it may be years before regrowth occurs.

Treatments for a recurrent tumor may include additional surgery, radiation therapy if the tumor was not previously radiated, or a form of local radiation if the tumor was previously radiated. There are also many clinical trials open to those with a recurrent tumor. Researchers are exploring the role of new drugs and new drug combinations, which may also be used. Several of these compounds are currently being evaluated in clinical trials.

#### PROGNOSIS

Prognosis is the medical term for a prediction of life expectancy. Keep in mind that these predictions are estimates. When your doctor talks with you about prognosis, s/he will take into account your age, the location of the tumor, grade of the tumor cells, whether your tumor has deletions of 1p and 19q and the amount of tumor removed during surgery. Low-grade oligodendrogliomas tend to be slow-growing tumors. Anaplastic oligodendrogliomas are more aggressive tumors that grow more quickly. Oligoastrocytoma growth generally depends on the percentage of astrocytoma in the tumor, as astrocytomas tend to grow more rapidly than oligodendrogliomas. Scientists continue to study the impact of natural biologic differences amongst all of these tumors and the role of various treatment plans.

If you would like detailed information about prognosis, we encourage you to feel comfortable asking your doctor about your expected outcome. Make your question direct and to the point. Your physician can provide you with prognostic and biological information specific to your tumor. When considering a therapy, ask your doctor how the recommended treatment will affect your prognosis.

Other questions you may wish to ask could include:

- What are the expected benefits of this treatment?
- What are the risks?
- What quality of life can you expect during and after this treatment?
- If this is an investigational treatment, how many patients with your tumor type have received this treatment and what were their results?

### 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 to find additional brochures, read about research and treatment updates, connect with a support community, join a local event and more.

We can help you better understand support resources available to you. Our team of caring professionals are available via email at abtacares@abta.org or via our toll-free CareLine at 800-886-ABTA (2282).

## **NOTES/QUESTIONS**

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## AMERICAN BRAIN TUMOR ASSOCIATION PUBLICATIONS AND SERVICES

#### **CARE & SUPPORT**

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

#### PUBLICATIONS

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

Tumor Types:

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

Treatments:

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

All publications are available for download in Spanish. (exception is marked \*)

#### **CLINICAL TRIALS**

TrialConnect®: www.abtatrialconnect.org or 877-769-4833

More brain tumor resources and information are available at www.abta.org.

#### AMERICAN BRAIN TUMOR ASSOCIATION

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

#### For more information contact:

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

To find out how you can get more involved locally, contact volunteer@abta.org or call 800-886-1281.

