Oligodendrogliomas & Oligoastrocytomas

American Brain Tumor Association®

Providing and pursuing answers

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Founded in 1973, the American Brain Tumor Association (ABTA) was the first national nonprofit organization dedicated solely to brain tumor research. The ABTA provides 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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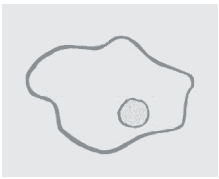
Oligodendrogliomas & Oligoastrocytomas

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 types of 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 were thought to be mixed glioma tumors, containing both abnormal oligodendroglioma and astrocytoma cells. In the 2016 World Health Organization classification, the diagnosis of Oligoastrocytoma is strongly discouraged. Nearly all tumors with features suggesting both cell types can be classified as either Astrocytoma or Oligodendroglioma using genetic testing. If they exist, true Oligoastrocytomas are very rare.

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 percent of primary brain tumors are gliomas, and about 10 percent of those gliomas are oligodendrogliomas. Oligoastrocytomas, historically accounted for about 3 percent of all gliomas. Biological 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.

In children, oligodendrogliomas are very rare. When they do occur, they tend to be in older children, comprising about 1.5 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, 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 oligodendrogliomas contain abnormal genetic material. The combined deletion of chromosomes 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.

The initial steps that change 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, the diagnosis did not "change." This is a very specific types of glioma tumor.

SYMPTOMS

Some oligodendrogliomas grow slowly and may be present for years before diagnosis. 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 current 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 your doctor conducts a neurological examination, he or she may suggest MRI or CT scans. The calcifications that are sometimes present in an oligodendroglioma may show up on a scan, which can help lead to the diagnosis. 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. MR perfusion scans may show an increase in blood flow to the tumor.

Although scans may give your doctors an educated idea of the tumor type, a biopsy (surgically removing a sample of the tumor tissue for a pathologist to examine) is the only way to confirm the diagnosis. Once the sample is examined, 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.

> The molecular tests for 1p/19q and IDH may take up to two weeks.

> 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.

A tumor grade 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 (WHO) 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 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.

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. This test and the test for IDH gene family mutation will take about two to three weeks for the results to be available

As you learn more about brain tumors, you will often see the word genetic, which 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 5 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 in the brain.

TREATMENT

Surgery

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 identify whether the tumor extends into vital areas of brain function.

> 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.

> "Awake" surgeries with testing of function during the operation are performed in certain circumstances.

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

Chemotherapy

Your doctor may talk with you about chemotherapy as part of your treatment plan. Oligodendrogliomas and anaplastic oligodendrogliomas tend to be responsive to chemotherapy. 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. Additionally, Bevacizumab (Avastin) is a targeted therapy that is sometimes used because it is an antibody against vascular endothelial growth factor (VEGF). VEGF is a signal protein produced by cells that stimulates the formation of blood vessels.

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. Steroids may be used to reduce edema (the buildup of fluid around the tumor) that is sometimes caused by the brain tumor or by other treatments. Antiepileptic drugs, also called AEDs, or anticonvulsant drugs, are used to control seizures. Antiemetic drugs prevent vomiting and help control nausea.

Radiation

Radiation is another treatment option that your doctor may suggest. If the tumor is a low-grade oligodendroglioma, your doctor will determine if radiation therapy will be beneficial as a part of your initial treatment. For high-grade tumors, radiation may be given at the time of diagnosis or deferred depending on other factors.

The type of radiation used, the dosage administered and the frequency of treatment is different for each patient. Most forms of radiation are aimed at the tumor, as well as a small area around the tumor. Intensity modulated radiation (IMRT) shapes radiation beams to the shape of the tumor. Stereotactic radiosurgery (SRS) aims converged beams of radiation at small areas of the brain. SRS is not usually used in the treatment of oligodendrogliomas, particularly early on, but it may be considered in certain circumstances. Radiation is usually given five days a week for five or six weeks.

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 weigh 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 remaining cells multiply and result in tumor regrowth. A tumor may recur as a higher-grade tumor; it may contain a greater percentage of anaplastic cells or the tumor may spread into the spinal canal. 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, he or she will take into account your age, the location of the tumor, grade of the tumor cells, whether your tumor has deletions of chromosomes 1p and 19q, and the amount of tumor removed during surgery. Low-grade oligodendrogliomas tend to be slow-growing tumors, while anaplastic oligodendrogliomas are more aggressive, fast-growing tumors. Tumor growth in tumors diagnosed as oligoastrocytoma may depend 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?

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PUBLICATIONS

About Brain Tumors: A Primer for Patients and Caregivers
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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.

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For more information contact:
8550 W. Bryn Mawr Avenue,
Suite 550, Chicago, IL 60631

800-886-ABTA (2282)

info@abta.org

www.abta.org