

Subependymal Giant Cell Tumor (SGCT) or Subependymal Giant Cell Astrocytoma (SEGA)

Subependymal giant cell astrocytoma (SEGA) occurs in about 1 out of every 10 individuals with tuberous sclerosis complex (TSC) and is more likely to develop during childhood and adolescence. SEGA is a type of brain tumor that occurs in individuals with TSC that is non-cancerous and is not malignant, but can still be very problematic. It is important to monitor for the occurrence of this brain tumor during childhood and until approximately the age of 21 years in individuals with TSC.

What is a SEGA or SGCT?

First, the word **subependymal** refers to the area below the ependyma (the membrane that lines the ventricles, or cerebral spinal fluid-filled spaces) of the brain. **Giant cell** refers to the very large, abnormal cells that are found with microscopic examination of the tumor. **Astrocytoma** refers to the type of tumor based on the most prevalent cell type. Historically, this tumor type has been classified as a slow-growing astrocytoma. However, a more appropriate term for this type of tumor is the **subependymal giant cell tumor**, or SGCT, since the cells in the tumor are of mixed types (not just astrocytes). You will see both terms used in the medical literature. We will use the term SGCT in this TSC Information Sheet.

The SGCT are found in the ventricles in the brain. These tumors are non-cancerous tumors, meaning they do not metastasize (spread to other parts of the brain or the body). However, the tumor can be problematic because it may grow so large as to block the flow of cerebral spinal fluid (CSF) within the brain, causing an increase in the pressure within the head and enlargement of the fluid-filled ventricles (a process known as hydrocephalus).

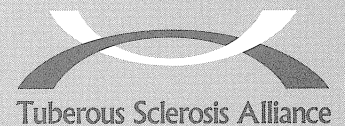
Typically, SGCT are very slow growing, but occasionally they may begin to grow more rapidly. It is not known what triggers the growth of a SGCT or why some individuals with TSC have a SGCT, whereas others do not. It is also not known why only some of the small nodules found on the surface of the ventricles, referred to as subependymal nodules (SENs), grow and become SGCT.

How do you diagnosis a SGCT?

Individuals with TSC should receive regular brain imaging, at least until adulthood and beyond that if medically necessary. If regular scanning is performed, then changes in the size of a SEN or the appearance of a SGCT can usually be noted at an early stage and appropriate follow-up and/or treatment initiated. SGCT can be observed on either MRI or CT scanning. Some individuals with TSC have been diagnosed with the disease only after they were diagnosed with a symptomatic SGCT.

What are the symptoms of a SGCT?

An individual with TSC who has a SGCT may initially have no signs or symptoms of having a brain tumor, only to develop symptoms when the tumor has grown large enough to block the flow of CSF and



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cause increased pressure. At this stage, some common signs are headaches, nausea, vomiting, increased frequency and/or severity of seizures, behavioral changes, and/or visual problems (blurred or double vision). The changes may be subtle, so it is important to be vigilant if an individual with TSC is known to have a SGCT. Signs of increased pressure could also be noted by a physician examining the eyes.

How do you treat SGCT?

Typically, any SGCT that is either causing symptoms and/or enlarging requires surgical removal. Although there are different philosophies on when to perform surgery to remove a SGCT, the NIH Consensus Conference report (Roach et al., 1999) recommends that any SGCT that is increasing in size or causing symptoms should be surgically removed. There are several different surgical approaches used by neurosurgeons to successfully remove this type of tumor, but radiation therapy should not be used to treat a SGCT (there are examples of radiation therapy given for a SGCT actually made the tumor grow and transform into a cancerous tumor).

A recent clinical study using a drug, Rapamycin, to treat SGCT shows great promise for the treatment of SGCT in individuals when surgical intervention is not possible. This study showed dramatic reduction in the size of the SGCT in those individuals who received the medication. Rapamycin has not been specifically approved for this use and is in clinical trials for other tumor growth in TSC (lung and kidney). Future clinical trials will help determine if this or similar drugs are useful to treatments for SGCT in TSC.

Will a SGCT regrow once it has been removed?

If a SGCT is completely removed, that SGCT will not regrow. However, there have been numerous cases in which another SGCT began to grow at a different location once the initial SGCT was removed. Also, there have been cases in which a SGCT was present in both ventricles, in other words, on both sides of the brain.

What is the danger in not removing a growing SGCT?

The danger in not removing a SGCT that is increasing in size is that it will eventually block the flow of CSF and cause a significant increase in pressure in the brain, leading to severe neurological and behavioral changes. The individual may have increased frequency and severity of seizures, drastic personality and behavioral changes, and even loss of vision. Severe headaches, nausea and vomiting may also be associated with a large SGCT. Before surgery to remove a SGCT was standard neurosurgical practice, many individuals with TSC died from the consequences of the tumor.

Who do I contact if I or my child have symptoms of a SGCT?

You should contact your physician as soon as possible. If you are not currently under the care of a neurologist, you should contact your primary care physician. Ultimately, you will probably be referred to a neurosurgeon.

How do I select a neurosurgeon?

Most neurosurgeons have specialties within the field of neurosurgery, so you should look for a neurosurgeon who has training in brain tumor surgery, and if possible, surgery for SGCT. When you meet with the neurosurgeon, you might ask the following questions:

- How many times have you performed surgery to remove a SGCT from other individuals with TSC?

- What was the age range of the individuals with TSC who you have operated on?
- What is the surgical approach you will use to remove the SGCT?
- What are the possible complications that might occur during and after the surgery?
- Once the surgery is complete, what will the recovery time be?
- Is there a chance that the same SGCT will regrow?
- How will this be monitored post-surgery?

References

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Roach ES, DiMario FJ Jr, Kandt RS, Northrup H (1999) Tuberous Sclerosis Complex Consensus Conference: Recommendations for diagnostic evaluation. *J Child Neurol* 14:401-407

Torres OA, Roach ES, Delgado MR, Sparagana SP, Sheffield E, Swift D, Bruce D (1998) Early diagnosis of subependymal giant cell astrocytoma in patients with tuberous sclerosis. *J Child Neurol* 13(4):173-7

Where can I go for more information?

National Brain Tumor Foundation
22 Battery Street , Suite 612, San Francisco, CA 94111-5520
www.braintumor.org
Telephone: 1-800-934-CURE

American Brain Tumor Association
2720 River Road, Des Plaines, IL 60018
<http://hope.abta.org/site/PageServer>
Telephone: 1-800-886-2282

Brain Tumor Society
124 Watertown Street, Suite 3H, Watertown, MA 02472
<http://www.tbts.org>
Telephone: 1-800-770-8287

The Childhood Brain Tumor Foundation
20312 Watkins Meadow Drive, Germantown, MD 20876
<http://www.childhoodbraintumor.org>
Telephone: 1-877-217-4166

Pediatric Brain Tumor Foundation of the United States
302 Ridgefield Court, Asheville, NC 28806
<http://www.pbtfus.org>
Telephone: 1-800-253-6530

National Cancer Institute, National Institutes of Health
Cancer Topics / "What you need to know about brain tumors"
<http://www.cancer.gov/cancertopics/wyntk/brain>

National Cancer Institute's Cancer Information Service
NCI Public Inquiries Office, 6116 Executive Boulevard, Room 3036A,
Bethesda, MD 20892-8322
<http://cis.nci.nih.gov/>
Telephone: 1-800-422-6237

National Institute of Neurological Disorders and Stroke, National Institutes of Health
Brain and Spinal Tumor Information Page
For health or medical questions and general information:
NIH Neurological Institute, PO Box 5801, Bethesda, MD 20824
<http://www.ninds.nih.gov/disorders/brainandspinaltumors/brainandspinaltumors.htm>
Voice: (800) 352-9424 or (301) 496-5751
TTY (for people using adaptive equipment): (301) 468-5981

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