Cavernous Angioma

Cerebral Cavernous Malformation

Information For Patients And Loved Ones

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**What is a Cavernous Angioma?**

Cavernous angiomas are clusters of abnormal blood vessels found in the brain, spinal cord, and, rarely, in other areas of the body. There are many names for this condition:

- cavernous angioma
- cavernous hemangioma
- cerebral cavernous malformation (CCM)
- cavernoma

A typical cavernous angioma looks somewhat like a raspberry, but it can range in size from microscopic to inches in diameter. It is made of multiple little bubbles (caverns) of various sizes, filled with blood and lined by a special layer of cells (endothelium). These cells are similar to those that line normal blood vessels, but the bubble-like structures of a cavernous angioma are leaky and lack the other layers found in a normal blood vessel wall. A cavernous angioma can cause seizures, stroke symptoms, hemorrhages, and headaches.

**Incidence**

Cavernous angiomas are estimated to occur in approximately 0.5-1% of the population, or in 1 in 100-200 people. Most people start having symptoms in their 20’s or 30’s. Cavernous angiomas can form later in life, so incidence rates and number of angiomas per person are higher among adults. Generally, more than 30% of those with cavernous angioma eventually will develop symptoms.

**Familial Cavernous Angioma**

For at least 20% of those with the illness, cavernous angioma is inheritable. This form of the illness is often associated with multiple cavernous angiomas. While familial cavernous angioma can happen in any family, it occurs at a higher rate among Mexican-American families. Each child of someone with the familial form has a 50% chance of inheriting the illness. Current research indicates that at least three genes can be implicated in the hereditary form. A mutation in any one of the three genes can lead to the illness.

**Sporadic Cavernous Angioma**

A solitary cavernous angioma may be present at birth or may develop later in life. If no other family members are affected, the condition is often not inheritable and is considered sporadic. This means that children of those with sporadic cavernous angioma may have no greater chance of having cavernous angioma than anyone else in the general public.
**Associated Venous Angioma**

Up to 40% of solitary cavernous angiomas may develop in the vicinity of another vascular anomaly called a venous angioma. The venous angioma, also known as venous malformation or developmental venous anomaly, usually does not create problems unless it is associated with a cavernous angioma. It may make surgery more difficult; the goal is not to disturb the venous angioma while removing the cavernous angioma.

**Symptoms**

A cavernous angioma may have no symptoms. The most common symptom is seizure, and a person who suffers from seizures is said to have epilepsy. There are many types of seizure. The type of seizure a person experiences depends, in part, on the location of the cavernous angioma. If a person has epilepsy and more than one cavernous angioma, it may be difficult to pinpoint which lesion is the cause of the seizures.

Cavernous angioma can cause neurological deficits such as weaknesses in arms or legs, vision problems, balance problems, or memory and attention problems. As with seizure, the type of deficit is associated with which part of the brain or spinal cord the cavernous angioma affects. Symptoms may come and go as the lesion changes in size with bleeding and reabsorption of blood.

Angiomas can bleed in a number of different ways:

- Angiomas can bleed slowly within the walls of the angioma and remain quite small. A small hemorrhage may not require surgery, but may be reabsorbed by the body. However, continued small hemorrhages in the same lesion often cause deterioration in function.

- Angiomas can bleed more profusely within the walls of the angioma. This can cause them to grow and put pressure on the surrounding brain tissue.

- Finally, angiomas may bleed through a weak spot in the angioma wall into the surrounding brain tissue. This is called an overt hemorrhage.

The risk of hemorrhage is dependent on the number of angiomas. The higher the number, the greater the chance of one or more hemorrhages occurring sometime over a lifetime. Unfortunately, cavernous angiomas that have bled are those that are the most likely to bleed again, particularly in the first two years after their initial bleed. It is also important to note that a hemorrhage in a cavernous angioma in the brain stem can be life-threatening.

Finally, those with cavernous angioma may experience headache. This seems to be true particularly when a lesion has oozed recently.
**Diagnosis and Treatment**

Cavernous angiomas are diagnosed most often when they become symptomatic. Although angiomas have been known since the 1930’s, they have not been reliably diagnosed until the advent of the MRI (magnetic resonance imaging) in the 1980’s. Previously, the illness may have been misdiagnosed as multiple sclerosis or as a seizure disorder with no known cause because the cavernous angiomas were not visible on angiogram and were not consistently visible on CAT scans. An MRI scan, with and without contrast and with gradient echo sequences, read by an experienced physician remains the best means of diagnosing this illness. The MRI scan may need to be repeated to assess change in the size of a cavernous angioma, recent bleeding, or the appearance of new lesions.

Most cavernous angiomas are observed for change in appearance, recent hemorrhage or clinical symptoms. Medications are available to treat seizures and headaches caused by cavernous angiomas. Surgery is advocated for cavernous angiomas with recent hemorrhage, those which are expanding in size, and in some cases, those which are causing seizures. Radiosurgery, by gamma knife, linear accelerator or new shaped beam techniques, is a controversial treatment that has been used on cavernous angiomas too dangerous to reach through traditional surgery.

**Surgery**

Cerebral cavernous angiomas are surgically removed (resected) using a craniotomy, or opening the skull. This is usually performed under general anesthesia, except in cases where mapping of the brain while awake is needed. Cavernous angiomas in the spine are removed using laminectomy, or unroofing, of the vertebrae. Surgery for cavernous angioma has been made safer using the operating microscope (microsurgery) and image guided surgical navigation (also known as computer-assisted or frameless stereotaxy) to reach the lesion with as little disruption to normal brain or spinal cord as possible.

Risks of any surgery, including cavernous angioma, include stroke, paralysis, coma or death, although these complications are rare with modern surgery performed by expert neurosurgeons. Surgery on cavernous angiomas in the brain stem and spinal cord is more risky, but these cavernous angiomas are more dangerous if left alone. Most patients leave the hospital within a few days and resume normal life within a few weeks of surgery. However, people with neurological deficits may require a prolonged period of rehabilitation.
Cavernous Angioma Statistics

1 in 100-200 people have at least one cavernous angioma.
At least 30% of those with a cavernous angioma eventually will develop symptoms.
At least 20% of those with cavernous angioma have the familial form of the illness.
Up to 40% of solitary cavernous angiomas may have an associated venous angioma.

Age at first diagnosis:
- Under 20: 25-30%
- Age 20-40: 60%
- Over 40: 10-15%

Primary symptom:
- Seizure – 30%
- Neurological deficit – 25%
- Hemorrhage – 15%
- Headache – 5%

Odds of your child having cavernous angioma:
- If you have sporadic cavernous angioma, your child may have a 1 in 200 chance (0.5%).
- If you have familial cavernous angiomas, your child may have a 1 in 2 chance (50%).

What We Don’t Know about Cavernous Angioma

While researchers continue to discover new facts about cavernous angioma every day, many important research questions remain.

- Genetic researchers and a growing number of other researchers are working to determine the cause of the illness and the mechanisms by which the defective blood vessels are formed.
- We don’t know most of the factors that lead to angioma bleeding and re-bleeding. Because we don’t know what causes a particular cavernous angioma to bleed, we don’t know how to reduce the risk.
- We don’t know how to remove a cavernous angioma without brain surgery. Less intrusive removal methods may allow for treatment of more lesions before they become problematic.
How Angioma Alliance Can Help

Angioma Alliance is a non-profit charitable organization created by people affected by the illness. Our website www.angiomaalliance.org provides information about the illness, networking opportunities, updates on research, and information about participating in ongoing research studies. Our email address is info@angiomaalliance.org. We distribute patient education materials to neurosurgeons, neurologists, and genetics professionals nationally.

Dr. Issam Awad, Professor of Neurosurgery, past chair of the Cerebrovascular section of the American Association of Neurological Surgeons, 51st President of the Congress of Neurological Surgeons, and NIH funded cavernous angioma researcher, serves as the chair of our scientific advisory board.

Angioma Alliance seeks to improve the quality of life for individuals with cavernous angiomas and their families through education, support, and promotion of research.

Our goals are:

• To ensure that those with cavernous angioma have access to materials written in layperson’s terms providing information about the illness.
• To provide opportunities for persons affected by the illness to learn of each other and provide support to each other.
• To promote research by offering additional information on a website, including summaries of the latest research results and news of research study enrollment opportunities.
• To help the public to become aware of cavernous angioma so that those affected by the illness will receive more understanding and support.

This brochure is not a substitute for obtaining competent medical advice. It is for informational purposes only.

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