American Association of Neurological Surgeons

Cavernous Malformations

Patient Information

This resource, developed by neurosurgeons, provides patients and their families trustworthy information on neurosurgical conditions and treatments.

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Cavernous malformations are clusters of abnormal, tiny blood vessels and larger, stretched-out, thin-walled blood vessels filled with blood and located in the brain. These blood vessel malformations can also occur in the spinal cord, the covering of the brain (dura) or the nerves of the skull. Cavernous malformations range in size from less than one-quarter inch to 3-4 inches. Cavernous malformations are also referred to as cavernomas, cavernous angiomas, cavernous hemangiomas or intracranial vascular malformations. The term angioma implies a propensity for growth that is associated primarily with the familial form of the illness.

Incidence and Prevalence

- The incidence of cavernous malformation is estimated at one in 100-200 people.
- Cavernous malformations account for an estimated 8-15 percent of all intracranial and spinal vascular malformations.
- A minimum of 30 percent of people with cavernous malformations will develop symptoms, most in their 20s or 30s.
- Cavernous malformations hemorrhage at an estimated rate of approximately 0.7-1.7 percent per lesion each year.
- At least 20 percent of those with cavernous malformation have the familial form of the illness (referred to as cavernous angioma). The familial form is associated with Hispanic heritage, multiplicity of lesions and a demonstrated propensity for growth of lesions. The latter two features are less characteristic of the sporadic form of the illness.
- If a parent has familial cavernous angioma, his or her child may have a 50 percent chance of developing this condition.
- As high as 40 percent of solitary cavernous malformations may have an associated venous malformation.
- Diagnosis by age: age 20 and younger - 25-30 percent; age 20-40 - 60 percent; age 40 and older - 10-15 percent.

Symptoms

A person with a cavernous malformation may experience no symptoms. When symptoms occur, they often are related to the location of the malformation and the strength of the malformation walls. The type of neurological deficit is associated with the area of the brain or spinal cord that the cavernous malformation affects. Symptoms may appear and subside as the cavernous malformation changes in size due to bleeding and reabsorption of blood. Any of the following symptoms may occur:

- Seizures
- Weakness in arms or legs
- Vision problems
- Balance problems
- Memory and attention problems
- Headaches

Diagnosis

Cavernous malformations are part of a group of lesions known as "angiographically occult vascular malformations." This means that they are not visible on an angiogram. Angiograms cannot visualize cavernous malformations because blood flows through these types of lesions slowly. The relatively milder symptoms from the lesion, even when ruptured, are presumed to be related to this state of relatively low blood flow.

Magnetic resonance imaging (MRI), with and without contrast and with gradient echo sequences remains the best means of diagnosing cavernous malformations. MRI scans may need to be repeated to analyze a change in the size of a cavernous malformation, recent bleeding or the appearance of new lesions.

Treatment

Asymptomatic Lesions

In general, lesions that are incidentally discovered should be followed with MRI scans annually for two years, then every five years thereafter. An MRI should be performed sooner if there is any clinical evidence of hemorrhage or new symptoms appear. Some patients may be prescribed anti-convulsant medications. This is an example of a subtype of AVM that may be monitored radiographically, specifically because the consequences of hemorrhage from these lesions are much less dire than those from classic AVMs or aneurysms.

Symptomatic lesions

Surgery should be considered for seizure control if: 1.) Seizures cannot be controlled through medication management; 2.) The cavernous malformation is in a low risk, easily accessible area of the brain; and 3.) It has been determined that the lesion is causing the seizures. If seizures are controlled through medication management, there may not be any compelling reason to perform surgery. In general, although seizures may indeed be cured by successful microsurgical removal, the primary goal of surgery is to prevent future bleeding and problems such as seizures that may be associated with it. Seizure control by itself is not justification for performing microsurgery on a cavernous malformation.

Surgery may be indicated in patients who have experienced one neurologically symptomatic hemorrhage from a lesion in a low risk, easily accessible area. For lesions in eloquent areas of the brain, surgical removal should be contemplated in the context of surgical risk to nearby brain tissue, balancing this risk against the risk of bleeding to that same tissue in the event of a second hemorrhage.

Surgical removal should be considered in patients with progressive neurological deficits, but such neurological deficits can worsen after surgery. Although brain or spine surgery may carry substantial risk, so may hemorrhage into nervous tissue. The risk of surgery must be balanced against the risk of no surgery, on an individualized, case-by-case basis.

Outcome

Most patients can leave the hospital a few days following surgery and resume normal life within a few weeks of surgery. Many patients can be cured without neurological deficit. Many patients with neurological deficits are able to regain their neurological baseline (condition at time of surgery) with therapy and may even show further improvements. Patients with neurological deficits may require a prolonged period of rehabilitation. The rebleeding rate of cavernous malformations is extremely variable. Some patients with malformations and one bleeding episode never experience a recurrence of their symptoms, while others experience frequent rebleeding.

Cavernous Malformation Resources

- Angioma Alliance

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