John Laidlaw Neurosurgery

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Cavernous angiomas

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SPECIAL POINTS OF INTEREST:

- **Cavernomas are clusters of thin walled vessels filled with low pressure blood.**
- **Familial cavernomas are thought to be inherited and are often multiple.**
- **Cavernomas usually ooze blood into the surrounding brain, but large bleeds are uncommon.**

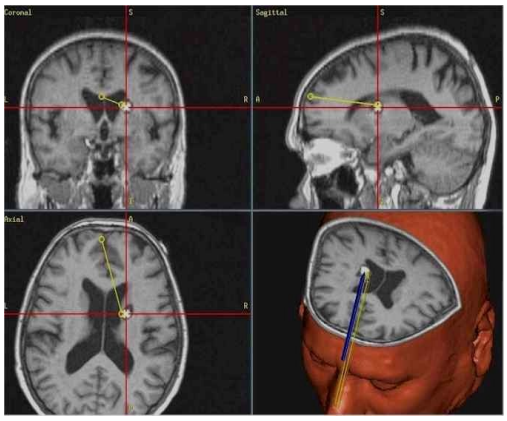
INSIDE THIS ISSUE:

What is a cavernoma?

A cavernous haemangioma (cavernoma) of the brain is a discrete lobulated mass of abnormal thin-walled blood vessels in the brain substance. Cavernomas can range in size from a millimetre to many centimetres. The collection of thin walled distended vessels are filled with slow-flowing blood, and often has the appearance of a mulberry.

Although there isn't any normal brain tissue between the vessels in a cavernoma, there is often evidence of old small bleeds causing staining (hemosiderin staining) and scarring (gliosis) in the surrounding brain.

Cavernomas are often suspected on CT scan, but are best demonstrated on MRI scan



which typically shows the dilated loops of vessels and the hemosiderin staining around the lesion.

Typically cavernomas don't have high-pressure arteries feeding them, but the blood enters them at low pressure from dilated capillaries and other small vessels. Therefore, because of slow blood flow, they usually cannot be seen on blood-vessel x-rays (angiograms).

There is often a large vein (venous angioma) near the cavernoma. Although this venous angioma is larger than normal veins, it serves the normal function of draining blood from the brain and in itself usually poses no risk to the patient (and requires no

Are cavernomas inherited?

Cavernomas may be congenital (i.e. you are born with them). However, there certainly have been some which have been demonstrated to first appear in later life, and some which have been shown to enlarge. They usually affect males and females equally.

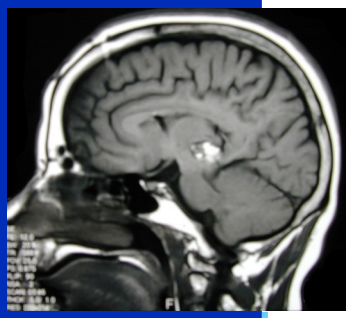
People with cavernomas are thought to be of 2 types: sporadic cases (with no known affected relatives) and familial cases (which seem to be inherited in an autosomal dominant pattern affecting half of all children).

Sporadic cases account for about half of all cavernoma patients, and most (85-90%) sporadic patients have only one cavernoma.

However, familial cases very commonly (up to 90% of familial cases) have multiple cavernomas in the person's brain. Familial cases may be associated with a defect on chromosome 7, and are more common in Hispanic people. Certain genes have been identified as related to this condition.

Relatives of people with multiple cavernomas are often offered MRI screening procedures, although it is best for these people to discuss the issues with cerebrovascular neurosurgeon first.

Cavernoma symptoms



This lady's midbrain cavernoma has been removed with excellent recovery.

Although cavernomas may be found coincidentally (e.g. when scanning someone who has a head injury), they usually become apparent when they cause symptoms. These symptoms include headache, epilepsy seizures, or neurological deficit (eg weakness, speech problems, numbness, visual problems, etc).

The headache and neurological deficits associated with cavernomas may be a result of the cavernoma bleeding in the brain (haemorrhage).

Although major haemorrhage is uncommon, most cavernomas ooze a small amount of blood in and around the vessels, staining and scarring the surrounding brain.

Surprisingly, the risk of haemorrhage and neurological deficit is the thought to be the same for small and larger cavernomas.

Fortunately, because of the low pressure of the blood in the cavernoma, most bleeds are relatively small and usually not fatal. The risk of a cavernoma having a significant bleed is thought to be quite small, probably less than 1% per year. However, cavernomas that have bled before and those in the brainstem have a higher risk of significant bleeding (some reports measure this risk in excess of 10% per year).

Some reports also suggest familial cavernomas may have a slightly higher risk of bleeding (around 1% per year) than sporadic cases.

What tests are required?

"Cavernomas often present with headaches, epilepsy seizures, or neurological deficits."

Most people with a cavernoma have the diagnosis made on MRI scan. The MRI scanner is a large machine, and the patient lies on a moving bed which slides into a tunnel while the scan is in progress. Often a small injection of dye (contrast material) is injected into the patient's hand during the scan.

Unlike CT scans, the MRI does not use radiation, but used a strong magnetic field and ultrasound to generate the images of the brain. Most people are surprised to find the scanning process very noisy, as the ultrasound generator causes a loud drumming noise.

Because of the magnetic field, people with pacemakers, surgical clips, or metal implants in their body often cannot safely have an MRI. However, for most people this is considered to be extremely safe.

Blood vessel x-rays (angiograms) are sometimes required just to run out other lesions (such as an AVM, which has high flow vessels). An angiogram requires an arterial injection into the groin and floating a fine tube into the arteries in the neck. Angiography is an invasive procedure and does have a small risk of serious complications including stroke, arterial injury and contrast allergies. The cavernoma itself does not show on the angiogram

Should a cavernoma be treated?

MRI scanner

The decision whether to treat a cavernoma or not is difficult, as multiple factors need to be considered. These include the patient's age and health, the site of the cavernoma, whether it has previously bled, and whether the patient has a significant seizure disorder.

Many cavernomas which are causing minimal symptoms, and many of those in deep areas of the brain which would make surgery vmore risky, are observed and not removed.

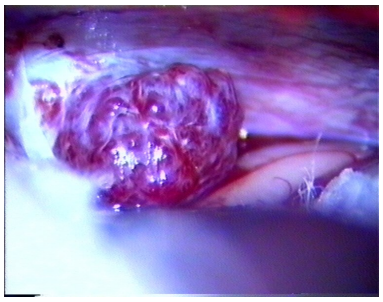
However, symptomatic cavernomas, particularly those that have had bleeds or cause major seizure problems, are often removed surgically.

Surgery for cavernomas

Microsurgical resection of a cavernoma involves operating on the brain, typically under general anaesthetic. Usually a computerised system (frameless stereotaxy) is used to precisely localise the cavernoma in the brain. Depending on the site of the cavernoma, a small window (craniotomy) is made in the overlying skull bone, and the brain exposed. Using the operating microscope a small cut is made in the brain over the cavernoma, and the cavernoma carefully removed. The tiny blood vessels are all coagulated to prevent bleeding. The bone is then replaced and the wound closed. This is very delicate surgery and should be performed by a experienced cerebrovascular neurosurgeon.

The risks of surgery are largely dependant on the site of the cavernoma, but include stroke, seizures, other neurological deficits, infection, and anaesthetic complications. The risks for each operation need to be fully discussed with the neurosurgeon prior to the surgery.

Despite these known risks, I have found that most cavernomas, even many of those in very sensitive areas of the brain (including some in the brainstem) can usually be removed with good results. Most patients who have uncomplicated surgery are up and walking in the evening of surgery and home 3-4 days later. Patients often need anticonvulsant (anti-epilepsy) medications for some time after surgery and shouldn't drive for 3 months. Of course patients with neurological deficits or other complications may require rehabilitation.



Cavernoma as seen under the operating microscope.

What if I chose no treatment?

" You need to know where in the brain your cavernoma is, and what the function of that part of the brain is."

It is very important that you have a full and frank discussion with an expert cerebrovascular neurosurgeon about your cavernoma. You need to understand which part of the brain it lies in, and what the function of this area is (e.g. does it control speech or movement in one limb, etc?). Using this knowledge, and having discussed the specific risks of surgical removal of your cavernoma with the surgeon, you can then decide whether to have surgery or not. It is important to understand that the risk of bleeding in the short-term is usually very small, so you should get plenty of information and not feel rushed in your decision.

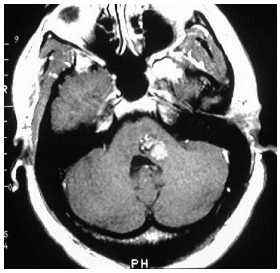
People who have had seizures from their cavernoma usually need anticonvulsant medication. They also need to take specific seizure precautions. Similarly, some people who have neurological deficits might not be able to drive, etc. However, I usually advise well people with a cavernoma to lead a normal life, and I don't usually place any restrictions on their activity.

I usually advise a clinical follow-up and often MRI scan every year or so for people with known untreated cavernomas. Of course, if they develop headaches, neurological deficits or seizures they must seek medical attention without delay.

Other treatments for cavernoma.

Surgical excision is the only proven treatment for cavernomas.

However, some clinics have been using radiosurgery (delivery of a large dose of focused radiation) on cavernomas. There have been mixed reports as to whether this radiosurgery is beneficial for cavernomas. Although radiosurgery does not need an open operation it does have risks of radiation injury to the brain. The other major problem is that we cannot tell if the radiosurgery has worked or not, and there is some argument about whether it actually reduces the risk of haemorrhage. Therefore, at this stage I would not advise radiosurgery for a cavernoma, and advise either microsurgery or observation.



This person is well, and we thought it safest to observe the cavernoma rather than operate.

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Associate Professor John Laidlaw is an experienced neurosurgeon in Australia and USA. His areas of particular interest and expertise include cerebrovascular surgery, skull-base surgery, brain tumour surgery, and surgery on the cervical spine and craniocervical junction.

Surgery, if appropriate, is usually performed using the most minimally invasive microsurgical techniques appropriate to the case. All surgical procedures are performed at The Royal Melbourne and Melbourne Private Hospitals.

All consultations (insured or uninsured patients, Parkville or Geelong rooms, or Royal Melbourne Hospital clinics) should be arranged through the Parkville rooms.

Cerebrovascular Neurosurgery

Cerebrovascular neurosurgery involves management of specific blood vessel abnormalities in the brain and spinal cord. These conditions include aneurysms, arteriovenous malformations (AVM), cavernous angiomas, Moyamoya disease, stroke, and vascular compression syndromes such as trigeminal neuralgia and hemifacial spasm.

Although all Australian-trained neurosurgeons have some training in this area, the recent introduction of special x-ray procedures (endovascular treatment and radiosurgery) and the medical management of stroke means that less cerebrovascular neurosurgery is done now than was done previously.

Therefore, as cerebrovascular neurosurgery is technically one of the most demanding areas of neurosurgery and it is becoming more difficult to develop and maintain these microsurgical skills, many neurosurgeons no longer treat cerebrovascular conditions, and refer such cases to expert cerebrovascular neurosurgeons. These cerebrovascular neurosurgeons have had specialized post-graduate training in this area. They continue to maintain and further develop their skills by performing cerebrovascular microsurgery on a regular basis

Cerebrovascular neurosurgeons need to work in institutions which have expert interventional neuroradiologists with specialized neuroradiology facilities. They also need to have close contact with radiosurgery specialists. The reasons for this are that many cerebrovascular conditions can be treated by either surgery, endovascular techniques or radiosurgery, and the cerebrovascular neurosurgeon needs to consult with these other specialists to determine the best treatment for each particular patient's condition.