

Treating Brainstem Cavernous Malformations

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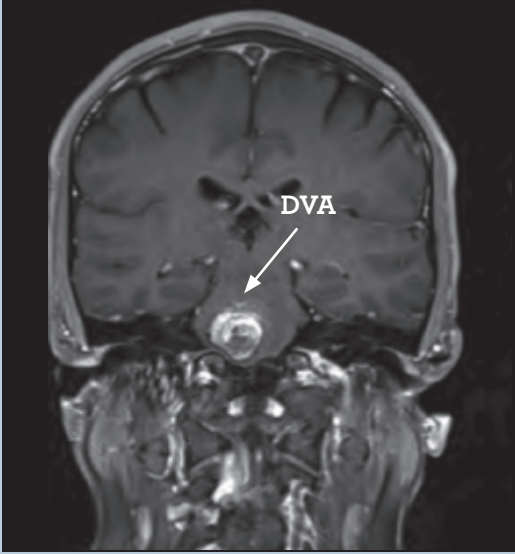
Cavernous malformations (CMs) are unusual collections of tightly packed, abnormal blood vessels that can occur anywhere within the brain or spinal cord. CMs in the brainstem are rare in the general population, but they pose the risk of significant neurological damage. Presenting symptoms for brainstem CMs vary depending on the location of the CM, but may include ataxia, aphasia, dysphagia, numbness or weakness of the face, arm or leg, seizures and brain hemorrhage.

Because of their delicate location, CMs within the brainstem are some of the most difficult lesions to treat in the nervous system. Some studies report a yearly risk of hemorrhage that ranges from 0.5 percent to higher than 2.7 percent¹. Consequently, the diagnosis of a brainstem CM in childhood carries a very high risk of bleeding during the child's lifetime. Most cases of CMs are sporadic, but there are some genetic forms of the disease in which patients have multiple CMs throughout the central nervous system. Familial cases, particularly in patients of Hispanic/Latino origin, may also occur.

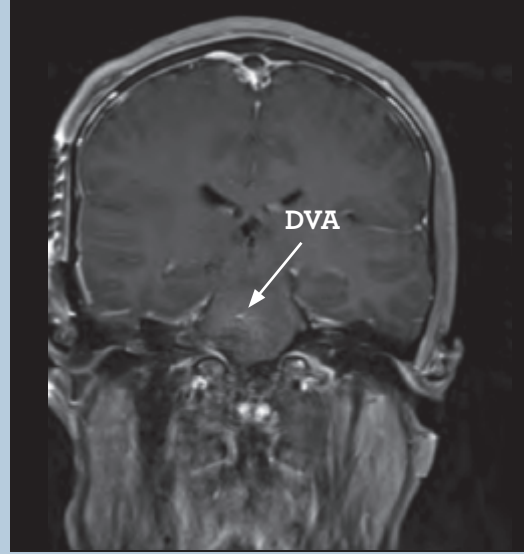
Brainstem CMs bleed at similar rates as CMs in other parts of the nervous system; however, brainstem CMs can become symptomatic even with small hemorrhages, as a result of their location near very high-density nervous system tracts and nuclei. Because of the concentration of critical structures within the brainstem, even small hemorrhages can cause catastrophic long-term neurological damage. Unfortunately, once neurological function is lost, it may not be recovered. Treatment should be considered for patients who have large symptomatic malformations or for those who experience repeated hemorrhages.

KEY INSIGHTS

- Cavernous malformations (CMs) are unusual collections of tightly packed, abnormal blood vessels that can occur anywhere within the brain or spinal cord.
- People who have brainstem CMs may gradually develop neurological deficits. Or an acute hemorrhage may be the first sign of a brainstem CM.
- Often, the best course of treatment is to observe the lesions until they cause significant neurological symptoms.
- If major neurological deficits occur, surgery remains the best treatment, and complete resection of the CM is curative.
- Because additional CMs can develop, long-term follow-up with repeat MRIs is important.

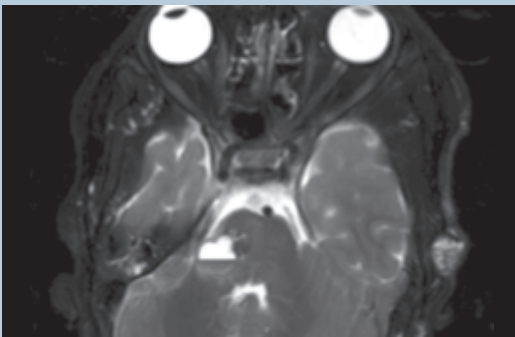


Pre-op

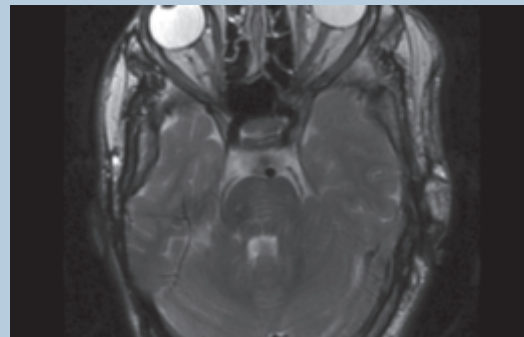


Post-op

Imaging of cavernoma resection showing preservation of developmental venous anomaly (DVA), which is almost always associated with a cavernomas. It is critical to spare the DVA because it drains normal surrounding nervous tissue.

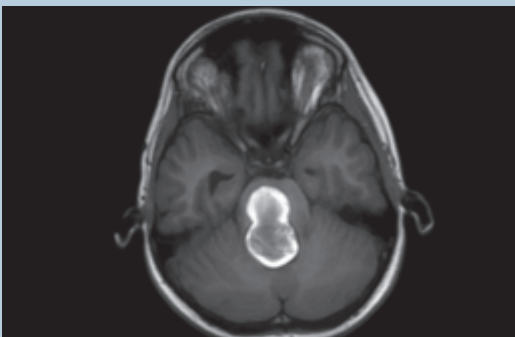


Pre-op

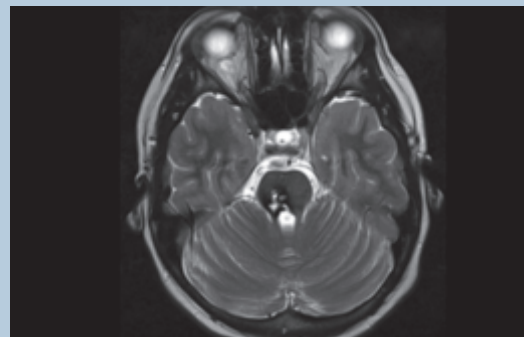


Post-op

An 8-year-old boy, who had been observed for years, became symptomatic and experienced facial weakness, dizziness and increased falls, so the lesion was removed via a pre-sigmoid approach without new neurological deficit.



Pre-op



Post-op

This 12-year-old patient had symptoms that slowly progressed. The sudden onset of quadraparesis necessitated surgery.

This view shows the same patient at three years post-op (now 15 years old).

Diagnosis

People who have brainstem CMs may gradually develop the neurological deficits that are associated with brainstem disorders, such as hemiparesis or cranial nerve deficits. Alternatively, an acute hemorrhage may be the first sign of a brainstem CM. In either case, an MRI will confirm the presence of the CM. Our multidisciplinary team assesses each patient to determine the optimal course of therapy.

Treatment

Because of the problematic location of a brainstem CM, many times observation is best. However, progressive neurological deficits or a catastrophic event may necessitate surgery. Before MRIs were developed, many patients with brainstem CMs were thought to have multiple sclerosis, because of the gradual, stepwise decline of neurological function that was associated with repeated occasional hemorrhages spread over many years.

At present, there are no endovascular options for treating these lesions, and radiation is rarely considered to be a reasonable option. Surgery remains the best treatment, and complete resection of the CM is curative. When deciding whether to observe the patient or to undertake surgery, we consider the following variables:

- The lesion's size and location (large or small, deep or superficial, the structures involved)
- Whether the CM is bleeding and/or increasing in size
- Whether the patient is experiencing increased neurological symptoms

Observation is appropriate if the lesion is small, not growing and located deep in the brainstem. If the lesion is large, superficial, growing, or rebleeding, and the patient's neurological symptoms are progressing, surgery will be considered. When a patient experiences an acute onset of symptoms from a hemorrhage, surgery may be the best option.

Because of the complexity of resecting brainstem CMs, our multidisciplinary pediatric neurovascular team consists of a pediatric neurosurgeon, a neurovascular surgeon, an otolaryngologist, and neurologists/electrophysiologists who provide intraoperative monitoring of critical neurological function during surgery. Postoperatively, our pediatric rehabilitation specialists manage ongoing rehabilitation.

Although complete resection of the CM is curative, additional CMs may develop elsewhere in the brainstem, so patients who have a CM history need long-term follow-up with ongoing MRI surveillance.

We welcome your questions and referrals.

¹ Fritschi J, Reulen H, Spetzler R, Zabramski J. Cavernous malformations of the brainstem: A review of 139 cases. *Acta Neurochir (Wien)*. 1994;130(1-4):35-46.

Author PROFILES

The authors are the founders of the Twin Cities Multidisciplinary Pediatric Neurovascular Consortium. This group brings together practitioners from multiple fields to obtain optimal outcomes for children who have complex neurovascular lesions, including brainstem cavernous malformations.



Patrick Graupman, M.D.

Patrick Graupman, M.D., is a pediatric neurosurgeon at Gillette who treats patients who have spina bifida, epilepsy, cavernous malformations of the brainstem, craniofacial disorders, hydrocephalus and complex movement disorders. In addition to his work with the Twin Cities Multidisciplinary Pediatric Neurovascular

Consortium, he has expertise in pediatric deep brain stimulation (DBS) as a treatment for dystonia and other complex movement disorders, and he participates in a working group of national and international experts in DBS.

He received his medical degree at the University of Minnesota School of Medicine, where he also completed a neurosurgery residency. He then finished a pediatric neurosurgery fellowship at University of Colorado Children's Hospital of Denver.

He is board-certified in pediatric neurosurgery and adult neurosurgery. His professional memberships include the American Society of Pediatric Neurosurgery and the American Association of Neurological Surgeons Section of Pediatric Neurosurgery, among other organizations.



Eric Nussbaum, M.D.

Eric Nussbaum, M.D., is a board-certified neurosurgeon. He is considered a national expert in the diagnosis and treatment of brain aneurysms and vascular malformations. Nussbaum leads the neurovascular neurosurgery program at United Hospital.

Nussbaum and his team at Minnesota Neurovascular and Skull Base Surgery provide consultation and surgical care for hundreds of patients from across the country who have complex brain tumors and brain vascular lesions. He works collaboratively with other specialists including neurointensivists, stroke neurologists, interventional neuroradiologists and other medical professionals to provide the best care for his patients.

He is among a small group of surgeons worldwide who specialize in brain aneurysm surgeries. His team has treated approximately 5,000 aneurysms, and he has personally performed more than 2,000 open microsurgical procedures for brain aneurysms.

Case STUDY

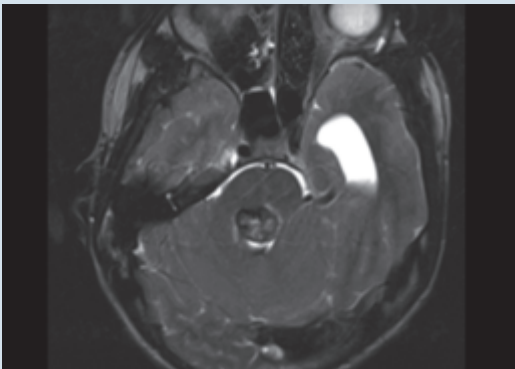
14-Year-Old Boy With Cavernous Malformation of Brainstem

This 14-year-old boy, who has mild cerebral palsy and a known small brainstem cavernous malformation (CM), was being followed by one of the authors. Because there was no change in lesion size and no progression was observed in his neurological symptoms, observation alone was recommended.

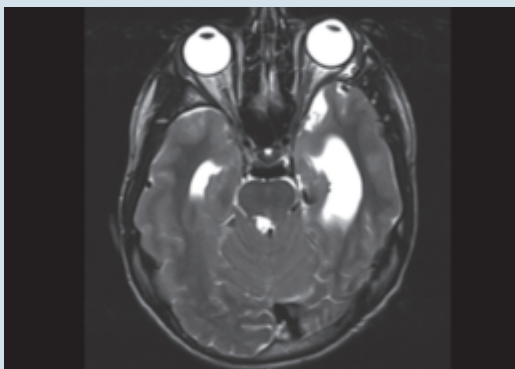
He then presented with a sudden onset of blurred vision, clumsiness, ataxia and weakness. Imaging showed that his known CM had doubled in size due to a hemorrhage. He was found to have ophthalmoplegia, mild hemiparesis and facial numbness.

The CM was resected completely, with care being taken to spare the associated developmental venous anomaly that drains the normal surrounding brainstem.

After a short inpatient rehabilitation stay, he was discharged to home with minimal (and improving) neurological deficits. He will continue to be seen for long-term follow-up and serial imaging.



Pre-op



Post-op

A Pediatric PERSPECTIVE

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A Pediatric Perspective Survey Inside!

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NEWS & NOTES

The Gillette Mankato Clinic Opened Sept. 15

Our new facility is located adjacent to the Mankato Clinic on the Wickersham Health Campus and is open Monday through Friday. Each week, Gillette holds clinics in pediatric neurology, pediatric orthopedics and pediatric rehabilitation medicine with providers rotating on a regular schedule. One to two times per month, physicians in pediatric craniofacial surgery, pediatric neurosurgery, pediatric spine orthopedics, sleep medicine and specialty pediatrics conduct clinics at the Mankato location. A family nurse practitioner and two registered nurses are on-site full-time Monday through Friday to coordinate care and refill intrathecal baclofen pumps.

Two New Physicians Joined Gillette in September

Todd Dalberg, D.O., who is board-certified in pediatrics, is part of Gillette's palliative care team. He practices at the St. Paul Clinic and St. Paul – Phalen Clinic.

Pediatric neurologist **Amanda Moen, M.D.**, takes care of children who have epilepsy and other neurological conditions. She works at the St. Paul Clinic and the Gillette Mankato Clinic.

Save the Date!

On Friday, December 12, 2014, Gillette will present "Management of Musculoskeletal Conditions in Children and Adolescents," an accredited continuing medical education course for clinicians. The conference will be held at the St. Paul RiverCentre from 8:00 a.m. to 5:00 p.m. Watch for complete conference and registration information in the coming weeks.