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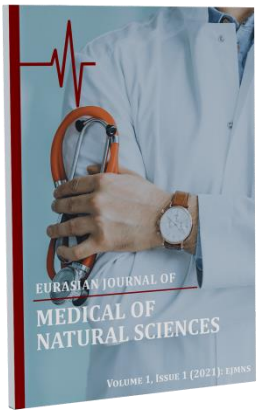

BRAIN ARTERIOVENOUS MALFORMATION

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ABSTRACT

Arteriovenous malformations (AVMs) are developmental anomalies of the vascular system characterized by tangles of poorly formed blood vessels, where feeding arteries are directly connected to a venous drainage network without an interposed capillary system. While AVMs can occur anywhere in the body, brain AVMs are particularly concerning due to the high risk of bleeding, which can cause neurological damage. This article reviews the pathophysiology, clinical presentation, and emphasizes the importance of the interprofessional team's role in managing these patients.

Objectives:

- Explain the pathophysiology of brain AVMs.
- Summarize the clinical presentation of brain AVMs.
- Describe the available treatment and management options for brain AVMs.
- Discuss strategies for interprofessional teams to enhance care and outcomes for patients with brain AVMs.

Introduction. Arteriovenous malformations (AVMs) are developmental anomalies of the vascular system, characterized by tangles of poorly formed blood vessels where feeding arteries are directly connected to a venous drainage network without an interposed capillary system. Although AVMs can develop anywhere in the body, brain AVMs are particularly concerning due to the high risk of bleeding from the abnormal blood vessels, which can lead to neurological damage.

Etiology. The etiology of brain AVMs is not well understood. Although the exact cause remains unknown, it is likely multifactorial, involving both genetic mutations and angiogenic stimulation (the physiological process of forming new blood vessels from pre-existing vessels). Some researchers suggest that AVMs develop in utero, while others propose that an angiopathic reaction following a cerebral ischemic or hemorrhagic event (subtypes of stroke) may play a primary role in their development.

Epidemiology In the United States, the incidence of AVMs is 1.34 per 100,000 person-years, though the actual prevalence is higher due to many cases being clinically silent, with only about 12% of AVMs becoming symptomatic. The mortality rate for patients experiencing a

hemorrhage is 10-15%, while morbidity ranges from approximately 30-50%. There is no gender predilection. Despite the presumed congenital origin of AVMs, the clinical presentation most often occurs in young adults.

Pathophysiology Arteriovenous malformations consist of a central vascular nidus, a tangle of arteries and veins without an intervening capillary bed. The feeding arteries drain directly into the veins through one or multiple fistulae. These arteries lack the normal muscularis layer, and the draining veins often appear dilated due to the high-velocity arterial blood flow shunted through the fistulae. AVMs can cause neurological dysfunction through three primary pathophysiological mechanisms. First, the abnormal blood vessels are prone to bleeding, leading to hemorrhage in the subarachnoid space, intraventricular space, or more commonly, the brain parenchyma. Second, in the absence of hemorrhage, seizures may occur due to the mass effect of the AVM or venous hypertension in the draining veins. Third, the "steal phenomenon" can cause slowly progressive neurological deficits, as the normal brain parenchyma is deprived of nutrients and oxygen, with blood bypassing the normal capillary bed to flow through the malformed arteriovenous channels.

History and Physical Examination

AVMs are clinically asymptomatic in about 15% of cases until a significant event occurs.

Between 41% and 79% of patients present with intracranial hemorrhage, making AVMs the second most common cause of intracranial bleeding after cerebral aneurysms, accounting for 10% of all subarachnoid hemorrhage cases. Children are more likely to present with hemorrhage than adults. These hemorrhages are typically intraparenchymal but can also occur in the subarachnoid space. Symptoms of hemorrhage include loss of consciousness, sudden and severe headache, nausea, vomiting, and potential sequelae such as seizures, hemiparesis, sensory loss on one side of the body, and language processing deficits due to local brain tissue damage. Minor bleeding may be asymptomatic. Most patients recover symptomatically following the cessation of bleeding as the damaged blood vessel repairs itself.

Seizures are reported as a presenting symptom in 15% to 40% of patients, with the risk increasing for AVMs that are cortical, large, multiple, or superficially draining. Seizures are typically focal, either simple or complex partial, but can secondarily generalize.

A progressive neurological deficit may develop in 6% to 12% of patients over a period ranging from a few months to several years. This is often attributed to vascular steal syndrome, mass effect, hemorrhage, or seizures. Symptoms include hemiparesis, visual disturbances, loss of sensation on one side of the body, and aphasia. Minor bleeding might occur without noticeable symptoms.

Headaches are commonly reported but do not have specific features that associate them with AVM and may be incidental.

Evaluation Brain AVMs are typically first identified through cross-sectional imaging such as computed tomography (CT) or magnetic resonance imaging (MRI). A combination of MRI and angiography is often used to plan treatment and predict the likely success and associated risks of surgical, endovascular, or radiological therapies.

Computed Tomography (CT): On a non-contrast CT scan, the nidus appears as a hyperdense area compared to the surrounding brain tissue, with possible evidence of enlarged draining veins and calcification. Despite many AVMs being large, there is typically no mass

effect or edema unless there is bleeding. Post-contrast CT, especially with CT angiogram, clearly shows the feeding arteries, nidus, and draining veins, producing a "bag of worms" appearance. Angiography can delineate the exact anatomy of feeding vessels and draining veins. In the acute setting of hemorrhage, CT sensitivity is reduced due to the compression of the nidus by the hematoma, necessitating more sensitive techniques such as MRI or angiography.

Magnetic Resonance Imaging (MRI): MRI is highly sensitive for locating the brain AVM nidus and any associated draining veins or distant bleeding events. The fast flow in tangled blood vessels generates serpiginous and tubular flow voids visible on both T1 and T2, but primarily on T2-weighted images. MRI can also show complications like previous hemorrhage, adjacent brain edema, and atrophy. Post-radiosurgery, MRI can evaluate the regression of the nidus volume, post-therapy edema, and radiation necrosis within the treatment area.

Angiography: Angiography remains the gold standard for diagnosis and treatment planning. It provides a precise evaluation of the nidus configuration, its relationship, and drainage to surrounding vessels. The presence of an associated aneurysm indicates a higher risk of hemorrhage. Contrast transit time, which relates to the flow state of the lesion, can provide critical information for endovascular treatment planning.

Treatment / Management

Treatment Modalities: Invasive management is recommended for younger patients with one or more high-risk features for AVM rupture. For older individuals without high-risk features, medical management is usually preferred. In these cases, anticonvulsants for seizure control and analgesia for headaches may be sufficient. Studies indicate that a history of previous rupture is a significant risk factor for long-term bleeding. Other factors include patient age, AVM location, presence of aneurysms, size, and other vascular features. Patients with AVMs and intractable epilepsy are also candidates for treatment.

Surgical Excision: Open microsurgical excision is the primary treatment for patients at high risk of hemorrhage.

The Spetzler-Martin Grade (SMG) scale is commonly used to assess the risk of surgical morbidity and mortality in brain AVMs. This composite score includes:

Nidus size (<3 cm, 3-6 cm, >6 cm; 1-3 points)

Eloquence of adjacent brain tissue (1 point for lesions in the brainstem, cerebellar peduncles, thalamus, hypothalamus, or language, sensorimotor, or primary visual cortex)

Venous drainage (1 point if any or all drainage is via deep veins, such as basal veins, internal cerebral veins, or precentral cerebellar veins)

A higher score indicates a greater risk of surgical morbidity and mortality.

Radiotherapy and Endovascular Embolization: These are valuable alternatives to surgical treatment for patients at high surgical risk and can also serve as adjuncts to surgical management.

Differential Diagnosis

The differential diagnoses of cerebral AVMs include:

- Carotid/vertebral artery dissection
- Cavernous sinus syndromes and thrombosis
- Cerebral amyloid angiopathy
- Cerebral venous thrombosis

- Dissection syndromes
- Fibromuscular dysplasia
- Intracranial aneurysms
- Migraine and cluster headaches
- Moyamoya disease
- Stroke
- Vein of Galen malformation

Prognosis

Various scoring systems are used to assess the morbidity and mortality associated with observation versus intervention for different types of cerebral AVMs. The primary ones include:

- Spetzler-Martin scale: Used for microsurgery
- Supplementary Spetzler-Martin scale: Also for microsurgery
- Pittsburgh radiosurgery-based AVM grading scale
- Toronto score: Used for microsurgery
- Buffalo Score: Used for endovascular treatment

Complications

The main complications associated with AVMs include:

- ✓ Intracranial bleed
- ✓ Mass effect
- ✓ Seizures
- ✓ Steal phenomenon
- ✓ Neurological deficits

Enhancing Healthcare Team Outcomes

The diagnosis and management of brain AVMs require an interprofessional team consisting of a neurosurgeon, neurologist, internist, and invasive radiologist. Follow-up care is typically managed by a nurse practitioner and primary care provider. The treatment approach for brain AVMs depends on factors such as size, location, patient age, and the AVM's rupture risk. While surgery is the primary treatment, embolization is another viable option. Patient outcomes are influenced by the AVM's size, symptoms, location, comorbidities, and mental status. Post-surgery complications are common, and many patients need extensive rehabilitation for recovery. The most significant risk factor for mortality is AVM rupture.

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