

Cerebro arteriovenous malformation presenting as recurrent epistaxis: a rare entity

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Abstract

An arteriovenous malformation is an abnormal tangle of blood vessels in the brain or spine. Some AVM's have no specific symptoms and little or no signs to one's life or health, while others cause severe and devastating effects when they bleed. Treatment options range from conservative watching to aggressive surgery, depending on the type, symptoms and location of the AVM.

Keywords: AVM- Arteriovenous Malformation, Brain, Spine

Introduction

Arteriovenous malformations (AVMs) are vascular abnormalities consisting of fistulous connections of arteries and veins without a normal intervening capillary bed. In the cerebral hemispheres, they

frequently occur as cone-shaped lesions with the apex of the cone reaching toward the ventricles. Nearly all AVMs are thought to be congenital. Supratentorial location is the most common (90%) [1].

Case Report

A 16 year old female patient presented to the emergency department with a history of six episodes of epistaxis noticed since 2 days prior to the admission to the hospital. There is no history of trauma, fever or joint pains. Past history revealed recurrent epistaxis with multiple blood transfusions from the age of 8 years and with a history of stroke with right sided hemiplegia and right sided UMN type of facial palsy at the age of fourteen years for which treatment was taken at a local hospital and only supportive treatment was given without imaging. On examination, child had severe pallor with right hemiparesis. Initial blood counts revealed microcytic hypochromic anemia. Child was advised imaging which revealed complex intracranial AV malformation with a large aneurysm and pressure erosion of the cribriform plate of the ethmoid bone and hypodense per vascular edema noted in posterior limb of left internal capsule.

MR angiogram of brain showed large vascular nidus (4.4*7.8*4.2cm) in left frontal region with extension along the floor of anterior cranial fossa, root of nose on left side, intraparenchymal hematoma (3.8*3.7cm) multiple cystic areas (3.8*4cm) in left high parietal region likely to be CSF fluid or arachnoid cyst and 2mm midline shift to the right side. Child was stabilised with supportive measures like blood product transfusion and was referred to a tertiary neurological center clipping of the feeding blood vessel was done where in after digital subtraction angiography study.

Discussion

AVMs of the brain are congenital lesions developing during the late somite stages between the 4th and 8th weeks of life. The lesion consists of persisting direct connections between the arterial inflow and venous outflow without an intervening capillary bed [2]. AVMs

arise from persistent direct connections between the embryonic arterial and venous sides of the primitive vascular plexus, with failure to develop an interposed capillary network [3,4,5].

Genetic variation may influence pathogenesis and the clinical course of brain AVMs [6] Identification of genetic polymorphisms associated with clinical course

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would help in stratifying risk and understanding the underlying biology. Molecular studies of brain AVMs have revealed an altered expression profile compared with normal tissue, including upregulated expression of genes involved in angiogenesis and inflammation [7]. Brain AVM patients homozygous for the interleukin (IL)-6-174G allele had a greater risk of ICH at presentation than IL6-174C carriers; a polymorphism in the inflammatory cytokine IL6 was associated with ICH presentation of brain AVM [8]. Local IL6 release by endothelial cells within the brain AVM nidus may, therefore, contribute to vascular wall instability by stimulating release and activation of matrix metalloproteases [9,10].

Types- There are several types of AVMs:

- Arteriovenous malformation – abnormal tangle of blood vessels where arteries shunt directly into veins with no intervening capillary bed; high pressure.
- Cavernoma – abnormal cluster of enlarged capillaries with no significant feeding arteries or veins; low pressure.
- Venous malformation – abnormal cluster of enlarged veins resembling the spokes of a wheel with no feeding arteries; low pressure, rarely bleed and usually not treated.
- Capillary telangiectasia – abnormal capillaries with enlarged areas (similar to cavernoma); very low pressure, rarely bleed and usually not treated.
- Dural AV fistula – direct connection between one or more arteries and veins into a sinus. The veins of the brain drain into venous sinuses (blood-filled areas located in the dura mater) before leaving the skull and traveling to the heart. Dural AV fistulas and carotid-cavernous fistulas (CCF) are the most common [1]

The most common presentation of an AVM is intracerebral hemorrhage (ICH). After ICH, seizure is the second most common presentation. Other presentations of AVMs include headache and focal neurological deficits, which may be related to steal phenomena or other alteration in perfusion in the tissue adjacent to the AVM, such as venous hypertension from arterialization of normal draining veins.

Diagnostic Evaluation- A computed tomography (CT) scan may be used as an initial screening tool for patients presenting with neurological sequelae related to unruptured or ruptured AVMs. This study can be used quickly to determine location of the lesion, acute hemorrhage, hydrocephalus, or areas of encephalomalacia from previous surgery or rupture. A non-enhanced CT scan may show irregular hyperdense

areas frequently associated with calcifications in unruptured AVMs and acute haemorrhage on plain CT scan with ruptured AVMs. With the addition of intravenous contrast material, a CT scan can demonstrate the nidus and feeding vessels or dilated draining veins.

Magnetic resonance imaging (MRI) is superior to CT scan in delineating details of the macro architecture of the AVM, except in the case of acute hemorrhage. These architectural features include exact anatomic relationships of the nidus, feeding arteries, and draining veins as well as topographic relationships between AVM and adjacent brain [11].

MRI and angiography in combination provide complementary information that facilitates understanding the three-dimensional structure of the nidus, feeding arteries, and draining veins. Complete cerebral angiography with multiple projections is a mandatory step in the preoperative evaluation of a patient with an AVM. Cerebral angiography can localize the nidus, the feeding arteries, and draining veins.

Many techniques are available for studying the functionality of cortical structures surrounding the AVM. These include the use of positron emission tomography, functional MRI, magnetoencephalography, and direct provocative testing of cortical function. Judicious use of these techniques will enhance safety of AVM therapy. Such information may allow the surgeon to tailor treatment modalities to increase the margin of safety during treatment and decrease periprocedural flow-related hemorrhagic or ischemic complications [1,12].

Clearly, one of the most important considerations in terms of decision making is the AVM itself. Location, size, and configuration (compact versus diffuse) of the nidus; the pattern and location of the feeding and draining vessels; and the association of abnormalities, including aneurysms, direct arteriovenous fistulae, stenosis, or occlusion of the venous draining system are all factors that must be taken into consideration to estimate not only the risk of surgical excision of a particular AVM but also the risk of no treatment. To help the neurosurgeon estimate the surgical risk, a number of classifications have been developed but the most commonly used classification today is that proposed by Spetzler and Martin [13].

Treatment- The currently used treatments for AVMs include microsurgical resection only, preoperative endovascular embolization followed by microsurgical

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resection, stereotactic radio surgery only, preprocedural endovascular embolization followed by radio surgical treatment, endovascular embolization only, and observation only. The ultimate goal for all of these modalities is cure for the patient; however, the only way to achieve cure is with complete obliteration of the AVM. Microsurgical resection, whenever it can be performed safely is the “gold standard” treatment for brain AVMs, and other methods of treatment must be measured against it. Hence, AVM's should also be considered as a possibility while ruling out the causes of epistaxis in children.

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