Vascular Malformations of the Brain

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Vascular malformations of the brain are classified into four principal groups: **arteriovenous malformations, cavernous malformations, capillary telangiectasis and venous angiomas**. Of these, the first two are the types associated with the risk of hemorrhage and the subsequent development of neurologic symptoms.

Arteriovenous malformations (AVMs) are responsible for approximately 5-10% (range 5-28%) of non-traumatic subarachnoid hemorrhages. Emmanuel, Luschka and Virchow first described AVMs in the mid-1800s. Olivecrona performed the first surgical procedure on AVMs in 1932. AVMs consist of an abnormal connection between arteries and veins, without any capillaries intervening (Figs. 1 & 2). In a normal brain oxygen enriched blood from the heart travels in sequence through smaller blood vessels going from arteries, to arterioles and then capillaries. Oxygen is removed in the latter vessels to be used by the brain. After the oxygen is removed, blood reaches the venules and later veins, which will take the blood back to the heart and lungs. When there is a AVM, blood goes directly from the arteries to the veins through abnormal vessels disrupting the normal circulation of blood. Although, the cerebral blood flow is markedly increased in this region of the brain due to the vascular malformation, with the lack of capillaries, tissue perfusion may be markedly reduced, resulting in chronic ischemia in that portion of the brain that contains the AVM.

They are considered to be congenital, and occasionally familial, but the genetic transmission of AVM, if there are any, are unknown, thus, they are not typically thought of as an inherited disorder, unless in the context of a specific hereditary syndrome, such as Von Hippel-Lindau disease or hereditary hemorrhagic telangiectasia. In 7-10% of cases, AVMs can coexist with saccular aneurysms. The most plausible theory for the origin of AVMs is congenital absence of the capillary bed in a region of the brain. The lack of capillaries leads to anatomic changes in the arteries and veins composing the AVM. The absence of capillaries causes shunting of blood

directly from the arteries into the veins, which causes the intraluminal pressure within the veins to be markedly elevated. This leads to ectasia and muscularization of the veins creating hybrid vessels with venous and arterial characteristics. Thus, the lesion contains large vessels with shunt related changes along with the original malformation per se (Figs. 1 & 2).

Although, more than 90% of AVMs are asymptomatic, when symptoms occur they are most commonly headaches and seizures. Other symptoms which can occur are a pulsing noise in the head, progressive weakness and numbness and vision changes, as well as debilitating excruciating pain. Males are affected twice as frequently as females, with the lesion typically being recognized clinically between the ages of 10 and 30 years. A more serious complication of AVMs is hemorrhage occurring either as an intracerebral hemorrhage, or as a subarachnoid hemorrhage. Intracranial hemorrhage is unpredictable in its occurrence, occurring at a rate of 2-4% per year. Although bleeding can occur at any age, it typically occurs in patients under 40-years-of-age.

Arteriovenous malformations (AVMs) occur both within the cranial vault and the spinal canal. They involve vessels in the subarachnoid space extending into the brain parenchyma or may occur exclusively within the brain parenchyma. They are composed of greatly enlarged blood vessels separated by gliotic tissue, often with evidence of prior hemorrhage. Some of the vessels will appear as arteries with duplication and fragmentation of the internal elastic lamina, while others will show marked thickening or partial replacement of the media by hyalinized connective tissue.

The most common region AVMs occur is in the territory of the middle cerebral artery, especially its posterior branches.

Large AVMs occurring in the newborn period can lead to congestive heart failure due to the shunt effects, especially if the malformation involves the great vein of Galen. These lesions may also obstruct the aqueduct of Sylvius and result in hydrocephalus.

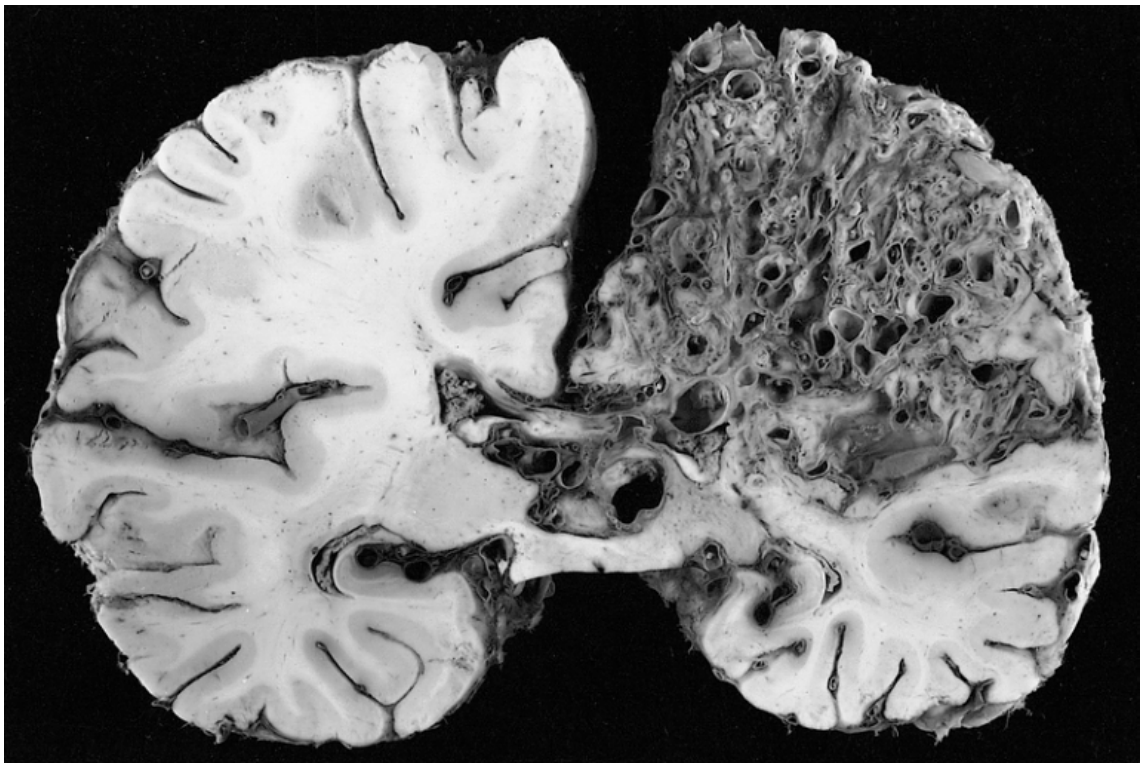

Fig. 1. This is a gross photograph taken at the time of neuropathological examination of the brain of a patient who died from complications of a large arteriovenous malformation of the parietal lobe. (en.wikipedia.org)

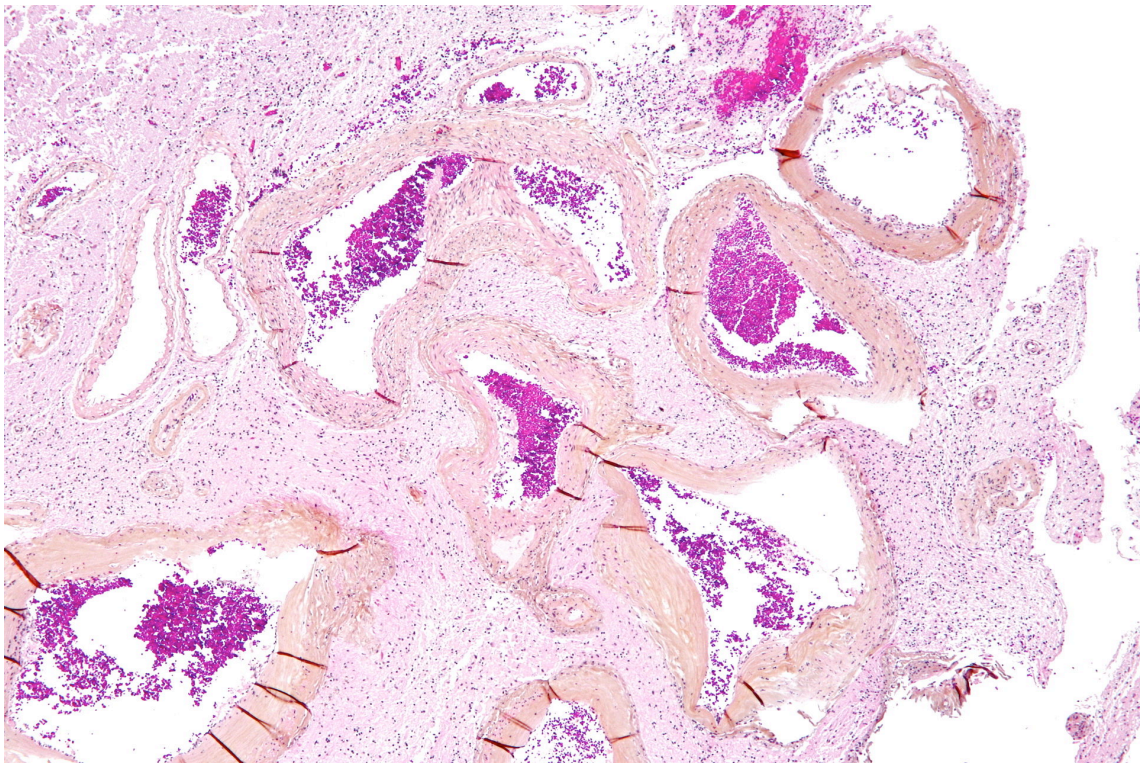

Fig. 2. This is a photomicrograph of a AVM in the brain. Note the greatly enlarged blood vessels separated by gliotic tissue showing evidence of recent hemorrhage (upper right side of photograph). (en.wikipedia.org).

Occasionally, intracranial AVMs communicate with extracranial arteries, usually branches of the external carotid, with the superficial temporal artery being the most common, forming **cirsoid aneurysms**. A cirsoid aneurysm refers to a group of dilated blood vessels resembling a varix. These aneurysm typically occur in the venous system, although they can occur in the arterial, as well as the lymphatic system (Fig. 3).

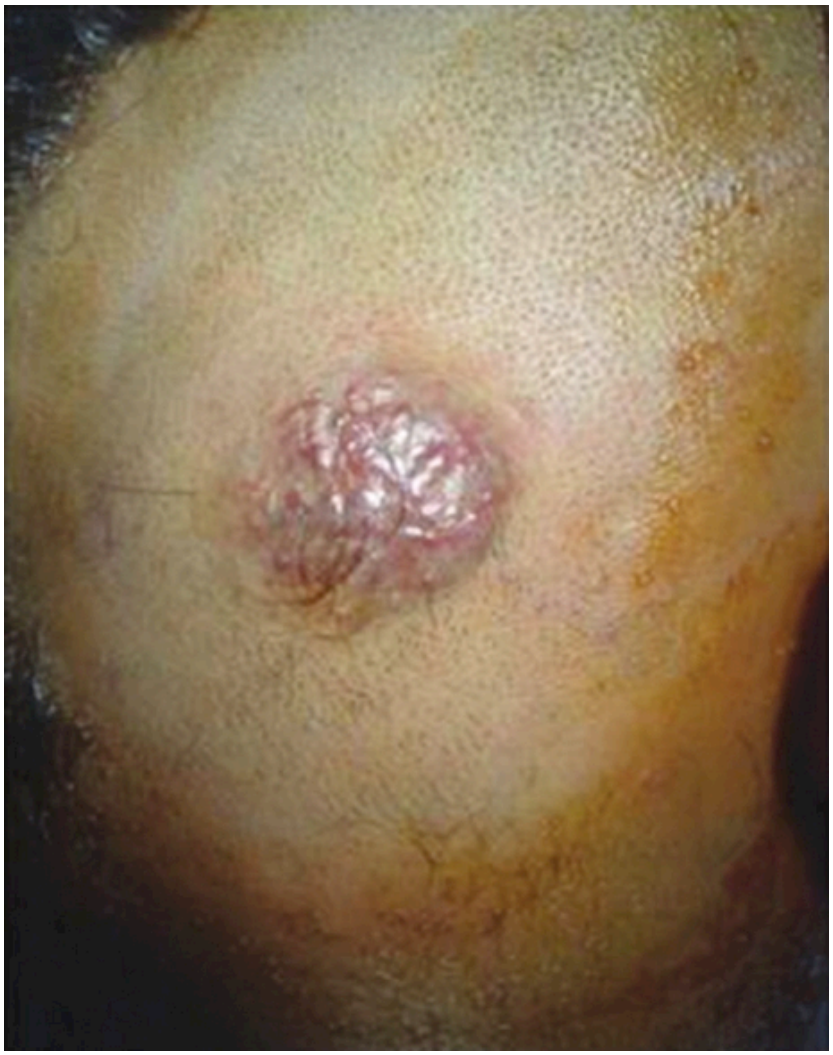

Fig. 3. This is a photograph of a cirsoid aneurysm of the scalp. (www.asianjns.org)

Cavernous malformations, also called **cavernous hemangiomas**, are compact, occasionally multiple lesions up to several centimeters in diameter, which may occur anywhere in the brain or leptomeninges, although, they occur most commonly in the cerebellum, pons, and subcortical regions, in decreasing order of frequency (Fig. 4).

They may present with seizures, headache and focal neurological deficits and, less commonly, with hemorrhage. However, many cavernous malformations never manifest any symptoms. Cavernous malformations are unique among vascular malformations for familial forms are relatively common, with a variety of identified genetic loci. Some autosomal dominant cerebral cavernous malformations are caused by mutations of the KRIT1 gene on chromosome 7q,

encoding a protein that interacts with the krev-1/rap1α tumor suppressor gene. Other familial cases linked to loci at 7p and 3q. Typically, when cavernous malformations present with multiple lesions they are often familial.

Cavernous malformations consists of greatly dilated, loosely organized vascular channels with thin, collagenized walls and are devoid of intervening nervous tissue, which distinguishes them from capillary telangiectasias (Fig. 5). They may show areas of calcification and even ossification. Often the vascular channels composing a cavernous malformation are occluded. Cavernous malformations may occasionally be associated with similar lesions in other organs, such as the kidney, liver, lung or skin.

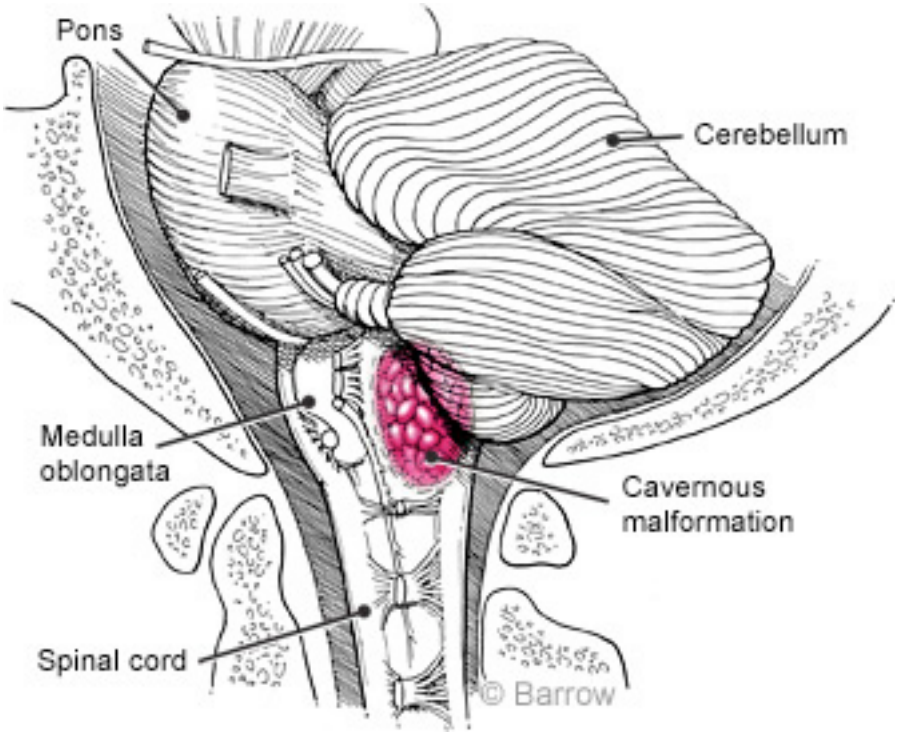

Fig. 4. This is a drawing of a cavernous malformation of the brainstem. (www.thebarrow.org)

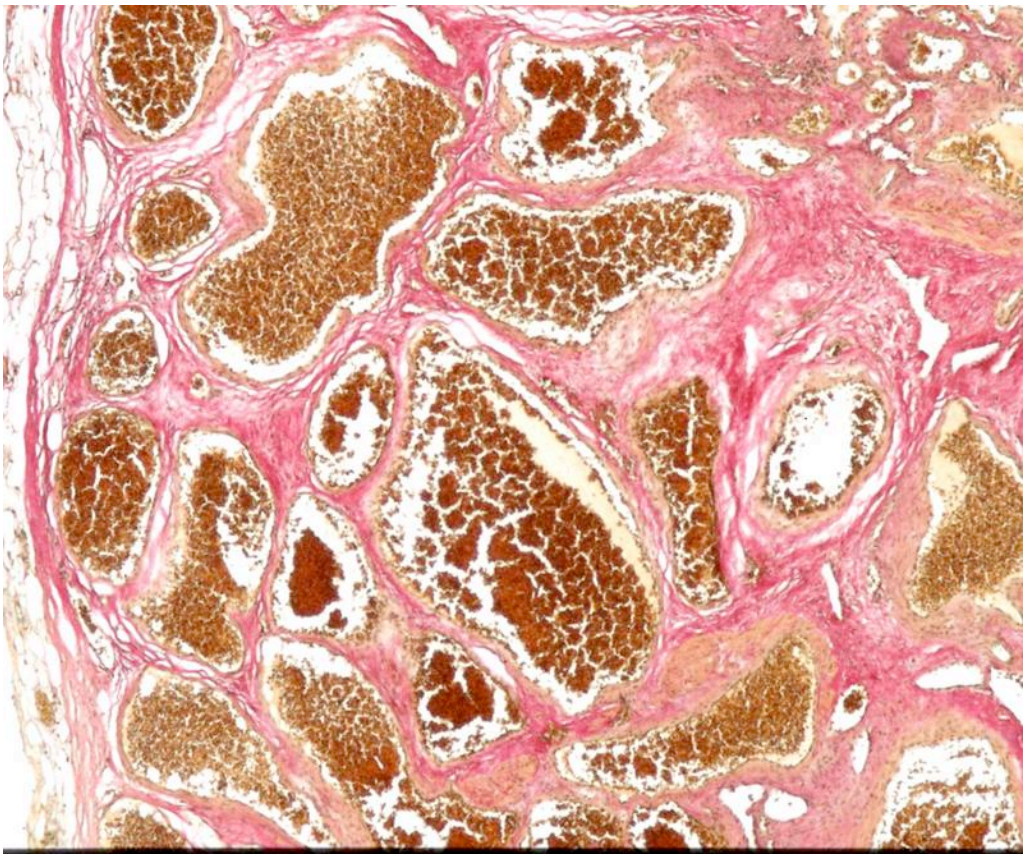

Fig. 5. This is a photomicrograph of a cavernous malformation of the brain. (en.wikipedia.org)