

**Case Report**

Open Access, Volume 3

**Cerebrovascular malformations in adults: What mode of revelation? About 53 cases****V Ndayishimiye\***; M Sabiri; O Jbara; M Banao; M Labied; G Lembarki; S Lezar; F Essodegui

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**Abstract**

Cerebrovascular malformations include a broad spectrum of intracranial blood vessel disorders, involving the arterial wall, capillary bed, venous and lymphatic systems. Their main associated risks are intracerebral hemorrhage, seizures and focal neurological deficits. We carried out a retrospective study over a period of 45 months (3 years and 9 months), ranging from April 01, 2018 to December 31, 2021; from the files of patients referred for magnetic resonance imaging (MRI) or CT scan cerebral exploration in the context of neurological symptomatology or as part of an assessment carried out for other medical or surgical purposes, with as judgment criterion the demonstration of a cerebrovascular malformation. We collected fifty-three (53) patients, with an average age of 47 years old and as extremes ages 18 years old and 70 years old. We noted a female predominance with a sex ratio of 1.7 in favor of female sex. The most of patients were explored with MRI in 85% of cases. Our patient history was dominated by arterial hypertension in 20% of cases and hemorrhagic stroke in 12% of cases. The symptomatology presented by the patients was mainly represented by headache and epileptic seizures in 20% of cases each, dizziness in 15% of cases. The cerebrovascular malformations were dominated by cavernomas in 54% of cases (complicated of bleeding in 27% of cases). Other cerebrovascular malformations were : aneurysm: 19%, arteriovenous malformations: 15% and developmental venous abnormalities in 12% of cases. MRI and CT scans are very important radiological modalities for diagnosing malformative cerebral vascular abnormalities. Their discovery is more often made during a radiological assessment motivated by another cause.

**Keywords:** Cerebrovascular; Malformation; Revelation; Adults; MRI; CT scan.

Received: Mar 29, 2022

Accepted: Jun 07, 2022

Published: Jun 14, 2022

Archived: www.jcimcr.org

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DOI: www.doi.org/10.52768/2766-7820/1888

**Citation:** Ndayishimiye V, Sabiri M, Jbara O, Banao M, Labied M, et al. Cerebrovascular malformations in adults: What mode of revelation? About 53 cases. J Clin Images Med Case Rep. 2022; 3(6): 1888.

**Introduction**

Cerebrovascular malformations include a broad spectrum of intracranial blood vessel disorders, involving the arterial wall, capillary bed, venous and lymphatic systems. Their main associated risks are intracerebral hemorrhage, seizures and focal neurological deficits.

The goals of our study are :

-To show the radiological characteristics in magnetic resonance imaging (MRI) and computed tomography of cerebrovascular malformations in adults, symptomatic or not.

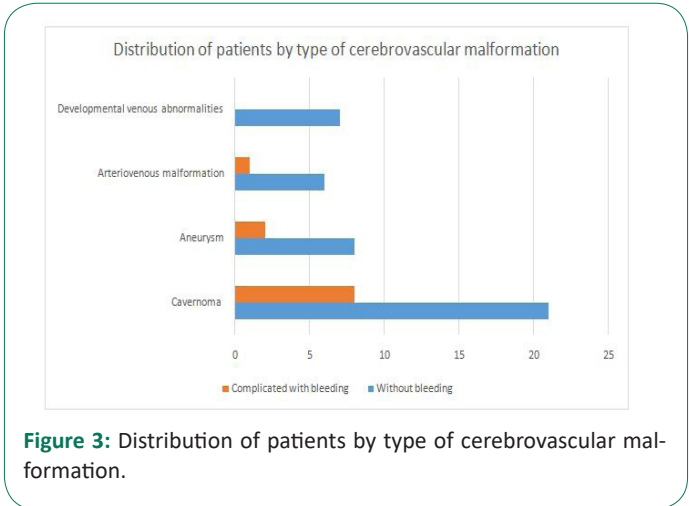
- Enlighten clinicians on the type of the lesions.

**Materials and methods**

We carried out a retrospective study over a period of 45 months (3 years and 9 months), ranging from April 01, 2018 to December 31, 2021 from the files of patients referred for MRI or CT scan cerebral exploration in the context of a neurological symptomatology or as part of an assessment carried out for other medical or surgical purposes.

The judgment criterion was the demonstration of a cerebrovascular malformation, whether or not it is related to the patient’s clinical symptomatology.

Data processing were performed using Excel software.



**Figure 3:** Distribution of patients by type of cerebrovascular malformation.

**Results**

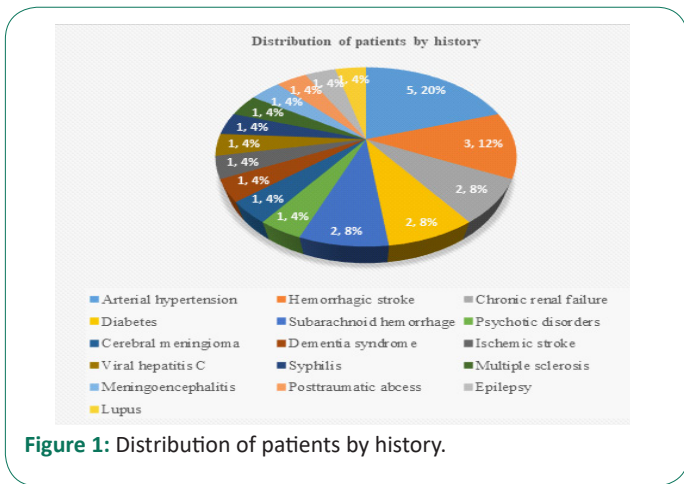
- Fifty-three (53) patients were collected.
- Average age: 47 years old, with the extremes of 18 years old and 70 years old.
- Gender: 34 women and 19 men.
- Exploration method: MRI n = 45, CT scan :8

**Discussion**

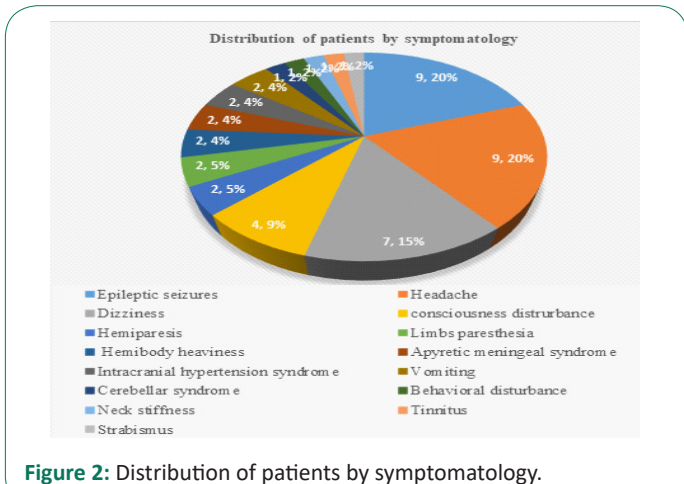
The classification of cerebrovascular malformations distinguishes between arteriovenous malformations (with plexiform nidus or with arteriovenous fistula), cavernous angiomas, capillary telangiectasias and venous development abnormalities (venous angiomas) [1].

The clinical manifestations of cerebrovascular malformations can be defined empirically, including clinical penetrance (in familial cases), age at clinical presentation, symptomatic presentation (accidental; headache or nonspecific symptoms; convulsions; hemorrhage; focal deficits, frequently by epilepsy). Non-neurological manifestations include skin lesions [2].

Cerebral cavernous malformations, also called cavernomas, are vascular abnormalities of the brain made up of clusters of abnormal capillaries surrounded by deposits of hemosiderin and a gliotic margin. The vascular system is filled with blood and is thrombosed to varying degrees [3]. Its precise prevalence is not known because the diagnosis can only be made by brain imaging or autopsy. Estimations from autopsy studies, clinical MRI studies, and studies performing brain MRI for non-clinical purposes suggest a prevalence of 0.16% to 0.9% [4]. The majority of cavernomas cases include a single lesion with or without an associated venous developmental abnormality. These are called sporadic cavernomas and are often asymptomatic and non-hereditary. The other type, called hereditary or familial cavernomas, is caused by an inherited autosomal dominant genetic mutation, associated with multiple lesions. Cavernomas can be found in several places in the central nervous system, supratentorial lesions being more common than infratentorial lesions [5]. The majority of cavernomas cases are asymptomatic but can result in severe neurological symptoms such as hemorrhagic stroke (30-40%), seizures (40-70%), headache (10-30%) and focal neurological symptoms (35-50%) [6]. Cavernomas



**Figure 1:** Distribution of patients by history.



**Figure 2:** Distribution of patients by symptomatology.

typically show heterogeneous signal intensity on conventional MRI images, representing the accumulation of hemoglobin degradation products at different stages. The T2-weighted and sensitivity (SWI) images clearly show a surrounding hypointense edge due to hemosiderin deposition from recurrent microhemorrhages; this is considered a characteristic finding of Cavemomas. SWI can also detect very tiny cavernomas, which are often present in familial cases [7].

About 3% of the adult population has one or more unruptured intracranial aneurysms. In a recent study from Japan, an unruptured intracranial aneurysm was found in 4.3% of 4,070 people undergoing magnetic resonance imaging of the Willis polygon for screening [8]. The occurrence of an intracranial saccular aneurysm is increased in certain hereditary disorders compared to the general population. Autosomal dominant polycystic kidney disease is the most common inherited disease associated with an intracranial saccular aneurysm. Other inherited disorders associated with a brain aneurysm include multiple endocrine neoplasia type I, hereditary hemorrhagic telangiectasia, Ehlers-Danlos syndrome type IV, Marfan syndrome, and neurofibromatosis type I [9]. Several theories have been developed about predictive factors of rupture, but the simple and useful geometric index, particularly suited to small aneurysms, is the aneurysm / vessel size ratio more commonly referred to as the size ratio and this depending on the location [10]. Angiography is a special resolution method of exploration and allows a dynamic study of cerebral vascularity. It is also therapeutic. It is an invasive and irradiating method that is no longer indicated for screening. CT angiography and MRI angiography and magnetic resonance angiography are used for this purpose [11,12].

Developmental venous anomalies (DVA) are the most frequently encountered cerebrovascular malformations, with an incidence of up to 2.6% reported in a series of 4069 brain autopsies. They are often discovered incidentally during routine brain imaging using computed tomography (CT) or magnetic resonance imaging (MRI). They are seen in both pediatric and adult populations, with a slight male predominance [13]. While the vast majority of DVAs are asymptomatic and follow a mild clinical course, there have been numerous reports of DVAs causing clinical symptoms, particularly through multiple different pathophysiologic mechanisms: focal neurologic deficit, headache, or seizure [14]. The detectability of DVAs largely depends on the imaging techniques used. However, they are characterized by very typical morphological imaging features. These are large collecting veins passing through the brain parenchyma in places where only capillaries and small veins are generally expected, and radially contributing veins resulting in a typical jellyfish head appearance. These veins drain blood from an atypical territory [15].

Arteriovenous malformations are a rare pathology. Prevalence and incidence are difficult to assess, but the prevalence for all modes of discovery is estimated to be between 10 and 15 per 100,000 population. In France, we discovered in theory 800 AVMs per year, including 300 hemorrhagic. It is a congenital or acquired malformation defined by the existence of an arteriovenous shunt without an intermediary of capillaries [16]. Unruptured cerebral arteriovenous malformations have an annual risk rate of hemorrhage of 1.5% to 3%, and a risk of death at the first hemorrhage of 10%, which increases with each episode of repeated hemorrhage. Certain angioarchitectural features are considered to be associated with the risk of future hemorrhage,

including posterior fossa or deep localization, the presence of associated aneurysms, exclusive deep venous drainage, and venous stenosis or ectasia [17]. They are considered to be both vascular lesions and mass lesions and should be imaged with computed tomography (CT) and magnetic resonance imaging (MRI), as well as complete catheter angiography, which should include injections of circulation of the external carotid artery. Computed tomography is best for identifying acute bleeding and the resolution, calcifications, and location of embolic material. MRI, in particular the T2 sequence, provides excellent correlation of components of AVMs with the surrounding parenchyma. Angiography obviously provides detailed imaging of the arteries and draining veins [17].

## Conclusion

MRI and CT scans are very important radiological modalities for diagnosing malformative cerebral vascular abnormalities. Their discovery is most often made during a radiological assessment motivated by another cause.

## Declarations

**Competing interests:** The authors declare no conflict of interest.

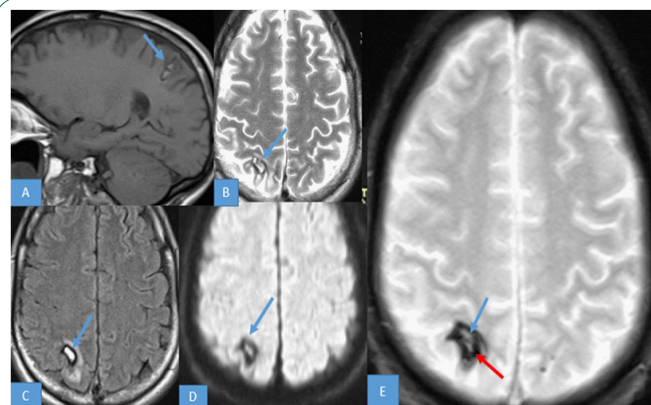
**Contributions from authors:** All the authors contributed to the conduct of this work. They also state that they have read and approved the final version of the manuscript.

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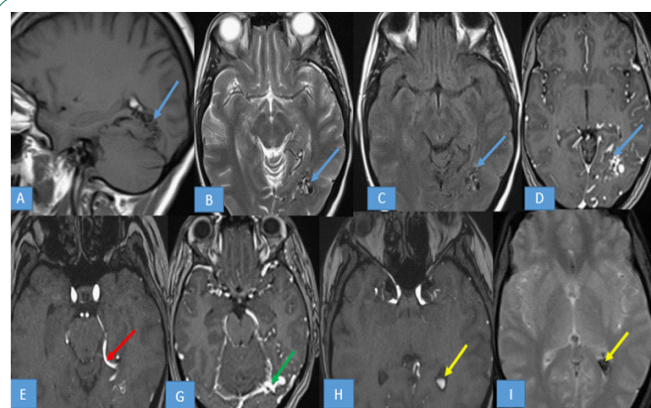
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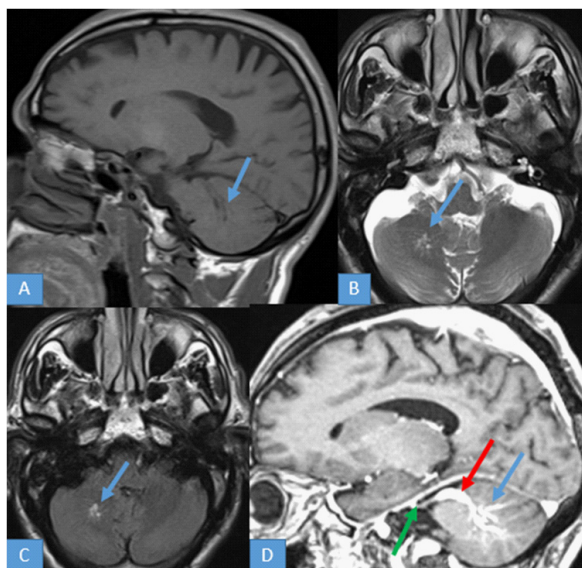
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**Imaging figure 2:** Cerebral MRI, T1 sagittal sequence (A), T2 axial, FLAIR, diffusion and gradient echo (B, C, D and E): polylobed right parietal lesion in heterogeneous hypersignal with the appearance of 'salt pepper' in T1, T2, FLAIR and diffusion (blue arrows) surrounded by a hypointense border more visible on the gradient echosequence (red arrow) related to a cavernoma in a 34-year-old patient with chronic renal failure on dialysis.



**Imaging figure 3:** Cerebralangiography: T1 sagittal (A), T2 axial, FLAIR and T2\* (B, C and I), T1 axial after injection of gadolinium (D and J) sequences, axial arterialangiographic sequences (E and H): Left occipital serpiginous structures with a central nidus strongly enhanced after injection of gadolinium (blue arrows) with afferent artery coming from left cerebral posterior artery (red arrow) and the efferent veins draining into the left lateral sinus (green arrow). Note a bleeding in the occipital horn of the left lateral ventricle (yellow arrows), related to an arteriovenous malformation complicated of bleeding in a 38-year-old patient complaining of an apyretic meningeal syndrome.



**Imaging figure 1:** Cerebral MRI, T1 sagittal (A), T2 axial, FLAIR (B and C) and T1 sagittal sequence after injection of gadolinium (D): right cerebellar serpiginous structures (blue arrows) confluent towards a common collector (arrow red) performing the appearance of the head of a jellyfish, draining into a subependymal vein opposite the brainstem (green arrow), related to a developmental venous anomaly (AVD) in a patient aged 58 year sold.