Management of intracranial arteriovenous malformations: Experience of the neurosurgery department of Specialty Hospital-Rabat.

ABSTRACT:

Cerebral arteriovenous malformations (cAVM) constitute an uncommon but serious neurovascular pathology due to the hemorrhagic risk linked to the natural evolution of the disease. Cerebral arteriovenous malformations (AVMs) are often revealed in young adults by several types of neurological symptoms, including epileptic seizures, recurrent headaches, progressive neurological deficits or intracranial hemorrhages.The aim of this study is to report the experience of the neurosurgery department, Specialty Hospital in RABAT; covering 30 consecutive cases of cerebral AVM treated surgically in our department. Highlight the epidemiological and semiological profile of this pathology; Highlight the benefit of microsurgical treatment, as well as certain prognostic factors compromising the therapeutic result. This is a retrospective study of 30 cases of operated arteriovenous malformation collected over a period of 5 years in the neurosurgery department of the Specialty Hospital in RABAT.

The total number of our patients is 30. The average age of patients is 40.5 years with extremes of 16 years and 65 years. Among the 30 patients, 19 are male and 11 female, for a sex ratio of 1.72. The average time to diagnosis was 3 months. Clinically, the main symptom remains headaches. Symptomatic treatment including analgesic and antiepileptic treatment. Curative treatment based on neurosurgery, embolization and radio surgery. The evolution was favorable in 91.6% with total recovery in 45.45% of cases while 54.5% of cases retained after-effects such as motor deficit.

INTRODUCTION :

- Cerebral arteriovenous malformation (cAVM) is a congenital lesion generally consisting of a cluster of abnormal vessels (the “nidus) which has formed around one or more arteriovenous shunts located in the cerebral vascular network [1]
- 14 per 10,000 individuals (0.14%) and appear at any stage, peak frequency between 30 and 40 years. [2,3]
- The most common clinical presentations of cAVMs: intracranial hemorrhage, epilepsy, chronic headaches and focal deficits [2,5].
- Management involves monitoring, microsurgical treatment, endovascular treatment and radiosurgery [1].
- The aim of this work is to report our experience and study the different epidemiological, diagnostic, therapeutic and evolutionary aspects of this anomaly.

MATERIALS AND METHODS :

- This is a retrospective descriptive study of 30 patient cases, admitted for a cAVM, carried out in the neurosurgery department at the Rabat Specialty Hospital, over a period between 2019 and June 2023.
- For each patient the following variables were studied:

  Epidemiological data.

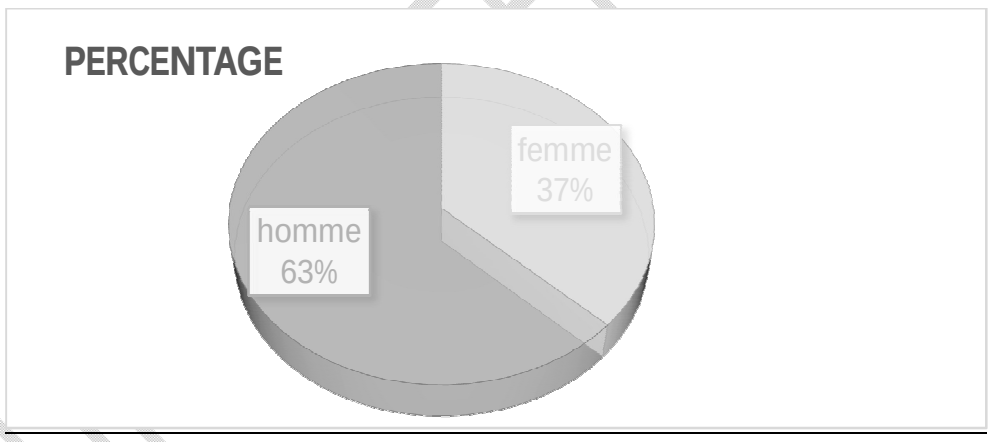
  Clinical and paraclinical information,

  Therapeutic and progressive modalities.
- The study included all patients who were hospitalized in the department for the management of a cAVM.

RESULT :

- During the study period, 30 patients presented a cAVM, representing a frequency of 6.6 cases/year. An average age of onset was 32 years.
- A sex ratio of 1.72 (19 men to 11 women).

Chart 1. Distribution according to sex



Graph 1. Distribution according to age group

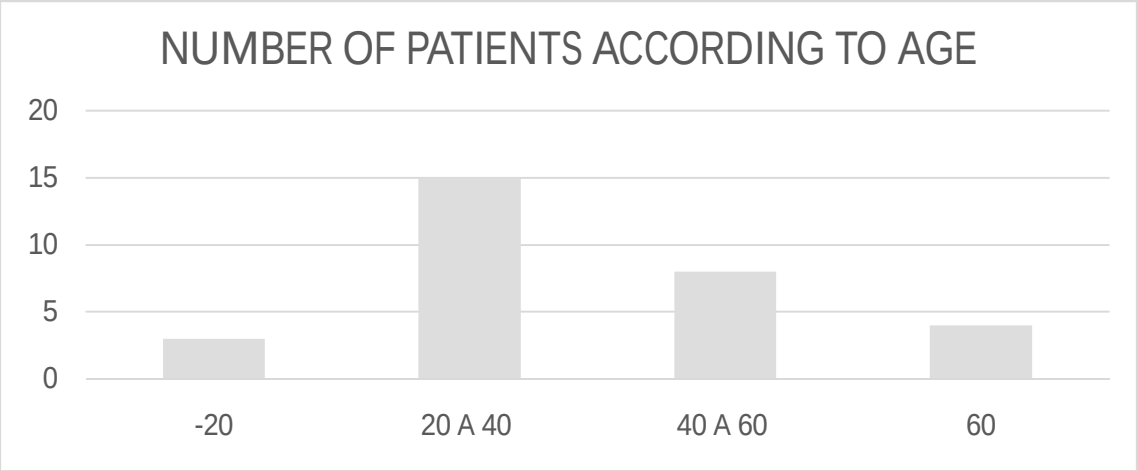


Table 1. Distribution of patients according to clinical data:

Clinical data	**Number and % of patients**
Headache	30(100%)
Disturbance of consciousness	15(50%)
Seizures	8(26.66%)
Motor signs	15(50%)
Aphasia/slow speech	5(16.66%)
Cranial nerve compression	6(20%)
Vision problems	2(6.66%)
Sensitive signs	1(3.33%)
Cerebellar SD SC	1(3.33%)

4/Radiological exploration:

- **<u>Brain computed tomography (CT)</u>**carried out in 95.8% of cases.

It showed signs in favor of a cerebral hemorrhagic accident in 25 patients or 83.33%, including 79.2% having isolated or associated cerebral hematomas and 9.52% having isolated ventricular hemorrhage.

It revealed hydrocephalus in 2 cases.

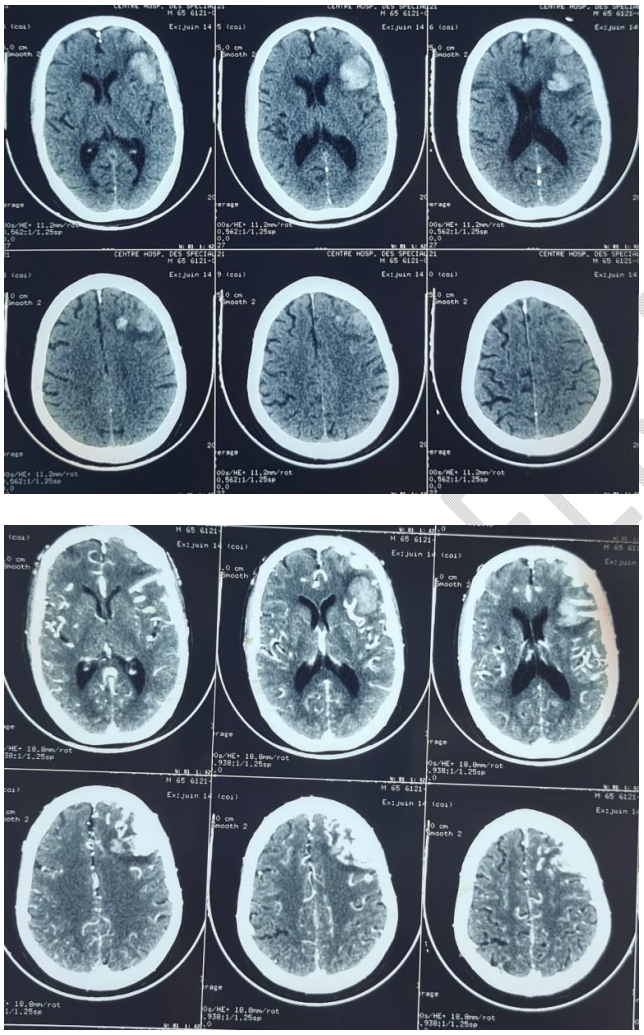


Figure (1): Brain CT with Agiographic sequences showing a left frontal AVM

- **Cerebral arteriography:**

- Confirmation of the diagnosis in 100% of cases.

- The AVMs were of superficial location in 75% of cases compared to 25% of deep location, of lobar location in 80% of cases, i.e. 24 cases, 12.5% of cases at the level of the posterior cerebral fossa and 7.5% of cases. at the level of the basal ganglia.
- The size of the nidus varies between 1 and 5 cm.
- The cAVM is supplied mainly by the middle cerebral artery in 50% of cases, the anterior cerebral artery in 33.3% of cases and the vertebro-basilar system in 16.7% of cases.
- Venous drainage of the nidus was superficial in 71.4%, deep in 23.8% and combined in 4.8%.

Figure (2): Right frontal AVM subtotally embolized preoperatively

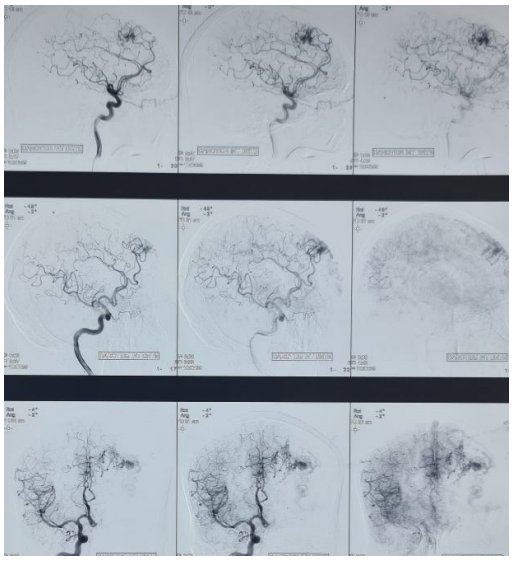
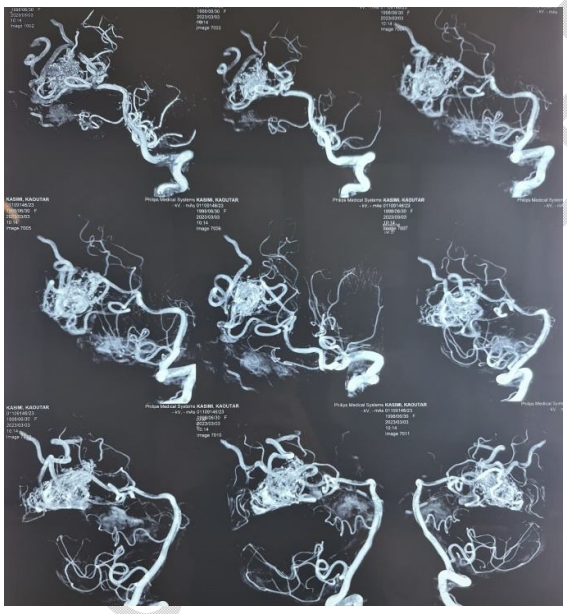


Figure (3): Cerebral angiography showing a left frontal AVM



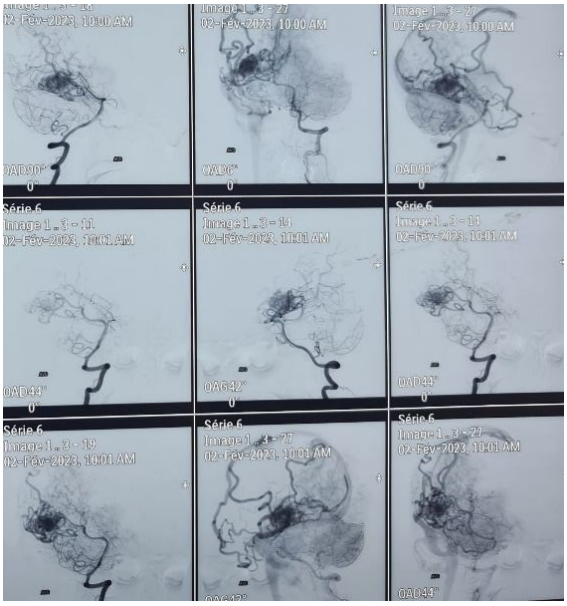


Figure (4): Cerebral angiography which shows a grade 2 right temporal AVM according to the Spetzler-Martin classification

Table 2. According to the classification of Septzler and Martin, the AVMs in our series are classified as follows:

Grade	**Effective**	**Frequency**
Grade I	1	3.33%
Grade II	8	26.66%
Grade III	19	63.33%
Grade IV	2	6.66%
Grade V	0	0%
Total	30	100%

5/Therapeutic support:

- The support arrangements were based on 2 components:

Symptomatic treatment including analgesic and antiepileptic treatment.

Curative treatment based on neurosurgery, embolization and radio surgery.

- 16 patients or 53.33% benefited from surgical treatment with total excision of the cAVM
- 4 patients only had evacuation of the hematoma since the AVM was deep in location.
- Embolization was carried out in 7 patients 24 hours before the surgical procedure, i.e. 23.33% of cases.
- Radio surgery in 36.66% of cases.

6/Evolution:

- The evolution was favorable in 91.6% with total recovery in 45.45% of cases while 54.5% of cases retained after-effects such as motor deficit.
- Complications were recorded in 8.4% of cases: postoperative meningitis in one patient and a sensorimotor deficit of the hemibody in the other.

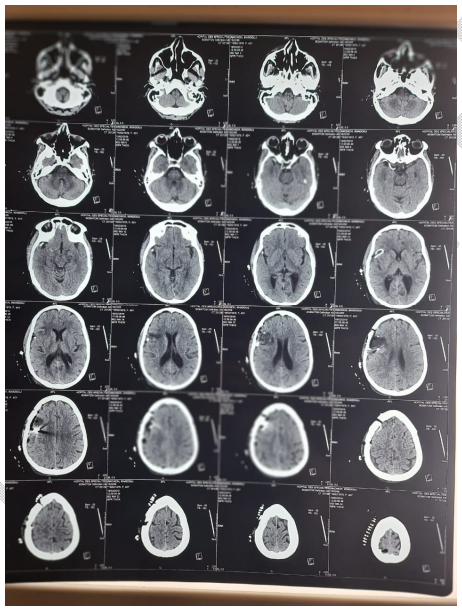


Figure (5): Postoperative cerebral CT of an excision of a right frontal AVM operated 24 hours after embolization

DISCUSSION :

- AVMs have an incidence which varies between 0.89 and 1.34/100,000 per year.
- A prevalence estimated at 0.94 per 100,000 inhabitants/year and a frequency of 14 per 10,000 individuals [6].
- They often become symptomatic in young adults between 30-40 years old according to Stapf et al [2].
- In a retrospective study of 701 patients, the average age was 31 years, which agrees with the average age in our series [7].
- AVMs are often revealed in young adults by several types of neurological symptoms.
- Intracranial hemorrhage is the most common mode of clinical presentation with a frequency between 30% and 82%; in our series we noted a frequency of 83.33% [2,8,5].
- The annual bleeding rate is estimated between 2% and 4% (Ondra et al., 1990; Kondziolka et al., 1995; Brown 2000) [2,5,9]. Itoyama et al found that the risk of rebleeding after a first hemorrhagic episode is 6.9% in the first year, 1.9% per year after 5 years and 0.9% after 15 years [9,10] .
- Recent longitudinal analyzes suggest that – apart from the criterion of initial hemorrhage, other topographical, angio-architectural and hemodynamic factors positively influence the hemorrhagic risk of an AVM [10,11].

The angioarchitectural factors are: deep location, unique deep venous drainage, presence of intra or juxta-nidal aneurysms, small size of the nidus, presence of venous stenosis and presence of venous reflux

The hemodynamic factor is represented by high pressure in the afferents and efferents of a cAVM [2,7,12].

- Several recent series suggest that the morbidity and mortality of ruptured cAVMs is lower than that of other causes of hemorrhage and only 2% of all intracranial hemorrhages are related to ruptured cAVMs [7,5,13]. ..
- Intracranial hemorrhage most often conditions the vital and functional prognosis due to its topography and its abundance [13]. Hemorrhages from cAVMs are meningeal in 30% of cases, parenchymal (23%), intraventricular (16%) and in combined locations in 31% of cases [13,14]. In our series we have parenchymal localization in 52.5% of cases, 9.5% isolated ventricular hemorrhage, and 38% combined localization.
- Crawford et al in his series of 217 cases report that patients who presented with a hemorrhagic accident, 62% of them did not present neurological disability, 25% had minor instability and 6% had major deficits with a risk of death estimated at 29% and a morbidity rate estimated at 45% [9].
- In our series we estimate that 41.7% of patients present neurological deficits.

- Epilepsy is the second most common symptom in patients with cAVM, according to the study by Ondra et al, 24% of patients with cAVM presented convulsive seizures which is consistent with the results of our series (26%) [2.9].As well as the discovery of AVM during the assessment of late-onset epilepsy[4]
- However, there are other modes of presentation that are documented, Pool et al. observed that apart from the initial symptoms of hemorrhage (42%) and epilepsy (33%), other symptoms such as neurological deficit (23%), headache (14%), aphasia (8%), or tinnitus (2%) [7.5]
- The imaging evaluation of cAVMs requires selective visualization of the different compartments of the lesion in order to decide on the therapeutic strategy [2,12].
- Cerebral angiography remains the reference method but non-invasive imaging techniques such as magnetic resonance angiography (MRA) provide interesting information.
- These two techniques make it possible to provide prognostic arguments on the hemorrhagic risk by the angio-architectural study [2,12].
- CT is most often reserved for the emergency context for the detection of a hematoma [12].
- Bharatha et al reported the rarity of multiple cAVMs in the same patient [4]. In the event of multiple lesions, underlying conditions such as hereditary hemorrhagic telangiectasia (Rendu-Osler / hereditary hemorrhagic telangiectasia), arteriovenous malformation syndrome, cerebrofacial arteriovenous metameric syndrome should be sought [4].
- The most used system for the classification of cAVMs is that of Spetzler-Martin which uses 3 criteria: the size of the nidus, the location of the cAVM and venous drainage. It assigns a score of 1 for small (<3 cm) cAVMs, 2 for medium-sized (3-6 cm) and 3 for large (>6 cm). The location of the nidus in a non-functional (0) or functional (1) territory. Venous drainage is scored as superficial only (0) or including drainage to deep cerebral veins (1). The score obtained makes it possible to divide the AVMs into 3 classes and to guide the therapeutic attitude. Class A includes Spetzler-Martin grades I and II; class B grade III and class C grades IV and V [1].
- Spetzler et al in their series, 79% of cAVMs <3cm, 29% of cAVMs between 3 to 6cm and 12% of cAVMs >6cm present intracranial hemorrhage [15]
- The prognosis of cAVMs also depends on their location; Willinski et al observed that hemorrhages seem to be more frequent for deep lesions [16,15].
- The main feeding artery is affected in the majority of cases, the sylvian artery (ACM) in 32%-57% of cases then in decreasing order the anterior cerebral artery (ACA) 28%-40% of cases then the vertebro-basilar system (SVB) 12% of cases (Masahiro et al., Turjman et al) [14,12]. The results found in our series are close to those of Misahiro et al with a frequency of 50% for ACM, 33.3% for ACA and 16.7% for SVB.

- Deep venous drainage is a factor that plays a role in the spontaneous and therapeutic prognosis of cAVMs according to a multi-varied study focused on the angioarchitectural parameters of 631 cAVMs [2, 10, 13, 11,12].
- The specific treatment of cAVM is a priori invasive and requires the application of one or the combination of several therapeutic techniques: endovascular embolization, surgical excision and/or stereotactic radiotherapy.
- Microsurgery constitutes the oldest therapeutic strategy with an average complete eradication rate of up to 97%, without or with pre-surgical embolization (stapf et al.,2002c) with an average overall postoperative morbidity of 8, 6% [2,9,15]
- Thus the size of the AVM and its location in the eloquent and/or deep zone constitute therapeutic prognostic factors for surgery alongside the Spetzler and Martin grade [15].
- Depending on the angio-architecture and the material used, complete obliteration can be achieved by embolization alone in 11%, 17%, or 40% of cases (Gobin et al., 1996; Meisel et al., 2002; Yakes et al. ., 1997) [8].
- Morbidity reported after endovascular treatment is 0.4% to 12.5% of cases and mortality is 0.4% to 7.5% [8].
- Radiosurgery constitutes the third aspect of the treatment of cAVMs; it is reserved for small cAVMs or residues not excluded after surgery or embolization and located in areas that are difficult to access [16].
- BCVA volume is also a major factor in radiosurgery failure [16]
- Determining the factors linked to the rupture of cAVMs and the predictive factors of response to each therapeutic means is imperative with regard to an adapted therapeutic strategy.

CONCLUSION :

- cAVMs are uncommon compared to other vascular malformations but are more serious due to the hemorrhagic risk linked to the natural evolution of the disease.
- Determining prognostic factors constitutes the tool for establishing an appropriate therapeutic plan.
- The latter most often requires consultation between a multidisciplinary team: neurosurgeon, neuroradiologist and radiotherapist.

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