

# Cavernous Malformation of the Central Nervous System; Diagnosis and Surgical Management

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## Abstract

**Background:** Cavernoma is grouped within vascular malformations and could affect the whole of the central nervous system. They are angiographically occult lesions, with a low flow dynamism. Their management is capital to stop the progression of several consequences such as epilepsy and neurological deficit.

**Material and Methods:** The work is a retrospective study for all operated patients with cavernous malformations. We have reviewed the charts of 42 patients, but included 27 in the present work that had complete studied parameters.

**Results:** The consequent topographical repartition was; 19 patients harboring cavernoma in the cerebrum, 4 patients in the brainstem, and 4 others in the spinal cord. We had 100% of gross total resection in the cerebrum and the spinal cord and 75% in the brainstem. We didn't have any cases of mortality or morbidity. We had heterogeneous data about long-term follow up but most of the patients had a regression of their postoperative complications, especially for cranial nerves for the brainstem location.

**Conclusion:** Microsurgical removal of cavernomas is an effective modality to avoid the onset of severe neurological deficit or further their progression without significant morbidity. The timing of surgery and appropriate technics must be performed to achieve such results. The reduction of other therapeutic modalities will prompt more anatomical studied and surgical approaches to resect more deep-seated malformations.

**Keywords:** Microsurgery, Cavernoma, Haemorrhage, Seizures.

## Introduction

One of the most known classifications of vascular malformation is the one established by a neuropathologist named William F McCormick, who described: angioma called also cavernous malformation (CM) as a unique entity among five classes of malformations (1). Both adult and pediatric populations could be affected by this focal vascular abnormality (2, 3) and the prevalence rate was estimated at 0.5 to 0.7 % of the general population (4). They are also called angiographically occult vascular malformations because of the absence of arteriovenous shunt and their low blood flow making than the catheter angiography diagnostic test negative in all cases (5).

One revealing symptom of these dynamic vascular lesions is seizures, which are encountered in 30 % of cases (6). Another usual occasional presentation is the hemorrhagic stroke that was found in 25 % of cases (7). In one work, (8) focal neurological was reported to be a presentation modality in 46% and headaches in 65% of cases (8). Some of these malformations are discovered incidentally in usual serial imaging, especially by the increased availability of radiological investigations. As it was reported, In up to 40% of cases, patients are asymptomatic (9).

Mainly, two forms of cavernous malformation were described; sporadic marked by isolated lesions and familial characterized by multiple localizations, an autosomal mode of occurring, and the presence of CCM multiple lesions in molecular investigations (10) (11). The mutations in KRIT 1 gene were analyzed in some studies and their percentages were raised in both forms of CM (10, 12).

The genes of CM encode for proteins that are expressed widely in the neural tissue and interfere with proteins of cell junction during angiogenesis(13). The cellular evidence of alteration of vascular development and endothelial permeability are the final results of this growth disturbance (13). Developing this field of research will have a great impact on the prognosis of these malformations and their morbidity (13, 14).

Related epilepsy to CM is a matter of debate and several surgical techniques were described as a curing method; lesionectomy, extensive lesionectomy, and even lobectomy especially for temporal localization. The improvement of the quality of life and significant control of seizures was noticed after the surgical resection of these vascular malformations (15). The process of decision-making for brainstem CM is specific compared to other situations, and this is due to planning the right indication in the appropriate time. their surgical resection could be achieved without significant mortality and good postoperative results (16).

We report our experience with the management of cavernous malformation of the central nervous system. the surgical modality was selected after the right process of decision making and the results and eventual complications were also reviewed.

## Materials and Methods

The retrospective work is about 43 patients presenting CM affecting the central nervous system managed in the department of neurosurgery (Mohamed Lamine Debaghine University hospital Algiers- Algeria). We have included 27 patients in the analysis that had the full useful data. The medical files, pathological reports were reviewed. the surgical technique adopted in every patient was also reported. Three compartments of the central nervous system were distinguished; cerebrum, brainstem, and spinal cord.

## Results

### 1. Cerebral topography

**Age and sex:** 11 patients of the studied subgroup were male, a slight female population predominance (15 patients) was noticed. the age of patients ranged between 14 to 62 years.

This anatomical location included the cerebrum, cerebellum, and the vermis .this topography was by far the most affected in our series by 19 patients . The parameters that were evaluated are: the mode of revelation, the size, the quality of resection, and immediate outcome . (Table I)

**Revelation mode:** The seizures were the most encountered symptom during disease revelation. 10 patients (52%) presented epileptic seizures in two modes; isolated in 7 patients and associated with other symptoms in 3 others. Headaches were encountered in 4 patients increased intracranial pressure was present in one patient with a hemorrhagic event in the posterior fossa. In one patient, consciousness disorders were related to an important ventricular hemorrhage. A cerebellar syndrome was noticed in one patient. The visual hallucinations, status epilepticus, and dysarthria were reported only one time in three other patients.

**Topography:** The most affected anatomical region was the temporal lobe (10 patients), among this subgroup; the mesial side was affected in 6 patients, and the neocortex in 5 patients. we have to precise that (03) three patients had multiple cavernoma (double). The frontal lobe was affected in two patients. The occipital and parietal lobes in one patient for each one. The Rolandic cortex was affected in one patient. We had two patients harboring vermian cavernomas and we had an exceptional ventricular location in another patient.

**Size:** The size of cavernoma was calculated on MRI. It ranges between 8 and 45 mm. most lesions size varied from 20 to 25 mm. (9 patients). One patient had a 45 mm-sized lesion.

**Surgical management:** All patients were operated on and had different technic depending on the preoperative evaluation and the clinical status. the technics that were adopted in this subgroup of patients were: lesionectomy ; where the cavernoma was resected selectively: this procedure was performed in 16 patients . An extended lesionectomy where the perilesional gliotic tissue of the cavernoma was additionally removed and even part of the lobe (cases with the hemorrhagic event), this was adopted in three patients. A combination of the previously cited technics was adopted in two patients. the third technic was lobectomy which was adopted in two patients, where they presented resistant epilepsy with an expanded network of seizures affecting the whole of the mesial region of the temporal lobe.

**Quality of resection:** We have achieved a total resection in all cerebral localization of cavernoma. This was confirmed by the intraoperative inspection of the surgical cavity and postoperative MRI during follow up.

**Outcome:** Good outcome was defined by an uneventful postoperative course. This was achieved in 14 patients. for the rest of patients; one patient had dysarthria after a surgical removal of left temporal (neocortex), that improved significantly at 6 month follow up. Another patient with a Rolandic cavernoma, had a hemiparesis that resolved partially one year after surgery. Some visual disorders were noticed in one patient (lateral hemianopsia). One patient presented seizures that were well controlled medically.

**Table 1:** Demographics of cerebral cavernoma (IICP: increased intracranial pressure, Sd: : syndrome, A: Adult, C: Child)

Pa-tient	Popu-lation	Revelation	Localization	Size (mm)	Surgical tech-nic	Resec-tion	Outcome
1	A	seizures	Temporal (mesial)	25	Lobectomy	Total	Good
2	C	Seizures	Temporal (neocortex)	20	Lesionectomy	Total	Good Dysarthria (improvement at 6 months)
3	A	Seizures	Temporal neocortex Temporal mesial	9.5 10	Lobectomy	Total	Good (Engel Ia)
4	C	- IICP - cerebellar sd	Superior vermis	21	Lesionectomy	Total	Good
5	A	Chronic headaches	Frontal	20	Lesionectomy	total	Good
6	A	Seizures	Temporal (mesial)	21	Lesionectomy	Total	Good (Engel Ia)
7	A	Temp seizures	Temporal (neo cortex)	18	Extended lesionectomy	Total	Good
8	C	Seizures	Central Tempo (mesial).	34 45	Temp; hematoma +lesion removal	total	Favorable
9	A	Cerebellar Sd	Superior vermis	12	Lesionectomy	Total	Good (no cerebellar Sd)
10	A	Seizures	Parietal (sub cortical)	25	lesionectomy	Total	Good
11	A	Consciousness disorder	Ventricular (Frontal horn)	8	Lesionectomy	Total	Good
12	A	Headache seizures	Frontal lobe	19	Lesionectomy	Total	Good
13	A	Previous hemorrhage (7 years before)	Rolandic cortex region	34	Lesionectomy	total	Hemiparesis resolved partially 1 year after
14	A	Seizures	Temporal (neocortex)	20	Lesionectomy	total	good
15	A	(Temporal epilepsy)	Temporo-mesial	16	Lobectomy	total	Good (no seizures)
16	A	Headache Visual hallucinations	Occipital	16	Lesionectomy	Total	Immediate postop (Visual hallucinations) than Improvement
17	C	Headaches Seizures	Multiple (Rolandic cortex, temporal mesial)	45 34	Hematoma removal + lesionectomy (temporal)	Total	Immediate (seizures)
18	A	Staut epilepticus	Frontal	22	lesionectomy	Total	Good
19	A	Dysarthria + epilepsy	Temporal (neocortex )	20	lesionectomy	Total	Good

## 2. Brainstem topography

Cavernoma affecting the brainstem in our group was found in 4 patients. The same parameters were evaluated except for the timing and the surgical approach. (Table 2).

**Revelation mode:** The most frequent symptom was hemiparesis; present in all patients with this topography. Cranial nerves were affected in three patients.

**Topography:** At this location, pons was the most affected part of the brain stem and this fact was present in three patients. In two patients the cavernoma or its hematoma had a posterior extension to the fourth ventricle and in the third case, a lateral extension to the cerebellopontine angle was noticed.

**Size:** The sizes of lesions of this topography ranged between 12 and 35 mm. two patients had a 12 mm cavernoma.

**Surgical management:** All the patients were operated on in a delayed mode (more than two weeks); this was due to selecting the right timing of surgery but also to the clinical status of patients that most of them tolerated the hemorrhagic event. Two patients were operated on 2 months after bleeding. One patient was operated on 10 days after the hemorrhagic event. We operated on the patient at this time because his clinical status was not stable. The last patient was operated on after one month.

**Quality of resection:** The brainstem is one of the most eloquent compartments and the resection must be well guided and conducted with appropriate strategy. We have achieved total resection in two patients. the first case was in a pontic cavernoma with a posterior extension to the floor of the fourth ventricle. the malformation was removed through a Telovelar approach; the contents were both liquid (hematoma) and solid and the removal was total. We have achieved also total resection in a peduncular cavernoma completely solid. we have adopted a subtemporal approach with a partial transection of the free edge of the tentorium. The resection was subtotal in the rest of the cases (2 patients ). The first case was also a pontine lesion that had more of a posterolateral extension, just under the emergence of the trigeminal nerve. A retro sigmoid approach was performed and the resection was subtotal and this was due to the poor visualization of some blind angles in the surgical cavity. The fourth case was pontine cavernoma that had a posterior extension. The surgery using a Telovelar approach allowed us to remove the hematoma without finding the mass of cavernoma. In this case, the surgery was early because the patient was in a poor clinical status.

**Outcome:** We had two patients with a good postoperative status without neurological deficit or cranial nerve disorders. in one case with a pontine localization, he had worsening cranial functions (III + VI) that improved significantly during long-term follow-up in the last patient with a more extensive lesion in the pons and bulbar regions, we had a good post-operative status with regression of hemiparesis and we noticed a partial palsy of cranial nerves (V.VI.VII). Total recovery of this deficit was observed later on follow-up.

**Table 2:** Demographics of brainstem cavernoma (CN : cranial nerve)

Patient	Age /sex	revelation	Localiza-tion	Size (mm)	Timing (After bleeding)	Approach	Quality of resection	Outcome
1	26/F	CN deterioration Hemiparesis	Pontine	12mm	1month	Retrosigmoid	Subtotal	Facial numbness
2	32/F	CN deterioration Hemiparesis	Pontine	18mm	2 months	Telovelar	Total	III nerve palsy (improvement)
3	18/M	Multiple CN deterioration (V VI VII) Hemiparesis	Pontine with bulbar (posterior)	35mm	10 days	Telovelar	Hematoma removal	Good Partial improvement of CN
4	43/M	Hemiparesis	Cerebral peduncle	12mm	2 months	Subtemporal Transtentorial	Total	good

### 3. Medullary topography

We had 4 patients who presented this topography. two with a cervical location. and two others in the conus medullari (Table 3)

**Revelation mode:** For the patients with cervical cavernoma; one of them complained of hemiparesis and cervical neuralgia, and the second had paresthesia associated with neuralgia in the upper limbs in the second subgroup, both of the patients presented a neurological deficit of the lower limbs. One patient, had additional legs neuralgia the other had more, urinary disorders and hypoesthesia.

**Size:** The size of cavernoma was relatively small compared to other locations. It ranges between 12 and 15 mm.

**Surgical management:** The patients were operated on after their admission to our department. The surgical strategy was ordinary using a posterior midline approach.

**Quality of resection:** In this group of patients, the resection was total in all cases. during surgery, the cavernoma was resected in a monobloc fashion. This was followed by a careful inspection of the surgical cavity, great care was considered during the phase of inspecting the gliotic tissue around the lesion, and this was due to the presence of important pathways surrounding the lesion.

**Outcome:** For the patients with cervical cavernoma, in one case the postoperative course was uneventful with a good regression of the neurological deficit, and this was noticed during follow up. In the second case, the evolution was favorable except for some residual paresthesia managed with appropriate medication. In the conus medullaris location, the evolution was excellent in one patient but it was marked by an improvement in terms of pain but he kept a motor deficit requiring rehabilitation.

**Table 3:** Demographics of spinal cord cavernomas.

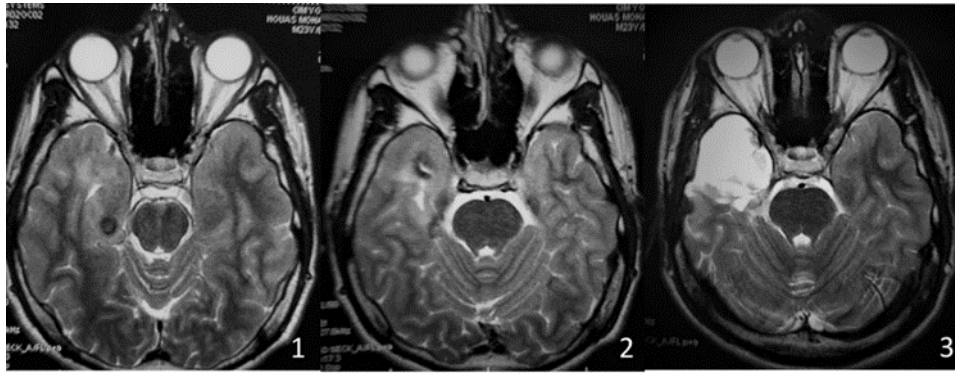
Patient	Age /sex	revelation	Localization	Size (mm)	Management	Outcome
1	A	Paraparesis	Conus medullaris	12	Total resection	good
2	A	Hemiparesis Cervical neuralgia	Cervical	13	total	Good
3	A	Paraparesia +sciatica	Conus medullaris	15	Total	Pain resolved Deficit remained stationary
4	A	Cervical neuralgia + paresthesia (right side)	Cervical	12	Total	Residual paresthesia

### Illustrative cases

#### Case1:

A male of 23 years old with a past medical history of treated hepatitis. The patient was referred to us by neurologists for management of disabling epilepsy; 5 to 10 seizures per day; which was not controlled by a double therapy. The seizures were complex, partial, and related to the mesial and neocortical cortex preceded with an aura and associated with a restriction of the visual field at the right side. The length of the seizure is 30 seconds and was followed by post-critic amnesia. The EEG confirmed the temporal origin of the seizures and a total suffering hemisphere from the epileptic developed network. The MRI (figure1) showed a double localization of cavernoma ; the first was neocortical, and the second was mesial affecting the body of the hippocampus(1+2). We have performed a classic temporal lobectomy that included the hippocampus and both lesions (3). The postoperative course was uneventful. We have obtained a good control of seizures during follow up and achieved a grade IA of Engel classification.

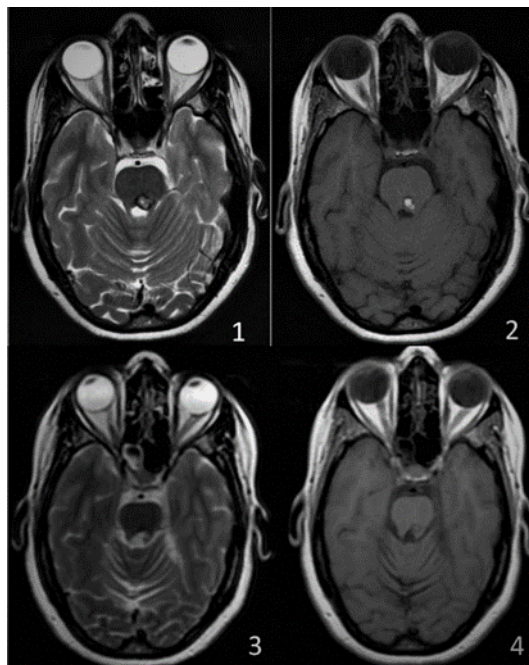




**Figure 1:** 1+2+3 MRI imaging of the first illustrative case;1: T2 AXIAL, shows a temporomesial cavernoma that affects the body of the right hippocampus.2: T2 AXIAL; a retro polar cavernoma on the right side .3: T2 AXIAL , the postoperative image of a temporal lobectomy including the resection of both cavernous malformations.

### Case 2:

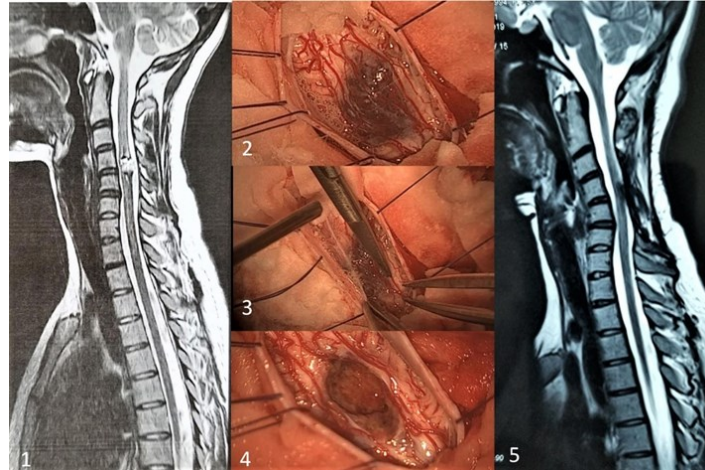
A 32 years old female started to present a left hemiparesis that was neglected by the patient. A few days before her first consultation, she had a partial third nerve palsy moreover she remained conscious and kept a good quality of life. The MRI (figure 2) showed a posterior pontine projecting cavernoma. The malformation was subependymal in the pontine triangle of the fourth ventricle, with left lateralization. the contents of the lesion were acute microbleedings. We have planned to operate on the patient three weeks after the hemorrhagic event. the lesion was amenable for surgery and the patient was symptomatic. The patient had a Telovelar approach for microsurgical removal of the malformation after the opening of the appropriate arachnoidal plans and the progression to the lesion in the ventricular floor. A slight discoloration of the ependyma guided the entry point to the malformation, where a small amount of blood was removed. Further exploration allowed us to remove the resting cavernoma with standard microsurgical technics and great care and consideration to the surrounding tissue. Careful hemostasis of the surgical cavity was then secured at the end of the surgery. In the immediate postoperative course, we have noticed a slight worsening of nerve palsy. At long-term follow-up, a clear improvement of the cranial palsy and the motor deficit with appropriate rehabilitation. The postoperative MRI showed complete resection of the cavernoma without any complications.



**Figure 2 :** Preoperative (1.2) and postoperative (3.4) MRI: 1: axial T2 sequence showing a pontine cavernoma projecting posteriorly to the floor of the 4<sup>th</sup> V. The lesion is heterogeneous with a hypointense ring (popcorn aspect) surrounding the mass. 2: A predominantly hyperintense testifying the acute nature of blood. notice the difference between the distances of dorsal aspect of the cavernoma and the floor of the ventricle, where in T1 looks thicker. T1 is the reliable sequence to plan on surgery. 3 (axial T2) 4(axial T1) postoperative imaging showing a complete resection of the malformation.

**Case 3:**

A female of 26 years old who was complaining for more than weeks of sensory disorders. The paresthesia and slight weakness were affecting the right hemi body. she had more cervicobrachial neuralgia that affected her daily activity. The MRI (figure3) showed a typical radiological aspect of a cervical cavernoma projecting toward the posterior columns of the cervical medulla, at the level of C3-C4. A posterior approach was performed through a targeted laminectomy after the opening of the dura and arachnoid, the microscopic view (image 2) shows the dorsal aspect of the cavernoma with a black to a reddish lesion within a swollen appearance of the spinal cord. A plan was developed between the lesion and the medullary tissue (image3) using micro scissors, bipolar and suction tube. At the end of the cavernoma resection (image 4), the hemostasis was performed using classic tools. We had a good long-term follow-up results with regression of the preoperative symptoms. The control MRI showed a total resection of the cavernoma without any abnormal features in the spinal cord.



**Figure 3 :** Preoperative MRI (1) , per operative microsurgical views (2+3+4) , postoperative MRI (5): 1: sagittal T2 sequence showing a cavernoma at the level C3-C4 ; with a posterior projection . the presence of the hypointense halo of hemosiderin around is typical for these malformations.2: per operative microsurgical view, showing the dorsal aspect of the cavernoma and the aspect of a swollen spinal cord at the level of the lesion .3 : beginning of microsurgical resection .4: the final view of the surgical cavity after the removal and a careful hemostasis .5: sagittal T2 showing a complete resection , without other abnormal signals in the spinal cord.

**Discussion**

CM are benign vascular abnormalities that affect the central nervous system by a percentage of 5 to 10 % (17). Despite this low frequency, serious neurological deficits such as seizures and hemorrhage are attributed to their bleeding event. The incidence of these lesions is up to 0.8 % of the general population(18), while in autopsies studies, it's estimated to be 0.4%(19). They are called also "angiographically occult malformation" and are marked in their architecture by enlarged and abnormal venous channels, without the interposition of brain parenchyma inside of the lesion's mass.

Both familial and sporadic forms were described in the occurrence of these lesions. In sporadic forms, the lesion is unique and no history of familial disease is reported by the patient. Most of our patients had a sporadic form. We had 24 patients presented with this form. One work(20), confirmed that during the onset of the familial form; Three protein-encoding genes (CCM1, CCM2, and CCM3) were identified to be the origin of this disease. A syndrome of familial CM was defined by the existence of several cavernoma for the same patient, the occurrence of cavernoma in at least two members of the same family, or finally the appearance of a mutation of one of the three previously mentioned genes(10, 12).

We had three patients with multiple cavernoma (double locations) in the supratentorial compartment of the brain. The main affected regions were the temporomesial and the Rolandic cortex. The age of these patients ranged between 17 and 23 years. We had two patients aged 17 years that could be included in the pediatric population. The serious clinical presentation of cavernomas is hemorrhagic stroke with a rate of 0.25 to 0.7% per year per person that didn't have a previous history of the hemorrhagic event(21).

The most frequent presentation in brain localization was seizures. In one study (10), epilepsy is encountered in about 55 %. We had 13 patients that presented this symptom. In this subgroup, epilepsy was the only symptom in 9 of them and associated with other symptoms such as dysarthria and headaches. We had two patients (10%) that had static cerebellar syndrome, and in both cases, the cavernoma was located in the superior vermis.

Three patients were complaining of headaches in a chronic form, and in all cases, this symptom was associated with seizures in two of them and visual hallucination in the third patient. One patient (5%) had a ventricular hemorrhage, and presented with consciousness disorders in a more alarming profile. This rate is much lower than the one that was reported in another work (25%)(22). We had one patient with a previous history of bleeding several years before the patient was evaluated during follow up and surgery was decided as a modality of treatment.

In brainstem topography, all of our patients had neurological deficits. The deterioration of the cranial nerve was noticed in 75% of cases. The clinical manifestations were associated together, except in one case where hemiparesis was the only symptom. The hemorrhagic event in this location is a capital parameter that must be considered during the process of decision-making in patients.

One author(23) established a system for grading brainstem cavernoma after the analysis of more than 300 patients operated with different localizations over the central nervous system. The scale was composed of two scored variables; the location, where the infratentorial, spinal cord and basal ganglia had a score of 2, whereas the supratentorial had a score of 1. The other variable was the focal neurological deficit (presence: 1, absence :0). They concluded to three grades from 1 to 3. 62 % of patients were included in this group, with a favorable postoperative outcome in 87% of them. 19% of patients belonged to grade 2, of the 79% had a good recovery at follow-up. grade 3 was the group where patients had a high risk of disability after surgery(23) to get more precisions and solve limitations of the previous study, another author(24) proposed a scale with five predictors that are; size (cm), crossing axial midpoint, developmental venous anomaly, age (years), and hemorrhage. the important utility of this scale is that it could be used by neurosurgeons as a tool in the selection of the right candidate for surgery with acceptable outcomes. They concluded also that malformations that cross midline are deeply located close to the tracts and nuclei and require more dissection for their complete resection and this will be a significant factor of morbidity (24). The utility of the scale was to help the neurosurgeon in the process of the right selection of patients for surgery. Both of these scales were not applicable in our present work because of the lack of data in the old medical files.

Radiological evaluation for these malformations was first reported in a study conducted on a familial form (4). The work was about the natural history including; interviews, examinations and serial MRI imaging of patients. The authors concluded that familial cavernomas are not stable lesions, requiring a well-conducted follow-up and their appearance is in a constant cycle of change, with acute and resolved hemorrhagic events. The other changing parameters are the number and the size of lesions. They reported that prophylactic surgery is not recommended for asymptomatic patients despite this dynamic profile of cavernous malformation. The classification was made up on the aspect of cavernoma of T1 and T2 sequences and defined four types from the first one where the hemorrhage is subacute to the fourth type where we can notice multiple microhemorrhage. recently, highly defined imaging technics were added to the preoperative evaluation; DTI (Diffusion Tensor Imaging) estimates the exact course and connectivity of different white matter tracts. The topography and distribution of fibers around the cavernoma will define also the appropriate surgical approach to get a complete resection(25).

A center of several nuclei and a zone of transition for nervous tracts, bleeding in the brainstem is almost always symptomatic (up to 100%) as reported in one work (26). the age of patients ranged between 18 to 43 years old. The pontine portion of the brainstem was the most affected in three patients (75%) and the last case (25%) was a lateral peduncular cavernoma. The reported percentages about the repartition of these malformations by some authors were; 14 to 15% in the mesencephalon, 42 % in the pons, 15% in the medulla(27). The size of cavernomas ranged between 12 to 35 mm. A crucial step in the management of brainstem cavernomas is the timing and the indication of surgery as it was debated in several works (28, 29).

The timing of surgery for brainstem cavernoma was previously debated in the literature(28-30), and the period of four to six weeks when the patient tolerates the bleeding is the appropriate one. we have operated on our patient between one and eight weeks after the symptomatic hemorrhagic event. Our attitude is similar to the one adopted by these authors. In a large series of patients, the authors (31) recommended that surgery for cavernoma in this location is beneficial within 6 weeks after the last bleeding. In another work(28), surgery was performed earlier in a delay of 3 weeks (ranged between 4 and 90 days) with acceptable morbidity and good outcome.

The surgical option is selected for symptomatic and accessible lesions; the term accessible includes all malformations that present pial or ependymal surface. It could be also an effective modality for these cases; patients with evidence of multiple bleedings and progressive neurological deficit, and lesions close to the cisterns or the fourth ventricle even if it's asymptomatic (32). In one work(33); two other parameters are considered for surgery, the extension of acute hemorrhage outside the capsule and the presence of a large shift effect of the malformation. Our patients presented approximately all of the previous characteristics; they were symptomatic, with a previous history of bleeding, and had surgically amenable cavernomas.



The surgical approach was selected after analyzing the position of the lesion. We consider first which part of the brainstem is affected, evaluate the depth of the malformation in the brainstem in a multiplanar way and finally estimate the closest pial or ependymal surface to the lesion. We have used the Telovelar approach in 50% for pontine posteriorly projecting cavernoma. One lesion had ependymal contact and the other was sub ependymal with a discoloration of the floor of the fourth ventricle, the lesion was mainly a hematoma with a small amount of tumor in the depth of the surgical cavity in one case, we had troubles with the removal of peduncular lesion close to the trigeminal entry zone. The exposure was performed through a retrosigmoid craniotomy. This was because of the presence of multiple blind spots that limited the total resection. The subtotal removal of the malformation was confirmed also in the postoperative imaging. The last case was a peduncular lateral projecting cavernoma with a pial surface. We have performed a subtemporal transtentorial approach. The lateral surface of the peduncle was entered in the lateral mesencephalic zone. We have also noticed an abnormal coloration of the pia and the cavernoma was under this surface and the resection was accomplished as elsewhere and totally. One of the effective methods of selecting the appropriate approach was the "two-points method" described in previous work (34). It requires the use of the MRI sections and the most appropriate image of the cavernoma. The first point is placed in the center of the lesion and the second point is in the margin of the lesion where it comes close to the ependymal or pial plans. A virtual line connecting the previous points and extending to the brain surface is made. The surgical approach is then selected according to the direction of the final line (34).

The postoperative course in our patient was marked by the dysfunction of cranial nerves functions that were already present during the last bleeding in almost all patients. We had facial numbness in one case of a pontine cavernoma which the origin could be the trigeminal nuclei or fibers manipulation. In two posterior pontine projecting cavernomas, we had partial palsies of the third nerve in one case, and a significant improvement of the nerve function and this was due to the removal of a large hematoma. At long term follow up, all patients improved their functions in all aspects. We didn't have important disabilities or mortality.

We had four cases of spinal cord cavernoma, three of them were females and their ages ranged between 26 and 62 years old. They had a similar distribution along the spinal cord where the cervical spine and conus medullaris were affected equally. The dorsal spinal cord was the most affected by a percentage of 55.2% (35). The patients of this group presented; paraparesis and sciatica for the conus medullaris location and they had both of them cervical neuralgia, paresthesia, and a slight hemiparesis for the cervical location. The revelation of these cavernous malformations in patients was by; motor deficit (63%), sensory disorders (65%), pain (27%), and 11% complained of bowel or bladder troubles (36).

The size of cavernomas was relatively small ranging from 12 to 15 mm. despite the reduced size of lesions, patients were symptomatic and this was due most probably to the highly eloquent small section of the spinal cord as it was reported in one work (37). The surgical option was planned for symptomatic patients with surgically amenable lesions. surgery aims to stop further neurological decline or other episodes of bleeding. We didn't have asymptomatic cases in this location.

All patients were operated on using a posterior approach through a targeted level laminectomy allowing the exposure to the upper and lower intact limits of the cavernoma. When the lesion had a pial surface, the dissection is started at this level with maximal precaution to the spinal parenchyma during the peritumoral dissection to reduce the risk of postoperative neurological impairment. In other cases where the lesion stayed under the pia, a well guided micromyelotomy is performed to gain access to the dorsal aspect of the lesion and the same principles of resection were applied as elsewhere. Anteriorly located cavernomas presents more challenge in their microsurgical resection, and this for their deep position and the proximity to the motor's fibers of the spinal cord. The approaches through the body vertebrae will induce instability and necessitate fixation. At the dorsal level, the difficulty is the ergonomic exposure of the ventral aspect of the spinal cord that imitates the watertight closure of the dura. Most of these surgeries are completed by the implantation of a chest tube in postoperative care. This maneuver can be the origin of an imperfectly closed dura(38). One team described a minimally invasive approach named the anterior to dorsal root entry zone myelotomy in a population of ten patients with ventral cavernoma in the cervical and dorsal levels of the medulla. A gross total resection was achieved in all of them with an acceptable postoperative result (39).

For the postoperative course in this subgroup, patients had good outcomes with a total improvement of paraparesis and hemiparesis and cervical neuralgia in two patients. in one case a conus medullaris location, we had a total regression of the lower limbs pain but the motor deficit improved significantly but not totally after additional physical therapy. in the last case of a cervical cavernoma, we had good improvement of symptoms but the patient had medication to control the residual numbness in the upper limb. Surgery remains a good option for the management of cavernous malformations in this location with more than 80% improving their preoperative symptoms after surgery, and 15.4% that stayed in the same preoperative status(40).

### Limits of the study

We have established some limits in our work. firstly, the heterogeneous aspect of the preoperative data; in radiological evaluation with the adequate protocol and sequences, limited the true grading of cavernoma in the brain and, brainstem and spinal cord locations using the appropriate scores as Zabramski and Lawton's group scales.

The study didn't include asymptomatic cases that were followed with serial imaging that could cover more the aspect of the natural history, especially for the spinal cord topography. The lack of precision about the long-term follow-up after surgery because of the absence of patients in their periodic clinical evaluations. The retrospective aspect of the study is also an additional factor that may limit the results.

## Conclusion

Central nervous system cavernoma are surgically cured lesions in an experienced team. The lack of other options for management favors more research in the anatomical and microsurgical aspects of these malformations especially the deep-seated ones in basal ganglia and brainstem to perform safe and effective removal. Much debate was raised around the natural history of these lesions which is another field to explore by larger and multicentric prospective studies. The results of surgery are acceptable in our work and allowed a good quality of life for patients after surgery. The preoperative and modern technology in monitoring will ameliorate the quality of resection and reduce the morbidity related to this field of microsurgery.

## Conflict of Interest

The authors declare no conflict of interest.

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