Multimodal treatment for temporobasal arteriovenous malformation - Case report

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Abstract

Background: Arteriovenous malformations (AVMs) are congenital lesions requiring multimodal approach.

Method: We report a case of a 25 years old woman, with ruptured temporobasal AVM, who required multimodal treatment, emphasizing on treatment options, advantages and disadvantages of each treatment chosen, encountered difficulties, technical considerations and outcome.

Results: The patient, admitted for the first time in comatose state, underwent emergent surgery with evacuation of an intraparenchymatal hematoma and decompressive craniectomy. After neurological recovery, the patient was thoroughly investigated and positive diagnosis of left temporobasal AVM was established. The patient underwent surgery with subtotal resection of the AVM, followed by Gamma knife stereotactic radiosurgery of the residual nidus. The outcome was favorable.

Conclusions: AVMs need complex treatment performed in a multidisciplinary team. Surgery is the treatment of choice in management of the AVMs. Gamma knife stereotactic surgery is required if a residual nidus is left in place following surgery. Definitive treatment in AVMs is mandatory because of the high risks of hemorrhage with high morbidity and mortality.

Keywords: arteriovenous malformation, AVM surgery, residual nidus, stereotactic radiosurgery

Introduction

Arteriovenous malformations (AVMs) are congenital lesions composed of a complex tangle of dysplastic arteries and arterialized veins connected by shunts. Within the abnormal conglomerate of vessels, called nidus, blood is drained through feeding arteries directly into draining veins, without any capillary bed. Arteries have a deficient muscularis layer and red draining veins are dilated and contain high flow oxygenated blood.

AVMs were described for the first time by Luschka and Virchow in the mid 1800s and Olivecrona performed the first surgical excision of an AVM in 1932. (1)

It is difficult to estimate the real incidence of AVMs, it usually ranges between 0.15 and 3%. (18, 26) Prevalence is
0.14%. (1, 9) AVMs represent 6% of intracranial lesions. The average age at diagnostic is 33 years, and 64% of AVMs are diagnosed before age 40. (9, 20) AVMs are the main cause of hemorrhagic strokes in young patients. (8)

The two main clinical presentation forms are rupture of the AVM with consequent hemorrhage (intraparenchymatal, intraventricular or subarachnoidal bleeding) and seizures. Other clinical manifestations are progressive neurological deficit secondary to the vascular steal phenomenon from de adjacent brain, hydrocephalus due to venous hypertension in the draining veins, neurological deficits secondary to mass effect of an enlarging AVM and increased intracranial pressure.

Treatment options of AVMs consist of surgery, endovascular embolization, stereotactic surgery or multimodal approach. Treatment must be individualized according to neurologic state at admission, general state, co-morbidities, age, characteristics of the AVM and therapies available in the hospital of admission.

**Method**

We report a case of a left temporobasal AVM treated into the Forth Department of Neurosurgery. We reviewed medical records, imaging, treatment and follow-up. Patient BS, woman, 25 years old, with no significant previous history prior current episode, was transferred from another department on the 29th of October 2009 with the following diagnostic: ruptured profound left temporal AVM.

**History:** sudden onset 8 days before admission in our department, with headache, vomiting and impaired consciousness. Patient was admitted in emergency in another department of neurosurgery with comatose state, GCS 6 points and right hemiparesis. Emergency CT scan showed a left frontotemporoparietal intraparenchymatal hematoma. Patient underwent emergent surgery, hematoma was evacuated and decompressive craniectomy was performed. Two days after surgery a 4 vessel angiography was performed which revealed a profound left temporobasal AVM. Patient was redirected to our department for further treatment.

On admission the patient presented impaired consciousness, GCS 9 points (M5, V2, O2), had right hemiparesis, hyperactive deep tendon reflexes right > left and bilateral Babinski sign. The cerebral CT scan revealed postoperative left temporal hypodensity, intraventricular hemorrhage (within both lateral ventricles) and decompressive craniectomy (Figure 1).

Cerebral IRM showed a left temporohippocampal AVM, with consequent involvement of left internal capsule and thalamus, with feeding arteries from left middle cerebral artery and left posterior cerebral artery and venous drainage into left transverse sinus (Figure 2).

Four vessels cerebral DSA (digital subtraction angiography) showed a left capsulothalamic AVM grade III Spetzler-Martin, with nidus measuring 3 cm in diameter, with feeding arteries from talamostratic arteries from left posterior communicating artery and left posterior cerebral artery, anterior choroidal artery and perforating branches from left segment M1 of the middle cerebral artery (Figure 3).

Pulmonary X-ray and electrocardiography were normal.
Figure 1 Cerebral CT scan, left temporal hypodensity, intraventricular hemorrhage, decompressive craniectomy
Figure 2 Cerebral IRM T2, left temporohippocampal AVM involving left internal capsule with feeding arteries from left middle cerebral artery and left posterior cerebral artery.

Figure 3 Four vessels DSA, left internal carotid artery, left capsulothalamic AVM with nidus measuring 3 cm in diameter, with feeding arteries from tamoostriate arteries from left posterior communicating artery and left posterior cerebral artery, anterior choroidal artery and perforating branches from left segment M1 of the middle cerebral artery.
Results

She received Dexamethasone 16 mg/d, Mannitol 20% 250 ml/d, Furosemid 40 mg/d, antiepileptic drugs, analgesics and intravenous hydration.

Neurological outcome was favorable, the patient regained consciousness, GCS 14 points, had severe right hemiparesis and dysphasia.

In order to determine whether intraarterial embolization was a valid option we asked for an interventional neuroradiology consult which contraindicated embolization because the AVM had no pedicles that can be catheterized with microcatheters of the equipment.

Gamma knife stereotactic radiosurgery was also contraindicated because the nidus includes the middle cerebral artery and comes into contact with left optic radiations.

Pre-anesthetic consult found a risk score ASA IV and anemia (Hg=8.6 g/dl).

On the 9th of November 2009 the patient underwent surgery. The patient was position supine, with roll under her left shoulder. The head was turned 45° to the right. We reopened the wound on the previous skin incision for left temporal bone flap and we incised the neodura mater in a stellate fashion. We performed opening of the left sylvian fissure, with visualization of the left middle cerebral artery and sylvian artery trifurcation. A tangle of vessels (nidus) was discovered in close vicinity with middle cerebral artery, surrounding it. The nidus was dissected free from the middle cerebral artery, which was left intact. Left internal carotid artery and left anterior choroidal artery were found and spared. A vascular clip was put on the feeding artery arising from left posterior cerebral artery. Subtotal removal of the AVM was performed, the part situated in the vicinity of the left middle cerebral artery. We copious irrigated the operative field with normal saline and performed carefully hemostasis. At the end of the operation normal brain pulsation were noticed. We performed wound closure with watertight duraplasty with pericranium and anchoring of dura mater, epidural drain, replacement and fixation of the bone flap conserved from previous surgery and skin suture.

The patient was extubated and awakened in the operating room. Immediate postoperatory examination revealed: efficient spontaneous breathing, right hemiparesis and dysphasia. The patient was taken to intensive care unit. The epidural drain was pulled out after 48 hours. During the following days dysphasia slowly recovered. Postoperatory outcome was favorable, the patient regained consciousness, GCS 15 points, she had right hemiparesis and presented no aphasia.

Histopathological exam confirmed the diagnosis of racemous hemangioma (arteriovenous malformation).

Postoperatory CT scan was normal (Figure 4).

Postoperatory 4 vessels DSA showed the residual nidus (Figure 5).

On 2nd of March 2010 (5 months after surgery) the patient was admitted for stereotactic irradiation of the residual nidus. Gamma knife stereotactic radiosurgery was performed, using a marginal dose of 18.5 Gy at the 60% isodose line, on a target volume of 1.4 cm³. Total irradiation dose was 0.7 Gy (1.3 J). Postirradiation outcome was favorable.

Four vessels DSA performed in June 2011, 16 months after stereotactic
radiosurgery, revealed complete obliteration of the residual nidus (Figure 6). Long term follow-up CT scan showed a normal postoperative aspect (Figure 7).

Neurological examination found right hemiparesis and no aphasia. The patient presented no seizure under Fenobarbital 100 mg/d. She was included in a kinetotherapy intensive program.

Figure 4 Cerebral CT scan, normal postoperative aspect

Figure 5 Four vessel DSA, left internal carotid artery, residual nidus
Discussions

The patient had a typical sudden onset of ruptured AVM. Hemorrhage is the most common form of presentation, 50-70% of patients harboring such malformations present with bleeding. (9, 12, 24) Bleeding occur most commonly intraparenchymatal, intraventricular, usually concomitant with intracerebral hematoma as a result of rupture into ventricles or pure intraventricular or rarely subarachnoid or subdural. (9) The patient had no history of seizure. Usually patients having seizure are investigated and the AVM is diagnosed before rupture.

Increasing age, initial rupture of the AVM, deep brain location and exclusive deep venous drainage are independent predictors of subsequent hemorrhage. Annual hemorrhage rates on follow-up ranged from 0.9% for patients without bleeding to 34.4% in people with ruptured and deep AVMs and with deep venous drainage. (6, 7, 27) Hemorrhage carries a 10% mortality and 30-50% morbidity rates with each bleed. (9)

The patient was admitted in emergency in another neurosurgical department. The first surgery was performed in order to save her life, this is why only evacuation of the intraparenchymatal hematoma and decompressive craniectomy for brain swelling were performed.

Although the postoperative outcome was favorable, and the patient had no further vital risk, an ethiopathogenic treatment was mandatory. (6) She was transferred in our department for definitive treatment. The need for an ethiopathogenic treatment is sustained by risks of further rupture.

The risk of hemorrhage from an AVM is 2-4% per year, therefore the risk of bleeding over the entire life is very high. (9) The risk
of bleeding is calculated with the following formula:

| Risk of bleeding (at least once) = 1 – (annual risk of not bleeding)^expected years of remaining life |

The annual risk of not bleeding is 1-annual risk of bleeding.

In our patient aged 25, with expected 52 years to live, for an average risk of hemorrhage of 3% per year, the total risk of bleeding at least once is 79%. (9)

More, the risk of bleeding during the first year after primary rupture is 6%. (16) The risk deceases over the next years, but remains significant for decades.

Risk of bleeding is higher in AVMs with previous rupture, deep and infratentorial locations. (12, 19)

At admission in our department the situation is different, because her life is not in danger and she could be thoroughly investigated prior definitive treatment.

Treatment options of AVMs, all available in our department, consist of surgery, endovascular embolization, stereotactic radiosurgery and multimodal treatment. (6)

In 1986 Spetzler and Martin proposed a grading system of AVMs, based on size of AVM, eloquence of adjacent brain and pattern of venous drainage. (25) Spetzler-Martin grading system is helpful in predicting surgical risk. (9, 16, 29) While in patients harboring grade I and II surgical risk is low, patients with grade III AVMs represent a more challenging decision-making dilemmas, because they carry a significant higher surgical risk. (16) In the algorithm of decision-making we must take into account other factors such as ruptured vs. nonruptured AVM, clinical presentation, patient’s age and co-morbidities.

Embolization can be used as single treatment or as neoadjuvant for surgery or stereotactic surgery. In order to be done it requires an interventional neuroradiology department with trained personnel. Embolization is a minimally invasive procedure and when used as a neoadjuvant technique it facilitates surgery, by reducing the volume of the malformation, decreasing the intraoperative bleeding, occluding deep or difficult to reach feeding arteries and associated aneurysms outside the operative field. (10, 28) Using embolization alone the rate of complete occlusion is only 10%, and even in completed occluded AVMs the recanalization reaches 50%, but endovascular techniques reduce the volume of the malformation with 75%. (10) Surgery must be performed within 3-30 days after embolization. If patient becomes symptomatic and he has no intracranial lesion requiring emergent surgery, the operation is postponed until he recovers. (9) Morbidity and mortality rates following embolization are low. (5, 11, 13, 14) Mortality is 1% and morbidity is 3.9%. (5, 11, 22) Long-term permanent neurologic deficits are encountered in 8.6% of patients. (14) Immediately following procedure, significant neurological complications occurred in 2.7-7.1%, yet a majority of deficits improve or resolve over time. (22, 28) Embolization carries a procedural risk related to AVM’s grade and number of occluded branches. (14)

Unfortunately microcathethers of the equipment were unable to catheterize the feeding pedicles. The AVM had feeding arteries from talamostriate arteries arising from left posterior communicating artery and left posterior cerebral artery, anterior choroidal artery and perforating branches from left segment M1 of the middle cerebral artery. Catheterization of such small branches requires very small catheters
and carries a significant risk of improper embolization of the parental artery with post procedural morbidity. The answer to this problem is acquisition of smaller catheters.

Stereotactic radiosurgery is used in small AVMs, with nidus measuring 2.5-3 cm. Stereotactic radiosurgery include Gamma knife radiosurgery, proton beam radiosurgery, and linear accelerators (LINAC). Advantages of stereotactic radiosurgery are: it is a non invasive procedure and it can be done in an ambulatory patient.

Principle of radiosurgery consists of directing radiation with narrow beam particles, consisting of 201 converged sources, given in a single session, to a predetermined target volume, with minimum risk of damage to surrounding brain.

Stereotactic radiosurgery causes gradual obliteration of the nidus. Radiation of vessels causes proliferation of smooth-muscle cells and increases extracellular type IV collagen producing, which leads to progressive stenosis and obliteration of the nidus and, in the end, to cellular degeneration and hyaline transformation. (4, 23) Because of the delayed obliteration of AVMs after radiosurgery, comprehensive long-term management and observational strategies are necessary. The risks associated with AVMs, including the risk of bleeding, persist during this period of time, therefore until obliteration the lesion cannot be considered cured. Obliteration of the AVMs following stereotactic radiosurgery occurs late, after 1-3 years following initial procedure. (4, 9) Imaging follow-up are recommended at six month intervals for the first three years to assess the effect of radiosurgery on AVM. Another disadvantage of stereotactic irradiation is the fact that it is limited to lesions measuring maximum 3 cm in diameter.

If obliteration does not occur after 3 years, the irradiation can be safely repeated. (15)

Dose planning is based on location and volume of the AVM and relation with the surrounding structures. Marginal doses typically range from 16 to 25 Gy in a single fraction.

Although the nidus measured 3 cm in diameter Gamma knife stereotactic surgery was contraindicated because of the relation of the nidus with the middle cerebral artery and left optic radiation. Inclusion of the middle cerebral artery into the target volume can lead to radiation artery wall lesions with stenosis and occlusion. Also vicinity of optic radiations limits using stereotactic surgery. Tolerable dose to the optic radiation is 8 Gy, a dose similar to anterior optic pathways. (17)

Surgery is the treatment of choice, because if complete resection can be performed provides cure of the malformation. (7) Total resection cannot be always done. Causes that can prevent total excision are: anesthetic problems during surgery (patients develop cardiocirculatory instability, e.g. bradycardia, hypotension), high grade Spetzler-Martin AVMs (staged surgery is a solution for these cases), portion of the nidus located in eloquent areas, deep nidus with deep feeding arteries that cannot be approached, preoperative rupture of the AVM (hematoma can compress the lesion, angiography is occult or it does not show all malformation, and after hematoma removal or clot dissolution the AVM reopens), intraoperative rupture (due to accidental early vein occlusion with severe life-threatening bleeding), difficult anatomy with passage vessels supplying
normal brain, which must be spared, included into a complex AVM.

Surgery for AVMs must follow several principles. Always perform a large bone flap, providing a wide exposure, completely circumscribing the AVM and vessels. Feeding arteries must be occluded first to hinder blood flow into the malformation. Before occlusion arteries must be carefully inspected. Passage arteries must be spared, because they supply normal brain, and improper sacrifice causes ischemia and postoperative neurological deficits. Identification of passage arteries can be done by dissecting the nidus controlateral to vascular supply, and arteries emerging from the AVM are followed back to the nidus. Draining red veins, containing arterialized blood should be sacrificed last. If a vein must be occluded in order to proceed with removal, it should be temporary clipped first and malformation observed. If swallowing of the AVM occurs the clip must be removed and vein must be kept until malformation is completed devascularized. Small vessels are occluded using bipolar electrocautery, but larger ones necessities vascular clipping. Dissection of the AVM must be done into the gliotic plane circumscribing the nidus. Previous rupture facilitates dissection because it creates a plane between malformation and adjacent brain. AVM have a conic form. The deep part of the nidus is most difficult to remove because it contains small high-pressure vessels. When the deep part is approached an attempt to recanalization of the AVM occurs. (16) After nidus removal is done, carefully hemostasis is achieved at a postoperative estimated blood pressure.

Difficulties of the present case are represented by passing middle cerebral artery. The sylvian fissure was split, middle cerebral artery trifurcation was identified, and the main truck was followed back to the nidus. Location of the AVM into the left temporal lobe poses a risk for worsening language function. Avoidance of language area can be done using functional MRI, electrocortical stimulation mapping and optical imaging of intrinsic signals. (2, 3, 21) In patients with high risk of language deterioration awake craniotomy with intraoperative language monitoring can be done. (3) Our patient presented dysphasia as a result of rupture of the AVM. She presented aphasia immediately after surgery due to postoperative edema, but she fully recovered language function. This particular location is prone for developing right inferior homonymous quadrantanopsia because the AVM comes in close vicinity with optic radiations. Our patient had no preoperative or postoperative visual field deficits. Right hemiparesis, in this particular location, can occur because the AVM is in close relation with the left internal capsule and during resection of the deep portion the capsule can be damaged or secondary to arterial occlusion. The AVM has feeding arteries from talamostriate arteries from left posterior communicating artery and left posterior cerebral artery, anterior choroidal artery and perforating branches from left segment M1 of the middle cerebral artery. Improper occlusion of passage arteries supplying normal brain results in motor deficit. In our case although all arteries of passage were spared, the motor deficit was a prior sequel from the first rupture of the malformation with intraparenchymatal capsule-lenticulo-thalamic hematoma. Internal capsule damage explains why the patient did not improve after hematoma removal. Other complications following
surgery such as rebleeding from residual nidus, normal perfusion pressure breakthrough, retrograde venous occlusion, ischemia, vasospasm, seizures or hydrocephalus did not occur in our patient. Because total removal of the AVM was not achieved with surgery we reconsidered adjuvant therapy. Further treatment is still necessary after subtotal removal because the risk of hemorrhage form residual nidus remains. By removing the part of the AVM circumscribing the middle cerebral artery Gamma knife stereotactic surgery became a valid therapeutic option. Resection of the part close to middle cerebral artery made it possible to exclude it from the target volume.

The particularity of the presented case is presence of residual nidus after surgery, needing adjuvant Gamma knife stereotactic surgery.

Conclusions
AVMs need complex treatment performed in a multidisciplinary team, including neurosurgeon, interventional neuroradiologist and stereotactic neurosurgeon. AVMs should be addressed to specialized neurosurgical centers, where all treatment modalities are available and multimodal approach can be done for optimal results. Surgery is the treatment of choice in management of the AVMs. Reconsideration of treatment options for residual nidus after surgery is recommended. Gamma knife stereotactic surgery is required if a residual nidus is left in place following surgery. Definitive treatment in AVMs is mandatory because of the high risks of hemorrhage with high morbidity and mortality.

Abreviations
AVM - arteriovenous malformation
DSA - digital subtraction angiography
GCS - Glasglow Coma Scale

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References


