Spetzler-Martin Grade III Arteriovenous Malformation of the Brain Associated with Cerebral Vascular Aneurysm: Report of Three Cases

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Summary. The AVM is a rare congenital developmental anomaly of cerebral vessels, occurring in less then 0.01% of the population. The incidence of AVM associated with aneurysms is approximately 8.9% in patients with AVM. Surgery, endovascular embolization and radiosurgery or combined modalities are used in clinical practice of AVM management. We present three unusual cases of high flow AVM associated with intracranial aneurysms. These patients were treated only surgically. Possible causes, clinical signs, management modalities and outcomes are discussed.

Keywords: cerebral aneurysm, cerebral arteriovenous malformation, surgical resection.

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INTRODUCTION

A number of authors have described cases of coexisting cerebral lesions, such as arteriovenous malformation (AVM) and tumors, and aneurysms of cerebral vessels in the same patient [1–10]. Most investigators have analyzed epidemiology, causes, pathophysiology, clinical signs, diagnostics and management problems of this complex pathology of cerebral vessels [11–13].

The AVM is a rare congenital developmental anomaly of cerebral vessels, occurring in less then 0.01% of the population [14]. The incidence of AVM associated with aneurysms is approximately 8.9% in patients with AVM [15].

Surgery, endovascular embolization and radiosurgery or combined modalities are used in clinical practice of AVM management. In our opinion, AVM surgery is very individualized and requires decision making by a neurosurgeon with significant experience in this complicated neurovascular lesion.

We present three unusual cases of high flow AVM (Spetzler-Martin grade III) associated with intracranial aneurysms. These patients were treated only surgically. Possible causes, clinical signs, management modalities and outcomes are discussed.

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ILLUSTRATIVE CASES

Case 1

A 40-year-old woman presented with a month long history of epilepsy. A magnetic resonance imaging (MRI) scan demonstrated AVM in the right parieto-occipital region. The right common carotid artery angiography and computer tomography angiography ((CTA), Fig. 1a) showed a high flow AVM (Spetzler-Martin grading scale III (size (S) 2, eloquence (E) 0, venous drainage (V) 1)) in the right parieto-occipital region and three aneurysms (Fig. 1b). AVM was fed by the middle cerebral artery (MCA) and partially by the posterior cerebral artery (PCA). The blood passed from AVM to the right sigmoid sinus, superior sagittal sinus and one vein drained to the Galenic system. Two aneurysms were detected in the right MCA circulation: first small in the proximal part of M1 and second in the bifurcation of MCA, of irregular form with double sac (flow-related). Third aneurysm was diagnosed in the communicating segment of the right internal carotid artery (ICA) (not flow-related).

It was decided to operate this patient in two stages and to excise AVM firstly. During surgery, a high flow AVM was detected. Feeder arteries were from the MCA and a few not significant feeders from PCA. Draining veins were coagulated finally. Postoperative angiography demonstrated AVM excision, MCA and ICA filling normally. The patient did very well after the operation and went home till the next stage.

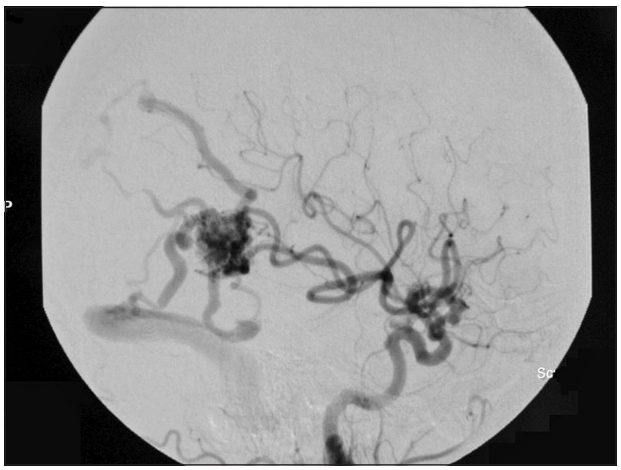

Fig. 1a. **Right common carotid artery angiograms and CTA showing AVM, which is fed by three feeders from the right MCA. Four draining veins are also observed: one is coming to sigmoid sinus, second – to the superior sagittal sinus and two – to the deep cerebral veins.**

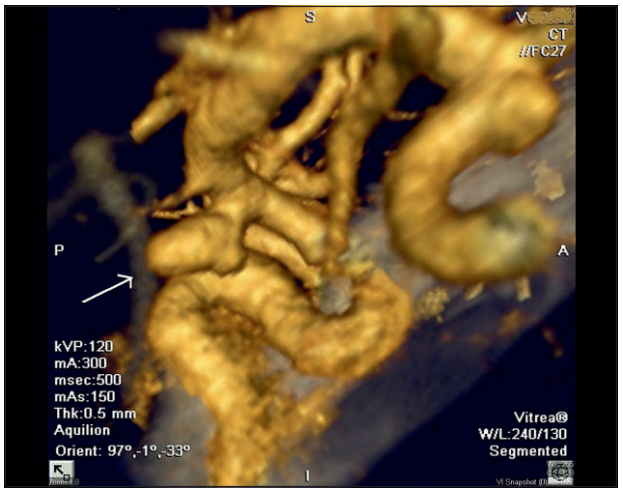

Fig. 1b. **CTA shows right side aneurysm of ICA, 14 mm in diameter (*arrow*).**

Fig. 1c. **Postoperative angiogram (after all surgeries), showing full AVM excision and clip ligation of aneurysms.**

All aneurysms were clipped after two months. In the course of the surgery, an ICA aneurysm was detected. It was clipped firstly to avoid accidental clipping of posterior communicating artery (PComA) and anterior choroidal artery (AChA). Two MCA aneurysms were clipped unremarkably. Postoperative angiogram showed complete clip ligation of aneurysms and AVM excision (Fig. 1c). Postoperative course went well too. The patient has been living independently for about 2 years after the operation without epilepsy.

Case 2

A 44-year-old man was admitted to our hospital with seizures. The seisures started 7 years ago. An emergency computer tomography (CT) scan showed AVM in the left temporal lobe. A MRI scan demonstrated AVM between posterior portion of superior and medial temporal gyrus in the left temporal lobe. The left common carotid artery angiography demonstrated high flow AVM in the left temporal lobe (Fig. 2a) and additional nidus from lateral posterior choroidal artery. CTA also showed a high flow AVM in the left temporal lobe and four varying MCA aneurysms. AVM was assessed as grade III according to Spetzler-Martin grading scale (S2E1V0). The AVM was fed by non-typical elongated and thick MCA from the beginning of M1 segment. AVM was draining to cortical veins. Four MCA aneurysms were diagnosed in association with AVM ((Fig. 2b) flow related).

This multiple pathology of cerebral vessels was operated in one stage. During surgery, we found high flow AVM and excised it. Three MCA aneurysms were clipped lastly. A small MCA aneurysm was eliminated by wrapping.

The acute postoperative period was complicated. Pneumonia and hypoxic encephalopathy were established. In our opinion, the causes of hypoxic encephalopathy were the loss of cerebral blood flow autoregulation and arterial hypotension. Postoperative angiograms showed removed AVM and clipped aneurysms (Fig. 2c). The patient became conscious, more euphoric than normal, regressive partial motor aphasia remained for two weeks after operation.

The patient has been living independently for about 1 year after the operation without focal neurological deficit and epilepsy.

Case 3

A 53-year-old man was hospitalized in our department after an episode of severe headache, without focal neurological deficit. The CT scan with contrast and sagittal MRI demonstrated a structure resembling an AVM in the frontoparietal region without subarachnoid hemorrhage. The left common carotid artery angiography and CTA scan showed high flow eloquent area AVM (III according to Spetzler-Martin grading scale (S2E1V0) and one aneurysm in the ICA communicating segment (Fig. 3a, 3b (not

Fig. 2a. **Left common carotid artery angiography: high flow AVM.**

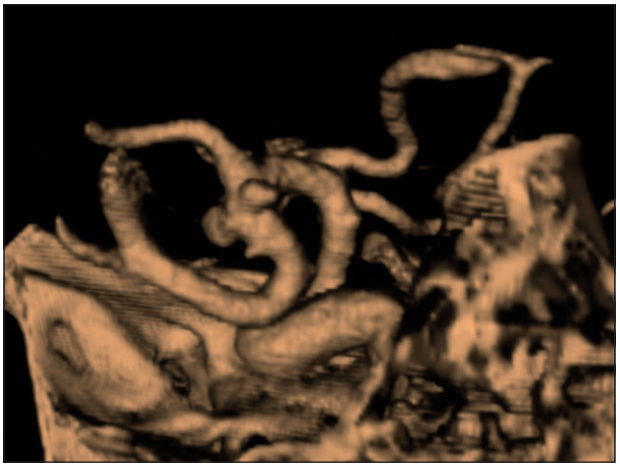

Fig. 2b. **MCA aneurysms on feeder (CTA).**

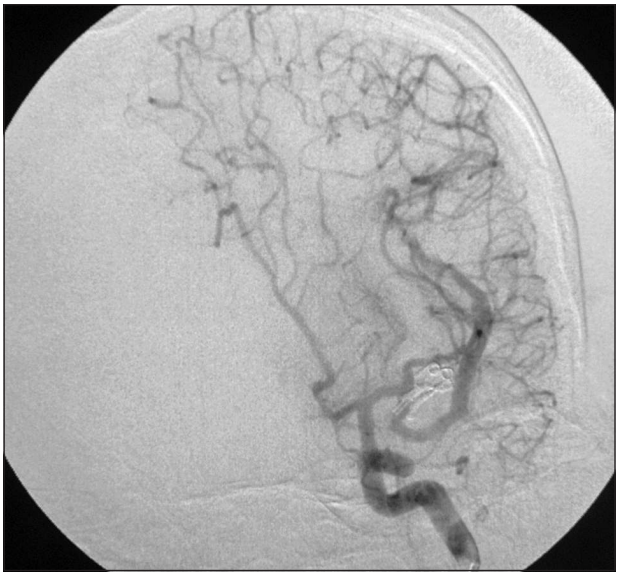

Fig. 2c. **Postoperative angiography showing full elimination of AVM and aneurysms.**

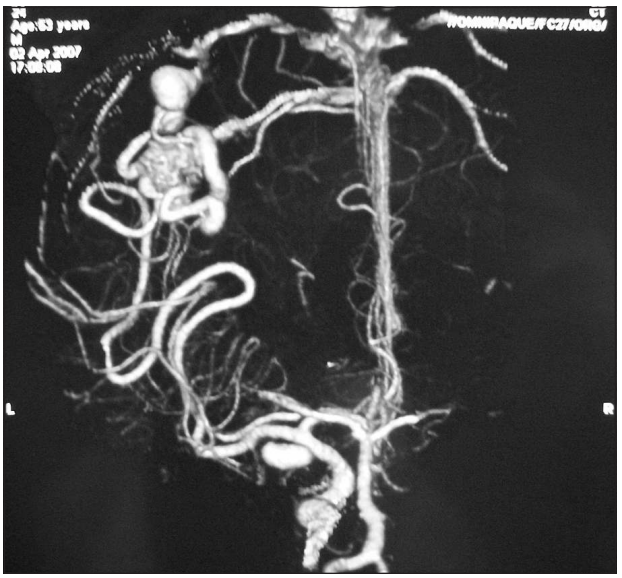

Fig. 3a. **CTA demonstrating high flow AVM in the eloquent motor area and left ICA aneurysm.**

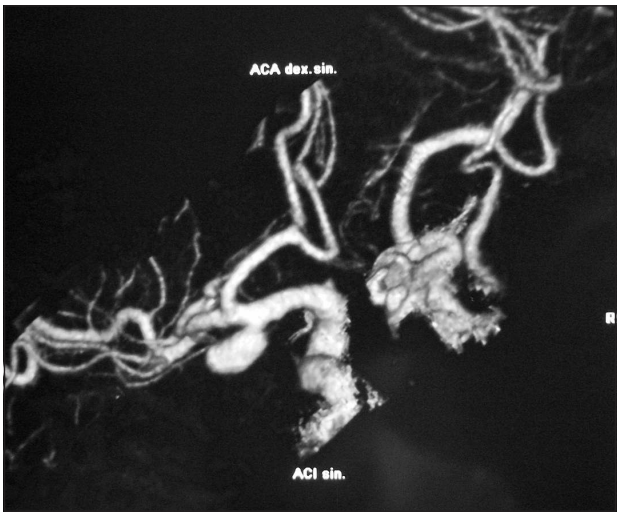

Fig. 3b. **CTA demonstrating high flow AVM in the eloquent motor area and left ICA aneurysm.**

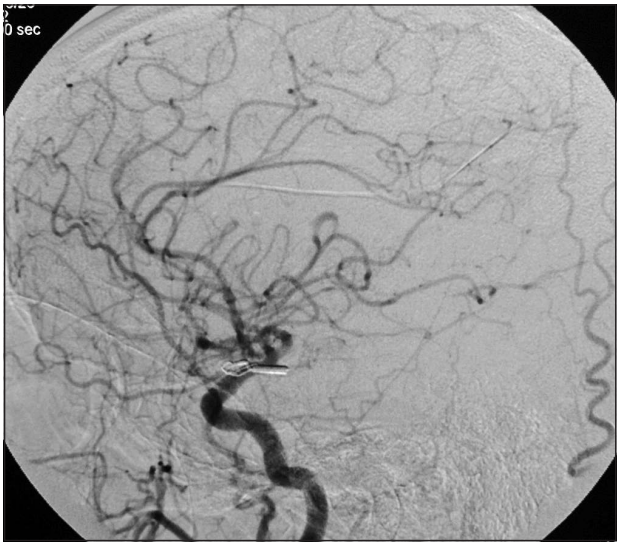

Fig. 3c. **Postoperative angiography with full surgical obliteration of AVM.**

flow-related)). The feeders were coming from MCA branches. Two draining veins were observable; they passed blood to superior sagittal sinus.

In the course of the surgery, AVM was removed from eloquent area. The ICA aneurysm was clipped to save PComA and AChA. Postoperative angiography showed normal view (Fig. 3c).

We observed right hand paresis (2 point) and a light dysphasia during an acute postoperative period. The patient was discharged with regressive right hand paresis (3 point), without additional neurological deficits.

The patient has been living independently for 3 years after operation without focal neurological deficits.

DISCUSSION

The risk of hemorrhage with coexisting aneurysm and AVM is about 7% per year, compared with 1.7% per year of AVM unassociated with aneurysm [16]. By Redekop et al. study, the bleeding rate was 5.3% per year and all hemorrhage arising from the AVM [11].

Three major theories are described in association of cerebral AVM with aneurysm: 1) the coexisting is a coincidence [17]; 2) congenital disturbance in vasculogenesis [18, 19], and 3) the hemodynamic stresses leading to development of an aneurysm in a feeder [20]. Multiple vascular disorders, such as multiple AVM, are associated with hereditary hemorrhagic telangiectasia, otherwise known as Osler-Weber-Rendu syndrome [21]. Moyamoya disease is associated with aneurysms that develop in a hemodynamically atypical area of brain [22].

The most acceptable classification of aneurysms linked to AVM's divide them into three groups: flow related, intranidal, and not-flow related [23]. Surgical practice as a treatment modality for AVM coexisting with intranidal aneurysms is unquestionable. The indications for the management of not-flow related aneurysms are the same as for incidental aneurysms. The vast majority of discussions are concerned with the flow related aneurysms. In our opinion, provided that major feeding arteries of AVM are exposed during the surgery, an associated aneurysm should also be approached surgically.

Halim et al. divided the patients suffering from AVM and associated aneurysms into six major groups according to clinical presentation: hemorrhage (38–50%), seizures (22–30%), focal deficit (6–10%), headache (8–15%), incidental (5–16%), and other (1%) [23]. Clinical manifestation depends on dominating lesion.

The symptomatic lesion must be treated first. Three major treatment modalities are used in treatment of AVM associated with aneurysms: radiosurgery, endovascular embolization and surgery. In all of our cases, the patients were treated only surgically. The patient has a risk of haemorrhagic complications of radiosurgically removed AVM's despite angiographic proof of their obliteration [25]. Presurgical embolizations reduce volume, blood flow and postoperative over flow in adjacent brain but, in our opinion, in many cases grade III AVM in eloquent area must only be treated surgically. Endovascular embolization has a high risk of AVM rupture, and is difficult to use in eloquent area. Embolization is limited in obliteration of grade III AVM, and is regularly used presurgically. Pre-surgical embolization is one of the choices in multimodality approach to deal with this complicated pathology. AN excellent microneurosurgical technique and perfect anesthesia are the best choices for the treatment of AVM. Two separate risks of complications arise from pre-surgical embolization in comparison with only surgical removal [26]. In the treatment of AVMs and aneurysms, associated to them, microneurosurgical technique is preffered to endovascular embolization.

We use presurgical embolization in our daily practice too. The group of grade III AVM is the most heterogenous with four different combinations of size, venous drainage and eloquence. The best results for only surgical treatment of grade III AVM's are S1E1V1 [27]. The most demanding surgery is grade III with S2E1V0. The presurgical endovascular embolization of this group is questionable in many cases. In our opinion, grade III AVM's with S2E0V1 is also acceptable for surgery alone. Vascular architectonic is very important in decision making for AVM's surgeries (compact nidus vs. diffuse architectonic).

The common cause of postoperative haemorrhage is residual AVM. The best way to avoid this huge complication is careful pre-surgical planning and safe and clear microneurosurgical technique of the operating neurosurgeon [28]. The perfusion pressure breakthrough is responsible for haemorrhage and cerebral edema even after precise haemostasis during the surgery of AVM. The microneurosurgical technique and excelent postoperative care in ICU is the best way to avoid this complication [29].

The published literature (Table) provides instances for various outcomes in the treatment of this vascular pathology, therefore a thorough meta-analysis is required for a deliberate treatment plan.

CONCLUSIONS

1. AVMs in company with cerebral aneurysms have high tendency for bleeding and they are dificult to manage.
2. Surgical approach for all combinations of Spetzler-Martin grade III AVMs is applied only by a skillfull and experienced surgeon. Vascular architectoninc is important too.
3. The multimodality treatment is sugested for the treatment of this complicated cerebral pathology.
4. The patient has two separate risks of complications if we suggest pre-surgical embolization in comparison to only surgical removal.
5. The surgery of AVM with aneurysms requires decision making by a neurosurgeon with significant experience.

Table. **Short literature review of treatment of AVM's associated with intracranial aneurysms.**

Authors	Clinical presentation	Cases	Treatment modalities	Early complication	Outcome	Conclusion
Henry A. Shenkin et al. 1971 [9]	ICH (AVM)	1	Only AVM surgery	Mild	Full recovery, independent	Upon removal of the AVM the aneurysm markedly decreased in size. Hemodynamic stress is an important factor of an aneurysm development on the feeding vessel.
Bizhan Aarabi et al. 1978 [7]	Seizures, progressive mental deterioration and urinary incontinence	1	AVM and aneurysm surgery at one stage	Severe	Full recovery, independent	Giant aneurysms may be resected if they cause pressure phenomena.
Jiro Suzuki et al. 1979 [6]	6 cases presented with SAH, 2 ICH and hemiparesis, 3 seizures	9	9 cases aneurysm surgery, 8 cases AVM surgery	NA	6 full recovery, independent, 1 partially disabled bur working, 2 unable to work but capable of self care	1) There are two possible sources of hemorrhage, 2) the mortality rate following conservative treatment is 60%, and 3) even when surgery is performed on either aneurysm or the AVM, there is danger of death from hemorrhage of the untreated lesions, it is desirable to treat both lesions in a single radical operation.
Manuel J. Cunha E Sa at al. 1992 [12]	ICH 62%, seizures 21%, headache 5%, mass effect in 8%	39	27 AVM and aneurysm surgery, 4 patients only endovascular treatment, 7 patients underwent multimodality treatment (surgery and endovascular).	NA	24 normal, 12 mild neurological deficit, independent, 3 dependent on other, moderate neurological deficit	Symptomatic lesion, when determined, is treated first. Whenever it is possible to safely exclude both the aneurysm and the malformation during the same operation without unreasonably increasing the risk of the procedure, this option is chosen.
Michel Piotin et al 2001 [24]	3 ICH (AVM), 12 SAH (ruptured aneurysms), 15 asymptomatic	30	30 underwent endovascular procedures. 5 patients both aneurysms and AVM were totally eradicated with embolization alone. Surgery on the AVM remnants after endovascular reduction n=2.	25 without complications, 2 mild, 3 severe	Normal 25, transient neurological deficit – 1, mild – 1, severe – 3.	Placement of GDCs is indicated in the majority of proximal aneurysms. Glue embolization of distal aneurysms can be part of the first step in the endovascular eradication of the AVM or can be the goal of the endovascular treatment if the characteristics (i.e., size, location) of the AVM do not allow for the anticipation of a total cure with embolization.
Mooseong Kim et al. 2006 [15]	4 ICH (AVM), 1 SAH (ruptured aneurysm)	5	Gamma-knife radiosurgery (GKS) for all patients. 1 patient GKS was applied after failed embolization	No complications	4 cases complete AVM and aneurysms obliteration after GKS, 1 case full aneurysm obliteration and partial AVM obliteration	In patients with AVM-associated aneurysms, treatment of the hemorrhage site should be performed firstly or simultaneously with AVM. Gamma Knife surgery is a possible method of choice for the treatment of an AVM with an associated intranidal aneurysm.
Joo Kyung Ha et al. 2009 [13]	Bleeding (headache, mental deterioration, dizziness) – 17; seizures – 3, 3rd nerve palsy – 1.	21	Gamma-knife radiosurgery for all AVM. For aneurysms treatment was used surgery (5), embolization (11) or observation.	NA	NA	AVMs associated with aneurysm have the characteristics of a low incidence, a high bleeding tendency and they are difficult to manage. Treatment with GKS and GDC embolization is considered to have a significant role in minimizing the neurologic injury.

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SPETZLER-MARTIN III LAIPSNIO GALVOS SMEGENŲ ARTERIOVENINĖS MALFORMACIJOS, ASOCIJUOTOS SU ANEURIZMOMIS: TRIJŲ KLINIKINIŲ ATVEJŲ APŽVALGA

Santrauka

Galvos smegenų arterioveninės malformacijos yra reta, įgimta galvos smegenų kraujagyslių patologija, kurios dažnis yra apie 0,01 % populiacijos. Apie 8,9 % arterioveninių malformacijų būna asocijuotos su intrakranijinėmis aneurizmomis. Chirurginis, enovaskulinis ir radiochirurginis gydymai arba jų kombinacija yra naudojami klinikinėje praktikoje, gydant arteriovenines malformacijas. Šiame straipsnyje pristatome tris retus greitos tėkmės arterioveninės malformacijos atvejus, asocijuotus su intrakranijinėmis aneurizmomis. Šie ligoniai buvo gydomi tik chirurginiu būdu. Galimos šios patologijos priežastys, klinikiniai požymiai, gydymo galimybės ir išeitys aptariamos straipsnyje.

Raktažodžiai: galvos smegenų aneurizmos, galvos smegenų arterioveninės malformacijos, chirurginis šalinimas.

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