SECg Staging System: A New Approach to the Management of Arteriovenous Malformations of the Head and Neck

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Objectives: Arteriovenous malformations (AVM) are the most troublesome vascular malformations to deal with. They tend to behave like low-grade malignancies with infiltrative and disruptive growth. Crucially, the clinical course of an AVM that has been improperly managed is usually characterized by a recurrence that is much more aggressive than the original disease. As in oncology, a comprehensive staging system is highly desirable and is to date lacking in the literature. The authors present a new comprehensive staging system.

Methods: A multicentric multidisciplinary team of experts in the field of vascular anomalies has created this new staging system. The SECg staging system defines the local extension of the disease (S1–S4), the vascular architecture of the malformation (E1, E2, E3), the severity of the symptoms (C0–C3) and the presence or absence of growth of the AVM (g+, g−).

Results: This staging system allows to address all the aspects of AVMs and, more importantly, to help building an appropriate, individualized treatment plan for affected patients. After being staged an AVM can be defined as (a) healable, (b) healable with predicted sequelae, or (c) unhealable. Then, the SECg system allows to outline (a) absolute indications, (b) relative indications, and (c) no indications for treatment. The purpose of the treatment (radical, palliative) is furthermore taken into consideration.

Conclusions: This multicentric, the SECg staging system that this multidisciplinary group of Authors has defined allows for a comprehensive staging of the disease which in turn has enabled to outline an algorithm to properly manage AVMs.

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Arteriovenous malformations (AVMs) are extremely difficult to treat effectively, and for the patient, difficult to live with. AVMs are congenital malformations of the vascular system that originate from aberrations in the normal development of the primitive vascular plexus. They are characterized by a nidus, which is probably an unregulated capillary bed. They are surrounded by a tortuous web of dysplastic vessels. The nidus enables a high flow shunt from the arterial to the venous system.[1] While not malignant, they are the most aggressive, destructive, and the most complex of all vascular anomalies. Treatment has centered on interventional procedures and aggressive and often complex surgeries. Rarely, if ever, is the AVM successfully contained after one procedure. They often persist and require numerous procedures to control the excessive shunting, as well as profound disfigurement.

Inherent to AVMs is a clinical behavior that resembles that of a low-grade neoplasm. They tend to expand, with occasional abrupt increases in growth. If untreated, they are frequently complicated by local ulceration, infection, and hemorrhage. They become very difficult for the physician to contain, and even more difficult for the patient to endure. Many AVMs are incorrectly diagnosed, as well as improperly managed. This can lead to numerous interventional treatments and complex and complicated surgeries which may make the condition worse. What often follows these interventions is a series of aggressive relapses that are often worse than the original disease.[2]

Numerous treatment regimens have been proposed but without a widely accepted staging system, it is not possible to evaluate and compare these regimens. Furthermore, outcomes analysis can only be possible with a uniform, widely accepted staging system. In this paper, the authors propose a comprehensive staging system that would standardize communication among different specialists and thereby allow for safer treatment planning. This staging system encompasses clinical, surgical-anatomical, and endovascular radiologic features.

MATERIALS AND METHODS

The SECg Staging System

The staging system is divided in 4 sections, named S, E, C, and G (Supplementary Digital Content, Table 1, http://links.lww.com/SCS/B332).

S represents the Surgical/Anatomical. Here the physical features of the AVM and the involvement of different anatomical structures are described.

S1: The AVM involves one single anatomical site such as the subcutaneous tissue, a muscle, or a bony segment. The AVM does not involve adjacent sites (Fig. 1).

S2: Two adjacent anatomical sites are involved. One example may be a preauricular AVM that also involves the underlying parotid gland (Fig. 2).

In both S1 and S2 stages, no vital structures are involved.

S3: The AVM infiltrates important structures such as the cheek, tongue, larynx, orbit, but not vital ones. For example, a cheek AVM that deepens into the plane of the facial nerve would be considered S3. Arteriovenous malformations infiltrating the orbital cone, the tongue or the larynx are other examples (Fig. 3).

S4: AVMs may involve vital structures such as the carotid artery, internal jugular vein and/or base of skull (Fig. 4).

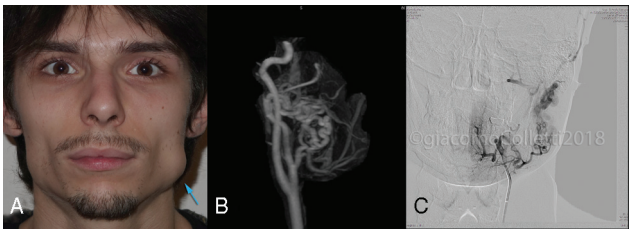

FIGURE 1. (A) Left intramasseteric AVM causing a pulsating and painful bulge. This makes this AVM a C1 stage. The mass has increased its size during the previous 6 months so it is a G+ one. (B) Angio CT scan of the same patient. The AVM is confined within the masseteric fascia and limited to the muscle. The AVM is a S1 stage. (C) Preoperative angiography: an arteriolo-venous architecture is present. This accounts for an E2 stage. The complete stage of this AVM is S1E2C1+. AVM, arteriovenous malformations.

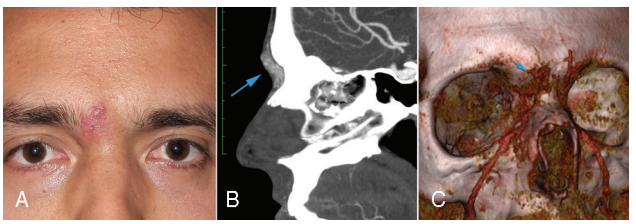

FIGURE 2. (A) AVM of the glabellar region. Discoloration, swelling and pulsations are present. Few episodes of spontaneous hemorrhage were reported by the patient (C2). The mass has significantly grown over the previous 6 months (G+). (B) Sagittal CT scan of the AVM. The malformation involves the region down to the bone (skin, subcutaneous tissue and procerus muscle). This makes this AVM a S2 stage. (C) 3D angio CT of the same patient. An arteriolo-venular architecture is present (E3). The comprehensive stage of this AVM is S2E3C2+.

E describes the endovascular characteristics of the AVM.

E1 are ArterioVenous AVMs.

E2 are ArterioloVenous AVMs (Figs. 1 and 3).

E3 are ArterioloVenular AVMs (Figs. 2, 4, and 5).

This classification essentially resumes the Houdart classification for intracranial AVMs (see further). C is used to address clinical features and or complications

C0: no symptoms nor complications are present. C1: the lesion is symptomatic but without complications: Pulsations (or thrills and bruits), Paresthesia and Visible Swelling.

C2: there are local complications: Ulceration, Infection, Hemorrhage.

C3: there are general complications: congestive cardiac failure, fatigue, or anemia.

G: describes the growth/progression of the AVM.

G-: AVMs have remained stable during the previous 6 months.

G+: AVMs have been progressive during the previous 6 months.

Exemplificative cases are illustrated in Figs. 1-5

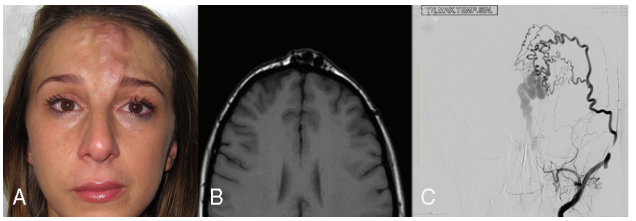

FIGURE 3. (A) AVM of the forehead. The AVM involves skin, subcutaneous tissue and muscle: this makes this AVM an S2. (B) Axial MRI of the same patient involving the forehead in its full thickness but sparing the bone. This, again makes the AVM S2. (C) Angiography demonstrating multiple arterial feeders draining into one single vein (E2). AVM, arteriovenous malformations.

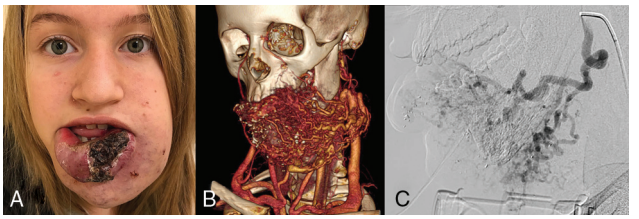

FIGURE 4. (A) Wide AVM of the inferior third of the cervico-facial region, entirely substituting the inferior lip. Radical resection of the AVM will surely procure a severe functional and cosmetic impairment. This makes this AVM an S3. The malformation was locally complicated by frequent hemorrhages, infections and ulcerations (C2). It rapidly increased its size in the previous 6 months (G+). (B) 3D angio CT of the same patient. The AVM involves the whole of the superficial third of the face. However, vital structures were spared (S3 and not S4). An arteriolo-venular pattern is present (E3). (C) Angiography confirms an arteriolo-venular pattern (E3). The comprehensive stage for this patient is S3E3C2+. AVM, arteriovenous malformations.

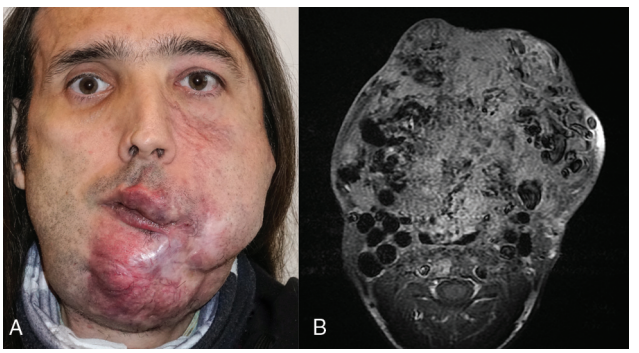

FIGURE 5. (A+B) Extensive AVM of the head and neck worsened by previous bilateral external carotid artery ligature (done elsewhere). Feeding arteries and draining veins coming from the vertebral circulation and infiltration of the skull base account for an incurable disease (S4). The AVM has worsened clinically in the previous 6 months (G+). A complex vascular pattern of arteries arterioles, veins and venulae compose the AVM (E3). Several episodes of hemorrhage, ulceration and infection were reported. An EKG and Cardiac US showed initial signs of left heart failure (C3). The overall stage of this AVM is S4E3C3+. AVM, arteriovenous malformations.

RESULTS

As seen previously the Authors have conceived and described a new staging system that encompasses all the features that, put together, allow to have a complete image and understanding of the AVM.

Moreover, by putting together the S, the C, and the G substages AVMs can be divided in:

Curable, Curable with predictable severe sequelae, Incurable; With Absolute Indications and With Relative (Or No) Indications (Supplementary Digital Content, Table 2, http://links.lww.com/SCS/B332). The comprehensive flowchart of proposed treatment protocol is illustrated in Fig. 6.

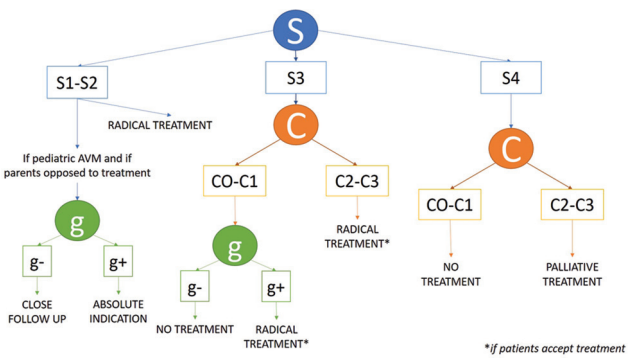

FIGURE 6. Protocol of treatment based on the staging system.

We have adopted with success this staging system in more than 100 patients. However, the description of the case series is beyond the scopes of the present paper and is the object of a future publication.

DISCUSSION

Arteriovenous malformations are rare, complex, and often aggressive lesions. They typically present as a small uncomplicated lesion, which, over time can become aggressive and often fatal. It is believed that the prevalence of AVM's is less than 1:2000 (thus making them a rare disease). However, this figure may be understated and the true incidence may be higher due to the use of incorrect nomenclature and misdiagnosis.[3]

The following factors mitigate for a comprehensive staging system.

Firstly, early diagnosis will result in appropriate early treatment which may, in turn, lead to control of the disease.[4]

Secondly, inappropriate diagnosis and treatment of an AVM may result in the lesion becoming aggressive which, in turn, will result in difficulty to control the disease. Moreover, these inappropriate treatments may result in local or general complications.[5,6]

Thirdly, the lack of a uniform, widely accepted staging system will prevent physicians from evaluating and comparing different treatments for AVM's. Many staging systems have been proposed but none have become widely accepted.

Finally, AVMs are remarkable because they are believed to be caused by impaired, excessive angiogenesis that closely resembles that which supports cancer growth.

We believe that once an accurate diagnosis is made, a comprehensive staging of the AVM that takes into account all of the relevant clinical features will lead to a greater chance of containment of the disease. Furthermore, a widely accepted staging system will allow for accurate comparison of the various treatment regimens.

As has become standard with malignant tumors, a reliable and widely accepted staging system, for AVM's, should allow for adequate evaluation of treatments. This will be accomplished, by first describing how far the disease has progressed; second, by facilitating better treatment planning; and, third, by properly inferring a prognosis.

Ultimately, this staging system should enable meaningful communication between clinicians of different specialties. This last aspect is crucial, since AVMs are often managed by a multidisciplinary group of physicians.

The first classification for AVMs was the Spetzler-Martin classification. It was published in 1986[7] by the American neurosurgeons Robert F. Spetzler and Neil A. Martin and was primarily used for intracranial AVM's. It is divided into 3 sections: Size of Nidus, Eloquence of Adjacent Brain, and Location of Venous Drainage. For each section a score is given to the AVM and a global score is obtained by summing the single score. Six grades are derived that correlate the AVM stage with the risk of surgical management. Grade I are small, superficial, surgically removable AVMs in non eloquent brain. Grade VI AVMs are inoperable. This classification is still used for intracranial AVMs but is unfortunately not applicable to extracranial lesions.

The first clinical staging system for extracranial AVMs was proposed by Robert Schöbinger, a Swiss vascular surgeon, during the 1994 workshop for the study of vascular anomalies (Mulliken book pg 880), which in 1996 became the International Society for the Study on Vascular Anomalies (ISSVA). The Schöbinger classification divides AVMs in 4 stages: (I) Quiescence; (II) Progression; (III) Complications; (IV) Decompensation. This classification has the undeniable merit of describing the natural history of AVMs which usually progress from stage I to III over time. However, it lacks any description of the features of the AVM. Kohout and others have proposed a modification of the Schöbinger in 1998.[5] Here the stages are as follows. Stage I: cutaneous blush, warmth; Stage II: bruit, audible pulsation, expanding lesion; Stage III: pain, ulceration, bleeding, infection; Stage IV: Cardiac Failure. The Schobinger classification, which has become widely adopted, lacks any description of the anatomical extent of the disease or the endovascular features.

On a parallel route, the first endovascular staging system was proposed in the literature by the French neuroradiologist Houdart et al in 1993.[8] This staging system was used to classify intracranial AVMs as: Type A: Arterio-Venous malformation; Type B: Arteriolo-Venous malformation; Type C: Arteriolo-Venular malformation. This "angioarchitectural classification" has a practical relevance for endovascular treatment according to the Authors.[8] They stated that while type A and B can be managed through the arterial and venous system, type C should be approached trans arterially only. In 2006 the Korean radiologist Ki Cho et al proposed a very similar classification for extracranial AVMs.[9] They described peripheral AVMs as follows: Type I: Arteriovenous fistula; Type II: Arteriolovenous fistula; Type IIIa: Arteriolovenular fistula without dilation; Type IIIb: Arteriolovenular fistula with tortuous dilation. All the "endovascular" classifications are aimed at describing the vascular anatomy of the AVM in order to guide an interventional approach. Here, many agents may be applied by using either a direct puncture technique into the nidus or a superselective catheterization technique to deliver embolotherapy or sclerotherapy agent selectively to the nidus of the lesions.

The transarterial approach has been preferentially used to embolize AVMs when possible, and either direct puncture or the transvenous approach is selected when important normal arterial branches arise in very close proximity to a malformation or when extreme arterial tortuosity or previous treatment (including surgical ligation and embolization of the feeding artery) preclude successful transarterial catheterization.[10] Based on results published by Cho,[9] the main target of type II AVMs is the venous component of the nidus. Therefore, the mainstay therapeutic approaches are transvenous and direct puncture with or without coil embolization of the venous component of the nidus to reduce the amount of ethanol and to stabilize the thrombosis within the large venous component. Only the transarterial approach is available for type IIIa AVMs because the fistula is too fine for direct puncture. Type IIIb AVMs can be treated properly via transarterial and direct puncture approaches.

The approaches used for mixed types are combinations of those used to treat the individual types, whereas transarterial approaches were more frequently used in mixed types IIIa/IIIb to assess a safer access and embolization. With advances in instrumentation and imaging, sclerotherapy and embolotherapy now play a significant role in the treatment of AVMs and have been found to be effective for certain types of AVMs as a primary mode of therapy.[11] Other Authors on the other hand suggest that endovascular treatment alone yields to very high relapse rates.[4] Recent researches, based on experimental settings, seem to demonstrate that embolization alone may have a dramatic proangiogenic effect[11] on AVMs. Thus, endovascular treatment is preferably part of a multidisciplinary "nidocidal" approach.

These vascular classifications do not describe any of the clinical features nor the feasibility of a surgical (or, as previously stated preferably, a combined endovascular and surgical) management of the AVM.[12]

In 2010, Richter and Suen, proposed an interesting classification divided as follows.[13] They coined a "TDS" system where T is the superficial extension of the AVM, D describes the depth of the malformation and S its growth rate. T1 involves 1 cervicofacial subunit, T2 involves 2 cervicofacial subunits, T3 involves 3 or more cervicofacial subunits and T4 are bilateral or multifocal AVMs. D1

AVMs involve skin and/or subcutaneous tissue only, D2 subcutaneous and muscle, D3 subcutaneous, muscle and cartilage or bone and D4 AVM involve the skull base or have intracranial extension. S0 are Schöbinger 1 AVMs, S1 are Schöbinger 2 and S2 are Schöbinger 3.

Although we applaud this work and agree with most of the points of this classification, we believe that in AVM management it is more crucial to determine if the disease involves anatomical structures that are functionally relevant or vital rather than to precisely establish their linear extent. Furthermore, we believe that the addition of endovascular characteristics will make our classification more universally relevant, especially when working with a multidisciplinary group of physicians. We also believe that the presence or absence of symptoms should be mentioned to aid in treatment planning.

Thus, putting together the S, the C, and the G substages we believe that AVMs can be divided in:

Curable, Curable with predictable severe sequelae, Incurable; With Absolute Indications and With Relative (Or No) Indications (Supplementary Digital Content, Table 2, http://links.lww.com/SCS/B332). The comprehensive flowchart of proposed treatment protocol is illustrated in Fig. 6.

We firmly believe that an endovascular staging should be incorporated into a comprehensive classification. Endovascular treatments can indeed be used to prepare the patient for surgery in small, superficial AVMs or to palliate vast, unresectable AVMs.

One final consideration concerns bony AVMs.[14] These are usually considered S1 because they are confined within bone. However, when they breach the periosteum they can become anything from S2 to S4, depending on the extension and the location of the disease.

CONCLUSIONS

To date, a comprehensive staging system (which is needed, for the aforementioned reasons) is lacking in the literature.

The SECg might help filling this void and enhancing communication between specialist and, ultimately, improve AVM management.

REFERENCES

1. Doppman JL. The nidus concept of spinal cord arteriovenous malformations. A surgical recommendation based upon angiographic observations. *Br J Radiol* 1971;44:758–763
2. Colletti G, Valassina D, Bertossi D, et al. Contemporary management of vascular malformations. *J Oral Maxillofac Surg* 2013;72:1–19
3. Hassanein AH, Mulliken JB, Fishman SJ, et al. Evaluation of terminology for vascular anomalies in current literature. *Plast Reconstr Surg* 2011;127:347–351
4. Liu AS, Mulliken JB, Zurakowski D, et al. Extracranial arteriovenous malformations: natural progression and recurrence after treatment. *Plast Reconstr Surg* 2010;125:1185–1194
5. Kohout MP, Hansen M, Pribaz JJ, et al. Arteriovenous malformations of the head and neck: natural history and management. *Plast Reconstr Surg* 1998;102:643–654
6. Visser A, FitzJohn T, Tan ST. Surgical management of arteriovenous malformation. *J Plastic Reconstruct Aesthetic Surg* 2011;64:283–291
7. Spetzler RF, Martin NA. A proposed grading system for arteriovenous malformations. *J Neurosurg* 1986;65:476–483
8. Houdart E, Gobin YP, Casasco A, et al. A proposed angiographic classification of intracranial arteriovenous fistulae and malformations. *Neuroradiology* 1993;35:381–385
9. Cho SK, Do YS, Shin SW, et al. Arteriovenous malformations of the body and extremities: analysis of therapeutic outcomes and approaches according to a modified angiographic classification. *J Endovasc Ther* 2006;13:527–538
10. Shin BS, Do YS, Lee B-B, et al. Multistage ethanol sclerotherapy of soft-tissue arteriovenous malformations: effect on pulmonary arterial pressure. *Radiology* 2005;235:1072–1077
11. Buell TJ, Ding D, Starke RM, et al. Embolization-induced angiogenesis in cerebral arteriovenous malformations. *J Clin Neurosci* 2014;21:1866–1871
12. Eivazi B, Werner JA. Management of vascular malformations and hemangiomas of the head and neck - an update. *Curr Opin Otolaryngol Head Neck Surg* 2013;21:157–163
13. Richter GT, Suen JY. Clinical course of arteriovenous malformations of the head and neck: a case series. *Otolaryngol Head Neck Surg* 2010;142:184–190
14. Colletti G, Frigerio A, Giovanditto F, et al. Surgical treatment of vascular malformations of the facial bones. *J Oral Maxillofac Surg* 2014;72:1326.e1–1326.e18