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CLASSIFICATION OF SPINAL CORD ARTERIOVENOUS MALFORMATIONS. A LITERATURE REVIEW

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The article presents an overview of classifications of arteriovenous malformations of the spinal cord, taking into account historical changes in the concepts of this vascular pathology of the spinal cord as clinical experience accumulates. Classifications of spinal arteriovenous malformations are very confusing and complex, and publications on this issue are quite few. We analyzed 33 scientific sources to describe the development of classifications of arteriovenous malformations of the spinal cord and selected the most important ones for practicing neurosurgeons and endovascular surgeons. We emphasize that a universal classification that could satisfy the entire scientific community has not yet been developed.

Key words: arteriovenous malformations of the spinal cord, classification, historical review

Introduction Spinal arteriovenous malformations (SpAVM) are a heterogeneous group of dysplastic lesions of vascular genesis that do not have blastomatous features and are located in the spinal cord, its roots, membranes, surrounding tissue, and vertebrae [1]. The prevalence of SpAVM varies from 1 to 3 cases per 1 million people per year [2, 3]. The majority of patients are men (60%), and the age of most hospitalized patients is 45–64 years. Significant clinical deterioration is observed in 72% of patients with all types of SpAVM within 4 years from the onset of the first symptoms [4]. The variability of such SpAVM characteristics as extent, localization, and vascular anatomy complicated their study and the choice of treatment strategy. The emergence of the SpAVM classification made it possible to orient clinicians to the features of the clinical picture and determine the prognosis of the disease. Despite the existence of classifications, the treatment strategy for vascular lesions of the spinal cord continues to be a subject of debate, since these are rare diseases that require complex treatment, which in itself can be dangerous for patients. The frequency of complications in the treatment of SpAVM, according to scientific literature, varies from 5 to 25% regardless of the method of treatment [5]. Systematization, determination of important anatomical characteristics that could indicate commonality or, conversely, differences, seemed to be a difficult task.

The first classifications Vascular malformation of the spinal cord was first described by Hebelt at autopsy in 1885 (according to SE Akopov, W. Schievink [6]), and to this day the ideas about this disease continue to change. In 1971, a group of authors G. Di Chiro et al., based on data obtained from selective spinal angiography, first divided SpAVMs into 3 large groups: type I - single arteriovenous shunts (currently called "dural arteriovenous fistulas"), type II - glomus, type III - juvenile [7]. A single arteriovenous shunt, or dural arteriovenous fistula (AVF), consists of an afferent vessel originating from the radicular artery and an efferent vessel draining into the radicular vein, which then extends intradurally and drains into the perimedullary venous network. Glomus-type SpAVM is localized intramedullary and is a glomus that has multiple afferents, including those originating from the anterior and/or posterior spinal arteries, and drains into the venous plexus of the spinal cord. The juvenile type of SpAVM involves damage to the entire sclerotome, including the spinal cord, dura mater, muscles, and soft tissues. In 1977, M. Djindjian et al. published an article in which they drew attention to the existence of a small group of SpAVMs that differ from all the others. These SpAVMs receive blood supply from the anterior spinal artery, but are located outside the spinal cord and its pia mater. They differ in their clinical manifestations from other SpAVMs [8]. Much later, in 1986, RC Heros et al. presented a case of treatment of a patient from Belgium, in whom the anatomy of SpAVM also did not correspond to any of the types described by G. DiChiro. RC Heros et al. analyzed their case and compared it with the data of M. Djindjian et al., after which they proposed to distinguish a new (IV) type of SpAVM [9]. B. Rosenblum et al. in 1987 presented data on 81 patients with SpAVM. They divided all SpAVMs into 4 types, and the first 3 types did not differ from those in the classification of G. DiChiro, and type IV consisted of intradural arteriovenous fistulas. In their work, in addition to vascular anatomy, the authors were able to compare the variants of the clinical course of the disease, the features of pathogenesis and the results of treatment in patients with dural and intradural AVFs. They put forward a hypothesis that dural AVFs are an acquired defect of the spinal cord vessels, and intradural AVFs are a congenital defect [10]. After 4 types of SpAVM were identified, according to the studies, it became possible to predict the course of the disease taking into account the type of lesion. Currently, it is believed that the prognosis of the disease depends on the duration of the existence of symptoms before the start of treatment. Patients under 60 years of age have a greater chance of restoring impaired functions. After complete occlusion of the spinal dural AVF (SpAVM type I), the progression of the disease stops in most cases, and with timely treatment, the severity of neurological disorders decreases in 90% of cases [11-13]. Hemorrhages and corresponding symptoms are not characteristic of SpAVM type I. In patients with SpAVM type II, the annual risk of hemorrhage is 4% [14]. It is worth noting that after the initial hemorrhage, the frequency of repeated hemorrhages is 10% within 1 month and 40% within 1 year. Mortality associated with SpAVM type II approaches 18% [15]. According to the analysis of long-term treatment results, clinical improvement is observed in 57% of cases, no dynamics - in 31%, deterioration - in 12% [14]. The frequency of hemorrhages associated with SpAVM type III is 2.1% per year. In patients with this type of lesion, after treatment, improvement occurs in 73% of cases, no dynamics - in 10%, deterioration of neurological symptoms occurs in 17% [16]. The frequency of hemorrhages in patients with SpAVM type IV is 2.5% per year. After treatment of this group of patients, improvement is observed in 68% of cases, no dynamics in 26%, and deterioration in 6% [17]. JJ Merland et al. [18], B. Gueguen et al. [19], KL Mourier et al. [20] drew attention to some differences in intradural AVFs and proposed a subclassification of type IV SpAVM, consisting of 3 subtypes. Subtype I is a short single AVF receiving blood supply from the terminal section of the anterior spinal artery and located most often in the projection of the conus medullaris or filum terminale; the draining veins are slightly dilated. Subtype II is an AVF with several afferents (they can depart from the anterior and posterior spinal arteries simultaneously) and medium blood flow intensity. Subtype III is a giant AVF, which, like the previous subtypes, has several afferents.

Modern classifications

The classification of scientists from the Barrow Neurological Institute (USA) has gained great popularity in the world. In 1992, JA Anson and RF Spetzler proposed to distinguish 4 types of SpAVM (Table 1), with the first 3 types of SpAVM remaining the same (i.e., they coincide with earlier classifications), and type IV was proposed to be perimedullary AVFs. To avoid confusion, the subtypes of type IV were reclassified: I was designated A, II – B, III – C [21]. In January 1997, the journal “Neurosurgery” published a work by YH Bao and F. Ling, who, based on an analysis of data from 80 patients, identified 5 large groups of SpAVM (Table 2). The proposed classification includes the previously described types of SpAVM, but is slightly different: the authors added paravertebral AVMs, which are characterized by damage to the paravertebral muscles, the opening of the nerve root and the epidural space of the spinal canal, and Cobb syndrome, which is manifested by damage to the spinal cord, dura mater, vertebra, paravertebral space and necessarily the skin within one metamere [22]. Cobb syndrome is sometimes associated with juvenile AVMs, which is not entirely true. Juvenile AVMs, intradural AVFs, and angiomas may be just a component of Cobb syndrome [23]. In accordance with the developed classification, an approach to the treatment of SpAVMs was proposed. According to YH Bao and F. Ling, most true intramedullary AVMs of the juvenile type are inoperable because the tangle of abnormal blood vessels fills almost the entire segment of the spinal canal, including the spinal cord itself. If their treatment is considered, they consider endovascular embolization to be the method of choice. Although recanalization and clinical recurrence after embolization are common, subsequent embolization as a final therapeutic alternative is acceptable [22].

In the treatment of glomus type SpAVM, a microsurgical method can be used, which is characterized by a higher radicality in the case of dorsal localization of the node. Preoperative embolization significantly reduces the severity of intraoperative bleeding and facilitates the removal of SpAVM. Radical removal of laterally located SpAVM without disruption of the spinal cord function, according to the authors, is impossible [22]. The main principle of treating intradural AVFs of all types is to block the arteriovenous shunt microsurgically or by embolization. Microsurgical disconnection of the fistula is more suitable for the treatment of intradural AVFs of type I, since the feeding artery (usually the anterior spinal artery) is too small and tortuous to perform superselective catheterization of the fistula. If embolization is chosen, YH Bao and F. Ling recommend using embolic materials such as coils, adhesives, or detachable balloons, which must be carefully delivered into the fistula to disconnect it. Type II intradural AVFs have larger afferents that are suitable for embolization with coils or adhesives. Type III intradural AVFs have wide afferents, which allows placement of a balloon or coils into the fistula with complete anatomical and clinical cure [22]. Since most of the feeding branches of dural AVFs are thin and tortuous, the authors recommend microsurgery for many patients [22]. In paravertebral AVMs and Cobb syndrome, treatment is aimed at eliminating the likely mechanism for the development of myelopathy. For example, YH Bao and F. Ling described a patient with multiple paravertebral fistulas who underwent only adhesive embolization because venous hypertension was the main mechanism of neurological deterioration. However, if other pathophysiological mechanisms are involved (e.g., spinal cord compression), a combination of microsurgery and embolization may provide more satisfactory results [22]. Based on their experience treating over 130 patients, RF Spetzler et al. proposed a modified classification of vascular pathologies of the spinal cord. The authors divided them into 3 main categories: neoplasms, aneurysms, and arteriovenous malformations. Hemangioblastoma and cavernous malformation were classified as vascular neoplasms. Spinal aneurysms not associated with AVMs were also classified as a separate category [24]. They are extremely rare and their pathogenesis is based on a lesion of a dissection nature [25, 26]. The authors reported only 2 cases in their practice. Arteriovenous malformations were divided into AVFs and AVMs (Table 3) [24]. New categories were included in this classification: extradural AVFs, intradural diffuse AVMs and intradural conus medullaris AVMs. Extradural AVFs are characterized by a direct connection between the extradural branch of the vertebral artery and the extradural venous plexus in the cervical spine, which leads to the development of a high-flow fistula, congestion in the epidural venous system, spinal cord compression and, as a result, progressive myelopathy. Intradural diffuse AVMs are intramedullary AVMs with subpial extension in several segments of the spinal cord. Intradural conus medullaris AVMs have multiple afferents. They have several direct AVFs (which originate from the anterior and posterior spinal arteries) as well as glomus-type foci (which are usually extramedullary and pial), but they may also have an intramedullary component. They are always localized in the conus medullaris and may be located along its entire length. It is worth mentioning that intradural ventral AVFs correspond to type IV of the early classifications, intradural dorsal AVFs to type I, extradural-intradural AVMs to type III, and intramedullary compact and diffuse AVMs to type II [27].

RF Spetzler et al. in the provided classification propose treatment tactics using endovascular or microsurgical methods depending on the type of malformation [24]. Extradural AVFs are mainly treated endovascularly, as are extradural-intradural AVMs. In the latter case, microsurgery can only be used to eliminate spinal cord compression. For intradural AVFs, microsurgical interventions or embolization are recommended. Regarding small and medium intradural ventral AVFs, the authors propose performing surgical intervention, while large fistulas are better suited for occlusion by embolization. The authors point out that embolization can be performed for intramedullary AVMs, but microsurgical removal remains the basis of their treatment. For conus medullaris AVMs, microsurgery and embolization can be used. The experience of RF Spetzler et al. in the treatment of malformations of this type showed that the best results are achieved by aggressive embolization followed by resection [24]. The modified classification of RF Spetzler et al. (2002) was seriously criticized. Thus, the French endovascular neurosurgeon P. Lasjaunias sent an open letter to the editors of the "Journal of Neurosurgery" and categorically rejected the possibility of combining cavernous malformations and hemangioblastomas into one group of "vascular neoplasms". He also considered it inappropriate to distinguish between the categories of “intradural AVMs of the conus medullaris” and stated that it is incorrect to divide intradural AVFs into ventral and dorsal. In addition, he raised the question of the anatomy of the anterior spinal artery, arguing that the latter is located subpially, not subarachnoidally, therefore, in his opinion, subarachnoid hemorrhages from this artery are impossible [28]. In 2002, G. Rodesch, together with P. Lasjaunias and other authors, proposed their own classification, taking into account the primary nature of the malformation (a node or fistula in which the angioarchitecture changes over time). All spinal AVFs were divided into micro- and macrofistulas, and genetic categories were identified: hereditary lesions, non-hereditary lesions [29]. Subsequently, the modified classification of RF Spetzler et al. (2002) and the genetic classification of G. Rodesch et al. (2002) did not become generally accepted, since each of them shifted the emphasis towards microsurgery or endovascular surgery [30]. Another important classification was developed by Yu. A. Zozulya and E. I. Slynko in 2005. Depending on the anatomical features of SpAVM, the authors divided them into intramedullary, perimedullary, dural, epidural, intravertebral and mixed. Angiostructural (inflow and outflow pathways) and hemodynamic features of SpAVM were identified [31]. The classification turned out to be large, but took into account many important nuances.

The newest classifications

In 2011, L. Rangel-Castilla et al., studying extradural AVFs (earlier they were first identified by RF Spetzler et al. (2002) [24]), drew attention to the fact that even within this group of AVFs there are differences. They described radiological and clinical features in 8 patients with extradural AVFs and divided these AVFs into 3 types: A, B1, B2. With type A extradural AVFs, arteriovenous shunting occurs in the epidural space, but venous drainage extends intradurally. The latter causes venous hypertension and spinal cord edema. With type B1, shunting occurs in the epidural space without intradural venous drainage, but causes compression of the spinal cord and/or nerve roots with myelopathy and/or radiculopathy. Extradural fistulas of type B2 are also limited to the epidural space and do not have an intradural draining vein, but do not compress the spinal cord or root, and therefore do not cause symptoms [32]. In the differential diagnosis of dural AVF (type I) and extradural AVF type A, it is important to consider that dural AVF has a shunt within the root cuff and drains directly intradurally, without filling the epidural space, while extradural AVF type A has an arteriovenous shunt that lies in the epidural space, and the intradural draining vein comes from a venous sac ("lake"), which is located extradurally.

In 2017, K. Takai, based on a historical analysis of the classifications of SpAVM, proposed a new classification taking into account the data of B. Rosenblum et al. (1987), JJ Merland et al. (1980), B. Gueguen et al. (1987), JA Anson and RF Spetzler (1992), L. Rangel-Castilla (2011) [30]. Extradural fistulas were classified as type V AVM, with subtype VA being with intradural venous drainage and subtype VB being without intradural venous drainage.

This classification is easy to use and corresponds to the principles of construction of generally accepted historical classifications, but it also has shortcomings, which are determined by the anatomical features of the blood supply to the spinal cord at different levels. In his early study, K. Takai (2013) indicates that dural AVFs at the thoracic level may differ from dural AVFs at the lumbar level [33]. If we follow the history of the development of classifications and the identification of new types and subtypes of SpAVM, it becomes obvious that as neuroimaging technologies develop, new anatomical variants of SpAVM, previously not described in the scientific literature, may be identified over time.

Conclusion

All classifications are the result of a great deal of work by scientists from all over the world. The complex vascular anatomy of the spinal cord, the very high variability of vascular defects of the spinal cord, and their rarity have not allowed the creation of a single classification that could satisfy all the needs of the medical community dealing with this problem. Due to the rapid development of endovascular surgery and microsurgery, future classifications should not only take into account the anatomical variants of vascular lesions of the spinal cord, but also allow predicting the outcome of treatment based on its algorithm. The available data indicate that the prognosis of the natural course of the disease is extremely unfavorable, so the treatment of vascular lesions of the spinal cord seems justified and even necessary in most cases. Further study of vascular defects of the spinal cord, accumulation of experience will create a basis for optimal systematization, determination of modality and successful treatment of this disease.

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