

Clinical characteristic of spinal vascular malformation in pediatric patients

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Abstract

Objective Seventy-two pediatric spinal vascular malformation cases were reviewed and the characteristics of their clinical symptoms, diagnoses, and therapies were analyzed.

Materials and methods A thorough overview was compiled examining patient sex, age, location, history, development, treatment, clinical, and anatomical results.

Results Spinal cord arteriovenous malformation was the most common (44.4%) subtype to be seen in these pediatric patients, while subdural perimedullary arteriovenous fistula (23.6%) was the second, followed by Cobb's syndrome (13.9%) and intramedullary cavernous angioma (5.6%). No spinal dual arteriovenous fistulae were found in infants. The highest incidence was seen during the infant and adolescent periods. Sixty-nine cases were treated by surgeries, embolizations, or a combination of both, and 71.5% of them had improved.

Conclusions Early diagnosis and treatment are required. Surgery and embolization, or a combination of the two, are the current candidates for treatment.

Keywords Pediatric · Spine · Spinal cord · Vascular malformation

Introduction

Spinal cord vascular malformation is a rare disease, and it accounts for 2–4% [1] of spinal diseases. The diagnosis and treatment of this disease have been significantly advanced following the development of magnetic resonance imaging

(MRI) and digital subtractive angiography (DSA), which have been widely used in current clinical settings. However, there is much controversy regarding this disease's categorization, etiology, pathophysiology, and treatment [2–4].

Nearly all the diseases have different clinical features when comparing between children and adults, which is due to the fact that children have their own anatomical and physical characteristics. As they develop, the clinical features of a disease change and tend to progress toward the presentation of an adult. Thus, the study of a childhood disease will benefit not only young patients but also adults.

Materials and methods

Of 1,168 spinal vascular malformation cases, 72 were children <14 years old and these cases were collected from 1986 to 2007. Pediatric patients were divided into five approximate age groups. Group 1 was the neonate period (0–≤1 year) from birth to 1 year old, which is the quickest growth period throughout life. Group 2 was the infant period (1–≤3 years), whereby the growth rate slows down and the baby can move and touch the environment by himself. Group 3 was the preschool age (3–≤7 years), when the intellectual capabilities of the child develop rapidly. Group 4 was the school age (7–≤12 years), when almost all the organs and systems develop as a “young adult,” except for the reproductive system. Group 5 was the adolescent period (12–≤14 years), which is the second most rapid growth stage after birth.

The classification of spinal cord vascular malformation used in the study was adopted from Ling and is listed in Table 1, which had been used in all our studies of spinal cord vascular malformation.

The functional assessment of the spinal cord used in the study was the modified Aminoff and Logue scale (Table 2), which was also utilized in our related studies.

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Table 1 Classification of spine and spinal cord vascular malformation

1. Intradural lesions
(1) Cavernoma
(2) SCAVM, I intramedullary, II perimedullary, III intra-perimedullary
(3) Perimedullary arteriovenous fistula (SMAVF), type I, type II, type III
(4) Spinal aneurysm
2. SDAVF
3. Intraspinal-vertebral canal epidural lesions
(1) Cavernoma
(2) Arteriovenous malformation
4. Lesions outside canal
(1) Perivertebral canal arteriovenous malformation (PVAVM)
(2) Perivertebral canal arteriovenous fistula (AVAVF)
5. Vertebral body angioma
6. Cobb's syndrome
7. Syndrome-accompanied spinal cord vascular malformations
(1) KTW syndrome
(2) ROW syndrome

On the basis of the modified Aminoff and Logue scale score of disability, the scoring of spinal function (Table 3) and the therapeutic effectiveness criteria (Table 4) were employed.

The treatment strategies employed in this study included both interventional and surgical therapies. The selection of methods for each case was individually decided according to his/her illness and the desire of his/her parents.

Results

Seventy-two pediatric (age <14 years) cases of spinal cord vascular malformations were included in the study. These

Table 2 Modified Aminoff and Logue scale score of disability

Classification of gait disturbance
Grade 0: normal gait and activity
Grade 1: leg weakness or abnormal gait, no restricted activity
Grade 2: grade I with restricted activity
Grade 3: requires cane or similar support for walking
Grade 4: requires walker or crutches for walking
Grade 5: unable to stand, confined to bed or wheelchair
Classification of micturition
Grade 0: normal
Grade 1: hesitance, urgency, or frequency
Grade 2: occasional urinary incontinence or retention
Grade 3: total urinary incontinence or retention
Classification of defecation
Grade 0: normal
Grade 1: slight constipation, react to laxation
Grade 2: occasional incontinence or severe constipation
Grade 3: total incontinence

Table 3 The spinal functional score

Excellent: normal or almost normal, total score ≤ 2
Good: slight functional disturbances, total score ≤ 6
Fair: moderate functional disturbances, total score 6–8
Bad: severe functional disturbances, total score 9–11

patients accounted for 6.16% of all 1,168 cases of spinal cord vascular malformations collected in our center and there were 47 boys and 25 girls (a boy-to-girl ratio of about 2:1). The onset age was an average of 9 years old. The main symptoms included disturbances of movement and sensory, bowel, and bladder disorders. From the moment of onset, 52 cases (72.7%) appeared to possess different kinds of movement disorders. In addition, 15 cases (20.8%) had difficulties in defecation and urination. Hypoesthesia was seen in four cases (5.6%). Pain in the neck, thorax, and back occurred in 16 cases. Also, six cases were accompanied with headache and two went into a coma. These last two cases suffered from subarachnoid hemorrhage as confirmed by neurological images.

Furthermore, there were additional rarely occurring symptoms. An 11-year-old boy was diagnosed with appendicitis due to his sudden onset of abdominal pain. Subsequently, an appendectomy was performed. After the operation, paraplegia and incontinence were noticed and spinal cavernoma was diagnosed. Another 6-year-old girl suffered from a sudden onset of abdominal pain that was co-presenting with paraplegia and incontinence. A 14-year-old boy who suffered from Cobb's syndrome (spinal angiomas) had his left side's extremities underdeveloped since he was 7 years old. However, no other spinal cord symptoms were observed. Finally, there were two cases that manifested tics in their extremities.

Most of the spinal vascular lesions presented with a sudden onset, which was a commonly seen pattern in 52.8% of pediatric patients in this series. In contrast, 30.6% of cases suffered from a progressive disease course. All of the cavernous angioma cases and 55.6% of the spinal cord arteriovenous malformation (SCAVM) cases in the study presented with a sudden onset. For the perimedullary fistula and Cobb's syndrome cases, the onset patterns (sudden versus progressive) were about equal.

The average onset age was 9 years old (1 month to 14 years) and the average treatment age was 11 years old. The age distribution of the series is demonstrated in Fig. 1. It can be observed that there was a reduction in morbidity between 3 and 4 years of age. In addition, there was a peak in the morbidity noticed at 1 year old.

Table 5 shows the disease distributions among the various age groups. Each disease state had a different morbidity for each age group.

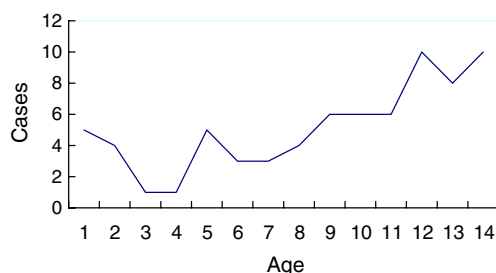
Table 4 Criteria of therapeutic effectiveness

Cure: total or more than 95% disappearance of the lesion, excellent spinal function
Improve: (1) score 3–5, (2) excellent spinal function at follow-up with more than 5% lesion residual
No change: score no change at pre- and post-treatment except excellent spinal function
Deterioration: score increase

The spinal cord can be divided into the following regions according to their blood supply: (1) cervical region, which includes all eight cervical segments; (2) upper thoracic region, which includes the first seven segments of the thoracic cord; and (3) thoracic and lumbar region, which includes all other cord segments from thoracic spinal nerve 8 (T8) to the conus. Lesions were found at the following regions in the series: 16 lesions were located in the cervical region, 15 in the thoracic region, and 41 in the thoracic and lumbar regions. The most involved segments were from T8 to L2, and there were 38 lesions found in these segments. Only two perivertebral arteriovenous fistula and one vertebral body angioma were located below the L2 segment. Table 6 shows the disease distributions among the different spinal regions.

Spinal cord arteriovenous malformations were often found in the thoracic and lumbar segments, although they could be seen throughout the various regions. In contrast, the perimedullary arteriovenous fistulas were concentrated in the boundary area between the thoracic and lumbar segments and the cervical region.

Of the 72 children with spinal vascular lesions, there were 32 cases of SCAVMs (44.4%), 17 perimedullary arteriovenous fistulas (23.6%), ten Cobb's syndromes (13.9%), four spinal cord cavernous angiomas (5.6%), three perivertebral arteriovenous fistulas (4.2%), three vertebral angiomas (4.2%), two epidural arteriovenous malformations (2.8%), and one spinal cord aneurysm in the study. Among the 17 perimedullary arteriovenous fistulas, three cases belong to type I, three to type II, and 11 to type III.

**Fig. 1** Age distributions of spinal cord vascular malformations in children

Satisfactory DSA data could be acquired for 45 cases among the 59 cases with subdural spinal cord vascular lesions (SCAVMs and superior mesenteric arteriovenous fistulas (SMAVFs), Cobb's syndrome). The arterial feeders of these lesions were identified as follows: six lesions were supplied by one feeding artery, 15 lesions by two feeding arteries, eight lesions by three feeding arteries, six lesions by four feeding arteries, six lesions by one feeding artery, and seven lesions by one feeding artery.

An aneurysm-like structure was found in 12 cases. Among these cases, seven were arteriovenous malformations, three were perimedullary arteriovenous fistulas, and two were Cobb's syndrome.

The treatment for pediatric spinal cord vascular malformation was almost the same as that for adults. Of 72 cases, 69 were treated by surgery, embolization, or a combination of both. The remaining three cases were not treated due to the refusal by the family.

Detailed clinical and imaging data could be obtained in 60 of 69 treated cases. According to the criteria for therapeutic effectiveness, there were 24 cases that had improved (40%), 31 with no change (51.7%), and five that deteriorated (8.3%) upon discharge. Importantly, there were no treatment-related deaths. Overall, there were eight cases treated by surgeries, 33 by embolizations, and 19 by both. The results of each procedure are listed in Table 7. The long-term follow-up results are not yet complete and will be published later.

The resolution of the lesions was achieved in 50% of treated cases according to DSA and/or MRI images. Fourteen percent of cases had less than 5% lesion area remaining as measured by these images. More than 5% of the lesion area remained in 36% of cases after treatment.

Discussion

Spinal and spinal cord vascular malformations are rare diseases and they account for only 2–4% of all spinal diseases. However, the morbidity is much less in pediatrics. Of 1,168 cases, 6.16% were examined in this study and complications from the disease include paraplegia and incontinence [5, 6]. Thus, it is important to pay attention to pediatric spinal cord vascular lesions.

There was no single case of spinal dural arteriovenous fistula (SDAVF) found in the study, although it is most commonly seen in adults, which accounts for 30–70% [7, 8] of all spinal vascular lesions. This phenomenon supports the view that SDAVF is an acquired disease rather than a congenital condition, in spite of some controversy.

Gerges Rodesch et al. have termed high-flow perimedullary fistula types II and III as macro-arteriovenous fistulas,

Table 5 Disease distributions among different age groups

Age group	Diseases, <i>n</i> (%)							
	Cavernoma	SCAVM	SMAVF	Spinal aneurysm	Epidural intracranial lesions	Perivertebral body lesions	Vertebral body angioma	Cobb's syndrome
Neonate		1 (20)	4 (80)					
Infant		2 (40.0)	2 (40.0)				1 (20.0)	
Preschool	1 (8.3)	6 (50)	4 (33.3)					1 (8.3)
School	2 (6.3)	15 (46.9)	6 (18.8)	1 (3.1)	1 (3.1)	2 (6.2)	1 (3.1)	4 (12.5)
Adolescent	1 (5.6)	8 (44.4)	1 (5.6)		1 (5.6)		2 (11.1)	5 (27.8)
Total	4	32	17	1	2	2	4	10

Table 6 Distributions of lesions among different spinal regions

Diseases	Region distributions (cases)			Total, <i>n</i> (%)
	Cervical, <i>n</i> (%)	Upper thoracic, <i>n</i> (%)	Thoracic–lumbar, <i>n</i> (%)	
Cavernoma	1 (25.0)	2 (50)	1 (25)	4 (100)
Spinal AVM	7 (21.9)	7 (21.9)	18 (56.3)	32 (100)
Perimedullary fistula	4 (23.5)	1 (5.9)	12 (70.6)	17 (100)
Spinal aneurysm			1 (100)	1 (100)
Epidural intracranial lesion		1 (50)	1 (50)	2 (100)
Perivertebral body lesion	1 (33.3)		2 (66.7)	3 (100)
Vertebral body angioma	1 (33.3)		2 (66.7)	3 (100)
Cobb's syndrome	2 (20)	4 (40)	4 (40)	10 (100)

Table 7 Treatment options for diseases

Diseases	Options				Total, <i>n</i> (%)
	No treatment, <i>n</i> (%)	Embolization, <i>n</i> (%)	Surgery, <i>n</i> (%)	Embolization+surgery, <i>n</i> (%)	
Cavernoma			4 (100)		4 (100)
Spinal AVM	2 (6.3)	20 (62.5)	4 (12.5)	6 (18.8)	32 (100)
Perimedullary fistula		7 (41.2)	1 (5.9)	9 (52.9)	17 (100)
Spinal aneurysm		1 (100)			1 (100)
Epidural intracranial lesion	1 (50)	1 (50)			2 (100)
Perivertebral lesion		3 (100)			3 (100)
Vertebral body angioma		1 (33.3)	1 (33.3)	1 (33.3)	3 (100)
Cobb's syndrome		6 (60)		4 (40)	10 (100)
Total	3 (4.2)	39 (54.2)	10 (13.9)	20 (27.8)	69 (100)

while the low-flow type I fistula was considered a micro-arteriovenous fistulas. This group suggested that the former was highly related to the hereditary hemorrhagic telangiectasia (HHT) [9, 10]. In our series, 80% (4/5) of patients younger than 1 year and 50% (2/4) between 1 and 3 years old suffered from perimedullary fistulas that were type II or III. Thus, it is implied that these children suffering from fistula might have HHT. The diagnosis of HHT in infants is not easy because classic symptoms of the disease appear later in development. Nevertheless, the perimedullary fistulas are often the first manifestation of the child's disease state, and it is reasonable to consider that children who suffer from perimedullary fistulas may also have HHT.

The incident of pediatric spinal vascular malformations had several features. There was a high incidence from birth to 2 years old followed by a reduction until 5–6 years old. The second highest rate was seen at age 12, and this was an explosive increase in rate. Most of spinal vascular lesions are congenital except SDAVF. In addition, vascular malformations are often formed at the third embryonic week [11]. The infant and adolescent periods are the two most developmentally rapid stages in human beings. The fast growth of the spine and relatively slow growth of the spinal cord leads to the imbalance of their blood supply. Hence, dramatic hemodynamic changes in blood flow and changes in the vasculatures might trigger the onset of these diseases. In addition, adolescent children have a greater chance to take part in activities. Thus, these factors might lead to higher incidence at these stages.

Moreover, the onset pattern in pediatric patients was different from that of adults. The sudden onset of the disease was common in the former group, while the slow progressive disease course was most commonly seen in the latter one [12]. In addition, there was no SDAVF, which is a slow progressive disease found in the study of pediatric patients that might lead to an increased incidence of sudden onset. Except for SDAVFs, the sudden onset rate of pediatric spinal vascular malformations (including SCAVMs, perimedullary fistula, spinal aneurysm, Cobb's syndrome, Klippel–Trenaunay–Weber (KTW) syndrome, Rendo–Osler–Weber (ROW) syndrome, and Roberston's syndrome) was 54.5% of patients, while the slow onset rate was 31.3% of patients. In adults, the rates were 33.5% and 44.4%, respectively [13]. These results clearly demonstrate that pediatric patients had a high sudden onset rate. The mechanisms for the sudden onset of the disease included the following: (1) intramedullary hemorrhage and subarachnoid hemorrhage, (2) acute infarction within malformations, and (3) hemodynamic changes from the malformations [14, 15]. Among these reasons for the sudden onset, hemorrhage was the most important cause. Mechanisms of slow onset include the following: (1) congestion in the veins of the spinal cord, (2) blood flow loss, and (3) mass effects of the

lesions [16]. Previous findings have demonstrated that the hemorrhage rate was higher in pediatric cases of spinal vascular malformations than that in adults [17]. Hence, this might explain the high sudden onset rate. But other reports have shown that intramedullary and subarachnoid hemorrhages in infant patients were not higher than that in adults, despite the high incidence of sudden onset [18]. It was reasonable to consider that significant changes of the spine and cord length proportions as well as changes of the vasculature played a role in the presence of the sudden onset pattern in rapidly growing children. In addition, the limitations of comprehensive and expressive abilities of children might inhibit their communications with parents and doctors. Thus, it was difficult to find their signs of discomfort until significant symptoms appeared. Accordingly, this led some slow progressive courses to appear as sudden onset.

Only 4.17% (two cases) of all cases in the study recovered after the onset but before treatment, while 27.78% (20 cases) partially recovered. But after treatment, the early recovery rate was 60%. Therefore, it is necessary to diagnose and treat these diseases as early as possible to achieve better outcomes.

Conclusions

Pediatric spinal cord vascular malformations exhibited many differences in clinical features compared to that of adults. The natural history of this disease is progressive deterioration without spontaneous improvement. Thus, early diagnosis and treatment of this disease is required. Surgery and embolization, or a combination of the two, are the current candidate approaches for treatment. After appropriate therapy, most patients improve in their condition or are cured.

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