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Perimedullary arteriovenous fistulas in children: report on six cases

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Perimedullary arteriovenous fistulas in children: report on six cases

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Abstract

Background Perimedullary arteriovenous fistulas (PMAVFs) are rare spinal lesions and even more uncommon in children.
Objective The aim of this study was to document rare occurrences of this type of arteriovenous malformation in six children treated at our institution.
Methods The clinical data, radiological findings, and treatment in six cases of PMAVFs were reviewed. Six patients with PMAVFs were managed at our institution over a 5-year period. The patients (four girls and two boys), ranging in age from 6 to 15 years, presented with initially fluctuating, and eventually permanent and progressive, sudden-onset paraparesis, sensory disturbances, and sphincter dysfunction. The duration of symptoms before diagnosis ranged from 1 week to 13 years.
Results All the patients underwent magnetic resonance imaging and spinal selective angiography, which demonstrated the characteristic imaging of an arteriovenous fistula. Embolization of the arteriovenous fistula was initially attempted in three patients with successful occlusion of the fistula in two. For the remaining cases, open surgery was performed, with complete occlusion of the fistula. There was no morbidity, regardless of the treatment performed. All the patients experienced neurological improvement after treatment.
Conclusions No specific clinical or radiological characteristic of PMAVFs in the pediatric population was observed when our series was compared with a general series. Early diagnosis and timing of the therapeutic intervention seemed to avoid the development of irreversible ischemic myeloradiculopathy and prevented hemorrhage. Treatment for PMAVFs is difficult to standardize because these are extremely rare lesions with different angioarchitecture configurations.

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Introduction

Spinal cord arteriovenous malformations (SCAVMs) are relatively rare lesions and constitute only 1% to 2% of vascular neurological pathological conditions [34]. They account for 3% to 12% of spinal space-occupying lesions [29, 50] and are about one tenth as common as intracranial vascular malformations [14, 49]. Only 13% of all cases of SCAVMs reported in various series were under 20 years of age [63].

According to their anatomical and pathophysiological factors, SCAVMs may be classified into four types: type-I SCAVMs are dural arteriovenous fistulas (AVFs); type-II

and type-III SCAVMs are intradural arteriovenous malformations (AVMs)—glomus and juvenile, respectively; and type-IV SCAVMs are intradural and extramedullary—so-called perimedullary arteriovenous fistulas (PMAVFs) [5, 32, 65].

PMAVFs were first described by Djindjian et al. [16] in 1977 as direct fistulous connections located on the pial surface of the spinal cord, between the anterior spinal artery (ASA) and/or posterior spinal arteries (PSA) and the medullary veins, without any intervening nidus. PMAVFs constitute about 4% to 38% of all SCAVMs [7, 9, 16, 18, 23, 24, 47, 60] and are even rarer in children. The estimated prevalence of PMAVFs in the pediatric population is around 0.5% to 3.25% [42]. Presentation in the first 2 years of life is exceedingly rare [9, 15, 31, 46, 53, 62, 69]. These lesions become clinically manifested in the third to sixth decades of life and do not demonstrate any gender preference [9, 19, 24, 47]. They are most commonly observed in the thoracolumbar spinal region [11, 13, 16, 25, 43, 47, 53], although cervical and thoracic PMAVFs have also been reported [25, 37, 47]. There is little information in the literature on PMAVFs in children. A search of the literature and a review of the reports cited in the literature revealed 45 papers, describing about 79 patients younger than 16 years, out of 389 patients with PMAVFs. Here, we report on six consecutive cases of PMAVFs in children treated at our institution.

Clinical material and methods

Patient population

The study group included all pediatric patients treated for PMAVFs at the Division of Pediatric Neurosurgery, University of São Paulo Medical School, between 2000 and 2005. Data from clinical records, radiological studies, and follow-up examinations were available in all cases. The clinical characteristics of the six patients are summarized in Table 1. One patient (case 1) has previously been published individually [40]. There were four girls and two boys, and the mean age was 11.6 years (range, 6–15 years).

The clinical presentation consisted of subarachnoid hemorrhage (SAH) in two cases and progressive paraparesis or tetraparesis in four cases. Initially, they all presented with both sensory and motor lower-limb disturbances of a vague nature. Intermittency of symptoms was followed by bladder dysfunction. The patients' neurological status before and after treatment was classified according to the following criteria: (a) normal, i.e., no motor, sensory, or sphincter deficits; (b) good, i.e., the patient was able to walk independently, with only a few restrictions on activities; (c) fair, i.e., crutches or a cane was needed for walking, with occasional urinary incontinence or retention; and (d) poor, i.e., the patient was unable to stand and had incapacitating urinary incontinence or retention. The interval between the initial symptoms and treatment range from 1 week to 13 years (mean=5.8 years). None of the patients had any previous history of trauma or spinal surgery.

Radiographic evaluation

Plain radiographs of the spine were obtained for all patients. One patient (case 1) had shown progressive spinal deformity since before she was 1 year old. Plain radiographs showed enlargement of the spinal canal in one case.

MRI was the initial examination on all patients, and vascular abnormalities were always seen as a large serpentine signal void. In one case, a giant venous aneurysm compressed the cord. The spinal cord signal was abnormal in three cases, and two patients presented with hemorrhage.

Selective spinal angiograms (SSAs) were performed using a transfemoral approach, and children younger than 12 years were treated while under general anesthesia. Thoracic–abdominal aortography was initially performed to locate the malformation and to reduce the amount of irradiation. Nonionic contrast medium was injected. Selective injection was done by hand. Angiograms were obtained using digital equipment at an exposure rate of two frames per second, prolonged for 6 s when abnormal vascularity was identified. Supine anteroposterior views were routinely

Table 1 Summary of clinical findings

Case	Age (years)	Gender	Time elapsed until diagnosis	Clinical presentation	Neurological status
1	6	F	5 years	PRM	Poor
2	14	M	13 years	SAH 4×	Poor
3	11	F	4 years	PRM	Fair
4	12	F	3 years	PRM	Fair
5	15	M	4 years	PRM	Poor
6	12	F	1 week	SAH 1×	Fair

PRM progressive myeloradiculopathy, *SAH* subarachnoid hemorrhage

obtained, with additional lateral views for radiculomedullary arteries. The radiological findings are summarized in Table 2

The main arterial supply was derived from the spinal arteries in all cases, except case 1, in which the PMAVF was supplied directly from two radiculomedullary arteries with no connection to the spinal cord vasculature. Three cases were supplied with more than one radiculomedullary feeder, and three cases presented with only one radiculomedullary artery. The number of feeder arteries ranged from one to four (average=1.8). The venous drainage was regional in all cases through the perimedullary pial veins. The drainage veins were severely dilated in two cases. These cases demonstrated venous aneurysm with the radiological features of an intraspinal space-occupying lesion.

The conus medullaris was involved in three patients, thoracic spinal cord in two, and cervical spinal cord in one. All PMAVFs were located ventrally to the medulla, except one case that was posterolateral. The cases were classified according to Anson and Spetlzer's [5] SCAVM classification: five patients as type IV-B and one patient as type IV-C (Table 3).

Treatment

Treatment was recommended on an individual basis and not in accordance with a protocol. The aim of the treatment was to occlude the AVF, with preservation of the normal spinal cord perfusion and function. For each patient, a discussion between the interventional neuroradiologist and the neurosurgeon led to decisions regarding the treatment. Initially, endovascular treatment was considered to be the first-line therapy for all cases, but depending on the arterial feeder (type, location, size, flow, and tortuosity) and the magnitude of the venous drainage, direct surgical occlusion of the fistula was performed instead. Initial endovascular therapy, using n-isobutyl-cyanoacrylate (NBCA) glue, was attempted in three patients, but total occlusion of the fistula was only successful in two cases. In the third case, because of

the tortuosity and long course of the ASA, endovascular occlusion of the AVF was unsafe and the patient underwent microsurgical occlusion of the fistula. In general, surgery was preferred for PMAVFs located at the conus or posteriorly on the spinal cord. Initial microsurgery was performed on three patients: two cases located at the conus and one case located posterolaterally on the spinal cord. Additional reasons for choosing microsurgical treatment without even attempting endovascular therapy were the tortuosity and long course of the ASA in two patients (cases 3 and 4) and high blood flow associated with a large aneurysm in one case (case 1). In case 2, the endovascular treatment failure was caused by difficulty in navigating to the point of the arteriovenous fistula due to the tortuosity of the ASA and by the risk of releasing the glue in a high-flow fistula. Surgical exposure of the lesions was performed through a posterior approach via laminotomy. Careful dissection was done to expose and correctly identify the feeder arteries, and then, the fistulous connections were clipped and closed. The fistula site was identified by the sudden change in the caliber of the vessel. Usually, there was an aneurysmatic dilatation on the venous side. The PMAVF was exposed through the rootlets of the cauda equine or by gentle rotation of the conus to view the ventral surface of the conus. No intraoperative spinal monitoring was performed in the present series. SSA was performed during the follow-up period.

Results

The clinical and angiography results before and after treatment are presented in Table 3.

Clinical results

Findings from the clinical follow-up were available for all the patients. The clinical follow-up was performed every year after the treatment, for at least 2 years. The neurological condition of all of these patients improved.

Table 2 Radiological findings

Case	Location	Situation	Artery	Feeders	Drainage
1	Conus	Anterior	RMA	Left-L1 Left-L2	Huge venous dilatation
2	Conus	Anterior	ASA	Right-L2 Right-T11 and right-T12	Venous ectasia
3	Conus	Anterior	ASA	Left-T8	Venous ectasia
4	Thoracic	Posterolateral	ASA+PSA	Left-T9 Right-T11	Venous ectasia
5	Thoracic	Anterior	ASA	Left-T8	Venous ectasia
6	Cervical	Anterior	ASA	Left-C6	Venous ectasia

ASA anterior spinal artery, *PSA* posterior spinal artery, *RMA* radiculomedullary artery

Type	Endovascular	Surgery	Results	Outcome neurological status
IV-C	−	+	Total occlusion	Fair
IV-B	+	+	Total occlusion	Fair
IV-B	−	+	Total occlusion	Good
IV-B	−	+	Total occlusion	Good
IV-B	+	−	Total occlusion	Good
IV-B	+	−	Total occlusion	Good

There were no new neurological deficits or any increases in existing deficits. A normal condition was seen in one patient, good results were observed in three patients, and fair results with moderate motor deficit and sphincter dysfunction were observed in two patients. The recovery was progressive over a period of 6 months. No cases of late deterioration were observed. None of the six patients suffered any serious neurological complications, and there were no deaths.

Therapeutic results

Three patients underwent initial therapy consisting of endovascular embolization, and complete occlusion of the fistula was obtained in two cases (a representative example after embolization alone is illustrated in Figs. 1 and 2). In one of these three patients, the endovascular therapy failed to achieve catheterization of the feeder artery. Four patients underwent surgical ligation of the PMAVF through laminotomy, with complete occlusion in all of them (a representative example after microsurgical occlusion is shown in Figs. 2, 3, 4, 5, and 6). There were no surgical

failures. No patients had to undergo reoperation.

Complete PMAVF occlusion was obtained in all patients. No early or late revascularization of completely treated PMAVF was observed. The patients demonstrated significant improvements in gait and bladder function after the surgery. No complications relating to either surgery or embolization for management of PMAVFs were demonstrated.

Postoperative SSA demonstrated total occlusion of the AVF in all cases. Angiography follow-up was performed 1 week and 1 year after treatment. The clinical status of all patients improved after the treatment.

Discussion

Clinical features

According to the anatomical, angiographic, and hemodynamic features of PMAVFs, they were subcategorized as three distinct types (1, 2, and 3) by Gueguen et al. [19] in 1987. They were later reclassified by Anson and Spetzler

Fig. 1 (Case 5) Angiography images showing selective injection into the left T8 intercostal artery. a An early arterial phase image revealing the enlarged left T8 posterolateral radiculomedullary artery and the ASA (arrow) feeding the fistula. b Late-phase angiography image showing filling of the enlarged, tortuous pial venous plexus, and medullary veins
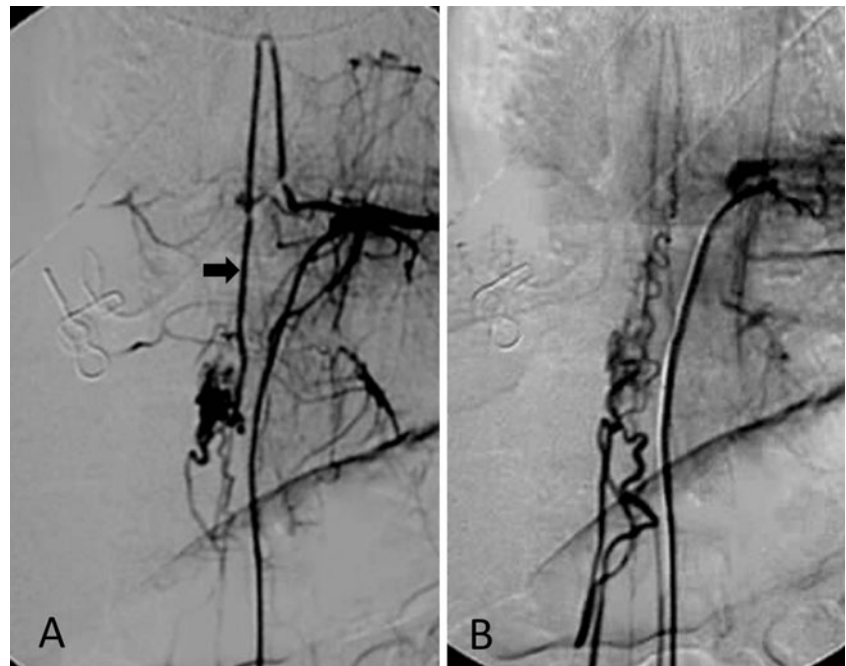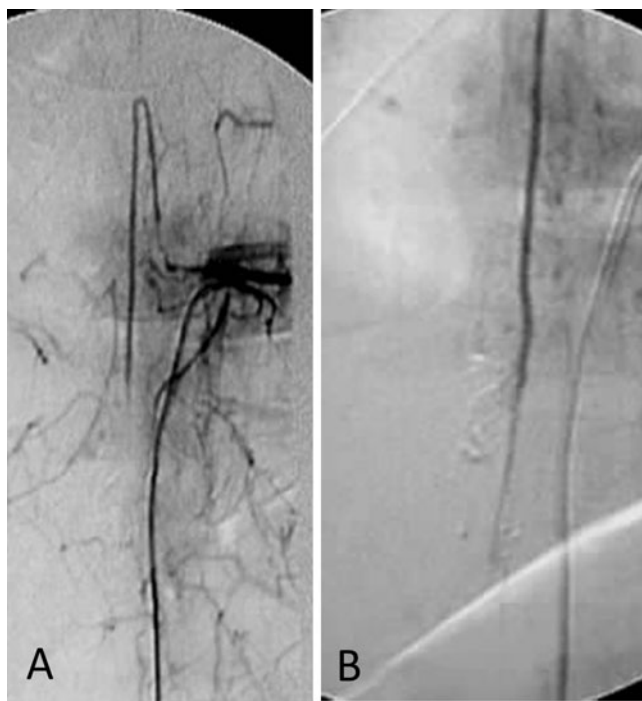
Fig. 2 (Case 5) After embolization: selective image of the radiculo-medullary artery originating from the left T8 intercostal artery. **a** Fistula occlusion with patent ASA up to the level of the conus medullaris. **b** Close-up image of the ASA showing fistula occlusion

Fig. 3 (Case 4) Sagittal T2-weighted MRI on the thoracic and lumbosacral spine showing serpentine flow voids in the subarachnoid space that represent enlarged blood vessels along the spinal cord and cauda equina

[5] in 1992 as SCAVM types IV-A, IV-B, and IV-C, respectively. In 2002, Spetzler et al. [65] classified PMAVFs as intradural ventral AVFs and maintained the three subtypes. Type IV-A consists of small and slowly flowing fistulas, fed by single artery, which is usually the end portion of the ASA, while the venous drainage is through minimally dilated, tortuous ascending perimedullary veins. Type IV-B consists of larger fistulas with one or more dilated arterial feeders, most frequently supplied by either the PSA or the ASA (or both), with drainage directly into an abnormally dilated and tortuous venous system containing a relatively high flow of arterialized blood. Type IV-C is nourished by multiple dilated arteries, with extremely rapid flow in the fistula and with giant dilated veins. The most frequent of the type-IV SCAVMs are types IV-B and IV-C [7].

The age of presentation is very controversial in the literature. Many larger series in the literature group all the patients together. The mean age of presentation has been found to range from 19.5 to 45 years [2, 13, 20, 21, 25, 33].

PMAVFs are reported relatively rarely in children. Their rate of occurrence has been found to range from 16.6% to 41.1% in general series [13, 16, 19, 48]. The ratio of pediatric-to-adult PMAVFs in the literature ranges from 0.1 to 0.5 [7, 16, 23]. Our study revealed that the greatest occurrence was during the second decade of life. Neonates and infants rarely present with PMAVFs [9, 15, 31, 46, 53, 62, 69].

Female predominance was present in our series (female-to-male ratio=2), which was the opposite of the findings in previous reports on pediatric series [42, 67].

As was observed in our series, these lesions can occur anywhere along the spinal axis but most commonly in the thoracolumbar region, particularly at the conus medullaris and, to a lesser extent, in the upper cervical region [2, 13, 21, 71]. They are most often located ventrally at the level of the conus medullaris or cauda equina [13, 25, 33, 47]. Their occurrence in this location may result from a disorder of the complex embryological development that underlies formation of the conus medullaris and its unique vascular anatomy [29]. The rate of occurrence of PMAVFs in the caudal region ranges from 58.8% to 100% in some series [13, 19, 20, 25, 57]. Conus medullaris lesions were separated into a different category because they are fed by multiple arteries, either ASA or PSAs (or both), or directly through radiculomedullary vessels [32, 40, 65].

The origin of PMAVFs is unknown. Although the etiology is not clearly determined in most instances, there are well-documented cases of both congenital and acquired type-IV SCAVMs [9, 11, 41, 61]. Most occurrences are presumed to be congenital lesions, as we could assume in our case 1. PMAVFs may be part of a more complex vascular malformative syndrome in 10.5% [41, 42], such as metameric Cobb syndrome, Weber–Osler–Rendu disease,

Fig. 4 (Case 4) Angiography images of selective injection angiography at left T9 (**a**) and right T11 (**b**) intercostal arteries, demonstrating filling of the dilated ASA, varix, and tortuous drainage veins. A sudden change in the caliber of the feeder artery demonstrates the fistula site (*arrow*). The venous system is dilated and tortuous, with sulcal veins, pial venous plexus, and medullary veins draining to the epidural venous plexus
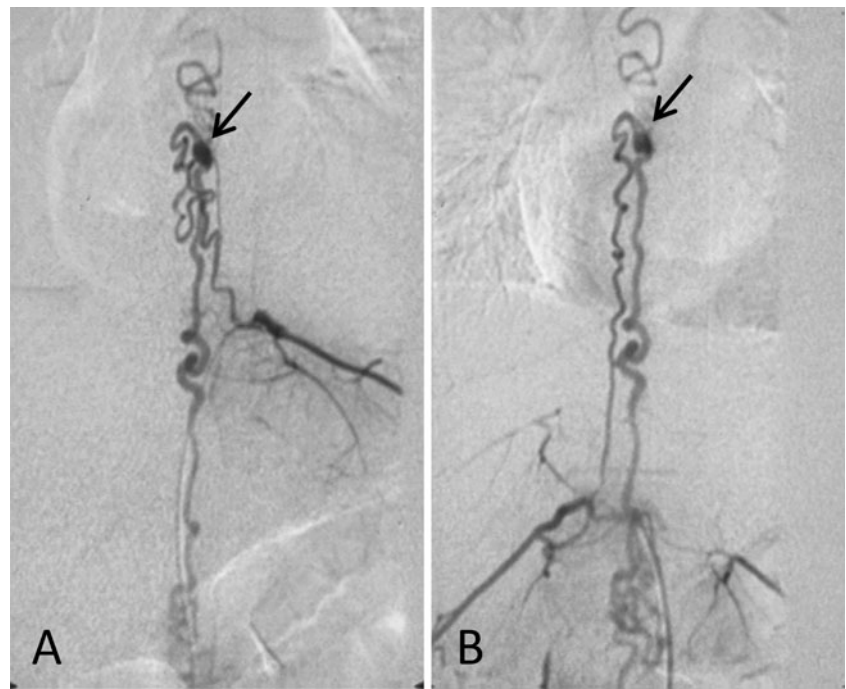
hereditary hemorrhagic telangiectasia, and Hirschsprung's anomaly [9, 15, 20, 44, 56, 57]. This occasional association with syndromes and genetic anomalies may corroborate their congenital origin [36, 57]. Associations with other congenital diseases have been reported, such as split cord type II malformations [73], lipomas of the conus medullaris [12, 75], and tethered spinal cord [25]. Acquired fistulas have been described following excision of conus medullaris ependymoma [9], removal of teratoma of the cauda equine [19], trauma [20, 41], and repeated lumbar myelography [70]. Medical histories may suggest an acquired etiology for spinal vascular malformation in 25% of the cases [8].

Although the majority of PMAVFs are considered to be congenital in origin, few cases are diagnosed during infancy [9, 15, 46, 53]. Congenital PMAVFs are rarely diagnosed during infancy and childhood, either because they do not cause symptoms or because the symptoms are misinterpreted [26]. Symptoms in infants may either be unspecific, like failure to thrive and psychomotor developmental delay, or be transient enough to be of concern to the child's parents. Early diagnosis is rarely made, and the resulting complications arising from the AVM may have devastating long-lasting neurological sequelae. Early diagnosis requires a high degree of clinical suspicion because SCAVMs represent an uncommon cause of progressive myeloradiculopathy. Unless there are acute neurological findings or congenital anomalies indicative of intraspinal pathology, the diagnosing of SCAVMs in children will remain very difficult and will often be delayed by an average of 4.9 years according to Scarff and Riegel [63], 9 years according to Mourier et al. [47], and 5.8 years as demonstrated in our series.

With recent advances in diagnostic neuroradiology, it may be expected that greater number of patients will be diagnosed at an early stage. We believe that both the rarity of these lesions and the physicians' unfamiliarity with the disease accounted for the delayed diagnosis in our patients.

Fig. 5 (Case 4) Intraoperative images showing: **a** fistula site (*arrow*) and varix dilatation, and **b** coagulation at the fistula site
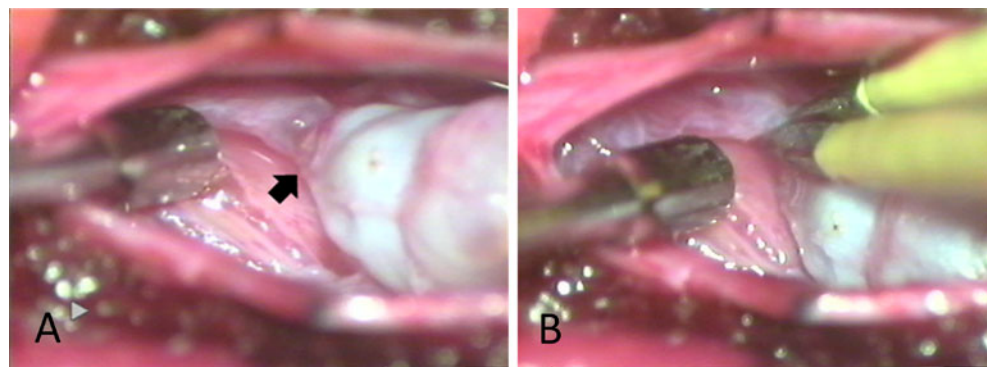
Fig. 6 (Case 4) Control angiogram obtained after surgery, demonstrating complete disappearance of the PMAVF: left T9 and right T11, postoperatively

Patients with PMAVFs usually have progressive or fluctuating neurological dysfunction; these features were observed in four cases reported in the present study.

It appears that ischemia due to shunting of blood into the malformation, i.e., the so-called vascular steal phenomenon, was the principal factor in provoking symptoms in these extramedullary malformations [1, 2, 9, 16, 22, 71]. Other pathophysiological processes involved in the genesis of neurological dysfunctions include venous hypertension, hemorrhage secondary to rupture of a feeder artery aneurysm, and the mass effect from large draining veins and varices [1, 25, 27].

Venous hypertension tends to be associated with spinal cord AVFs and is most likely responsible for development of progressive myeloradiculopathy [1, 9, 22, 32, 71]. Gradual worsening of myelopathy due to increased venous pressure and/or venous varix-induced spinal cord compression has been demonstrated by some authors [9, 23, 40, 42]. Acute onset of symptoms seems to be more frequent in children [42]. Based on a report on two cases, Tender et al. [70] speculated that the venous hypertension may be correlated with deficiency of the medullary venous system.

Congestive myelopathy may occur with vasogenic edema, which may progress to subacute necrotizing myelopathy, also known as Foix–Alajouanine syndrome [27]. Direct cord or nerve root compression by hypertrophied vascular structures was clearly evident in one of our six cases and in four cases reported in the literature [10, 30, 68]. Hida et al. [23] reported the presence of varices compressing the spinal cord in 45% of their cases. Varices are thought to be generated by two factors: venous hypertension and lack of intraparenchymal tissue pressure [23].

The initial symptoms may be leg weakness, sensory disturbance, back pain, or sphincter disorders [2, 22, 57, 71]. Patients with cervical PMAVFs may have episodic headache at the beginning of the history [74].

The majority of these patients present with progressive neurological deficit, although subarachnoid hemorrhage (SAH) may precipitate the symptoms, usually with sudden onset [19, 20, 25, 57, 62]. The incidence of hemorrhage is about 10–40% [19, 20]. Hemorrhage seems to be more frequent with types IV-B and IV-C lesions [47], and cervical type IV-A is probably related to high mobility of this segment of the spine [24, 62]. Hemorrhage may occur in type IV-C in up to 50% of the cases [48]. It may be suggested that flow plays a role in the development of hemorrhage in PMAVFs, given that types IV-A and IV-B bleed only infrequently. Aneurysmal dilatations, which are usually venous, occur in about one third of intradural spinal AVMs and may be associated with a higher percentage of hemorrhagic presentation. Rodesch et al. [58] reported that children with SCAVMs had a greater tendency to bleed than adults did (70% vs. 45%). SAH alone may produce headache and meningeal irritation that is indistinguishable from SAH of intracranial origin, and even hydrocephalus may result [25, 31, 37, 57]. The bleeding mechanism is unknown. Venous impairment probably occurs due to high flow and venous hypertension [57]. Intraparenchymal hematoma is very uncommon but represents an ominous prognosis [26].

The neurological examination may range in results from entirely normal to complete transverse myelitis. A high percentage of patients have some impairment of bladder control. A correct diagnosis is much more likely to be made in cases with a sudden onset, bloody spinal fluid, and neurological findings indicating some sort of myelopathy. Complaints typical of spinal lesions in older children and adults, such as radicular pain, sensory loss, and bladder and bowel problems, are usually not evident in infants and small children [10].

Diagnosis

PMAVFs are uncommon disorders associated with considerable difficulty in diagnosing them. MRI is the preferred diagnostic method for screening examinations on suspected SCAVMs and should be complemented by SSA. The MRI findings depend on the flow through the lesion, size of the draining veins, effect on the underlying cord, and presence

or absence of hemorrhage [22]. MRI on PMAVFs shows intramedullary and perimedullary serpiginous structures of signal voids produced by rapid blood flow on spin-echo images. The dilated subarachnoid vessels should not be mistaken for cerebrospinal fluid pulsation artifacts. PMAVFs are sometimes associated with intramedullary signal changes related to hemorrhage, ischemia, or venous congestion in the spinal cord [27]. MRI has become indispensable for defining the precise relationship with the spinal cord. This is extremely useful in selecting treatment options, planning operative strategies, and assessing the risk of the intervention.

SSA remains the gold standard for characterizing the angioarchitecture of spinal vascular malformations, although spinal arteriography is not without risk. Only spinal angiography can accurately demonstrate a PMAVF, by showing the high-flow shunt and the absence of interposed nidus between the arterial and venous sides of the vascular lesion. SSA is required to identify the various arterial feeders, their size and tortuosity, the flow, the exact location of the shunt and its morphology, and the venous drainage. It is important for the strategy of endovascular treatment to identify the exact number of arterial feeders. In our series, the mean number was 1.8, i.e., more than one feeder, just as indicated by other reports [42, 59]. Most of our cases were classified as type IV-B, and there was no association with genetic syndromes. As indicated by the literature, PMAVFs are more prone to associate with other genetic syndromes than other types of SCAVMs are [20].

The fistula site in cases of PMAVF may be identified by abrupt changes in the caliber of a blood vessel at the transition from feeder artery to draining vein [23].

Selective angiograms may guide the choice of the best approach towards embolization of the malformation. Occurrences of varices and/or shifts of the ASA greatly favor the diagnosis of PMAVF, rather than intramedullary AVM [23]. Thrombosis of PMAVFs may be precipitated by diagnostic angiography [6].

According to Mascalchi et al. [39], magnetic resonance angiography may help to detect intramedullary or perimedullary vessels that are occult or barely detectable by conventional MRI. It is a useful complement to MRI for detecting and characterizing spinal vascular malformations before digital SSA. It may provide information about residual or recurrent flow in abnormal peri- and intra-medullary vessels. An early follow-up examination is recommended, and this enables prompt detection of treatment failure and serves as a reference for follow-up examinations. Mourier et al. [47] cautioned that because of the small size and the nature of type-IV-A SCAVMs, neither MRI nor routine angiography may suffice to clearly demonstrate them. In their experience, tomo-myelography and angio-tomography played an important role in diagno-ses. Multidetector CT angiography is a recent imaging technique that can provide high-resolution and high-contrast images. This can locate the feeder vessels and the fistula, and greatly reduce the amount of time required for conventional angiography [35].

Treatment

Precise understanding of the angioarchitecture and patho-physiology of these lesions, combined with increasing imaging sophistication, endovascular capabilities, refined microsurgical techniques, intraoperative somatosensory and motor evoked potential monitoring, and intraoperative angio-grams, has resulted in an increasing percentage of successful elimination of these challenging lesions [32, 42, 73].

Several case series documenting the treatment options for these malformations have been published. The aim of the treatment is to obliterate the fistula while preserving the normal arterial supply to the spinal cord. Multidisciplinary treatment planning is mandatory, and it requires knowledge of the natural history of these vascular lesions. The decision as to which therapeutic method to use should be made by both the operating and the endovascular surgeons [32]. The treatment seeks to occlude the AVFs either surgically or by means of embolization. Closure of the proximal side of a feeder artery may not be enough to achieve complete cure of the AVF because many shunts are supplied by more than one feeder, as was shown in our series.

The currently recommended therapies include emboliza-tion alone using PVA particles [23], *n*-butyl-cyanoacrylate (NBCA) glue [13, 15, 51, 59, 72], balloons [54, 55, 57], platinum coils with or without silk [20, 48, 62], onyx [31], cellulose acetate polymer solution [66], direct conven-tional microsurgery alone, and combined embolization and microsurgery [23, 48].

Embolization presents the great advantages of continu-ous intraoperative angiographic monitoring and of absence of direct maneuvers on the parenchyma of the spinal cord. However, difficulties may rise from the small size of the pedicles and from the possibility of selective occlusion of the AVF in relation to the functional spinal arteries. Occlusion of these feeders, by means of embolic materials, may be dangerous because it is difficult to control the deposition of embolic material. In many instances, it is very difficult to advance the microcatheter close to the fistula site because of the size and tortuosity of the arterial feeders [23, 27, 47]. In one child of the present series, we had to abandon the endovascular procedure because of difficulty in catheterization and in navigating the microcatheter through the narrow and long course of the arterial feeder of the fistula. When navigation through tortuous medullary arteries is not possible, transvenous techniques may allow access to the draining varix [72]. Complications relating to

the transvenous route include thrombosis of the vein and pain relating to irritation of the adjacent nerve roots.

PVA embolic particles are safe and effective if appropriately sized particles are used [23]. However, particulate embolic material is not suitable for use in large direct fistulas because the particles may pass through the arteriovenous shunt. Under these circumstances, the emboli may filter the venous drainage, enlarge the fistula, and, in some instances, produce a massive hemorrhage [57].

NBCA glue is the most powerful embolic material, but this agent must be deposited accurately at the fistula site so as not to obstruct the perfusion of normal spinal cord tissue. PMAVFs of sizes ranging from small to giant have been successfully occluded by means of glue embolization [28, 51]. Polymerizing glue is considered by some authors to have a low recanalization rate, although the possibility that glue might pass through the fistula remains present, especially in relation to high-flow shunts [9].

Use of platinum coils with or without silk may produce complete occlusion of the shunt, although they are considered to be poor thrombogenic materials. Appropriate delivery and tight packing of the coils are necessary.

Balloon occlusion of AVFs is suitable in these lesions because of the angioarchitecture, with their single AV shunt. Balloon procedures seem to be safe and efficient regardless of the AVF location. The advantages of balloon occlusion are that when the caliber of the feeder artery is enlarged, balloons are easily carried by the flow to the fistula, even in very tortuous vessels, and they can be repositioned. The complications relating to balloon use that have been reported include migration of detachable balloons, infection with septicemia, and arterial rupture [20]. Deflation of the balloon with reopening of the fistula requires careful follow-up.

With regard to surgery, the difficulties basically relate to the location of the AVF, when it lies against the anterior aspect of the spinal cord [54]. A modified posterolateral approach for lesions involving the anterolateral pial surface of the spinal cord was proposed by Martin et al. [38]. An anterior cervical approach involving corpectomy at two levels was successfully used by Hida et al. [24], while complete corpectomy by means of thoracotomy was advocated by Anderer et al. [4] and lumbar corpectomy was used by Vitarbo et al. [73] in one case. A transmedullary approach was used by Giese et al. [17]. Surgery is indicated for AVFs when they are located at the conus medullaris, although complication may arise due to manipulation of the conus [9]. A sudden change in the caliber of the feeder artery going to the venous side demonstrates the fistula site. The aneurysmal dilatations are usually venous.

The treatment strategy can be based on the classification of the PMAVFs, and therefore, understanding of the angioarchitecture and pathophysiology of these vascular malformations is crucial for the treatment to be successful.

Separate subtypes of PMAVFs may necessitate different or combined forms of therapies [7, 9, 13, 20, 47].

An additional point to be considered with this fistula is the compressive effects and possibility of thrombosis of the large draining veins after occlusion of the fistula. Thrombosis of the collateral venous drainage or varix, or propagation of the thrombus, may compress the spinal cord or roots [20, 22, 48].

Dual treatment by means of embolization and a surgical operation is of value, particularly in the special circumstance where the malformation also causes compression of the spinal cord [16, 40, 48]. Compression of the spinal cord by large venous accumulations can be relieved through the operation. Most authors have considered that embolization is the first-line treatment method. Nonetheless, this procedure should be performed in specialized centers because there is a serious risk of ischemia of the spinal cord and worsening of the clinical neurological conditions. Although embolization of type IV-C is the recommended means of treatment, some reports have indicated good results through open surgical occlusion of the fistula [22, 40, 61]. Most PMAVFs in children are high-flow fistulas, and endovascular embolization should be the first choice for therapy [42, 67].

The importance of monitoring somatosensory cortical evoked responses secondary to peripheral nerve stimulation has been stressed by some authors [52]. Somatosensory evoked potential is used routinely and is quite useful as an aid in treating these vascular lesions. This method only measures dorsal column function, and anterior and/or lateral column transmission may be lost without a change in the evoked potentials. Ideally, techniques allowing monitoring of descending as well as ascending cord function would be more informative.

Intraoperative spinal angiography or color Doppler sonography may be of fundamental importance for identifying the fistula site [64]. Provocative testing with amytal and lidocaine may be used to find the most appropriate position for the catheter tip, and this may make it possible to detect the spinal cord arterial feeder. This procedure for younger patients has been performed under general anesthesia, and the provocative tests were evaluated using somatosensory evoked potential [20].

A very severe spinal deformity was identified at presentation in one case (case 1; reported elsewhere [40]), and in this case, the deformity was already present before treatment. Spinal deformity may also be correlated with postoperative complications in dealing with PMAVFs at pediatric ages [42].

SCAVMs are considered to be severe lesion with poor natural histories and prognoses if left untreated [3, 18, 59, 60]. Aminoff and Logue [3] reported that half of their patients became unable to walk within 3 years. PMAVFs

may also be associated with a poor prognosis if left untreated. Gueguen et al. [19] reported that if the intradural fistulas were left untreated, there would be a gradual progression to a disabling partial medullary syndrome, resulting in complete spinal transection within 7 to 9 years. The outcomes for type IV SCAVMs are as follows: types IV-A and IV-B are likely to respond well to surgery, but our knowledge is limited in terms of endovascular therapy, and type IV-C is likely to fare poorly from surgery but should have a better outcome from endovascular therapy [42, 45].

In our series, the neurological symptoms improved or regressed completely in all cases. Although we achieved good results, albeit with a delay in diagnosis, we think that early diagnosis is essential, along with removal of the shunt before there has been any progression to severe neurological deficits. The quality of the improvement is just as essential as early diagnosis and therapeutic action. Patients with severe disability within a short period showed the worst postoperative outcome.

Conclusions

There are relatively little data in the literature addressing the presentation of this disease in children. PMAVFs are rare and even uncommon lesions in children. In this study, we outlined the clinical course and treatment provided for six children.

The patients' ages ranged from 6 to 16 years (mean age= 11.6 years). The gender distribution comprised four females and two males. Four patients presented at admission with progressive myeloradiculopathy and two patients with subarachnoid hemorrhage. All the patients presented with severely compromised neurological status. The duration of symptoms prior to diagnosis ranged from 1 week to 13 years (mean time=4.8 years).

The diagnostic SSA study showed that most of the cases were fed by the spinal arteries (either ASAs or PSAs), but one case was nourished directly by radiculomedullary arteries that were not connected with the spinal arteries. Classification according to the Anson and Spetzler method demonstrated that five cases were graded as type IV-B, while one case was graded as type IV-C. All the children presented with some neurological impairment on admission. The treatment method was decided on a case-by-case basis, in conjunction with the endovascular therapist. Three patients were initially treated using embolization, with successful occlusion of the AVF in two cases. Three other patients and the case in which embolization failed were successfully operated on, with no morbidity. All the patients improved from their previous neurological status, regardless of the treatment method. Delayed diagnosis was apparently one of the factors that limited full recovery of

the neurological status. We believe that our technical limitations accounted for the delayed diagnosis in our patients. Although this experience at our unit produced good results, larger numbers of cases should be evaluated in order to establish a rational treatment method. The rapid evolution in endovascular techniques will probably avoid open surgery for the majority of these vascular spinal lesions. It is difficult to generate new guidelines for treating these lesions because of the small number of patients, the heterogeneity of the SCAVMs, and the significant evolution in endovascular technology.

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