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Case Report

Single hole, high-flow, spinal cord peri-medullary arteriovenous fistula (PMAVF) in a child, treated with transarterial embolization: A case report☆,☆☆,★

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ABSTRACT

Peri-medullary arteriovenous fistula (PMAVF) is a rare spinal vascular malformation that manifests as progressive neurologic deficits or hemorrhage in the spinal canal. We report a case of high-flow PMAVF in a child, with a single feeder artery and a large venous pouch, which was successfully treated with transarterial endovascular intervention. A 2-year-old boy was referred with a progressive 2-year history of myelopathy. The MRI revealed a large venous pouch at the midthoracic spinal cord with segmental surrounding edema. A spinal angiogram confirmed high-flow PMAVF with a single feeder artery from the anterior radiculomedullary artery, draining into the peri-medullary vein. The patient underwent transarterial embolization at the distal feeder artery, resulting in gradual motor strength improvement. PMAVF is classified as type IV spinal vascular malformation, usually presenting as a large, high-flow fistula with multiple feeders, although there was only one in this case. PMAVFs are intradural and may cause severe neurologic deficits due to mass effect, venous congestion, or hemorrhage, hence requiring prompt treatment. Treatment options for PMAVF include microsurgery, endovascular intervention, or a combination of the 2. Endovascular intervention with coil or liquid embolic material is considered first-line treatment for IVc PMAVF, and effective in type IVb with good clinical outcome. PMAVF is a rare spinal vascular malformation commonly manifesting as severe neurologic deficits but has the potential of favorable outcomes with endovascular therapy. This case demonstrates a unique

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angioarchitecture of high-flow PMAVF with a single feeder artery and large venous pouch, treated successfully with endovascular therapy.

Introduction

Spinal vascular malformations are rare and understudied diseases [1]. Peri-medullary arteriovenous fistula (PMAVF) or spinal AVF type IV is an abnormal direct connection between an intradural artery and vein within the spinal canal, without a nidus [2]. It has a significant impact, with patients typically presenting with progressive myelopathy, intraspinal, epidural, or subarachnoid hemorrhage, and spinal deformities [1–3]. Advances in endovascular techniques in recent decades have allowed effective treatment of PMAVF with good clinical outcomes. We report a case of high-flow PMAVF with a single feeder artery and a large venous pouch in a 2-year-old boy, successfully treated with endovascular intervention.

Case report‘

A 2-year-old boy was referred to our hospital with progressive bilateral weakness of the lower extremities for the past 2 years. The parents reported that the child's lower extremities were less active relative to the upper extremities, with the condition worsening within recent months. The patient was also unable to control his urination and defecation. The patient had no history of trauma or fever. He was born full term with an uneventful delivery. A family history of similar symptoms was denied. Neurological examination revealed spastic paraparesis with motor strength of 2/5 on both legs. The Babinski sign and clonus were positive bilaterally. Complete blood count, liver function, kidney function, and electrolytes were within normal limits. Magnetic resonance imaging with gadolinium contrast showed a large, dilated vascular structure at the level of thoracic vertebral bodies 5-7, compressing the spinal cord. In the caudal segment of the mass, the peri-medullary vein appeared dilatated (Fig. 1). The patient was then referred to an angiographic suite for further evaluation.

The patient then underwent a spinal angiography, which confirmed a high-flow arteriovenous fistula with a single feeder artery (anterior radiculomedullary artery), branching from the left seventh thoracic segmental artery. The fistula was connected to a large venous pouch which was found to drain into the serpiginous dilated peri-medullary vein (Fig. 2). After careful consideration, we decided to perform transarterial embolization with liquid embolic material.

A guiding catheter 6F (Chaperon 6F, Microvention Inc.) was navigated in front of the left seventh thoracic segmental artery ostium. Superselective injection of the microcatheter demonstrated a high-flow fistula through a single feeder artery (Fig. 3). We performed embolization using ethylene vinyl alcohol copolymer (EVOH) (ONYX 18, Medtronic inc). Due to the high flow of the fistula, the injected ONYX rapidly entered the fistula. To avoid overfilling the venous pouch, the microcatheter was positioned slightly more proximal, and injection of embolic material was performed as a small bolus. The internal flow through the fistula gradually slowed, and the EVOH was continuously injected until the fistula and venous pouch were not visible (Fig. 4).

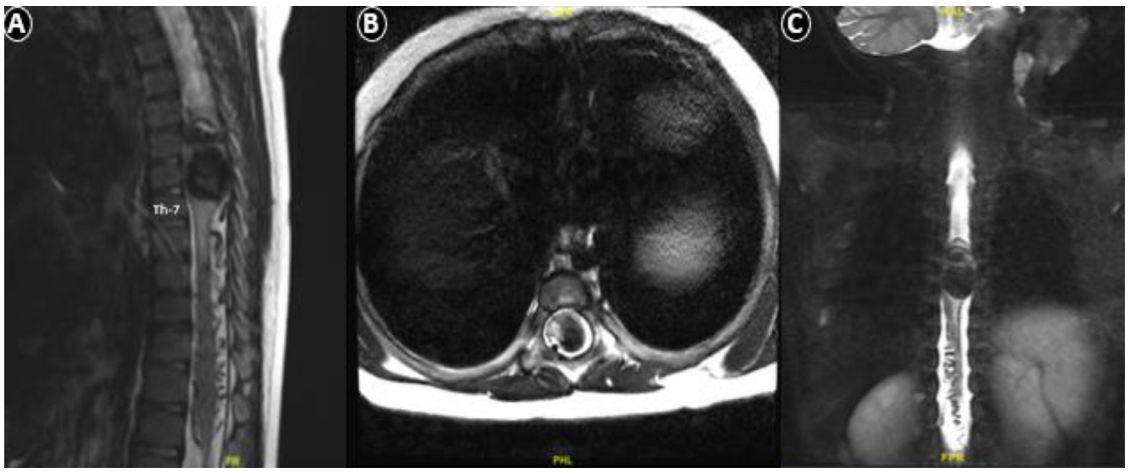

Fig. 1 – Magnetic resonance imaging (MRI) of the spine. (A) Sagittal T2WI demonstrated an abnormally dilated vascular structure at the midthoracic spinal cord, suggestive of a venous pouch, with serpiginous dilatation of peri-medullary veins on the caudal side. (B) Axial T2WI MRI showed that the venous pouch compressed the spinal cord to the right side. (C) Coronal STIR MRI view.

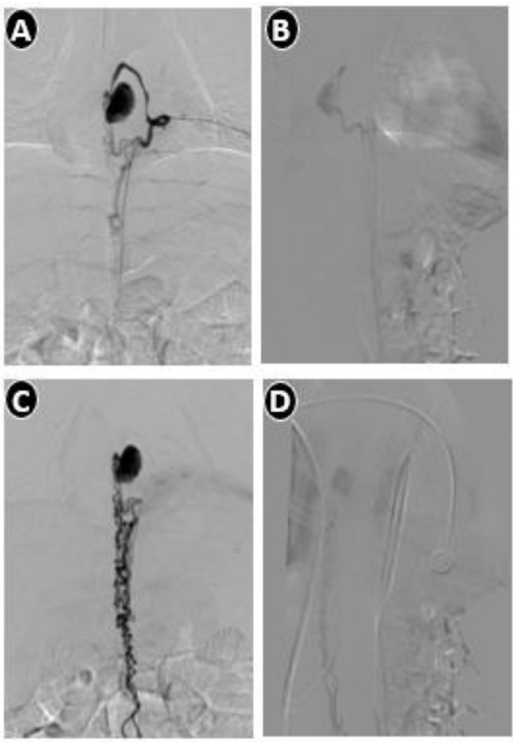

Fig. 2 – Angiogram of the left seventh thoracic segmental artery injection. Anteroposterior view (A) and lateral view (B) during the arterial phase, and anteroposterior view (C) and lateral view (B) during the venous phase.

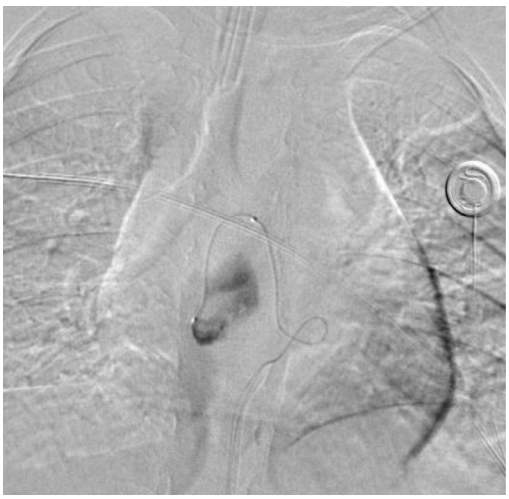

Fig. 3 – Superselective angiogram via microcatheter demonstrated a high flow fistula filling into the venous pouch.

After the procedure, the patient did not experience any new neurological deficits. The motor strength of both legs immediately improved to a score of 3 bilaterally. At the 2-week post-procedure follow-up, his motor strength improved, and the patient was able to walk with assistance. However, the autonomic function remains impaired. Follow-up MRI revealed

Fig. 4 – Angiogram via guiding catheter after embolization shows occlusion of the fistula and venous pouch.

thrombosis and involution of the venous pouch. The serpiginous dilatation of the peri-medullary vein and spinal cord edema were also attenuated.

Discussion

Spinal vascular malformation is a rare condition that accounts for 2%-3% of all spinal cord masses and is responsible for some unknown cases of myelopathy [3]. There are various classifications of spinal vascular malformations. The most common classification by Rosenblum divides spinal vascular malformations into four categories, which are type I (Dural arteriovenous fistula (AVF), type II (Glomus arteriovenous malformation (AVM)), type III (Juvenile AVM), and type IV (Perimedullary AVF) [4–6]. Type I is the most common, accounting for 80%-85% of all spinal vascular malformations, while type IV is rare [4]. PMAVF is then further classified into type IVa if there is a single feeder and a small fistula, type IVb if there are multiple feeders and moderate AVF, and IVc if there are multiple feeders and large AVF [2,4,7].

This patient presented with a 2-year history of progressive spastic paraparesis, impaired sensation, as well as urinary and fecal incontinence. This is due to the direct artery-to-vein, high-pressure blood flow, which leads to hemodynamic stress, chronic remodeling, and formation of a venous pouch that consequently results in the compression of spinal cord structures. In addition, flow into the peri-medullary veins will impair venous outflow and cause venous congestion. Since the lower thoracic region generally has less outflow than the cervical region, lesions in this location, as in this case, will be more susceptible to venous congestion [2]. Other causes of neurologic deficit in PMAVF include vascular steal and hemor-

rhage in the subarachnoid, epidural, or hematomyelia, especially in cervical lesions [2,4].

On an angiogram, the fistula is identified by a change in diameter from a smaller diameter artery to a larger diameter vein. In this case, because of the high flow of fistula, the transition was marked by the formation of a large venous pouch. PMAVF in the ventral area of the spinal cord is generally subpial and is supplied by the anterior spinal artery, whereas if it is subarachnoid, it is generally supplied by the posterolateral spinal artery [5]. This case was unique, because the large fistula was supplied by only 1 feeder artery, which was anterior radiculomedullary artery, and drain into a large venous pouch.

Microfistula PMAVF (type IVa) generally occurs at an older age and often has an idiopathic or traumatic aetiology. On the other hand, macrofistula PMAVF (types IVb and IVc) are more common in children and are associated with genetic syndromes such as Hereditary Haemorrhagic Telangiectasia (HHT), or less commonly Klippel Trenaunay-Weber or Cobb's syndrome [2]. HHT is an autosomal dominant disease caused by mutations in the endoglin or novel kinase gene, with a broad clinical manifestation that includes cutaneous telangiectasia, recurrent epistaxis, and arteriovenous malformations involving various organs, such as lungs, gastrointestinal tract, liver, genitourinary tract, brain, and spinal cord [5,8]. In this case, we did not perform genetic testing for HHT because the patient did not present with classic HHT clinical manifestations and had no family history.

The patient's progressive neurologic deficit emphasizes the need for prompt treatment. The treatment options in PMAVF include microsurgery or endovascular therapy [2]. Type IVa PMAVF is generally treated with microsurgery, because of the difficulty of navigating into fistulas with a small diameter. Type IVb can be treated with a combination of microsurgery and endovascular therapy, whereas type IVc is generally treated with endovascular therapy alone because of the difficulty and risk of surgery [7,9,10].

The goal of endovascular therapy is to shut the fistula without occluding the anterior spinal artery. Embolization is generally performed in a transarterial manner with the aim of closing the fistula, distal feeder artery or proximal draining vein, but avoiding closure of the proximal portion of the feeder artery or distal draining vein [3,7,11]. Proximal feeder artery occlusion increases the risk of recanalization, while occlusion of the venous drainage at an overly distal site will increase the risk of bleeding [3]. There is a case report showing the effectiveness of venous pouch embolization using a cotton candy-like injection technique [5].

The objective in treating perimedullary arteriovenous fistulas is to achieve total occlusion of the arteriovenous communication, while preserving the normal venous drainage and arterial supply. There is still ongoing debate on the optimal approach for treating perimedullary arteriovenous fistulas (AVFs). Both endovascular and open surgical approaches have been proposed as therapeutic options for this condition. The small size of the feeding arteries makes it difficult to position the catheter tip optimally for superselective fistula embolization, which is a challenge for embolizing a type IVa PMAVF. With the advancement of microcatheter technology, a greater number of lesions can now be effectively treated using endovascular methods. When using a standard posterior approach, it is easily possible to access a dorsally located type IVa PMAVF. Extra caution is taken while dealing with ventral fistulas located above the conus medullaris. The spinal cord's dorsal or dorsolateral region comprises PMAVFs of type a and type b, which are nourished by PSA branches. Through the utilization of a posterior surgical method, it is possible to observe and separate the fistula. This also pertains to ventral type A and type B PMAVFs situated at the filum terminale or conus medullaris. PMAVFs with ASA feeders are located on the ventral or ventrolateral surface of the spinal cord. A posterior surgical technique can still provide access to the lateral and ventral parts of the spinal cord in order to treat ventrolateral fistulae. However, fistula sites located in the median fissure or entirely on the ventral side of the PMAVFs are more challenging to treat and typically require more complex anterior or anterolateral methods [12].

The choice of embolic material includes liquid embolic agents, coil, or a combination of both [7]. The use of particle embolic material is not recommended because of the temporary effect and higher rate of recanalization, while the use of balloon is difficult in term of navigation through a small and tortuous feeder [5]. Coils have an advantage in high flow fistulas, as they are more stable, while ethylene vinyl alcohol copolymer (EVOH) has benefits especially when access of the microcatheter to the fistula is difficult. Both have a risk of occlusion of the anterior or posterior spinal arteries. In this case we opted for transarterial embolization by closing the fistula on the distal feeder artery side using EVOH (ONYX 18, Medtronic inc). N-butyl cyanoacrylate (NBCA) plus tantalum powder is another liquid embolic material that is also frequently used.

The embolization was initially performed by pushing EVOH as small bolus. The high flow in the fistula carried the bolus into the venous pouch and helps to slow the flow through the fistula. After the flow had slowed, continuous injection was performed to occlude the fistula. We avoid overfilling the venous pouch with embolic material as it can cause a persistent mass effect to the spinal cord.

To achieve optimal treatment of PMAVFs with endovascular therapy, it is necessary to perform embolization of the vein at the site of the fistula. Surgeons have used different embolic materials to achieve this objective, including balloons, cellulose acetate polymer solution, platinum coils (with or without silk), and coils. These materials are currently the most commonly employed. Coils are the optimal option for pediatric high-flow PMAVFs due to their increased difficulty of removal and enhanced stability compared to adhesive materials. Nevertheless, it is crucial to consider the impact of the bulk on the spinal cord during the coiling process [13].

When a lesion is located at a distance from the microcatheter tip, it is advisable to employ Onyx® -18 and its lower viscosity formulation. This is because they allow the liquid embolic agent to travel a greater distance and penetrate deeper into the lesion, resulting in a more effective embolization. In addition, Onyx® is as effective whether used alone or in conjunction with other embolic agents for the treatment of peripheral lesions. Lastly, Onyx® possesses physical attributes that include its thickness, ability to not stick to surfaces, delayed process of forming polymers, ability to adapt to any vas-

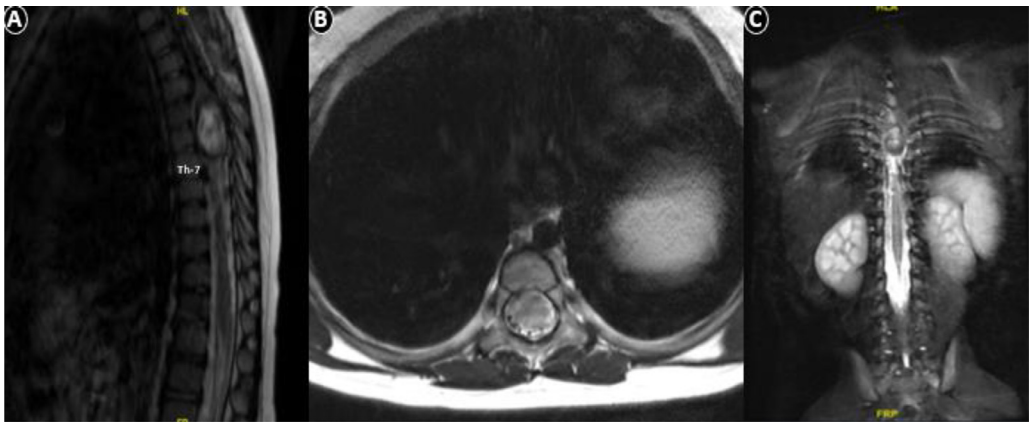

Fig. 5 – Follow-up MRI 2 weeks post-procedure. (A) Sagittal T2WI MRI, (B) Axial T2WI at the same level as Figure 1, (C) Coronal STIR MRI, revealed involution and thrombosis inside the venous pouch. The serpiginous dilatation of the peri-medullary vein and spinal cord edema were also attenuated.

cular shape, and excellent visibility under fluoroscopy. These properties provide numerous benefits when Onyx® is utilized in peripheral applications [14].

Due to the small size of the fistula, the patient's young age, and the fast rate of blood flow through the fistula, we decided to employ EVOH.

After the procedure, the patient had improved motor strength of both legs (strength assessment score of 3), but there was no improvement in micturition function. This is in line with the results of Antoinetti et al who also found that PMAVF therapy resulted in better improvement of gait and ambulation than micturition [9]. This suggests that PMAVF therapy should ideally be performed prior to the development of micturition symptoms and signs. PMAVF subtype also determines the postprocedural clinical outcome. Mourier et al found that complete occlusion of PMAVF type IVa and IVb resulted in improvement of symptoms in 50% of patients, whereas in type IVc, improvement was found in 100% of cases [15]. Apart from subtype, duration of neurologic deficit, bleeding history and presence of spinal cord compression also affects clinical outcome [11]. Complete resolution of symptoms did not occur immediately in this case considering the long-standing process before he was admitted, but the patient's relatively young age and the absence of genetic syndromes provided the potential for clinical improvement as in Fig. 5.

After 2 years of undergoing an embolization interventional procedure, the patient's motoric condition, urinary incontinence and development are still good. Genetic counseling for this patient already considered provided to the patient due to genetic hereditary and nonhereditary feature of PMAVF.

Conclusion

Peri-medullary arteriovenous fistula (PMAVF) is a rare spinal vascular malformation that commonly manifests as severe neurologic deficits but has the potential to have a favorable clinical outcome with endovascular therapy. This case demonstrates a unique angioarchitecture of high-flow PMAVF with a single feeder artery and large venous pouch. Endovascular therapy with EVOH can be effective in cases of high-flow PMAVF, and early recognition of this condition is paramount to improve the patient's clinical outcome.

Ethics statement

Ethical approval and consent to participate. This study was conducted according to the guidelines of the Declaration of Helsinki. The subject described in this case report has provided consent for publication from his mother.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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