Pediatric perimedullary arteriovenous fistula of the conus medullaris supplied by the artery of Desproges-Gotteron

Case report

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The cone artery, or artery of Desproges-Gotteron, is sometimes seen arising from the internal iliac artery. The authors describe a case of a symptomatic perimedullary arteriovenous fistula (AVF) of the conus medullaris in an 8-year-old boy who presented with a protracted history of urinary difficulty and severe sudden-onset right lumbosacral pain that evolved to severe paraparesis with compromise of the sphincter muscles. The spinal AVF, which was supplied by the cone artery and a thoracic radiculomedullary artery that joined at the fistula site in a large partially thrombosed varix, was completely occluded with Onyx liquid embolic. The patient’s clinical condition improved rapidly after embolization. As shown in this patient, urgent endovascular embolization of spinal AVFs can be very rewarding, even in patients with severe neurological presentation. The artery of Desproges-Gotteron appears to be a rare arterial variation. To the authors’ knowledge, this is the first pediatric case of a conal AVF supplied by this artery. (http://thejns.org/doi/abs/10.3171/2012.12.PEDS12363)

Key Words • artery of Desproges-Gotteron • conus medullaris • Onyx embolization • pediatric • spinal arteriovenous malformation • vascular disorders

**Abbreviations used in this paper:** AVF = arteriovenous malformation; AVM = arteriovenous malformation; HHT = hereditary hemorrhagic telangiectasia; MRA = MR angiography.

**Case Report**

**History and Examination.** This 8-year-old boy presented with a protracted history of urinary complaints, including difficulty in starting micturition that progressed over a 6-month period. Magnetic resonance imaging evaluation was scheduled but not performed. Suddenly the boy had severe right lumbosacral pain. He was admitted to the pediatric department in a small community hospital, where he was managed based on a working diagnosis of Guillain-Barré syndrome. However, his condition progressed over 2 days to severe paraparesis with compromise of the sphincter muscles.

Three days after he had demonstrated symptoms of spinal cord injury, rather than peripheral neuropathy, MRI
of the spine was performed, which showed a conus medullaris AVM with a partially thrombosed, large venous ectasia causing cone compression and ascending cord edema. Flow voids extended from the T-9 to L-1 vertebral bodies, and abnormal cord signal extended for several levels, far along the midthoracic spine (Fig. 1 left). The patient was transferred to our neurosurgery department.

Upon admission, the patient was suffering from significant lumbar pain and required opiates. His sphincter muscles were compromised and a urinary catheter had been placed. Neurological examination revealed complete plegia of all muscle groups in the lower limbs with the exception of the left quadriceps muscle (2/5), left tibialis anterior muscle (3/5), and left extensor hallucis longus muscle (1/5). He demonstrated a sensory level to pinprick sensation at L-4 on the right side, bilateral areflexia, bilateral flexor plantar response, and lax anal tone.

Urgent spine MRA performed on the day of admission confirmed that the vascular malformation was primarily supplied by a dolichoectasic cone artery (artery of Desproges-Gotteron) and by a thoracic radiculomedullary artery (Fig. 1 right). Two feeders converged into a single fistula in the largest venous varix. Endovascular treatment was planned for the next morning.

Procedure. With the patient under general anesthesia, a 5-Fr introducer sheath was placed at the right femoral artery. Using a 5-Fr Simmons catheter, we obtained a selective angiogram of the left iliac artery (Fig. 2A). Angiography confirmed a dolichoectasic cone artery feeding a high-flow perimedullary fistula in accordance with the MRA findings (Fig. 2B). With road mapping, co-axial navigation of a microcatheter through the inverted hairpin cone artery was successful in reaching the fistula site (Fig. 2C). From this point, injection of 2.2 ml of Onyx 34 (ev3, Inc.) allowed occlusion of the cone artery feeder, partial occlusion of the patent partially thrombosed venous varix, and then retrograde occlusion of the distal thoracic radiculomedullary feeder (Fig. 3). Angiographic evaluation of the radiculomedullary artery confirmed complete occlusion of the AVF (Fig. 4).

Posttreatment Course. Immediately after the embolization, the child experienced significant improvement in his lumbosacral pain and very mild improvement in his left quadriceps muscle weakness. He was transferred to a rehabilitation facility where he rapidly attained neurological improvement, regaining sphincter control as well as the ability to walk independently with canes only 6 weeks after endovascular embolization. Magnetic resonance imaging studies performed after 2 months showed thrombosis of the venous varix, disappearance of the rich perimedullary venous plexus seen on admission MRI, and complete resolution of spinal cord edema (Fig. 5 left). Magnetic resonance angiography performed at the 2-month follow-up confirmed complete exclusion of the vascular malformation and the embolized feeders (Fig. 5 right).

On neurological examination 6 months after endovascular embolization, the patient demonstrated significant improvement in muscle strength for all muscle groups of the lower limbs: iliopsoas muscles bilaterally 4/5, quadriceps muscle on the right side 4/5, quadriceps muscle on the left (–4/5), tibialis anterior muscle –3/5, and extensor hallucis longus muscles bilaterally –3/5. He had normal pinprick sensation bilaterally, areflexia on the right side, and a bilateral flexor plantar response.

Discussion

Intramedullary AVMs, perimedullary AVFs, and dural AVFs are all included under the rubric of spinal AVM, depending on their anatomical location and blood supply. We describe an unusual case of symptomatic perimedullary AVF of the conus medullaris supplied by the cone artery and a thoracic radiculomedullary artery that merged at the fistula site in a large, partially thrombosed varix in an 8-year-old boy. The spinal fistula was completely occluded with Onyx embolic agent, and the patient’s condition improved rapidly.

Perimedullary or spinal cord AVFs are intradural vascular malformations located superficial to the cord, similar in nature to brain AVFs. Although rare, they are a well-recognized entity first described by Heros et al., with direct communication between spinal cord arteries and veins and without any intervening vascular network or extension within the cord itself. The angioarchitecture of these lesions is usually less complex than that of nidus-type AVMs.
Based on the classification proposed by Rodesch et al., our patient presented with a macro-AVF. These are high-flow direct shunts fed by one or multiple spinal cord arteries and ending directly in a giant venous ectasia that is characteristic of these lesions, with secondary drainage into the extrinsic and intrinsic venous system of the cord.

In the pediatric population, macro-AVFs are often the first sign of HHT. The diagnosis of HHT may be difficult to confirm in children, because typical signs usually appear later in the life cycle. Evaluation of our patient's family tree failed to reveal a relative with known or suspected HHT. However, the lack of family history does not rule out the possibility of a spontaneous mutation in an affected patient.

There is little information in the literature on spinal cord AVFs in children. A search of the literature and a review of the reports cited in the literature, as performed by Matushita et al., revealed 45 papers describing approximately 79 patients younger than 16 years.

Rodesch et al. presented their 20-year experience in the management of spinal cord AVFs. Out of 10 pediatric AVFs, 6 were thoracolumbar macro-AVFs. Subanalysis of these macro-AVFs showed that 86% were embolized and 67% were finally cured. The authors concluded that embolization of spinal cord AVFs using glue is a safe treatment, comparing favorably with other approaches. Matushita et al. documented 6 cases of pediatric AVF; embolization was initially attempted in 3 patients, with successful occlusion of the fistula in 2. For the remaining cases, open surgery was performed, with complete occlusion of the fistula. These authors concluded that treatment for AVFs is difficult to standardize because the lesions are extremely rare with a unique angioarchitecture. Descriptions of Onyx embolization of a pediatric spinal AVF in the literature are rare. However, we believe the...
use of this stable and more controllable embolizing agent will become more widespread in the management of this entity.

Complementary surgical procedures have been advocated to totally eradicate the shunt even after embolization with glue. We agree with Rodesch et al. that surgical removal of the embolized AVF is unnecessary in view of the long-lasting clinical results achieved with liquid embolic agents in brain and spine malformations. Our patient was considered for surgery mainly because of the mass effect of the venous varix draining the AVF; however, we favored a conservative approach after confirming rapid clinical improvement with endovascular management. The patient’s clinical improvement after embolization was explained by resolution of the spinal venous hypertension with the associated cord edema and epidural venous plexus engorgement, as demonstrated on follow up MRI. Although the venous varix sustained complete thrombosis, clinical improvement did not correlate with any significant reduction in varix volume. It can be argued that a reduction in the pulsatile effect of the varix mass acted as a secondary explanation for the resolved cord edema.

Most experience with the endovascular management of perimedullary AVFs involves the use of different embolizing agents, notably cyanoacrylate glue. However, we preferred to use Onyx in accordance with growing experience in using the liquid embolic for endovascular procedures and specifically with the good results obtained in the embolization of brain AVFs. In our pediatric patient, once we had advanced through the distal cone artery, Onyx was intermittently injected until the draining varix was proximally filled. The thoracic radiculomedullary artery was subsequently filled retrograde, thereby completely occluding the fistula with a very controlled injection.

As can be concluded from this case, urgent endovascular embolization of spinal AVFs can be rewarding, even in patients with a severe neurological presentation.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References

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