Case Report

Remote Intramedullary Hemorrhage in the Thoracic Spinal Cord Secondary to a Perimedullary Arteriovenous Fistula of the Distal End of Conus Medullaris Mimicking Filum Terminale Arteriovenous Fistula

Abstract

Perimedullary arteriovenous fistulas (PMAVFs) of the conus medullaris are rare and usually manifest with progressive myelopathy secondary to venous congestion resulting from retrograde arterialization of the draining vein into the spinal cord. We present a rare case of conus PMAVF presenting with remote intramedullary spinal cord hemorrhage in the thoracic cord. A 37-year-old woman was transferred to our institute due to sudden severe pain in the left lower leg and weakness of the lower extremities following progressive paresthesia of the lower extremities. Magnetic resonance imaging of the thoracic and lumbosacral spine revealed spinal cord congestion extending from the conus medullaris to the level of T6 with intramedullary hemorrhage at the level of T8–9 on the left side of the spinal cord. There were abnormal serpiginous intradural flow voids along the anterior surface of the spinal cord extending from the level of L2 to the lower cervical with venous varix at the level of T8–9, probably being the source of hemorrhage. Spinal angiography confirmed conus PMAVF at the distal end of the conus medullaris supplied by the sulco-commissural artery arising from the enlarged anterior spinal artery originating from the left T11 intercostal artery with cranial drainage through the dilated anterior spinal vein into the tortuous perimedullary veins up to the lower cervical level. The patient underwent successful endovascular treatment with N-butyl cyanoacrylate and had gradually improved until being ability to walk independently without residual pain of the left lower leg. We speculated that an increased venous flow into a varix may be considered an important risk factor of hemorrhage.

Keywords: *Conus medullaris, filum terminale arteriovenous fistula, intramedullary hemorrhage, perimedullary arteriovenous fistula, Type IV spinal cord arteriovenous malformations*

Prasert Iampreechakul, Anusak Liengudom, Punjama Lertbutsayanukul[1], Somkiet Siriwimonmas[2], Sirintara Pongpech[3]

Departments of Neurosurgery and [1]Neuroradiology, Prasat Neurological Institute, [2]Department of Radiology, Bumrungrad International Hospital, [3]Department of Radiology, Division of Interventional Neuroradiology, Ramathibodi Hospital Medical School, Mahidol University, Bangkok, Thailand

Introduction

Spinal cord arteriovenous malformations have been classified into four subtypes including Type I, spinal dural arteriovenous fistulas (SDAVFs); Type II, intramedullary glomus malformations; Type III, extensive juvenile malformations; and Type IV, intradural perimedullary arteriovenous fistulas (PMAVFs). Type IV spinal cord arteriovenous malformations have been further divided into three subtypes including Type IVa, small or low-flow arteriovenous fistula (AVF) supplied by a single arterial branch of the anterior spinal artery (ASA); Type IVb, intermediated-sized fistula supplied by multiple arterial feeders; and Type IVc, giant high-flow fistula fed by several feeding vessels of the ASA and posterior spinal artery.[1]

Intradural extramedullary AVFs were first described by Djindjian *et al*.[2] in 1977 and later were classified as Type IV PMAVF, direct communication of the intrinsic arterial supply of the spinal cord and a vein without an intervening small-vessel network, by Heros *et al*.[3] in 1986. Based on the modified classification of spinal cord vascular lesions by Spetzler *et al*.,[4] they classified Type IV lesions as intradural ventral AVFs which are located ventrally and in the midline.

PMAVFs at the level of the conus medullaris are rare and classified as Type IV lesions and presented with either subarachnoid hemorrhage (SAH) or, more commonly, progressive myelopathy secondary to venous hypertension.[5,6]

Address for correspondence:
Dr. Prasert Iampreechakul,
312 Rachawithi Road, Khwaeng Thung Phaya Thai, Bangkok 10400, Thailand.
E-mail: bangruad@hotmail.com

We described a case of PMAVF of the conus medullaris with remote intramedullary spinal cord hemorrhage in the thoracic cord. The pathogenesis of thoracic intramedullary hemorrhage caused by conus PMAVF in our case was discussed.

Case Report

A 37-year-old woman complained of progressive paresthesia of the lower extremities for 3 months. She went to the local hospital and was treated with some medicines without improvement. She had no history of any injury. Two weeks later, the patient was hospitalized to the same local hospital with sudden severe pain in the left lower leg and weakness of the lower extremities. She also developed urination incontinence requiring urinary catheterization.

Magnetic resonance imaging (MRI) of the spine was performed and showed an abnormal T2 signal representing spinal cord congestion extending from the conus medullaris to the level of T6. There were abnormal serpiginous intradural flow voids along the anterior surface of the spinal cord extending from the level of L2 to the lower cervical with suspecting two venous varices at the level of T8–9 and T10. At the level of T8–9, there was abnormal heterogeneous signal intensity on T1- and T2-weighted image on the left side of the spinal cord, representing intramedullary hemorrhage [Figures 1 and 2]. The patient was diagnosis of ruptured spinal cord arteriovenous malformations and was transferred to our institute and admitted for further investigation and management. The neurological examination revealed the evidence of
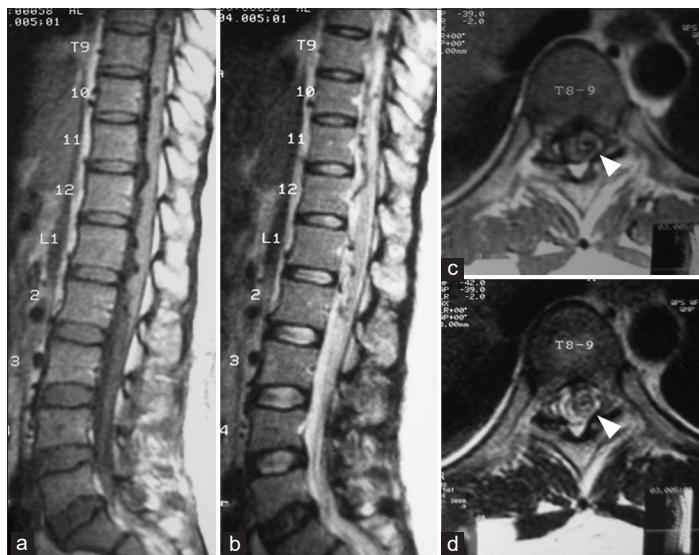

Figure 1: Sagittal (a) T1-weighted and (b) T2-weighted images of the thoracolumbar spine reveal serpiginous intradural flow voids along the anterior surface of the spinal cord extending from the level of L2 to the mid-thoracic. Axial (c) T1-weighted and (d) T2-weighted images at the level of T8–9 demonstrate abnormal heterogeneous signal intensity (arrowheads) on the left side of the spinal cord, probably representing intramedullary hemorrhage

Figure 2: Sagittal T1-weighted images of (a) the cervical and (b) thoracic spine show intradural flow voids (arrowheads) along the anterior surface of the spinal cord extending from the lower thoracic to lower cervical level. (c) Sagittal T2-weighted images of the thoracic spine demonstrate two venous varices (arrows) at the level of T8–9 and T10. Axial T2-weighted images at the level of (d) T6-7, (e) T7-8, and (f) T9-10 reveal abnormal hypersignal intensity within the spinal cord, representing spinal venous congestion
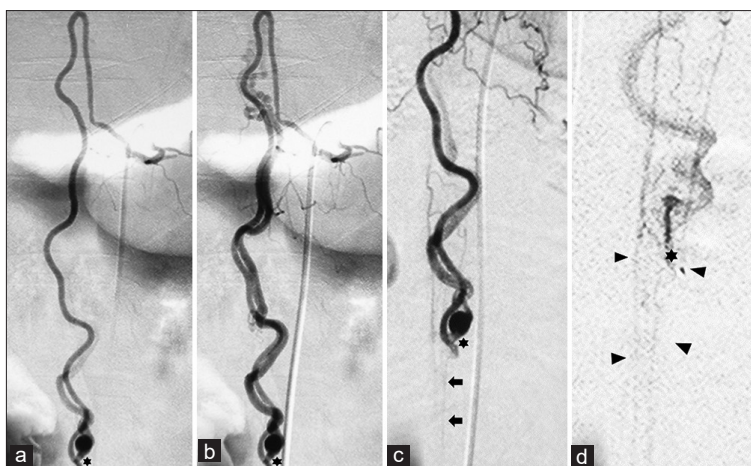
Figure 3: Anteroposterior views of the left T11 intercostal artery angiography in (a) arterial and (b and c) venous phases reveal a perimedullary arteriovenous fistula (asterisks) of the distal end of the conus medullaris at the level of L2, supplied by the enlarged sulco-commissural feeder arising from the enlarged anterior spinal artery with cranial drainage into the dilated anterior spinal vein. There is a venous dilatation (curve arrows) at the proximal draining vein. The normal-sized ASA (arrows) distal to the fistula is noted. (d) Selective angiography with microcatheter through the ASA clearly demonstrates the fistulous point (asterisk) located above the arterial basket of the conus medullaris forming from the ASA and posterior spinal arteries (arrowheads).
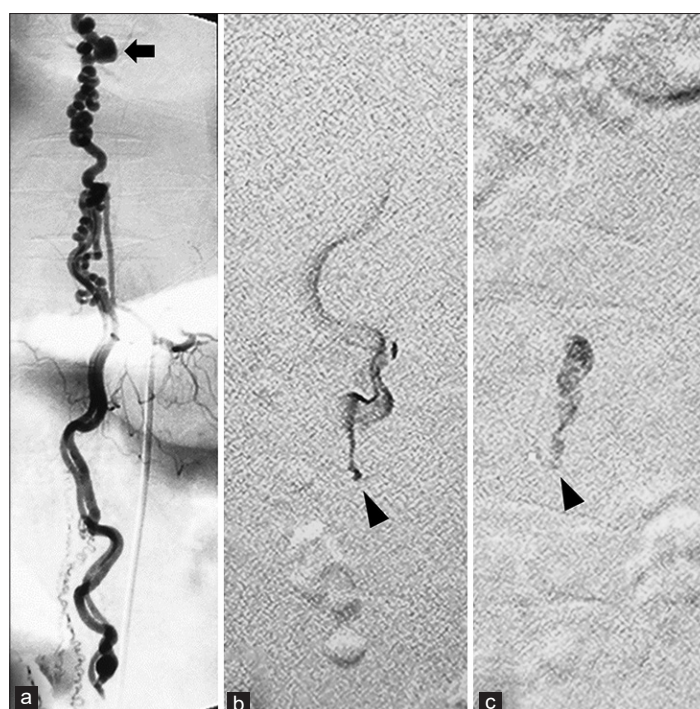

Figure 4: (a) Anteroposterior view of the left T11 intercostal artery angiography in the venous phase reveals the large venous varix (arrow) at the level of T8–9 pointing to the left side, probably corresponding with the area of intramedullary hemorrhage. (b) Oblique view of selective angiography with the microcatheter through the enlarged left sulco-commissural artery clearly demonstrates the fistulous point (arrowhead) and proximal draining vein. (c) During embolization, the glue cast can occlude the fistula (arrowhead) and the venous pouch of anterior spinal vein

spastic paraparesis (muscle strength 4/5), impairment of proprioception, hyperreflexia, and presence of Babinski sign in the lower extremities.

Spinal angiography was obtained and demonstrated a PMAVF of the distal end of the conus medullaris at the level of L2, supplied by the enlarged sulco-commissural feeder arising from the enlarged ASA originating from the left T11 intercostal artery with cranial drainage through the dilated anterior spinal vein (ASV) into the tortuous perimedullary

veins up to the lower cervical level. There was a venous dilatation at the proximal draining vein [Figure 3]. The venous phase of the left T11 intercostal artery angiography disclosed the large venous varix at the level of T8–9 pointing to the left side, probably corresponding with the area of intramedullary hemorrhage [Figure 4a]. Due to the enlarged ASA, we decided to proceed with endovascular as the first choice. We used Magic microcatheter 1.2 Fr (Balt, Montmorency, France). The microcatheter was navigated
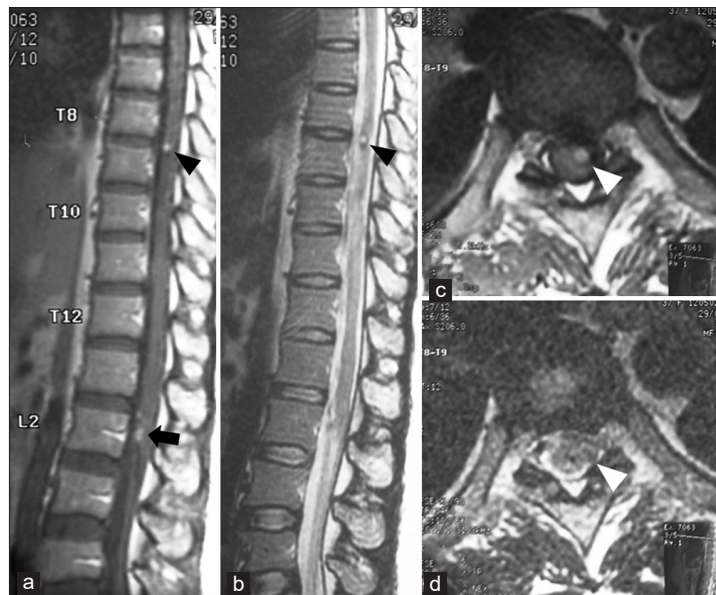
Figure 5: Magnetic resonance imaging of the thoracic spine obtained 2 months after endovascular treatment. At the level of T8–9 on the anterolateral cord, (a) Sagittal and (c) axial T1-weighted images show hypersignal intensity, and (b) sagittal and (d) axial T2-weighted images demonstrate hypersignal intensity surrounding with hyposignal intensity (black and white arrowheads), probably indicating thrombosed venous aneurysm. There are multiple small hypersignal intensity foci (black arrow) at the level of L2 just above the distal end of the conus medullaris, probably representing thrombosed venous pouch.
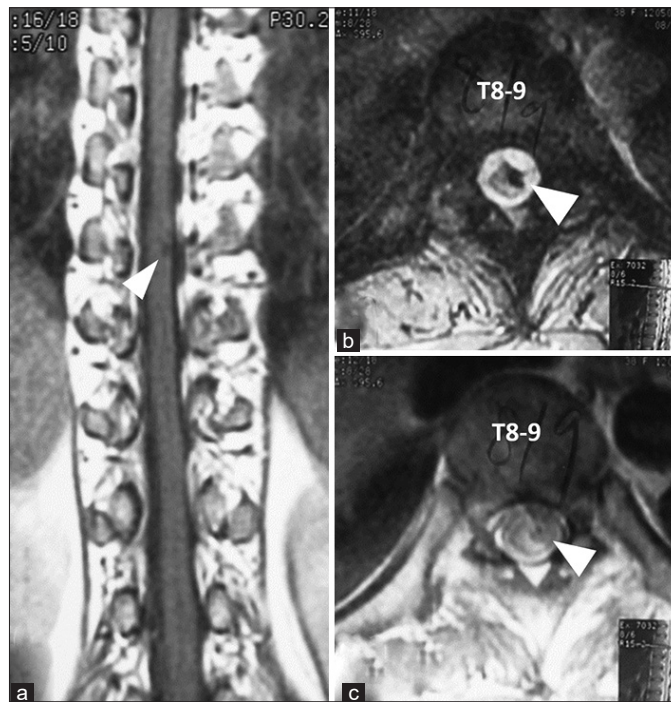

Figure 6: Magnetic resonance imaging of the thoracic spine obtained 2 years after endovascular treatment. (a) Coronal T1-weighted, axial (b) gradient-recalled echo T2*-weighted, and (c) proton density-weighted images demonstrate hyposignal intensity (arrowheads) at the level of T8–9 on the left anterolateral part of the spinal cord, probably corresponding to hemosiderin

through the ASA and the tip of microcatheter could be wedged into the enlarged left sulco-commissural artery just proximal to the fistula.With heparinization, transarterial embolization with N-butyl cyanoacrylate (NBCA) through the ASA was successfully performed with reaching the venous pouch of ASV [Figure 4b and c]. A mixture of NBCA and an oil-based contrast agent (Lipiodol Ultra Fluid; Guerbet, Aulnay-sous-Bois, France) was prepared in proportions of 1:0.7 ratio of NBCA to Lipiodol. Spinal angiography after embolization confirmed complete obliteration of the fistula and preservation of the ASA. To prevent further venous thrombosis, the patient received the prophylactic anticoagulation after the procedure.

Figure 7: Spinal angiography obtained 3 years after endovascular treatment. Anteroposterior views of the left T11 intercostal artery angiography (a) with and (b) without subtraction demonstrate the normal size of the anterior spinal artery (white arrowheads) with a characteristic hairpin turn (black arrowhead) without recurrence of the fistula

MRI of the thoracic and lumbar spine, obtained 2 months after endovascular treatment, showed the disappearance of intradural flow voids and thrombosed venous aneurysm at the level of T8–9 on the anterolateral cord and above the distal end of the conus medullaris [Figure 5]. The patient had gradually improved until being ability to walk independently without residual pain of the left lower leg 6 months later. Bladder function had completely recovered at 1 year after treatment. MRI and magnetic resonance angiography of the thoracolumbar spine obtained 2 years after embolization revealed complete obliteration of the fistula and significant resolution of spinal cord congestion. At T8–9 level on the left anterolateral part of the spinal cord, there was hyposignal intensity on T1-weighted, gradient-recalled echo T2*-weighted, and proton density-weighted images, probably corresponding to hemosiderin [Figure 6]. Spinal angiography, obtained 3 years after endovascular treatment, demonstrated the normal size of the ASA without recurrence of the fistula [Figure 7].

Discussion

Type IVa perimedullary fistulas are typically slow-flow lesions and usually located on the ventral surface of the conus medullaris or filum terminale.[7] At the level of the conus medullaris, the ASA may form an anastomotic basket with the posterior spinal arteries (PSAs) via anastomotic branches. The arterial basket of the conus medullaris consists of 1 (unilateral) or 2 (bilateral) arterial branches circumferentially connecting the ASA and PSAs.[8] In our case, the fistula was located at the level of L2. Therefore, it is difficult to differentiate between filum terminale AVF (FTAVF) and PMAVF at the

distal end of the conus medullaris. Angiographic pattern of conus PMAVF in our case was similar to FTAVF, which was located ventrally at the midline and supplied by the ASA with cranial drainage into the perimedullary veins without intervening nidus. Using selective angiography with the microcatheter through the ASA, we can identify the arterial basket of the conus medullaris and found that the fistula was located above the arterial basket of the conus with the presence of the PSAs and normal-sized ASA distal to the fistula. In addition, hemorrhagic events have never been reported from FTAVF.[9-11]

Conus PMAVFs usually manifest by progressive myelopathy or acute nonhemorrhagic paraplegia.[12] Our case initially presented with progressive paresthesia of the lower extremities secondary to venous congestion and subsequently developed sudden severe pain in the left lower leg from intramedullary hemorrhage. Conus PMAVF in our case was supplied by a single feeder from the ASA. Therefore, it should be classified as intradural ventral Type IVa AVF which is a slow-flow shunt. However, we speculated that this fistula should be considered as a relatively high-flow fistula due to markedly enlarged feeder and draining vein. The high pressure can cause multiple venous varices. A high-flow fistula in our case may produce high pressure in the venous varix, embedded into the spinal cord parenchyma, at the level of T8-9 leading to intramedullary hemorrhage. A high-flow fistula in our case may produce high pressure in the venous varix at the level of T8–9 leading to intramedullary hemorrhage.

Similarly, hemorrhage from SDAVFs is usually rare and may occur as SAH from the fistulas in the cervical and craniocervical region.[13-15] Intramedullary hemorrhage

or hematomyelia caused by SDAVFs is extremely rare. A previous study was reviewed in the literature of SDAVFs with intramedullary hemorrhage and showed only six cases. All but one of the SDAVFs had venous varices of draining veins, being the source of hematomyelia.[16]

Type IVa PMAVFs can be treated by surgery, endovascular treatment, or combined approaches. The goal of treatment is complete obliteration of the fistula with preservation of normal arterial supply to spinal cord. The key to complete occlusion is obliteration of the proximal vein.[5] In Type IVa PMAVFs located at the level or below the conus medullaris, surgical treatment has been the preferred method of treatment with higher complete obliteration rates and low rate of recurrence.[6] However, some authors suggested that it was easy to operate the fistula on filum terminale but difficult on the conus medullaris.[17] Endovascular treatment should be considered as second-line choice because of the difficulty in navigating a microcatheter through the long and tortuous course of the thin ASA; the possibility of reflux of the liquid embolic material into the ASA; the risk or tearing, dissecting, thrombosis, or vasospasm of the ASA during embolization; concerning about recanalization of the fistula; and requiring expertise and experience in neurointerventional procedure.[9-11] According to a review about treatment on spinal cord PMAVFs by Ji *et al.*,[18] they found that endovascular treatment is more effective in high-flow PMAVFs, leading to a good outcome.

In the present study, we decided to try endovascular treatment as the first choice because there were the accessible dilated ASA and the sulco-commissural artery. The important factor for the successful transarterial embolization is an introduction of the tip of microcatheter in a more stable and distal position to the shunt point. During embolization with NBCA, the safety margin for glue reflux was short. The glue should close the fistula without reflux into the ASA. In addition, the glue should be stopped just the proximal draining vein for avoiding anterograde venous occlusion. The safety margin is related strictly to the anatomy of the ASA and the posterior curve into the sulco-commissural artery that supplies the AV shunt. We cannot allow any embolic material refluxes more than short segment of this posterior curve which will immediately arrive in the ASA axis.

Even though the NBCA cast occupied only in the localized area (only in the sulco-commissural artery and proximal draining vein), the prophylactic anticoagulation was used in our case because the fistula is quite large and amount of NBCA injected into the dilated proximal draining vein could further create too extensive thrombosis within the rest of perimedullary vein and disturb the normal spinal cord drainage.

Conclusions

The authors reported an extremely rare case of conus PMAVF presenting with remote intramedullary hemorrhage secondary to ruptured venous varix, confirmed by imaging studies. This fistula was relatively high flow due to markedly enlarged feeder and multiple venous pouches. We speculated that an increased venous flow into a varix may be considered an important risk factor of hemorrhage.

Consent

The patient has given consent to be enrolled and has her data published.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Borden JA, Wu JK, Shucart WA. A proposed classification for spinal and cranial dural arteriovenous fistulous malformations and implications for treatment. J Neurosurg 1995;82:166-79.
2. Djindjian M, Djindjian R, Rey A, Hurth M, Houdart R. Intradural extramedullary spinal arterio-venous malformations fed by the anterior spinal artery. Surg Neurol 1977;8:85-93.
3. Heros RC, Debrun GM, Ojemann RG, Lasjaunias PL, Naessens PJ. Direct spinal arteriovenous fistula: A new type of spinal AVM. Case report. J Neurosurg 1986;64:134-9.
4. Spetzler RF, Detwiler PW, Riina HA, Porter RW. Modified classification of spinal cord vascular lesions. J Neurosurg 2002;96:145-56.
5. Barrow DL, Colohan AR, Dawson R. Intradural perimedullary arteriovenous fistulas (type IV spinal cord arteriovenous malformations). J Neurosurg 1994;81:221-9.
6. Cho KT, Lee DY, Chung CK, Han MH, Kim HJ. Treatment of spinal cord perimedullary arteriovenous fistula: Embolization versus surgery. Neurosurgery 2005;56:232-41.
7. Oran I, Parildar M, Derbent A. Treatment of slow-flow (type I) perimedullary spinal arteriovenous fistulas with special reference to embolization. AJNR Am J Neuroradiol 2005;26:2582-6.
8. Martirosyan NL, Kalani MY, Lemole GM Jr, Spetzler RF, Preul MC, Theodore N. Microsurgical anatomy of the arterial basket of the conus medullaris. J Neurosurg Spine 2015;22:672-6.
9. Giordan E, Brinjikji W, Ciceri E, Lanzino G. Arteriovenous fistulae of the filum terminale. J Neurointerv Surg 2018;10:191-7.

10. Iampreechakul P, Tirakotai W, Wangtanaphat K, Lertbutsayanukul P, Siriwimonmas S. Filum terminale arteriovenous fistula in association with degenerative lumbosacral spinal canal stenosis: Report of 3 cases and review of the literature. World Neurosurg 2020;138:231-41.

11. Chanthanaphak E, Pongpech S, Jiarakongmun P, Kobkitsuksakul C, Chi CT, Terbrugge KG. Filum terminale arteriovenous fistulas: The role of endovascular treatment. J Neurosurg Spine 2013;19:49-56.

12. Krings T, Lasjaunias PL, Geibprasert S, Hans FJ, Thron AK, TerBrugge KG, *et al*. Classification of spinal vascular malformations. Neuroradiol J 2009;22;97-106.

13. Kinouchi H, Mizoi K, Takahashi A, Nagamine Y, Koshu K, Yoshimoto T. Dural arteriovenous shunts at the craniocervical junction. J Neurosurg 1998;89:755-61.

14. Aviv RI, Shad A, Tomlinson G, Niemann D, Teddy PJ, Molyneux AJ, *et al.* Cervical dural arteriovenous fistulae manifesting as subarachnoid hemorrhage: Report of two cases and literature review. AJNR Am J Neuroradiol 2004;25:854-8.

15. Kai Y, Hamada J, Morioka M, Yano S, Mizuno T, Kuratsu J. Arteriovenous fistulas at the cervicomedullary junction presenting with subarachnoid hemorrhage: Six case reports with special reference to the angiographic pattern of venous drainage. AJNR Am J Neuroradiol 2005;26:1949-54.

16. Iampreechakul P, Lertbutsayanukul P, Siriwimonmas S, Jittapiromsak P, Tantivatana J, Niruthisard S. Persistent central neuropathic pain caused by intramedullary hemorrhage from spinal dural arteriovenous fistula: A case report and literature review. Austin J Anesthesia Analgesia 2019;7:1076.

17. Mourier KL, Gobin YP, George B, Lot G, Merland JJ. Intradural perimedullary arteriovenous fistulae: Results of surgical and endovascular treatment in a series of 35 cases. Neurosurgery 1993;32:885-91.

18. Ji T, Guo Y, Shi L, Yu J. Study and therapeutic progress on spinal cord perimedullary arteriovenous fistulas. Biomed Rep 2017;7:214-20.