Endovascular and Neurovascular Surgery for Spinal Vascular Malformations

Xianli Lv
Editor

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Preface

Spinal vascular malformations have a complex anatomical structure. Since their low incidence, it is generally well known to scholars. Their natural courses often lead to hemiplegia, bowel and bladder dysfunction, and a high disability rate. If treated properly, the patient's prognosis can be significantly improved. There has been a significant breakthrough in the surgical treatment and endovascular treatment of spinal vascular malformations. This made me think of editing a monograph on spinal vascular malformations, bringing together clinical and basic research on spinal vascular malformations. It is hoped that the research progress of spinal vascular malformations can be promoted, and breakthroughs can be made in basic and clinical aspects after appearance of this book. As far as I know, this is a monograph on *Endovascular and Neurovascular Surgery for Spinal Vascular Malformations*. Previously, there was a monograph on *Vascular Anatomy of the Spinal Cord*, which was published by Springer, editor-in-chief of Armin K. Thron. Together with this book on spinal vascular malformations, Springer Press will become more complete in spinal vascular anatomy and lesions. I thank all experts and scholars who have contributed to spinal vascular malformations to complete this monograph together. This will be a very interesting monograph because their contribution is rich and diverse, and their viewpoints are diverse.

Beijing, China
March 6, 2023

Xianli Lv

Commemorative of Academician Zhongcheng Wang

Prof. Zhongcheng Wang (20th, Dec, 1925 – 3 0th, Sep, 2012), Academician of Chinese Academy of Engineering

Acknowledgment

Thanks to Prof. Zhongxue Wu for his great contribution to Chinese Endovascular Neurosurgery, my great teacher, who is a human being of immense qualities, a man of incredible energy, and an extraordinary individual whom we will always love and admire.

Dr. Karel terBrugge (on the left), former Head of the Division of Neuroradiology and Site Chief of Medical Imaging at Toronto Western Hospital, and Dr. Xianli Lv (on the right) in 2012, Beijing, China

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About the Editor

Xianli Lv MD, is an associate professor of endovascular neurosurgery, at the Department of Neurosurgery, Beijing Tsinghua Changgung Hospital, School of Clinical Medicine, Tsinghua University, Beijing, China. He is selected for the "2023 Career-long and yearly Top 2% Global Scientists List" released by Stanford University in the United States. His research focuses on neuro-endovascular therapy of intracranial aneurysm, cerebral arteriovenous malformation, intracranial dural arteriovenous fistula, spinal vascular malformation, and pediatric cerebrospinal vascular malformation. He has one national invention patent and one utility model patent to his credit and has authored 195 peer-reviewed scientific articles and edited 10 books. His h-index is 31 in 2024 (Scopus). He is an editorial member of *Interventional Neuroradiology*, *Stroke and Vascular Neurology*, *Journal of Neuroradiology*, *The Neuroradiology Journal*, and *World Journal of Radiology*. He is also a deputy editor of Neuroscience Informatics and Frontiers in Neurology. Dr. Lv is a member of the World Stroke Organization (WSO) and the World Federation of Interventional and Therapeutic Neuroradiology (WFITN). He was featured on the cover of World Journal of Radiology in April 2022 and in June 2024, and journal of *China Science and Technology Achievements* in August 2023.

Vascular Anatomy of the Spine and Spinal Cord

1

Eilat Sapirstein, David Felzensztein, Eyal Hendler, and Eyal Itshayek

Abstract

This chapter reviews the arterial and venous anatomy of the spine and spinal cord, notably the intersegmental artery and its cranial and caudal derivatives: the vertebral, supreme intercostal, and sacral arteries.

Keywords

Artery · Vein · Anatomy · Spinal cord · Spine · Embryology

1.1 Introduction

The blood supply of the spine and spinal cord is highly complex and variable. Therefore, thorough knowledge of vascular anatomy and embryology is critical for understanding the pathophysiology of spinal vascular lesions and diseases. In addition, it serves as an essential tool for planning safe surgical and endovascular interventions.

Variability in the vasculature is frequently encountered; thus, the spine surgeon and endovascular interventionalist must be familiar with the possible anatomic variations before undergoing extensive diagnostic or therapeutic spinal procedure. Understanding the vascular supply to the spinal cord is especially important when considering different surgical approaches to avoiding devastating complications such as spinal cord ischemia or infarction.

This chapter provides a detailed description of the standard anatomical arterial supply and venous drainage of the spine and spinal cord, as well as embryological background and anatomic variations one must consider while considering surgical and endovascular interventions.

1.2 Embryology of the Spinal Vasculature

1.2.1 Embryology of Arterial Supply of the Spine and Spinal Cord

The embryological development of the arterial and venous system of the spine and spinal cord consists of four stages. The vascular system is fully established before 6 months of age, and there are no significant changes after that in the spinal cord vasculature.

The first stage, "the primitive segmental stage," occurs between 2 and 3 weeks of gestation. During this stage, 31 pairs of segmental vessels arise from the paired primitive dorsal aorta to supply 31 somites (segmental masses of the

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mesoderm) [1]. The primitive segmental artery divides into dorsomedial and dorsolateral branches and forms a capillary network on the surface of the developing neural tube. Each segmental artery supplies blood to the corresponding ipsilateral metamer—consisting of the future skin, muscle, bone, spinal nerve, and spinal cord [2, 3].

In the second stage, which occurs during 3–6 weeks of gestation, known as the "initial stage," arterial anastomoses continue to develop, as well as the formation of venous channels. This stage of development is susceptible to maldevelopment and malformations, and it is the stage in which the genesis of vascular malformation occurs and persists to adulthood [1].

From the sixth week to the fourth month of uterine life, the "transitional stage" occurs, in which the classic adult pattern of vascular supply will progressively form. Each longitudinal ventral neural axis provides several arterial branches, which run longitudinally and deep within the ventral sulcus and supply the anterior two-thirds of the spinal cord; these vessels are perforators that will become a part of the intrinsic arterial supply. The primitive ventral longitudinal arteries eventually fuse into a single ventral longitudinal vessel, the future anterior spinal artery. Posteriorly, two channels coalesce from a pial network on the lateral and dorsal surfaces to form the posterior spinal arteries. This process's alteration can lead to duplication and fenestration of the developing ASA [4].

During vasculogenesis, the future segmental arteries give rise to the radicular artery, which initially contributes to the anterior and posterior spinal arteries. By the end of the fourth embryological month, variable regressions of the segmental vessels take place, leaving about 4 to 8 anterior branches of the radicular arteries to supply the entire axis via the anterior spinal artery and from 10 to 20 posterior branches that supply the posterior spinal cord via posterior spinal arteries [5]. Variable segmental artery regression also leads to the formation of the vertebral arteries, thyrocervical trunks, and costocervical trunks in the cervical region and the iliac arteries and median sacral artery in the lumbar region. Regression is more prominent in the caudal region, leaving a single ARA at the lower thoracic and three upper lumbar regions that enlarge to supply the cord at this level—this artery is known as arteria radicularis magna or "artery of Adamkiewicz."

The last stage, the "terminal stage," occurs after 4 months of development in which the blood vessels mature and become more tortuous [1].

1.2.2 Embryology of the Venous Supply of the Spine and Spinal Cord

The first stage of venous development begins at the fourth gestational week. During this stage, two paired longitudinal veins, the anterior and posterior cardinal veins, are formed as the common cardinal vein. Concurrently, paired supracardinal and subcardinal veins develop inferior to the common cardinal vein and connect via transverse channels (the azygos, renal, and iliac veins). Simultaneously, the paravertebral plexus forms around the developing spinal cord, and the cardinal veins connect to the paravertebral plexus via transverse intersegmental veins [6, 7].

As the fetus matures and develops, the vertebral vein develops from the transverse intersegmental veins. The right supracardinal vein persists as the azygos vein, and the left supracardinal becomes the hemiazygos and accessory hemiazygos veins. The future internal and external jugular veins arise from the anterior cardinal veins, and the remnant of the right posterior cardinal vein becomes the arch of the azygos.

The azygos, hemiazygos, and accessory hemiazygos veins drain into the superior vena cava through the arch of the azygos vein [6, 7].

When compared to the arterial system, the venous system demonstrates high variability, especially concerning location and distribution.

1.3 Vascular Anatomy

1.3.1 Arterial Blood Supply

1.3.1.1 Arterial Supply to the Spine and Spinal Cord

Arterial blood to the spine and spinal cord can be divided into extrinsic and intrinsic or macrocirculation and microcirculation. Each spinal level has its unique vessel anatomy, with normal anatomic variations depending on the spinal level [5].

The branching anatomy of the arterial blood supply can be schematically considered as follows: a central arterial trunk (vertebral artery, aorta) gives rise to a spinal/segmental artery, which branches into radiculomedullary artery (ventral radicular artery) and radiculopial artery (ventral or dorsal artery) that give rise to a pial network, which connects the paired posterior or single anterior spinal arteries (Fig. 1.1).

Macrocirculation

The arterial supply to the spinal column derives mainly from the segmental arteries. The origin of the segmental arteries depends upon the spinal level. In the cervical region, they arise from the vertebral arteries, the ascending cervical arteries, the deep cervical arteries, and the occipital and ascending pharyngeal branches of the external carotid artery. The thoracic and upper lumbar spines originate in pairs from the posterior aspect of the descending aorta via intercostal and subcostal branches, from branches of the costocervical trunk, and from the internal thoracic artery of the subclavian artery. In the lumbosacral region, they arise from branches of the internal iliac artery (mainly the iliolumbar and lateral sacral arteries) and the median sacral artery (a branch of the aorta at the level of the bifurcation) [8]. There are extensive anastomoses between the segmental arteries providing collateral flow [9].

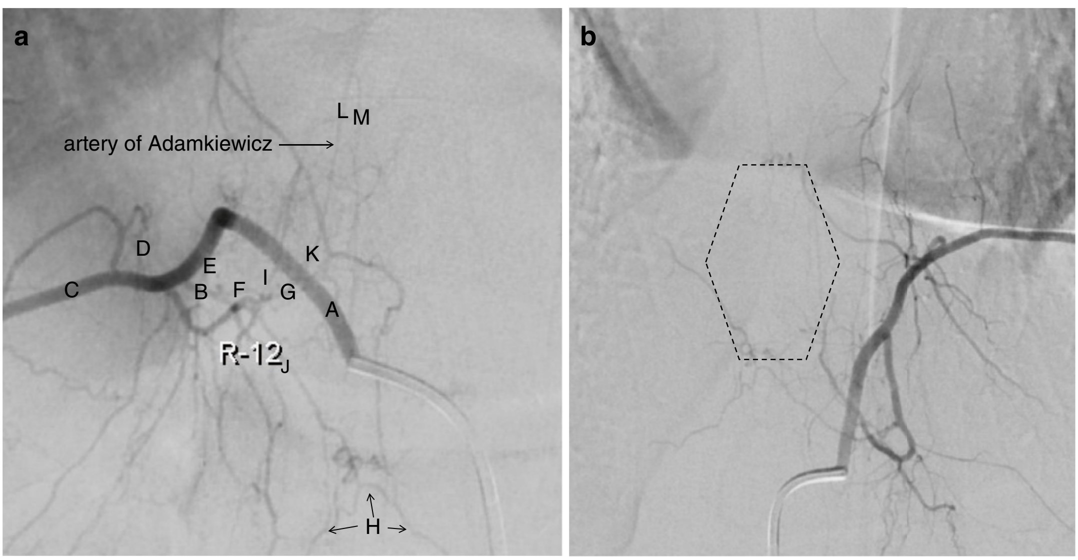

Fig. 1.1 (**a**) Right T12 segmental artery injection. (A) Segmental artery. (B) Dorsal spinal artery. (C) Intercostal/muscular artery. (D) Pretransverse anastomotic network. (E) Dorsal division of the dorsal spinal artery. (F) Ventral division of the dorsal spinal artery. (G) Radicular artery. (H) Ventral epidural arcade. (I) Nerve root sleeve dural branch of the ventral division dorsal spinal artery. (J) Dural branch of the ventral division dorsal spinal artery. (K) Radiculopial artery. (L) Radiculomedullary artery. (M) Anterior spinal artery. The artery of Adamkiewicz arising from the right radiculomedullary artery at T12 level. Notice the characteristic hairpin turn as the artery of Adamkiewicz joins the anterior spinal artery (M). (**b**) The catheter in the left T6 segmental artery and a hexagon-shaped multilevel anterior epidural arcade (retrocorporeal artery)

The segmental arteries continue their course posteriorly and laterally along the vertebral body, dividing into three significant trunks—lateral or ventral trunk, middle or dorsal trunk, and medial or spinal trunk. They enter the spinal canal via the intervertebral foramen. Each trunk further divides into anterior and posterior spinal canal arteries that supply the vertebral bony and ligamentous structures and a radicular artery that supplies the dura and nerve root at each level. The radicular arteries that supply the dura and nerve roots are called radiculoradial or radiculomeningeal arteries. At several levels but not all, the radicular artery gives branches that follow the anterior and posterior nerve roots termed radiculomedullary arteries. The radiculomedullary arteries further divide into anterior and posterior branches that supply the ASA and PSA, respectively [5].

Anterior Spinal Artery

The anterior spinal artery (ASA) arises at the level of the foramen magnum from the vertebral arteries originating from the first part of the subclavian artery. It passes along the anterior sulcus of the spinal cord and descends to the level of the conus medullaris.

Due to its long course and the significant structures it serves, additional arterial supply is required to reinforce its blood supply, which is supplied via the anterior radiculomedullary arteries. There are three major regions of blood supply to the anterior spinal artery from the radiculomedullary arteries: the cervicothoracic, midthoracic, and thoracolumbar regions. The most critical and dominant anterior radiculomedullary artery is the artery radiculomedullaris magna, also known as the artery of Adamkiewicz (AKA). This vessel almost always arises in the thoracolumbar region, between T8 and L2, and in 80% on the left side [9]. The territory of blood supply by the ASA includes the anterior two-thirds of the spinal cord [5, 10].

Posterior Spinal Artery

The two PSAs originate at the level of the foramen magnum from the ipsilateral vertebral or posterior inferior cerebellar arteries. They pass along the right and left posterolateral surfaces of the spinal cord. Around 10–20 feeders from the posterior radiculomedullary arteries supply the PSA at various levels. The anatomical distribution of the blood supply of the PSA includes the posterior third of the spinal cord [10].

Pial Plexus

The ASA and PSAs form end anastomosis at the level of the conus medullaris. Along the entire spinal cord along its surface, another extensive arterial network is found termed pial plexus or "vasocorona." It is responsible for supplying the periphery of the spinal cord [4].

Microcirculation

Microcirculation, also known as the intrinsic spinal cord supply or spinal cord perforators, consists of two groups of arteries arising from the anterior and posterior spinal arteries. These branches can be divided into centrifugal (from the center of the cord toward the surface, central) and centripetal (from the cord surface toward the center, peripheral) systems. The ASA is the main contributor to the centrifugal system, and both ASA and PSA contribute to the centripetal system. The centripetal system supplies mainly the white matter and the centrifugal gray matter of the spinal cord [4, 11].

The central system consists of about 200–400 central arteries (aka sulcal or sulcocommissural arteries) originating from the ASAs. These vessels travel in the anterior median fissure and penetrate the sulcus to enter the central gray matter. They branch centrifugally into small arteries that run toward the white matter. The sulcocommissural system supplies most of the spinal cord gray matter and the ventral half of the white matter [5].

The peripheral system consists of radial perforating arteries originating from the posterior spinal arteries and the pial plexus and penetrating the white matter. These centripetal arteries supply the posterior part of the posterior gray matter and the outer half of the white column [2].

1.4 Venous Blood Supply

The venous drainage of the spine and spinal cord can be divided, similarly to the arterial system, into an intrinsic system, which runs in proximity to the centrifugal arterial system, and an extrinsic system, which runs in proximity to the centripetal arterial system, and extradural component.

1.4.1 Intrinsic Venous System

The intrinsic system is composed of a network of radially arranged venous channels [6]. Capillaries from the medial parts of both sides of the spinal cord drain into ventral and dorsal sulcal veins. The number of ventral sulcal veins varies over the length of the spinal cord and increases over the thoracolumbar spine. Capillaries from peripheral gray matter and white matter travel toward the periphery forming radial veins; when reaching the surface, they form a venous ring that eventually drains into the external system. A longitudinally oriented intrinsic venous system is also present, interconnecting the radial veins [6, 9].

1.4.2 Extrinsic Venous System

The extrinsic system arises at the level of the spinal pia mater and includes the pial venous networks, the longitudinally oriented extrinsic venous system, and radicular veins.

The pial network, as mentioned earlier, drains the sulcal veins.

The anterior and posterior spinal veins represent the longitudinal venous system at ventral and dorsal surfaces in the midline of the spinal cord. The anterior median spinal vein accompanies the ASA and drains the sulcal veins and the veins of the ventral fissure. The posterior spinal veins (posterior median and posterolateral) accompany the PSA and receive blood supply from the radial veins of the dorsal spinal cord. Up to three ventromedial venous channels are present in the ventral cervical and upper thoracic regions. Three anterior cervical and upper thoracic longitudinal channels converge in the lower thoracic area, continue as a duplication, and unite proximal to the lumbosacral area. There is triplication in the thoracic region on the cord's dorsal surface, and the vessels converge at the cervicothoracic and thoracolumbar junctions [5, 6, 9].

The anterior and posterior median spinal veins drain into the radiculomedullary veins, which accompany the anterior or posterior spinal nerve root: the great anterior radiculomedullary vein (GARV) is the most prominent vein draining the anterior thoracolumbar spinal cord. In the intervertebral foramen, the radiculomedullary and radicular veins exit through the dura and drain into the epidural vertebral venous plexus via the spinal nerve venous channels [6].

1.4.3 Extradural Venous System

The vertebral venous plexus (Batson plexus) is divided into three divisions: the internal and external vertebral and the basivertebral plexuses [6].

The internal vertebral venous plexus (anterior and posterior) is located epidurally within the vertebral canal and continues superiorly to communicate with the cranial venous system. It includes two anterior and two posterior internal plexuses. The anterior (front of the vertebral bodies) and posterior (surrounding the posterior elements) external vertebral venous plexus surrounds the vertebral column, and the basivertebral veins run horizontally within the vertebral body [9].

Cervical, thoracic, lumbar, and sacral intervertebral veins connect the external and internal vertebral venous plexuses and communicate with various extraspinal longitudinal veins depending on the vertebral level.

Drain occurs via the vertebral, deep cervical, and jugular veins at the cervical level, ultimately into the superior vena cava. The thoracic and lumbar intervertebral veins connect to the azygos system, the ascending lumbar veins, and the superior vena cava. The ascending lumbar veins communicate with the inferior vena cava via lumbar segmental veins at the lumbosacral level. The sacral veins empty into the lateral sacral and internal iliac veins.

Valves exist only in the radicular branches draining the spinal cord. These valves prevent congestion of the spinal cord and cannot be found in other components of the venous system.

1.5 Anatomic Variations

The vasculature of the spinal cord is highly variable in size and structure due to differences in embryological development and proliferation, metabolic requirements, and blood flow demand. This section presents some of the most common anatomic variations one must consider.

Vertebral artery variants—the vertebral artery usually arises from the posterosuperior aspect of the first part of the subclavian artery. The most common variant is for the left vertebral artery to originate off the aortic arch between the left common and left subclavian artery, with a 2–5% prevalence [12]. An anomalous left VA arising directly from the aortic arch typically enters the C4 or C5 foramen transversarium, resulting in a longer course of VA in the neck. Aberrant right VA is an extremely rare anomaly.

Arterial duplication is dividing an artery into two distinct vessels with different courses. Fenestrations are a single arterial lumen that divides into two parallel channels following the same course. The frequency of duplication of the VA is around 0.72% in cadavers [12]. Other VA pathologies exist, including tortuosity, elongation, and kinking [12].

Artery of Adamkiewicz variants—75% of the time, the artery inserts into the dura and ASA at the level T9–T12. Fifteen percent inserts into the dura and ASA at the level T5–T8, and 10% of the time, the artery is inserted into the dura and ASA at the level L1–L2 [13].

Variants of ASA and AKA junction:

- Type 1: a small-caliber ASA joining a larger-caliber AKA. These vessels form a large ASA descending from that level. This variant is considered an AKA dominant circulation, and disruption of the AKA in patients with this circulation could lead to severe neurological damage.
- Type 2 variant is where the ASA and AKA are the same calibers, and when joining together, the caliber of the ASA does not increase (non-dominant AKA) [10].

Duplication of the ASA—until C5/C6 level is typical.

Variation in the number and origin of the radicular arteries.

Variations in the origin of PSAs—posterior spinal arteries can originate from either the VA or the PICA and with a symmetrical or asymmetrical configuration. The intracranial segment of the VA is the most common origin [14].

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Types of Spinal Vascular Malformations

2

Manas Panigrahi, Gudipati Ananta Ram, and Jitender Chaturvedi

Abstract

Spinal vascular malformations are often complex problems with varied clinical presentations. In this chapter, we will briefly overview common clinical presentations of patients with spinal vascular malformations after reviewing the normal vascular anatomy of the spine and then outline different types of vascular disorders, and possible treatment options. Catheter angiography remains the gold standard investigation for diagnosis and planning of treatment. Therapeutic options, treatment success rate and limitations are briefly described in particular.

Keywords

Spinal vascular malformation · Extradural · Dural · Intramedullary · Spinal dural AV fistula · Spinal angiography · Anterior spinal artery (ASA) · Posterior spinal artery (PSA) · Spinal arteries · N-Butyl cyanoacrylate · Endovascular Treatment

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2.1 Introduction

Spinal dural arteriovenous fistulae (sDAVF) are rare vascular lesions in neurovascular disease spectrum. The spine and spinal cord constitute just 1–2% of all vascular neurological disorders [1]. However, more than 80% of these spinal vascular pathologies are included within the umbrella disorder of sDAVF, making it the most common spinal vascular pathology [1]. Myelopathy is the usual presentation of this disease, though sensory and sexual symptoms are also seen. Treatment options consist of endovascular embolization, surgical disconnection, or a combination of both [2]. Surgical anatomy of arterial supply and venous drainage of the spinal cord is also discussed.

2.1.1 Endovascular Anatomy of Spinal Cord Arterial Supply and Venous Drainage

Before understanding the pathophysiology of shunting behind the origin of sDAVF, it is vital to understand the surgical anatomy of spinal cord blood supply. Various segmental arteries supply the spinal cord at multiple levels. Segmental arteries originate from the vertebral artery, thyrocervical and costocervical trunks, supreme intercostal artery in cervical and

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upper thoracic region, descending aorta in lower thoracic and median sacral artery in sacral regions.

The segmental arteries are named, e.g., D10 segmental artery that supplies D10 vertebrae. Segmental arteries are not named based upon the level of origin. Segmental artery gives three branches-spinal, dorsal and ventral. Sometimes a small trunk for the dorsal and spinal branches called dorsospinal trunk is present. The dorsal branch gives medial and lateral musculocutaneous branches which supply para spinal musculature, posterior aspect of lamina and spinous process. Osseous branches from segmental artery are also known as ventral somatic branches, supply ventrolateral surface of vertebral body. Ventral branch is nothing but the intercostal or lumbar segmental arteries. Superior intercostal artery. This is the most cephalad intercostal artery arising from the aorta, ascending to supply upper thoracic vertebral levels. Supreme intercostal artery arising from the costocervical trunk providing radicular arteries to the C7 and C8 levels.

Spinal branch gives Anterior epidural, radicular, and posterior epidural (prelaminar) arteries. Anterior epidural branches (dorsal somatic) connect with opposite and adjacent arteries, and form a diamond-shaped (retrocorporeal) anastomotic network. Posterior epidural branch contributes to the prelaminar anastomosis and supplies the dura and bone of the lamina. Based upon their depth of penetration in cord, these spinal radicular arteries are named [3] as radicular (radiculomeningeal), radiculopial, and radiculomedullary. A spinal radicular branch supplying the dura and the nerve root as a radiculomeningeal artery is present at each segment. At some of these levels, the radicular artery is enlarged as it also gives supply to the spinal cord via a radiculomedullary artery (Average, 6) which has ascending course to the ASA. Similarly, it may supply the ipsilateral PSA via a radiculopial artery, with predominant pial plexus supply (range10–20). The *largest radiculomedullary artery* is the famous artery of Adamkiewicz usually originating from T9–T12 (in 75% of cases, it originates between T8 and L2 but may go as high as T3 and as low as L4 to get origin). This artery, which most commonly originates from the left side (80%), is easily identified on angiogram, with a typical hairpin loop and a large descending along with smaller ascending branch. Longitudinal arterial trunks that extend along the long axis of the spinal cord surface are Anterior spinal artery and two posterior (or posterolateral) spinal arteries. The anterior spinal artery (ASA) is typically formed at the level of the foramen magnum by the confluence of descending branches of the intracranial segments of the vertebral arteries. Due to its long course, the ASA requires additional arterial supply via anterior radiculomedullary arteries at cervicothoracic, midthoracic and thoracolumbar. The two Posterior spinal arteries (PSAs) also originate at the level of the foramen magnum by branches of the ipsilateral vertebral or posterior inferior cerebellar arteries. Multiple anastomoses exist between the ASA and PSAs; these include the circumferential vasa corona vessels (pial network), the intramedullary vessels (i.e., sulcocommissural arteries), and the basket anastomosis in the region of the conus medullaris.

In relation to the transverse process, there are two longitudinal anastomotic plexuses, i.e., *pre-* and *post-transverse longitudinal anastomosis.* The dorsal branch of segmental artery communicates with these pre-transverse anastomoses. Post-transverse longitudinal anastomosis communicates freely with posterior intercostal arteries and lumbar arteries. These pre- and post-transverse longitudinal anastomoses also communicate freely with each other.

In lumbar spine, only up to L4 segmental artery, the aorta is the origin. L5 segmental artery originates from iliolumbar artery and median sacral artery. Lateral sacral arteries arising from internal iliac artery supplies sacral nerve roots.

Multiple anastomoses connect the left and right PSAs across the midline (posterior plexus)-rope ladder. PSA lies on either side of posterolateral surface of cord, medial to the posterior root entry zone.

Intrinsic arterial network of spinal cord: The ASA will give sulcocommissural arteries (centrifugal) to supply mainly the spinal cord gray matter, and both the ASA and PSA will give a pial network with small perforating (centripetal) vessels, to supply the white matter.

Venous drainage of the cord runs in three types of veins: internal cord veins, longitudinal veins and radiculomedullary veins.

Radiculomedullary veins follow the anterior and posterior roots. Internal cord veins form centrifugal pattern of drainage from central to periphery.

The short peripheral veins follow a radial course ending in the coronary plexus. The perimedullary venous (coronary) network interconnects the longitudinal spinal trunks (posterior and anterior median spinal veins).

Trans-medullary veins are veins that traverse the cord parenchyma and form anastomosis between the anterior and posterior surface veins in midline.

Longitudinal veins are anterior median and posterior medial spinal veins (usually 3 in number with constant posterior median spinal vein). Interestingly, the junction of median spinal veins with the radiculomedullary veins is an obtuse angle, giving it an often quoted *"coat hook" appearance.* It is important to make sure that this "coat hook" is not mistaken with *"hairpin"* of the artery of Adamkiewicz on angiogram. Radiculomedullary veins exit the dura, usually with the nerve root, may join radicular veins and empty into the internal vertebral venous plexus of the extradural system. The radicular and bridging veins are the key link between intradural and extradural venous systems. Intervertebral (emissary/foraminal) veins link internal vertebral (epidural) and external venous plexus. Intervertebral veins via segmental veins finally drain into azygos, hemiazygos venous systems or jugular vein.

2.1.2 Pathophysiology

Shunting is the main pathophysiological mechanism leading to symptomatology of sDAVF. Direct shunting of blood from radiculomeningeal artery to radicular vein leads to venous congestion in valveless spinal cord veins. This venous hypertension causes symptom onset. Secondary to spinal arterial perfusion pressure drop, spinal cord undergoes ischemic changes, and symptoms start to appear. In the terminal stage of the disease without treatment, necrotizing myelopathy sets in, which is irreversible and devastating. Usual site seen for this shunting is the thoracolumbar region, i.e., the site of sDAVF. Etiology is idiopathic in majority of the cases, though post-surgical and post-traumatic cases have also been reported in the literature [4]. Cervical cord is an extremely rare site for sDAVF to develop.

2.2 Classification and Types

We use a classification described by Keisuke Takai as it is according to the sites (dural, intradural, and extradural) and types (AVF and AVM) of AV shunts [5].

2.2.1 Type I Dural AVF

Most commonly appearing at thoracolumbar region, type I represents the most common (~70%) type of spinal vascular malformations. Progressive compressive myelopathy is the usual presentation. When located at the level of CVJ or upper cervical spine, the lesion may present at sub arachnoid hemorrhage. These myelopathic changes occur secondary to venous hypertension and intra-medullary venous stagnation, owing to a fistula between the dural branch of segmental artery and radicular vein. Anatomically, this fistula resides at the level of dural root sleeve. More than half of the cases worsen to a level of permanent disability within 24–36 months [6], if left untreated. Treatment consists of surgically disconnecting the draining vein of the fistula, intradurally. Surgical success is around 98% if the draining vein is identified properly [6]. Other treatment option is endovascular obliteration of the draining vein at proximal location. Complete obliteration is essential to get minimal recurrence (up to 20% overall recurrence is noticed in various series) [7]. It is important to learn that endovascular treatment is contraindicated when anterior or posterior spinal artery is supplied by radicular artery.

2.2.2 Type II Intramedullary Glomus AVM

When a parenchymal or intramedullary lesion has a nidus-like cranial AVM, it is classified as type II. Intramedullary hemorrhage, in addition to more common congestive myelopathy or SAH, can also be the presenting symptom. This lesion has multiple feeders from anterior as well as posterior spinal artery.

Embolization is the first line of treatment, keeping in mind that complete obliteration may never be achieved and both complete and partial obliterations reduce the recurrences of symptoms by decreasing the re-bleed rates [8]. Usual surgical excision of the nidus is obviously too risky to be advised. In presence of inadequate obliteration after embolization or presence of angiographic high risks of embolization, a guided and planned surgical excision may be offered.

Radiosurgery is a third modality, which has been explored in these lesions. Stereotactic radiosurgery has been used by researchers with documented success in motor symptom improvement and partial obliteration of AVM nidus [8].

2.2.3 Type III Intramedullary Juvenile AVM

When lesions appear metameric, i.e., multiple vascular malformations appearing from the same spinal metameric segment, it is sine qua non for diagnosing this variant. Adolescent and young adults are the usual sufferers. Fortunately, these are rare variants. Complete obliteration is rarely observed and never a target in planning treatment of this variant. Terminating the progression of neurological deficits is considered the success of treatment as genetic mutations are involved in pathophysiology of the disease.

2.2.4 Type IV Perimedullary AVF

In the absence of intervening nidus, direct arteriovenous communication on the spinal cord pial surface is the hallmark of this variant. Perimedullary fistula form more often on ventral surface of the spinal cord and typically have a midline location.

There are three different subtypes of this variant ranging from IVa to IVc depending on the size, level of blood flow and venous drainage. Type IVa single arterial feeder to the vein with slow flow. Type IVb has multiple dilated arterial feeders of intermediate size, while type IVc has multiple dilated arteries and ectatic draining veins. Type IVc has association with HHT, i.e., hereditary hemorrhagic telangiectasia, and appear in younger patients, more often.

Type IVa perimedullary AVFs are more amenable to microsurgery. Type IVb and IVc respond favourably to endovascular treatment due to multiple feeding arteries and shunt volume. IVb may require surgery for failed endovascular treatment.

2.2.5 Type V Extradural AVF

Congestive myelopathy is the clinical presentation, often indistinguishable from type I commonly located at lumbar level with frequent bilateral epidural arterial supply. Distinction is done by angiographically proving a direct fistulous communication between an extradural artery and vein. Extradural AVFs were subdivided into Subtypes Va- with intradural venous drainage and Vb without intradural venous drainage.

Type A has a retrograde parenchymal drainage with a perimedullary vein, which goes intradural and leads to venous congestion leading to symptoms. Treatment consists of treating extradural venous lake and this intradural perimedullary vein. If the draining vein is large enough to give an access, transvenous embolization can be done, if there is single draining vein. With multiple draining veins, meticulous coagulation and separation of the epidural venous lake is advised type Vb extradural AVFs are more amenable to endovascular embolization.

2.2.6 Conus Medullaris AVM

This variant is a subtype of type II where a nidus is clearly visible, added as a separate variant since last two decades. As indicated by the name,