Spinal AVM

Moderators: Dr. M.M.Singh
Dr. Vivek Tandon
Presentor : Mansukh

Introduction

- Vascular malformations of spinal cord are a rare clinical entity, representing 5% of all primary spinal cord lesions, with arteriovenous malformations(AVM) & cavernous malformations being the most common.
- They are relatively heterogeneous with distinct angiographic pattern and associated with specific etiology, pathophysiology, natural history and even a different therapy.

Historical perspective

- In 1888 -Gaupp :1st to describe a spinal dural AVM from a postmortem study.
- In 1910 –Fedor Krausse :1st attempted to resect lesion .
- Sir charles elsberge : 1st successful therapeutic operation for a spinal AVM in 1914 .
- 1926, Foix and Alajouanine reported the syndrome of subacute necrotic myelopathy associated with rapidly progressive onset of paraplegia and subsequent death.
- In 1960 with introduction of selective spinal angiography vascular anatomy of spinal AVMs became more clear .

Vascular anatomy

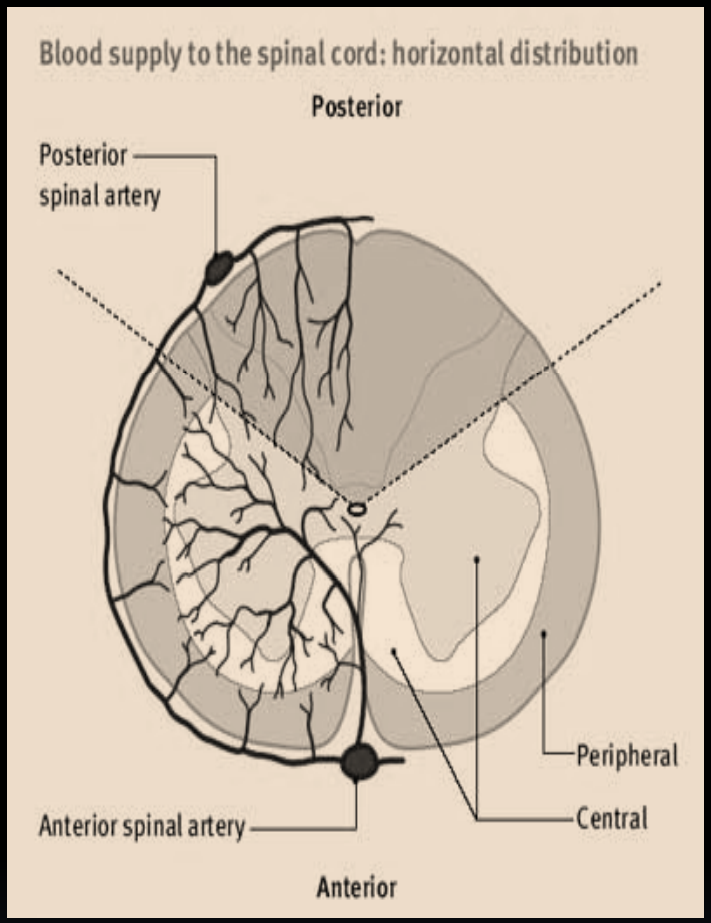
Arterial anatomy –

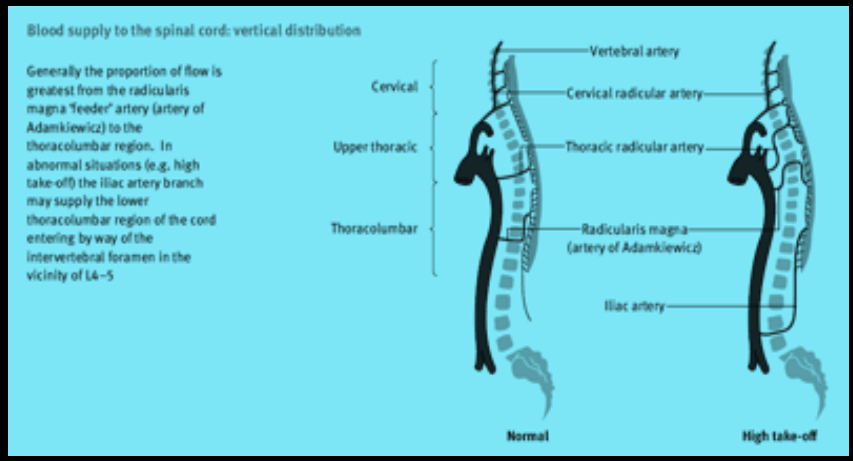
(1) Medullary arteries

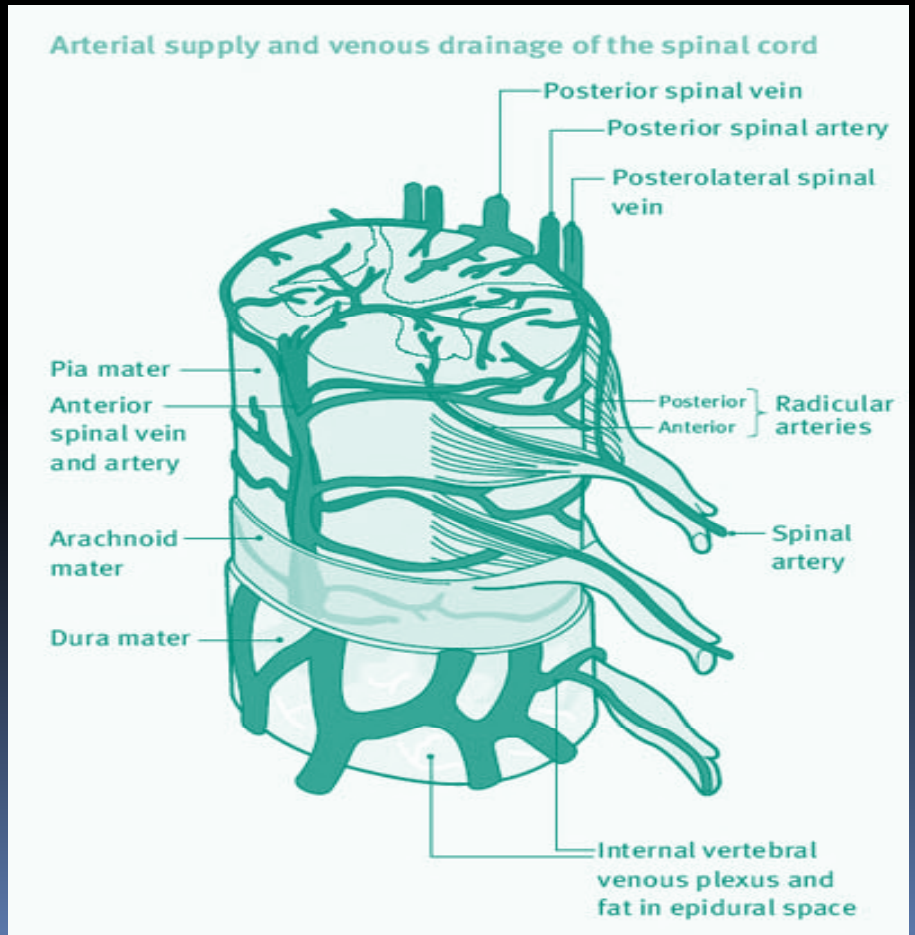
- Two arterial networks (a)Anterior (b) Posterior
- Both system supplied by medullary artery
- Upper thoracic –watershed area

(2) Radicular arteries-supplies dorsal / ventral nerve roots

(3) Dural arteries – dural root sleeves and spinal dura


Blood supply to the spinal cord: horizontal distribution
Posterior
Posterior spinal artery
Peripheral
Central
Anterior spinal artery
Anterior


Blood supply to the spinal cord: vertical distribution
Generally the proportion of flow is greatest from the radicularis magna 'feeder' artery (artery of Adamkiewicz) to the thoracolumbar region. In abnormal situations (e.g. high take-off) the iliac artery branch may supply the lower thoracolumbar region of the cord entering by way of the intervertebral foramen in the vicinity of L4–5
Cervical
Upper thoracic
Thoracolumbar
Vertebral artery
Cervical radicular artery
Thoracic radicular artery
Radicularis magna (artery of Adamkiewicz)
Iliac artery
Normal
High take-off


Arterial supply and venous drainage of the spinal cord
Posterior spinal vein
Posterior spinal artery
Posterolateral spinal vein
Pia mater
Anterior spinal vein and artery
Posterior
Anterior
Radicular arteries
Arachnoid mater
Spinal artery
Dura mater
Internal vertebral venous plexus and fat in epidural space

Venous drainage

- Spinal cord drained by radial veins which carries blood from parenchyma to surface where they empty into the sulcal vein & coronal venous plexus in piamater on the cord and via medullary vein to epidural venous plexus .

Classification

Niimi and Berenstein in 1999:

a) Spinal vascular lesion –

spinal dural avf

spinal extradural and paraspinal avf

b) Spinal cord vascular lesion -

AVF / AVM

metameric / non metameric

c) Spinal cord telangiectesias

d) Cavernous vascular malformation

Endovascuar treatment of spinal vascular malformation
Neurosurg clin N Am 10:47 -71,1999

Classification

- Anson and spetzler in 1992
- **Type I. Dural (intradural or extradural) AVF**
  (also referred to as Type I spinal AVM or as "angioma racemosum venosum," nidus, or true AVM)
- **Type II. Glomus AVMs**
- **Type III. Juvenile AVMs** (nidus usually intramedullary)
- **Type IV. Direct spinal AVF**

Classification of spinal arteriovenous malformation and implication for treatments: BNI Q 8 :2-8 1992

- **YURI P. ZOZULYA, EUGENE I. SLIN'KO, AND IYAD I. AL-QASHQISH, (2006)**
  I. Intramedullary
  II. Intradural or perimedullary
  III. Dural
  IV. Epidural
  V. Intravertebral
  VI. Combined

Spinal arteriovenous malformations: new classification and surgical treatment
Yuri P. Zozulya, et al Neurosurgical FOCUS May 2006, Vol. 20, No. 5: 1-17.

Extradural AVF

- Rare vascular lesion
- Abnormal communication between an extradural radicular artery and epidural venous plexus.
- Progressive myelopathy / radiculopathy occasionally epidural hematoma
- Diagnosis- MRI / Angiography

Extradural AVF

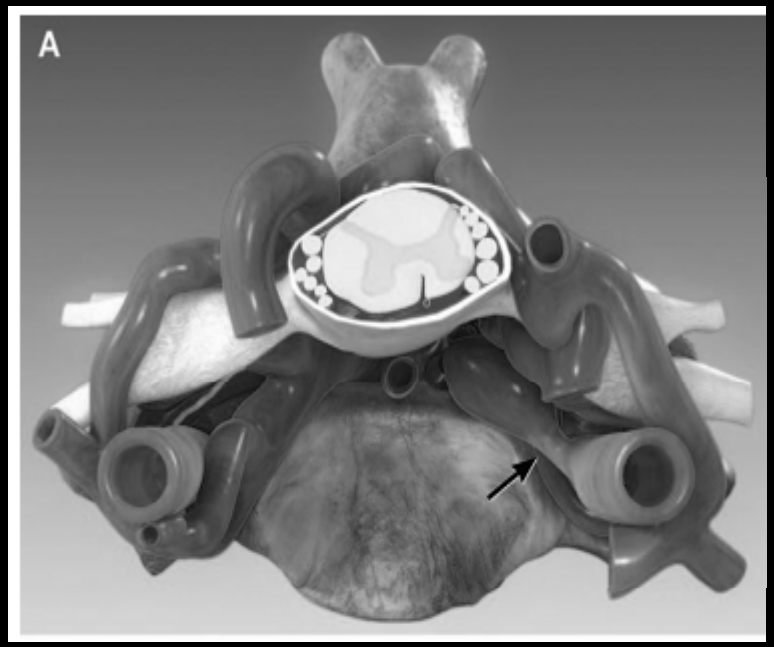
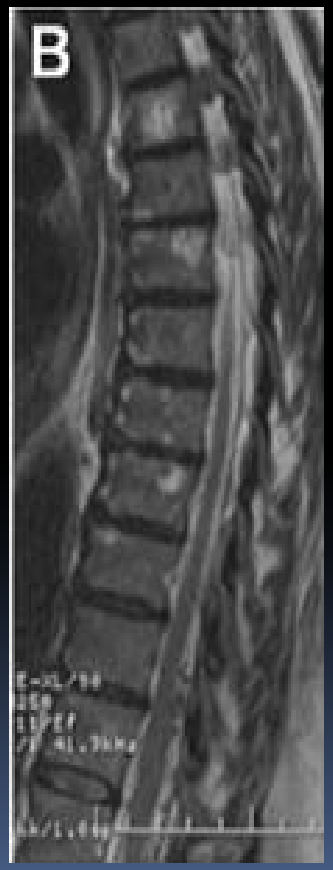


Fig A- axial view demonstrating an extradural avf along perforating branch of left vertebral artery

Fig B – posterior view showing that engorgement of epidural vein can produce symptomatic
mass effect on adjacent nerve root and spinal cord

- On Angiography : high flow lesion
- Type 1 AVM or dural AVF can be easily confused with a spinal extradural AVF.



Treatment

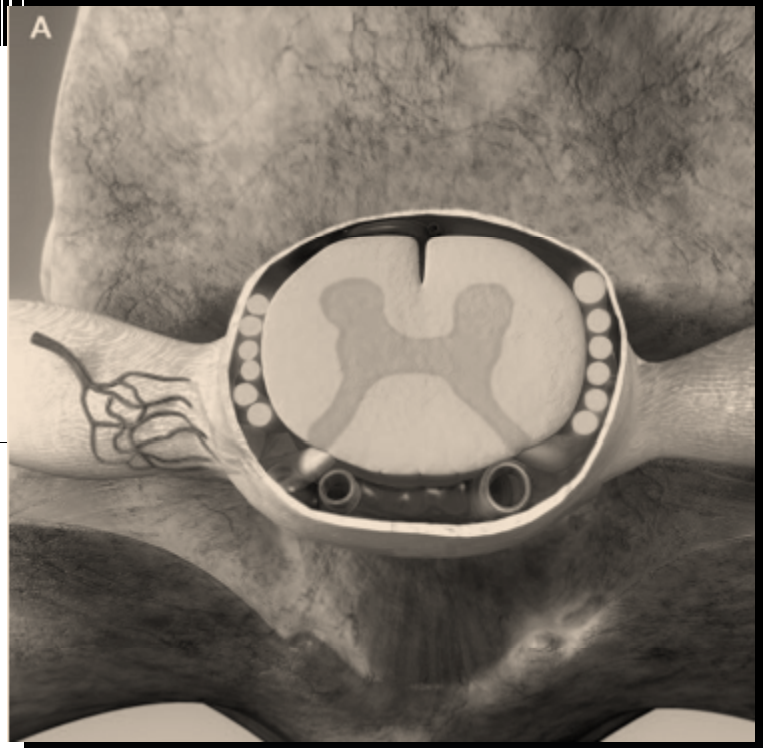
- Extradural AVF treated by endovascular treatment with embolisation of arterial feeder .

Intradural Dorsal AVF :type - 1

- Type –I spinal AVM also known as spinal dural AVF
- Most common type AVF : – 80 - 85 %
- Pathophysiology - slow flow fistula
  Between dural branch of radicular artery and intramedullary vein

- Clinical presentation
  Age – middle age (40 – 60 year)
  Male 90 % > female 10 %
  Site – thoracic spine
  Progressive myelo-radiculopathy or cauda equina syndrome
  Aggravated by raised intra abdominal pressure
  Sub arachnoid haemorrhage rare
  Acute deterioration unlikely
- Two sub type – Type -1a : single arterial feeder
  Type-1b : two or more arterial feeder

Dorsal Intradural AVF :type - I



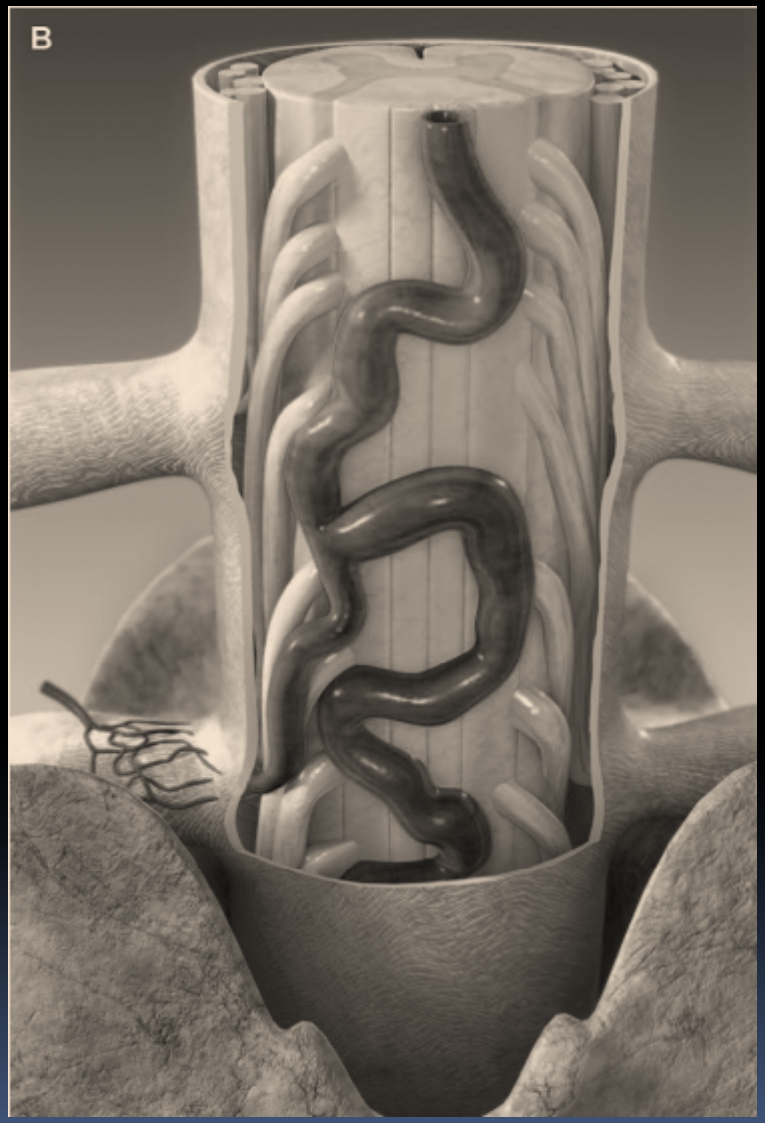
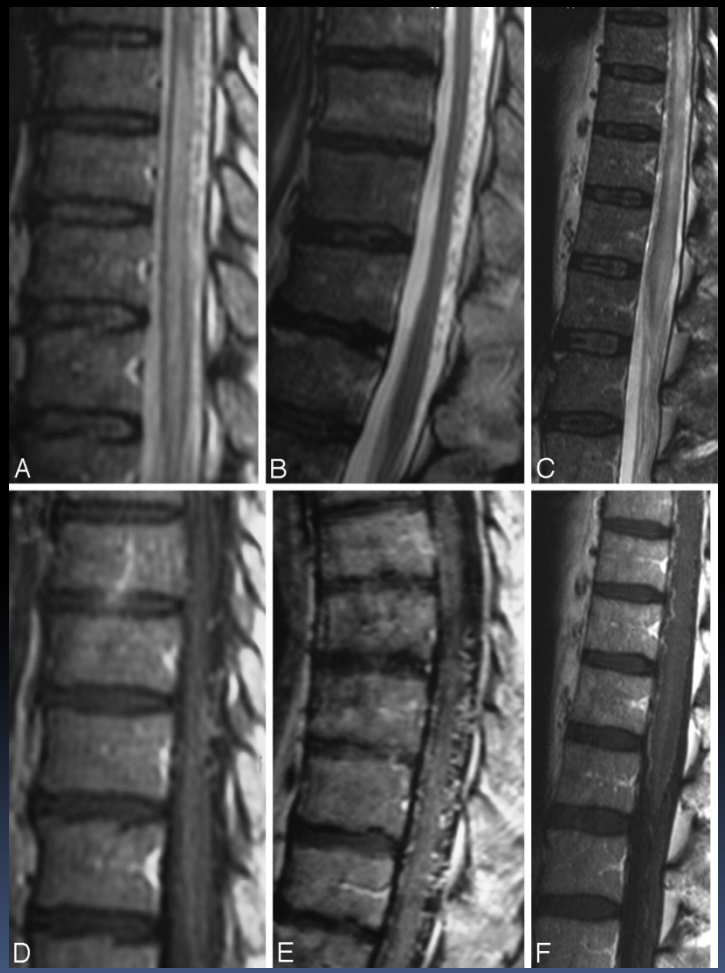


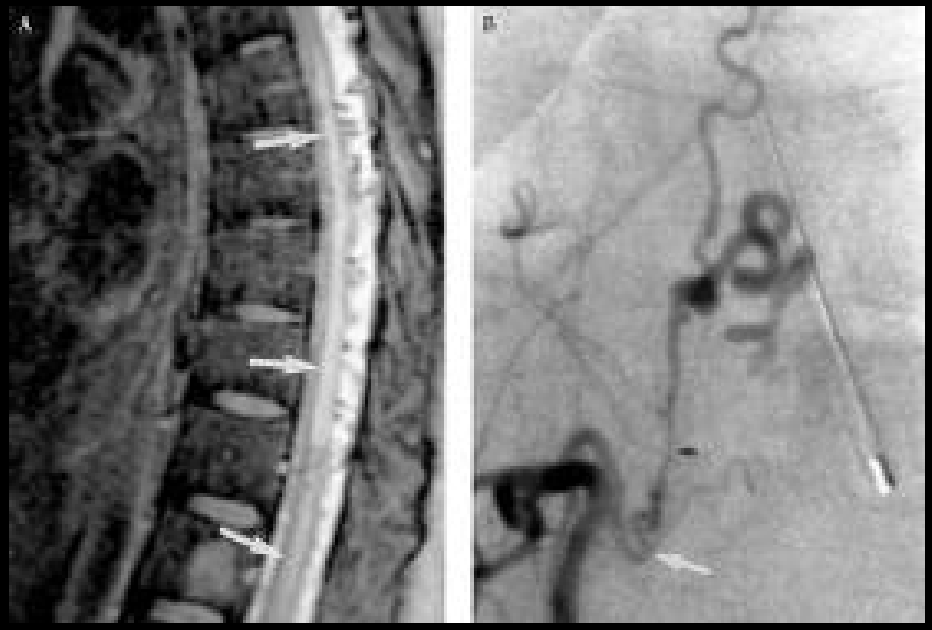
Fig A intradural dorsal AVF demonstrating abnormal redicular feeding artery along nerve root on rt side

Fig B posterior view showing dilatation of coronal plexus .

Diagnosis



MRI : the Perimedullary vein expansion look like serpiginous flow voids on the dorsal surface of the spinal cord, most often in the middle and lower thoracic spine.
spinal cord edema and thickening are typical



Angiographic studies

- Reveal expanded radiculomeningeal arteries, a vascular conglomerate in the region of the intervertebral neural foramen shunted into the expanded perimedullary veins.
- Nidus characterized by slow blood flow
- The blood flow in spinal cord arteries is also slower

Treatments

- Goal : isolation and obliteration of the fistula and draining veins.
- Best treated surgically:
  - Direct surgical ligation of arterial feeder
  - Low morbidity
  - High success rate
- Embolization of the feeding vessel, via endovascular techniques
  - Less invasive /same sitting /early rehabilitation

Dural AVMs

- Two variants of surgical technique

1) Occluding the malformation in the dural leaf of the spinal nerve root or cutting off the feeding vessels immediately outside the root.

2) occluding the radicular vein, which provides retrograde blood shunting from the AVM into the perimedullary veins.

Intradural Ventral AVF: type IV

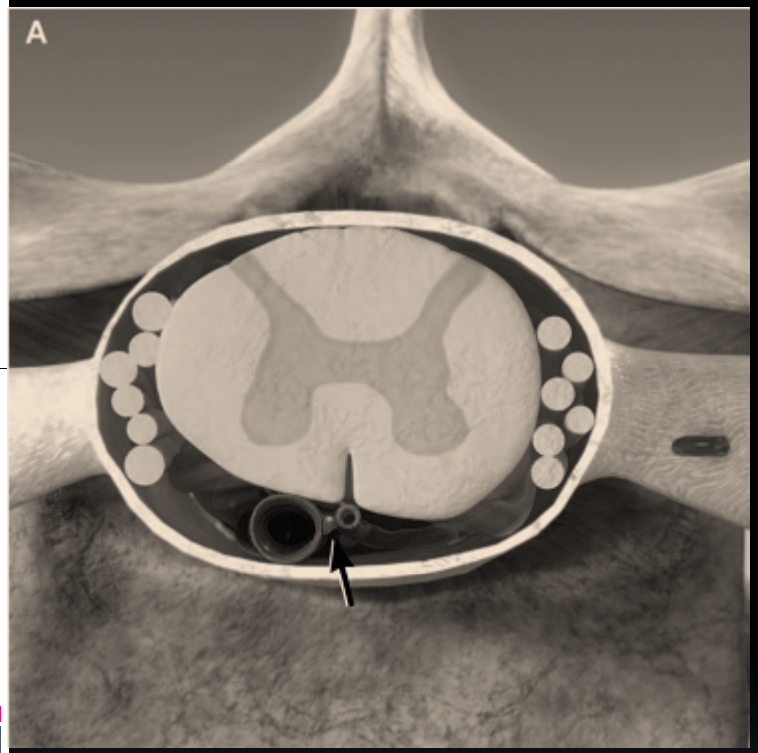
- Rare lesion , not true AVM but instead Pial AVF
- First describe by Djindjian & coworker as Perimedullary AVF
- Heros & associates introduce term Type – iv
- Direct fistula between ASA and draining vein
- Merland's Sub classification

  IV-A : smallest shunt , slow blood flow ,less venous hypertension

  IV- B : larger shunt , higher blood flow & greater degree hypertension

  IV-C giant : very high degree blood flow, with significant hypertension

Ventral intradural AFV: type –IV



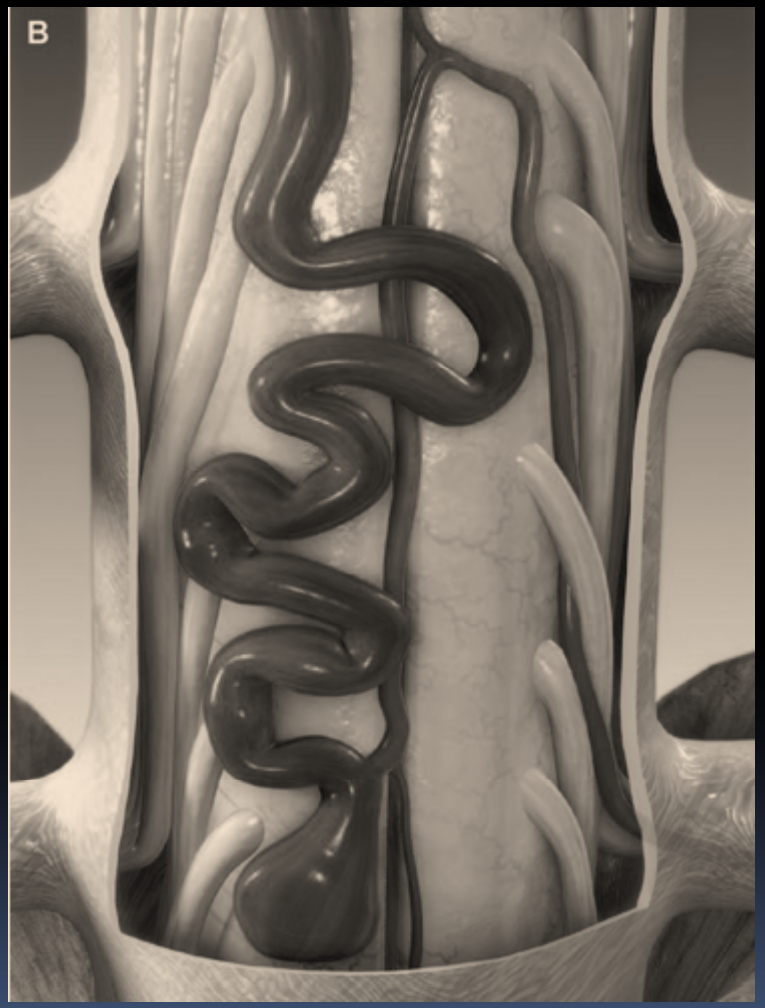
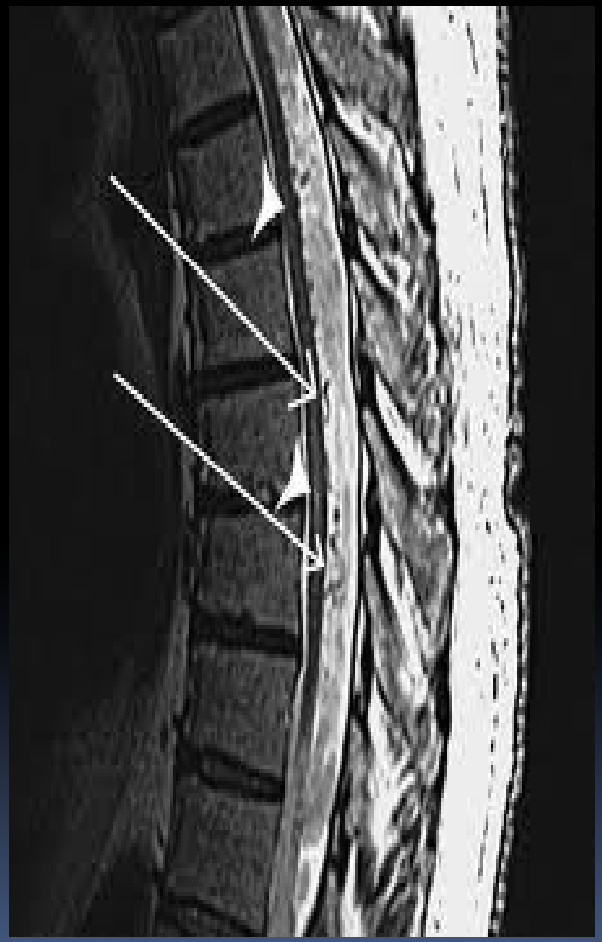


Fig A:demonstrating ventral intradural AVF ,a midline lesion derived from fistulous connection b/w ASA & coronal venous plexus

Fig B anterior view showing fistula along anterior aspect of cord

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Ventral intradural AVF: type:IV

Clinical presentation

- Younger age
- Progressive myelopathy due to compression , venous hypertension or hematoma
- SAH and acute neurological deterioration

Treatments –

- IV-A:surgery
- IV-B:embolisation
- IV-C:combination of endovascular ablation, followed by surgical excision of retained elements.

Perimedullary AVMs

- Occlusion of the feeding vessels right at the nidus as the first step, then cut off the draining perimedullary veins & perform total resection of the AVM

- During this procedure, try to preserve the pial vascular plexus of the spinal cord.

Intramedullary AVM

- Also known as type –2 AVMS, glomus AVM, classic AVM, angioma racemosum arteriovenous.
- True AVM of spinal cord closely resemble to intracranial AVMS
- 15-20 % of all spinal AVM
- Male = female
- Age -earlier , 2nd or 3rd decade of life
- Associated with other vascular anomalies (13 %– 37 %)
- Site –: uniforn distribution ,
  cervical (30%) , thoracolumabar (70%)
- Clinical presentation
  - acute (35 %) due to haemorrhage
  - progressive myelopathy (50 %)
- Two type – 1) compact, 2) diffuse

Intramedullary AVM : Type II

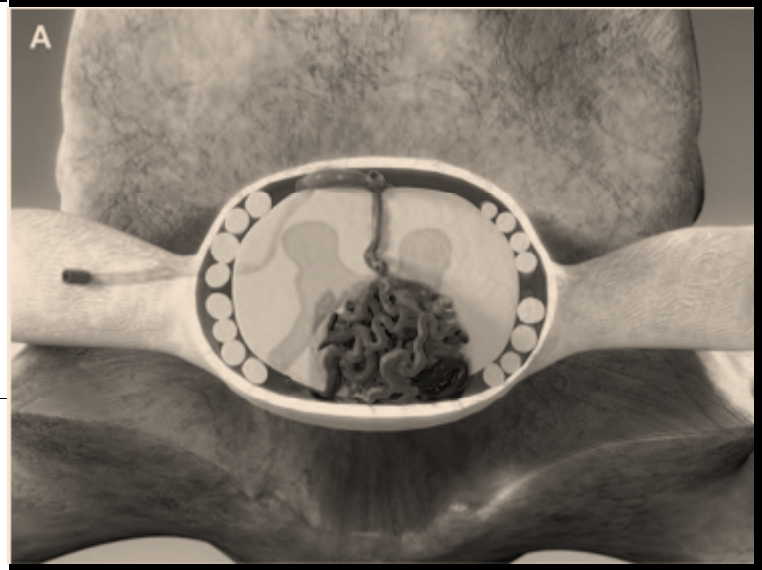
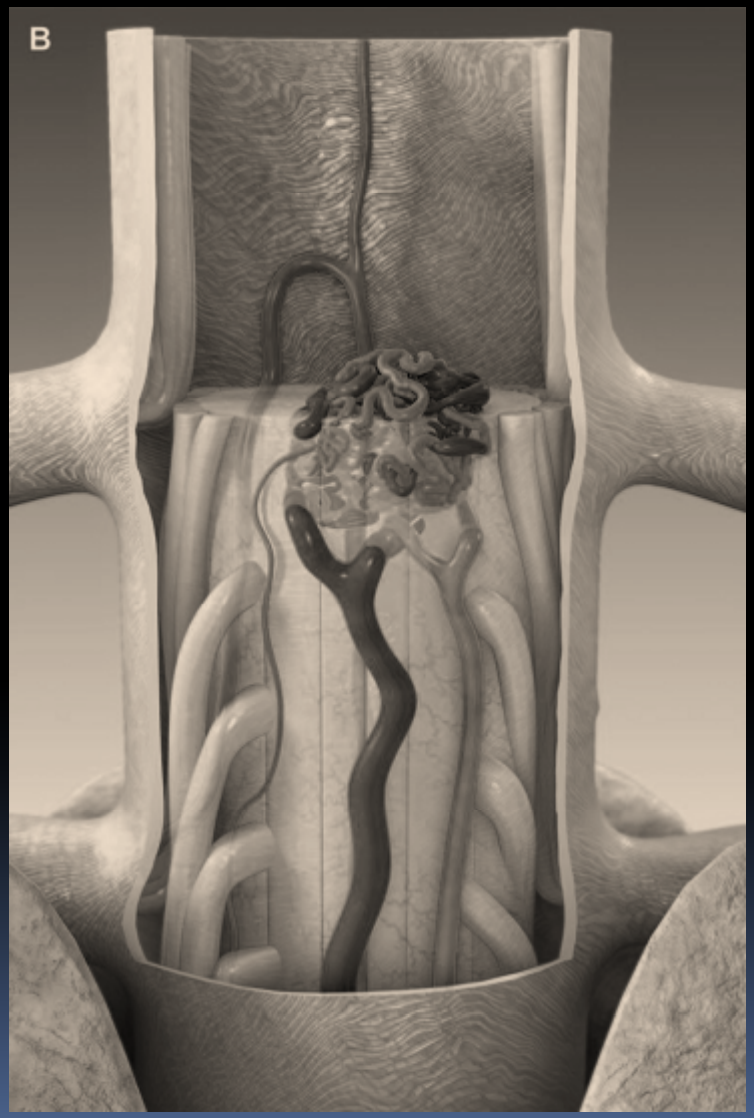
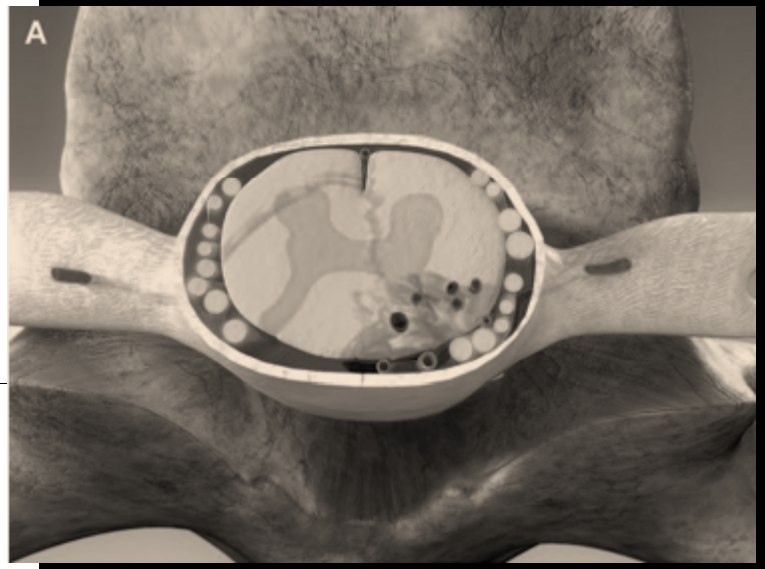


Fig :A- axial view showing compact intramedulary AVM with feeder from anterior spinal artery

Fig : B – posterior view showing additional feeding branches from posterior spinal artery



Intramedullary AVM:Type II



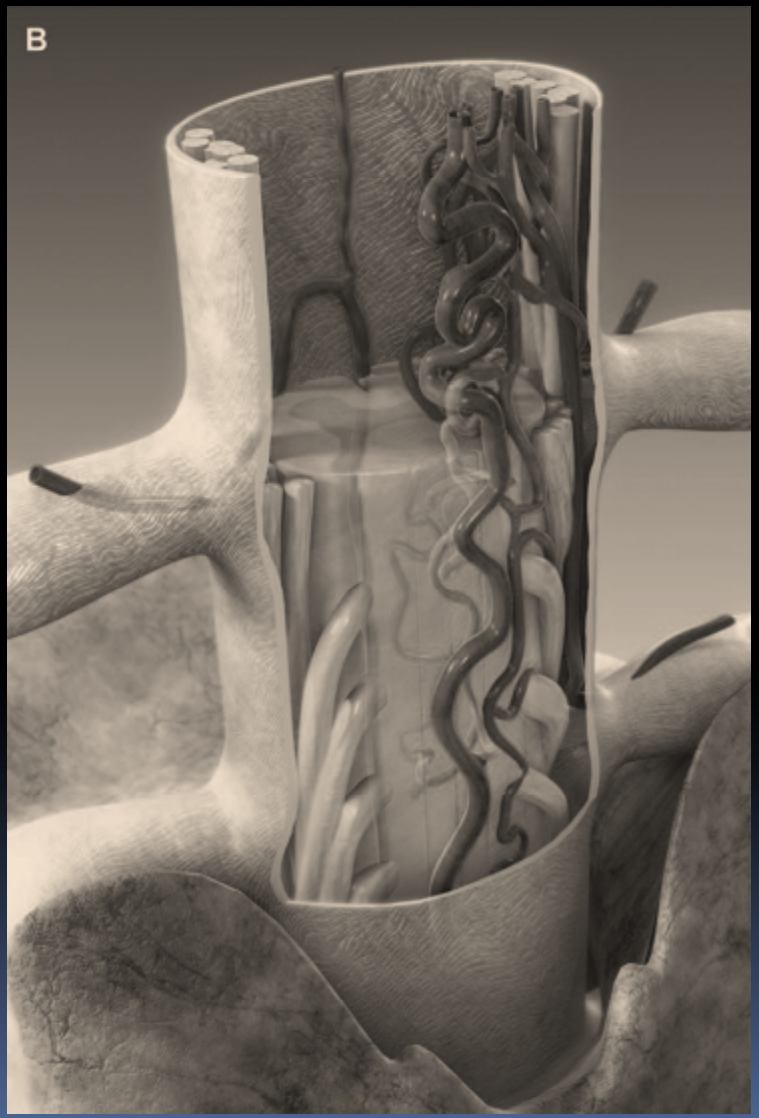


Fig :A-axial view demonstrating a diffuse intramedullary AVM with area of intervening neural tissue

Fig :B – oblique posterior view demonstrating the loop of AVM causing in and out of spinal cord

Intramedullary AVM Type- II

Diagnosis-

- MRI – Hematomyelia /cord expansion /SAH
- Spinal angiography –

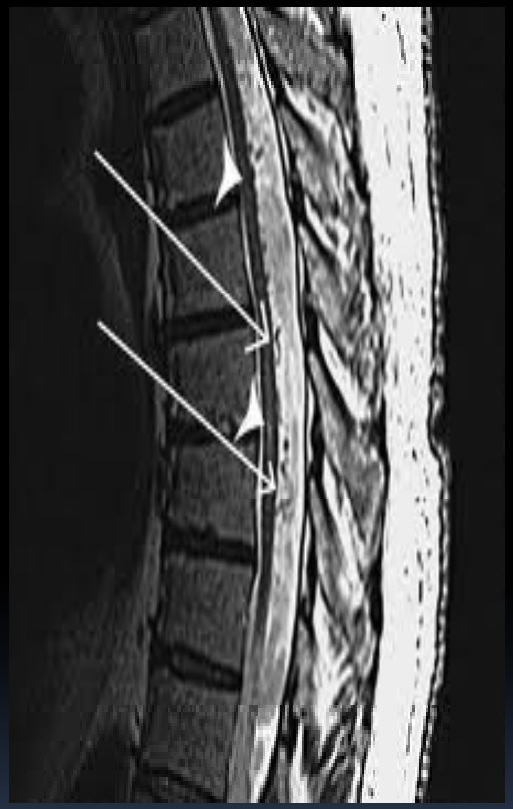
  - Multiple feeding vessels from ASA & PSA
  - High pressure , relatively low resistance and high blood flow

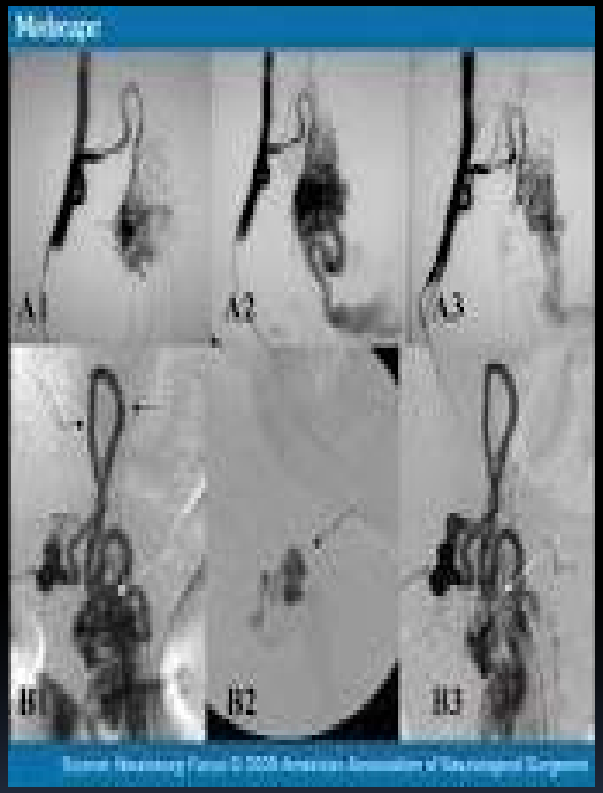
Managements

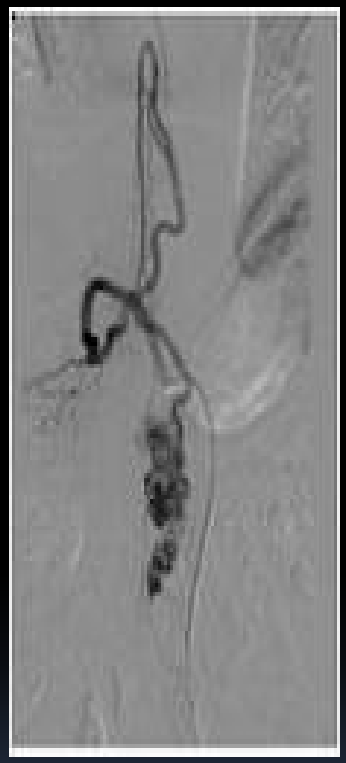
(A) Embolisation followed by surgery

(B) Radiosurgery for residual / non embolised lesion

Type -II







Intramedullary glomus AVMs

Two variants of nidus resection

1) Isolate the vessels near the nidus and coagulate, then dissect the nidus and resect
2) The vessels are cut off in the nidus itself during its separation, and resection of the nidus.

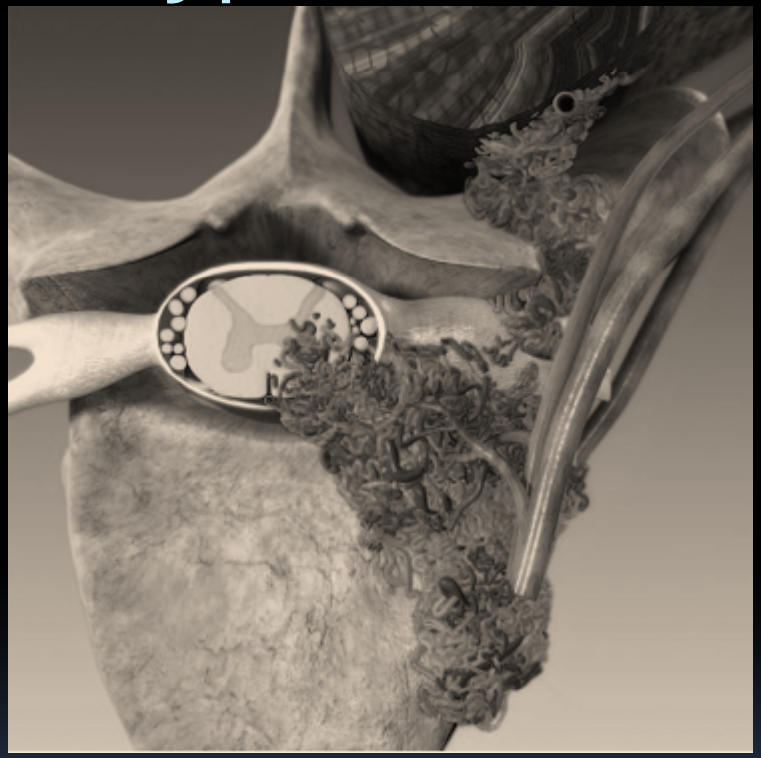
Extradural – Intradural: AVM Type-III

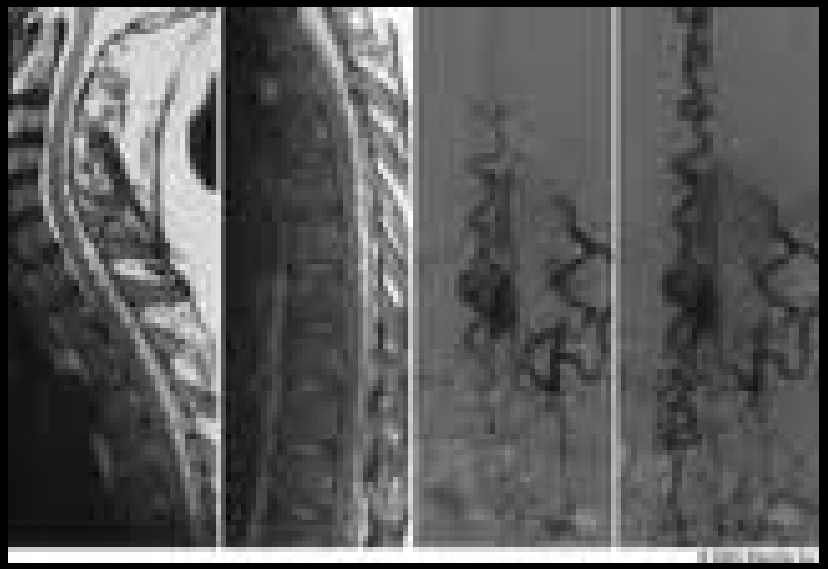
- Juvenile AVMs, Metameric AVMs
- Extremely rare lesion
- Complex lesion involving skin ,vertebra, and spinal cord
- Congenital lesion – inborn errors of vascular embryogenesis
- Anatomically lesion feed by multiple enlarge medullary artery via anterior and posterolateral spinal artery and voluminous arteriovenous nidus that completely fills the thecal sac .Nidus has neural tissue within its interstices.

Clinical presentation –

- Male = Female
- Onset – earlier age
- Site – uniform distribution along spinal axis
- Associated with other vascular anomlies
- Pain and progressive myelopathy related to compression and arterial steal

Extradural – Intradural: AVM Type-III





Treatment

- Primary extradural lo**c**alization -endovascular technology
- Mainly intradural location and spinal cord compression -combination of endovascular and microsurgical methods

Conus AVMs

- Conus AVM are location specific and have feature of both AVMs & AVFs . They can be both perimedullary and intramedullary with diffuse border
- C/F : progressive myelo- radiculopathy & occasional SAH
- Spinal angiography s/o multiple arterial feeder , more then one nidus , and complex venous drainage
- Treatments – combination of endovascular & surgical approach because of the possible pelvic disturbances, only performed occlusion of feeding vessels, leaving the malformation in situ.

Summary of treatments

Occlusion of the feeding and draining vessels and malformation resection

- Intramedullary glomus AVM
- Perimedullary AVM
- Epidural AVM
- Combined AVM

Indications for occluding only the feeding vessels

- Dural AVM
- Intramedullary diffuse AVM
- Combined AVM
- Conus medullaris AVM

Combining surgical intervention with endovascular embolization

- High flow AVM and numerous large feeding vessels running into it
- After the endovascular embolization a mass effect due to AVM blood flow remains.

Embolic agents

- Particulate materials
  - Poly vinyl alcohol(150-250micro)
  - Gelfoam
  - Sponge microparticulate
- Balloon occlusion
- Liquid agents
  - N-butyl cyanoacrylate
  - ethylene vinyl alcohol copolymer

Stereotactic radiosurgery

- Single high dose SRS
- 20 to 30% rate of occlusion.
- Hypofractionated irradiation
- Internal fiducial markers and image-guided radiation allow stereotactic irradiation for spinal disease with real-time verification and an accuracy of ±1 mm for every 0.03 seconds

Stereotactic Irradiation for Intramedullary AVMs Ten Years' Experience
Hokkaido University Collection of Scholarly and Academic Papers JAPAN

Thank you