SPINAL VASCULAR MALFORMATIONS

VAN HALBACH, M.D.
Neurovascular Medical Group
Interventional Neuroradiology
University of California San Francisco

Anson-Sptezler Classification

- Type I: Spinal Dural Fistula
- Type II: Intramedullary AVM
- Type III: Juvenile AVM
- Type IV: Perimedullary AV Fistula

KISS CLASSIFICATION SCHEME

- Keep

KISS CLASSIFICATION SCHEME

- Keep
- It
KISS CLASSIFICATION SCHEME
- Keep
- It
- Simple

KISS CLASSIFICATION SCHEME
- Keep
- It
- Simple
- Sir

KISS CLASSIFICATION SCHEME
- Keep
- It
- Simple
- Stupid!

SPINAL CORD MALFORMATIONS
- INTRAMEDULLARY AVMs
- PERIMEDULLARY FISTULAE
- SPINAL DURAL FISTULAE
- EPIDURAL AVMs
- METAMERIC MALFORMATIONS
- CAVERNOUS HEMANGIOMAS
SPINAL CORD MALFORMATIONS

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INTRAMEDULLARY AVMS ROLE OF ENDOVASCULAR TX

- CURATIVE USING PERMANENT AGENTS SUCH A LIQUID ADHESIVES OR ETOH
- PRESURGICAL ADJUNCT
- PALLIATIVE FOR PROGRESSIVE NEUROLOGICAL DECLINE, OCCLUDE HIGH RISK ANATOMY
- METAMERIC

28 Y.O. MALE WITH ACUTE QUADRAPARESIS
28 Y.O. MALE WITH ACUTE QUADRUPARESIS
Spinal Cord AVM

- 32 M
- Acute paraparesis, impaired sensation and position sense
- Dorsal thoracic spinal cord hemorrhage
- AVM at T-5, PLSA supply
- Preop embolization (microspheres)
Left T7 PLSA embo (microspheres)

JUVENILE TYPE INTRAMEDULLARY AVMS
SPINAL CORD MALFORMATIONS

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PERI-EXTRAMEDULLARY AVF

- FISTULA ON CORD SURFACE
- SPINAL OR RADICULAR ARTERIES
- NO NIDUS
- PRESENTS WITH PROGRESSIVE MYELOPATHY OR HEMORRHAGE
- GIANT (TYPE 3) ASSOCIATED WITH COBB’S SYNDROME (20%) OR RENDU OSLER WEBER (20%)

38 Y.O. FEMALE WITH FOUR MONTH HISTORY OF PARAPARESIS, PROGRESSIVE WEAKNESS, URINARY INCONTINENCE AND CONSTIPATION
9 y.o. with acute paraplegia while skiing
THE SIZE OF THE FISTULA IS NOT CORRELATED WITH SYMPTOMS

PERIMEDULLARY FISTULAE
ROLE OF EMBOLIZATION

• CURATIVE IN MOST TYPE 2 AND 3 (MEDIUM AND LARGE TYPE)
HALBACH ET AL
NEUROSURGERY
33(6)972-980 1993

PERIMEDULLARY FISTULA TYPE 3
TRANSVENOUS EMBOLIZATION

PERIMEDULLARY FISTULA TYPE 3
TRANSVENOUS EMBOLIZATION
65 Y.O. PRESENTED WITH BACK PAIN, LOSS OF BLADDER CONTROL, WEAKNESS IN LOWER EXTREMITIES, AND COGNITIVE DYSFUNCTION. A RECENT ONSET OF ACUTE BACK PAIN WARRANTED A SPINAL TAP + SAH. A MIDDLE CEREBRAL ARTERY ANEURYSM WAS CLIPPED

POSSIBLE CAUSES OF SPINAL SAH
- Posterior fossa aneurysm
- Vertebral artery dissection
- Intramedullary AVM
- Spinal artery aneurysm
- Perimedullary fistula
- Epidural AVM
- Trauma
- Tumor
SOLITARY PERIMEDULLARY FISTULAE
SPINAL CORD MALFORMATIONS

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...and there was a certain man there that had been eight and thirty years under his infirmity...

...Jesus saith to him, “Arise, take up thy bed and walk.” And immediately the man was made whole: and he took up his bed and walked.

John 5:5-9

SPINAL DURAL AVF’S

• NIDUS IN DURA
• DRAINAGE TO MEDULLARY VEINS
• SLOW FLOW
• PAIN, RADICULOPATHY, PARESIS
• NO HEMORRHAGE
• OLDER PATIENTS
• SYMPTOMS EXACERBATED BY EXERCISE AND STANDING

SPINAL DURAL FISTULAES
SLOW SHUNTS, DILATED VEINS

Coronal view of veins
Radicular vein
Radicular artery
Medullary vein
Radicular-medullary dual artery
PATHOPHYSIOLOGY
VENOUS HYPERTENSION

• INTRAOPERATIVE PRESSURE
  60-87.5% OF MEAN ARTERIAL PRESSURE
  HASSLER ET AL
  J NEUROSURG
  1989


SDAVF: UCSF Demographics

• 62 patients with SDAVF
• Mean age 62 years (26-83 years)
• 49 male : 13 female
• Range in time to diagnosis: 1 week to 6 years
• Frequent misdiagnoses: DDD, spinal cord tumor, Guillan-Barre syndrome, ALS, PVD, diabetic neuropathy
• SPINAL VASCULAR MALFORMATIONS
• 94 consecutive pt. with SDAVF
• 19 women, mean age 63
• All had weakness limiting exercise/activity at presentation range - mild to paraplegia
• Mean time from sx to onset to dx \ 23 months

Past Medical History
Contributors to delay in SDAVF diagnosis
Lumbar disc disease
Prostatic hypertrophy
Alcoholic neuropathy
Diabetic neuropathy
Peripheral vascular disease
Lumbar arachnoid cyst
Polio
Leg trauma
ALS*
Multiple sclerosis*
Spinal cord tumor*

SDAVF: Initial Symptoms

<table>
<thead>
<tr>
<th>Symptom</th>
<th>n</th>
<th>%</th>
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<tbody>
<tr>
<td>Lower extremity weakness</td>
<td>33</td>
<td>52</td>
</tr>
<tr>
<td>Lower extremity paresthesias</td>
<td>19</td>
<td>30</td>
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<tr>
<td>Back pain</td>
<td>15</td>
<td>24</td>
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<tr>
<td>Urinary dysfunction</td>
<td>4</td>
<td>6</td>
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</tbody>
</table>

SDAVF: Symptoms at Diagnosis

<table>
<thead>
<tr>
<th>Symptom</th>
<th>n</th>
<th>%</th>
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<tbody>
<tr>
<td>Lower extremity weakness</td>
<td>58</td>
<td>92</td>
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<tr>
<td>Lower extremity paresthesias</td>
<td>41</td>
<td>65</td>
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<tr>
<td>Back pain</td>
<td>19</td>
<td>30</td>
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<tr>
<td>Urinary dysfunction</td>
<td>52</td>
<td>83</td>
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<tr>
<td>Impotence</td>
<td>14</td>
<td>22</td>
</tr>
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SDAVF: Location

- Angiographic findings: fistulas from C1 to sacrum (internal iliac artery branches)
- T11 most common site (n = 6)
- 33 left-sided, 25 right-sided, 1 bilateral
- 3 patients had multiple SDAVFs

MOST SPINAL DAVFS ARE AT THE THORACO LUMBAR SPINE

COMPLETE ARTERIOGRAPHY IS ESSENTIAL
SDAVF: Lumbar Injections and Acute Symptomatic Presentation

- 3 patients with previously undiagnosed SDAVF acutely developed paraparesis following injection of medications into the lumbar epidural space
- Retrospective review of pre-injection lumbar MRIs demonstrated subtle signs of fistulas; post-injection showed more obvious signs

56 y M with BLE paresthesias

Before Steroid Injection After Steroid Injection
56 y M with BLE paresthesias

R T11 Intercostal Artery Injection

SDAVF: Treatment Strategy

- Surgical Therapy:
  - laminectomy
  - open dura
  - clip or coagulate site of AV shunt: vein turns blue
  - postoperative digital subtraction angiogram
- Endovascular Therapy:
  - favorable architecture for microcatheter access
  - no spinal artery at fistula site
  - permanent agent (liquid adhesive, glue, n-BCA)
  - n-BCA → AV fistula site + intradural vein + artery

SDAVF: Therapeutic Results

- Patients were evaluated for gait disturbance and urinary function using the Aminoff scale both before therapy and at follow-up (n = 47)

<table>
<thead>
<tr>
<th></th>
<th>Age (y)</th>
<th>Aminoff 1</th>
<th>FU (m)</th>
<th>Aminoff 2</th>
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</thead>
<tbody>
<tr>
<td>Endo n=16</td>
<td>66±1</td>
<td>3.4±1.7</td>
<td>39±33</td>
<td>2.5±1.6</td>
</tr>
<tr>
<td>Surg n=15</td>
<td>67±10</td>
<td>3.9±1.4</td>
<td>35±44</td>
<td>3.2±1.6</td>
</tr>
<tr>
<td>Combo n=16</td>
<td>60±10</td>
<td>3.4±1.2</td>
<td>49±59</td>
<td>3.5±1.4</td>
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</table>
SDAVF: Therapeutic Results

- Significant improvement in both motor function and micturition was noted in both endovascular and surgical groups (Student’s t-test)

<table>
<thead>
<tr>
<th></th>
<th>Motor</th>
<th>Urinary</th>
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<tr>
<td>Endovasal</td>
<td>0.004</td>
<td>0.006</td>
</tr>
<tr>
<td>Surgical</td>
<td>0.02</td>
<td>0.03</td>
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**Spinal Dural Fistulae Radiographic Presentation**

**MRI** (n=28)
- T2 hyperintensity: 24 (86%)
- Vascular Flow Voids: 21 (75%)
- Cord Enhancement: 15 (54%)
- Cord Enlargement: 6 (21%)
- Cord Compression: 1 (4%)
- Atrophy: 1 (4%)
- “Spinal cord tumor”: 2

**Myelography** (n=31)
- Vascular Filling Defects: 29 (94%)
- Cord Enlargement: 2 (6%)
- Cord Compression: 1 (3%)
DILATED VEINS ON MRI SEEN IN 50-75% OF MRI EXAMS
THE MOST SENSITIVE MRI FINDING IS INCREASED SIGNAL ON T2

MYELOGRAPHY SCREENING FOR SPINAL DAVF'S
- Good scouts
- Large contrast dose
- Small focal spot / fast films
- Narrow field overhead
- Supine Thoracic films
- Watch for layering
- CT myelo
- window appropriately

SURGICAL ENDOVASCULAR

SPINAL ARTERIOGRAPHY FOR SPINAL DAVF
- Every intercostal, lumbar injection
- Costo and thyrocervical trunks
- Bilateral vertebral artery injection
- Internal iliac injections
- Internal and external carotid injections
- Distal aortic injection for middle sacral artery
- Late filming

KEEP LOOKING UNTIL YOU FIND IT!
SDAVF: 48M
- 7 month h/o R leg numbness and R foot burning
- Retrospective vague h/o difficulty voiding
- Intermittent bilateral LE weakness
- Still ambulatory
SPINAL DURAL FISTULAE
ROLE OF EMBOLIZATION
• CURATIVE IN MOST CASES WITH PERMANENT AGENTS
• CONTRAINDICATED WHERE SPINAL ARTERIAL SUPPLY ARISES FROM FEEDING PEDICLE (MINORITY OF CASES)

SDAVF + spinal artery = no embo!
progressive paraparesis
SDAVF + spinal artery = no embol
69 M, progressive paraparesis

SDAVF: Therapeutic Results
• Endovascular and surgical patients had similar outcomes, with 2 significant differences:
  – Shorter hospitalization in endovascular patients 3.1±2.6 days versus surgical patients 9.8±2.7 days
  – Improved micturition in endovascular patients versus surgical patients (p=0.05) in the second decade of the study
• Patients receiving a combination of endovascular and surgical therapy did not have significant improvements in motor or urinary function

Spinal Dural Arteriovenous Fistula: Follow-up
• Immediate improvement!
• Immediate neurological decline → heparin
• Improvement → normal or plateau
• No improvement → reevaluate (MRI, A/G)
  – residual fistula
  – 2nd fistula (rare!)
• Improvement then decline → reevaluate (MRI, A/G)
• MRI at 3-6 months

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CONCLUSIONS

• Embolization can play a curative role in most spinal dural fistulae, larger and giant perimedullary fistulae, and selected intramedullary arteriovenous malformations.
• Surgery is ideal for small perimedullary fistulae.
Both embolization and surgery carry a high success rate, low morbidity, and both result in improvement in motor, gait and micturition. Endovascular treatment results in shorter hospitalization times than surgery.

Extraspinal Arteriovenous Fistula
- 43 M
- 2 month hx: progressive quadriplegia
- Abnormal MRI
Extraspinal Arteriovenous Fistula
- 43 M
- 2 month hx: progressive quadriplegia
- Abnormal MRI
- Tentorial/Incisural dural AVF with arterialized venous drainage to C-cord
- Endovascular cure: transarterial n-BCA
- Marked clinical, radiographic improvement after

Extraspinal AVMs/AVFs
- Location of A→V shunt:
  - Epidural
  - Paraspinal
  - Cranial
- Venous drainage affects spinal cord
- Rx: disconnect shunt to relieve venous congestion