Vascular malformation (VM) ABC

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Disclosure

• Research grant
  – Siemens Medical
  – Object Research System
  – Bracco Diagnostic
  – Biotronik
  – TVA medical

• Consultant
  – Cook Medical
Basic principles

• Use an appropriate terminology
  – Mulliken classification + new ISSVA classification

• Always correlate imaging findings with clinical history and examination
  – Vascular anomalies clinic

• Multidisciplinary management
  – IR (diagnosis, intervention)
  – Plastic surgery, dermatology, ENT, internal medicine
CLASSIFICATIONS

Hamburg classification 1993
(surgeons, pathologists)
Truncular and extratruncular lesions
VM/LM/AVM/combined

ISSVA Classification 1982/1996
(clinical)
tumors and malformations:
slow Flow and high Flow

New ISSVA classification
March 2014
## Hamburg Classification of vascular malformations

<table>
<thead>
<tr>
<th>Predominant Type</th>
<th>Lesion Form</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td><strong>Truncular</strong></td>
</tr>
<tr>
<td>Arterial</td>
<td>Aplasia or Obstruction</td>
</tr>
<tr>
<td></td>
<td>Dilatation</td>
</tr>
<tr>
<td>Venous</td>
<td>Aplasia or Obstruction</td>
</tr>
<tr>
<td></td>
<td>Dilatation</td>
</tr>
<tr>
<td>Lymphatic</td>
<td>Aplasia or Obstruction</td>
</tr>
<tr>
<td></td>
<td>Dilatation</td>
</tr>
<tr>
<td>Arteriovenous shunt</td>
<td>Deep</td>
</tr>
<tr>
<td></td>
<td>Superficial</td>
</tr>
<tr>
<td>Combined / Mixed</td>
<td>Arterial and venous</td>
</tr>
<tr>
<td></td>
<td>without shunt</td>
</tr>
<tr>
<td></td>
<td>Haemolympathic with or</td>
</tr>
<tr>
<td></td>
<td>without shunt</td>
</tr>
</tbody>
</table>

**Belov S. Semin Vasc Surg 1993; 6:219**
Extratruncal vs truncal

• Extratruncal
  – Defect occurring early in the embryogenesis
  – Mesenchymal cell memory (proliferation)
  – No or few connections with normal vascular system

• Truncal
  – Defect occurring later
  – Hypoplasia or dilatation of the vascular system

Lee-BB-JVD-2013
# 2014 ISSVA classification

<table>
<thead>
<tr>
<th>Vascular tumors</th>
<th>Vascular malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td>Simple</td>
</tr>
<tr>
<td>Locally aggressive or borderline</td>
<td>Combined °</td>
</tr>
<tr>
<td>Malignant</td>
<td>of major named vessels</td>
</tr>
<tr>
<td>Capillary malformations</td>
<td>CVM, CLM, LVM, CLVM CAVM* CLAVM* others</td>
</tr>
<tr>
<td>Lymphatic malformations</td>
<td>Details</td>
</tr>
<tr>
<td>Venous malformations</td>
<td>Details</td>
</tr>
<tr>
<td>Arteriovenous malformations*</td>
<td></td>
</tr>
<tr>
<td>Arteriovenous fistula*</td>
<td></td>
</tr>
</tbody>
</table>
## Anomalies of major named vessels

(aka "channel type" or "truncal" vascular malformations)

<table>
<thead>
<tr>
<th>Affect</th>
</tr>
</thead>
<tbody>
<tr>
<td>lymphatics</td>
</tr>
<tr>
<td>veins</td>
</tr>
<tr>
<td>arteries</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Anomalies of</th>
</tr>
</thead>
<tbody>
<tr>
<td>origin</td>
</tr>
<tr>
<td>course</td>
</tr>
<tr>
<td>number</td>
</tr>
<tr>
<td>length</td>
</tr>
<tr>
<td>diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm)</td>
</tr>
<tr>
<td>valves</td>
</tr>
<tr>
<td>communication (AVF)</td>
</tr>
<tr>
<td>persistence (of embryonal vessel)</td>
</tr>
<tr>
<td>Syndrome</td>
</tr>
<tr>
<td>-----------------------------------------------</td>
</tr>
<tr>
<td>Klippel-Trenaunay syndrome</td>
</tr>
<tr>
<td>Parkes Weber syndrome</td>
</tr>
<tr>
<td>Servelle-Martorell syndrome</td>
</tr>
<tr>
<td>Sturge-Weber syndrome</td>
</tr>
<tr>
<td>Limb CM + congenital non-progressive limb hypertrophy</td>
</tr>
<tr>
<td>Maffucci syndrome</td>
</tr>
<tr>
<td>Macrocephaly - CM (M-CM / MCAP)</td>
</tr>
<tr>
<td>Microcephaly - CM (MICCAP)</td>
</tr>
<tr>
<td>CLOVES syndrome</td>
</tr>
<tr>
<td>Proteus syndrome</td>
</tr>
<tr>
<td>Bannayan-Riley-Ruvalcaba syndrome</td>
</tr>
</tbody>
</table>
Vascular malformations

- Arteries
- Veins
- Lymphatics
- Capillaries

no cellular proliferation
no regression
Investigation

• Clinical examination combined with DUS
  – Coloration
  – Softness
  – Thrill, expansion with valsalva
  – Vessel dilatation, venous reflux
  – Limb overgrowth
  – CM (angioma)
  – Tissular destruction
Infantile hemangioma

- Infantile hemangioma
  - Growth 0-1 year
  - Stabilization 1-2 year
  - Regression 2-5 year
  - Glut 1 +
  - Conservative management
  - Propanolol, interferon, vincristin for complicated cases
Congenital hemangioma

- **RICH** (rapidly involuted congenital hemangioma)
  - Completely grown at birth
  - Regression 12-14 months
  - Glut –

- **NICH** (non-involuted congenital hemangioma)
  - Completely grown at birth
  - No involution
  - Growth during teenage
  - Glut –

- Hemangioendothelioma
- Tufted angioma
- Intramuscular hemangioma ?
Intramuscular hemangioma
= NICH?

- Intra muscular vascular tumor
- Hypervascular
- No AV shunting
- Surgery
Hemangioendothelioma

- F 46 ans
- Firm nodules since birth 1st 2\textsuperscript{nd}, 3rd finger and wrist slowly growing
Hemangioendothelioma
hemangioendothelioma

- Embolization
  - Particle
  - Onyx
- Surgery
- Vascular tumors in an adult patient & growth = biopsy
Capillary malformation

- Port wine stain
- No pulsation
- No mass
- Rule out AVM
- Associated syndrome
  - Struge Weber
  - KT
  - PKWS
Venous malformation

- Low flow
- Most frequent
  - Head and neck 40%
  - Body 20%
  - Limbs 40%
- Expansion
  - Valsalva
  - Dependent position
- Bluish coloration
Investigation

- Coagulation for large VM
  - 88% have a localized intravascular coagulopathy (LIC)\(^1\)
- Doppler ultrasound
- MRI if a treatment is considered or diagnostic unclear
- TDM not contributive
- Angiography not necessary +++

Venous malformation

- Doppler ultrasound
  - Low flow
  - Hypoechoic
  - Compressible venous dilatation
  - Phlebolitis
  - Evaluate feasibility of needle guidance for sclerotherapy
Compressibility & Doppler
VM & MRI

- Best examination for extension
- T2 (STIR), T1 and T1 fat sat post gado
Invasive treatment

• Sclerotherapy
  – Failure of conservative treatment
    • Pain
    • Aesthetic
    • Bleeding
    • Oropharyngeal compression

• Rarely surgery
  – Intramuscular
  – FAVA
Sclerotherapy

- Ethanol
- STS 3% (foam air + lipiodol)
- Fluoroscopic and ultrasound guidance
- Session 6-8 weekss
Foam-STS

Efficacy foam-STS > STS liquide

Post intervention

- NSAID/corticoid
- Pain medication
- LMWH if suspicion of foam migration in central venous & truncal lesions
VM follow-up

- Sclerotherapy every 6/8 weeks

**Clinical & DUS examinations 3-6 months after last sclerotherapy**

- Satisfactory
  - Stop ± consolidation MRI

- Partial improvement or failure with residual target lesions
  - Resume sclerotherapy

- Failure no residual target lesions
  - MRI Sclerotherapy or surgery
3D modeling

Caty V et al. Europ Radiol 2013
Lymphatic malformation

- Cystic cavity lined by an endothelial layer filled by a lymphatic fluid
  - ML macrocystic (> 2cm³)
  - ML microcystic (< 2cm³)
  - Mixed lesion (micro-macro)
  - Mixed lesion lymphatic and venous
- Present at birth
- Growth childhood-teenage
Lymphatic malformation

- Doppler ultrasound
  - Macrocyst with septation
  - Doppler
    - No flow in the cyst
    - High resistance flow in septa
  - Microcystic lesions infiltration of soft tissue by hyperechoic or heterogeneous mass
Sclerotherapy Bleomycin
Microcystic lymphangioma
Bleomycine

15 mg bleomycine in 15 ml nacl + 5ml contrast
Arterio-venous malformation

- High-flow malformation
  - AV-shunting
  - Nidus
- Congenital
  - Expansion
    - Teenage
    - Pregnancy
Schobinger classification

(Clinical staging system to grade the evolution of AVMs)

Stage 1: Quiescent
- Pink-bluish stain
- Warm
- Arteriovenous shunting (DUS)
Schobinger classification

Stage 2: Expansion

Stage 1

+ Darkening blush stain
+ Pulsations
+ Thrill
+ Bruit
+ Tortuous/tense veins
Schobinger classification

Stage 3: Destruction

Stage 2

- Steal
- Distal ischemia
- Dystrophic skin changes
- Ulceration
- Bleeding
- Persistent pain
- Tissues necrosis
- Soft tissues and bones changes
Schobinger classification

Stage 4: Decompensation

Stage 3 +

- High output cardiac failure
High flow vascular nidus

- Arterial side
  - High velocities and diastolic flow

- Venous side
  - Arterialization of venous flow

- Evaluate flow imbalance
  - Arterial feeder
  - Venous drainage
  - Normal arteries distal to AVM +++
Investigation

- MRI
  - Flow void
  - T1-T2
  - 4D angio
  - High resolution
  - steady state
CT-angiography

Arterial aneurysm and bone destruction
Embolization (ethanol)
Adapt your strategy to patient symptoms

- Venous congestion or haemorrhage
  - Aggressive on the arterial side before occluding the vein

- Tissue necrosis due to capillary shunting
  - Aggressive on the venous side

- Rely on Doppler ultrasound
Correlation between Doppler and DSA
Follow-Up
Klippel trenaunay

- Limb hypertrophy
- Cutaneous angioma
- Venous and or lymphatic
- R/O hypoplasia deep venous system
- Sclerosis of varicose vein
Parkes Weber

- AVM
- Cutaneous angioma
- Limb hypertrophy
AVM evolution: Boston Study: n=272


Progression MAV stade 1

Risk of Progression

- Stage IV (1.0%)
  - Stage III (21.6%)
  - Stage II (21.2%)

- Childhood (43.8%)
  - Childhood + Adolescence (82.6%)
  - Childhood + Adolescence + Adulthood (100.0%)

- Stage IV (1.4%)
  - Stage III (40.4%)
  - Stage II (40.8%)

- Stage III (47.9%)
  - Stage II (50.7%)
Recurrence embo versus surgery

(Boston study n=272)

• Mean FU: 8.9y ±5.2y
  – Recurrence all patients 93%

• Predictor of recurrence:
  – Embo vs surgery (combined or not with embo)
  – Shobinger stage at treatment

• Resection
  – Recurrence rate = 81%
  – Time to recurrence: 42.7% > 1y

• Embo alone
  – Recurrence rate = 98%
  – Time to recurrence: 14.4% > 1y

• Selection bias +++

Conclusion

• Make the diagnosis
• Clinical examination, Doppler, MRI
• Treat symptoms
• Multidisciplinary approach
• IR pivotal role
  – Must be involved in clinical evaluation and imaging work-up and follow-up