The ISSVA Classification System for Vascular Malformations

F Rohlffs, K Ivancev, W Yakes
University Hospital Hamburg, Germany
Yakes Vascular Malformation Center, Denver, Colorado, US

Disclosure
Speaker name:
Fiona Rohlffs

✓ I do not have any potential conflict of interest

Vascular Malformations

- Extraordinarily difficult lesions
- Seen throughout all populations 1%
- Protean manifestations
  - Heart failure
  - Bleeding
  - Ulceration
  - Disfigurement
- Every organ system and location
- Extreme confusion in the world literature

Multiple Classification Systems

ISSVA Classification is the only complete system to determine vascular malformation / vascular Tumor
International Society for the Study of Vascular Anomalies
Biennial international workshops, officially founded in 1992
Based on histologic diagnosis on the cellular basis
work started by John Mulliken in 1976

Benign vascular tumors 1

Infantile hemangioma / Hemangioma of infancy — see details
Congenital hemangioma — GNAQ / GNA11
Rapidly invovling (RICH) *
Non-invovling (NICM)
Partially invovling (PICH)
Tufted angiomata - *
Spindle-cell hemangioma — IDH1 / IDH2
Epithelioid hemangioma — P63
Pyogenic granuloma (also known as lobular capillary hemangioma) — BRAF / RAF / GNA14
Others — see details
### Simple vascular malformations I

**Capillary malformations (CM)**
- Nevus simplex / salmon patch, “angel kiss”, “stork bite”
- Cutaneous and/or mucosal CM (also known as “port-wine” stain)
- Non-syndromic CM
- CM with CNS and/or ocular anomalies (Sturge-Weber syndrome)
- CM with bone and/or soft tissues overgrowth
- Diffuse CM with overgrowth (DCAMO)
- Retinocutaneous CM
- CM of MIC-CAP (microcephaly-capillary malformation)
- CM of MCAP (megalencephaly-capillary malformation-polymicrogyria)
- CM of CM-AVM
- Cutis marmorata telangietectica congenita (CMT)
- Others
- Telangiectasia
- Hereditary hemorrhagic telangiectasia (HHT) (ENG, IT1, IT2, JH7, JH17, JH18)
- Others

**Malignant vascular tumors**
- Angiosarcoma
- Hemangioendothelioma
- Papillary hemangioendothelioma (PHEO), Dabeka tumor
- Composite hemangioendotheliomas
- Pseudomyogenic hemangioendothelioma
- Polymorphous hemangioendothelioma
- Hemangioendothelioma not otherwise specified
- Kaposi sarcoma
- Others

### Simple vascular malformations II

**Lymphatic malformations (LM)**
- Common (cystic) LM
- Microcystic LM
- Macrocytic LM
- Mixed cystic LM
- Generalized lymphatic anomaly (GLA)
- Kasabach-Merritt syndrome (KMS)
- LM in panniculitis disease
- Channel type LM
- “Acquired” progressive lymphatic anomaly (also called acquired progressive “lymphangiomatosis”)
- Primary lymphedema
- Others

### Simple vascular malformations III

**Venous malformations (VM)**
- Common VM
- Familial VM cutaneous-mucosal (VMCM)
- Blue rubber bleb nevus (Beanz) syndrome VM
- Glomuvenous malformation (GVM)
- Cerebral cavernous malformation (CCM)
- Familial intracranial vascular malformation (FICM)
- Venous venous malformation (formerly venous hemangioma)
- Others

### Simple vascular malformations IV

**Arteriovenous malformations (AVM)**
- Sporadic
- In HHT
- In CM-VM
- Others

**Arteriovenous fistula (AVF) (congenital)**
- Sporadic
- In HHT
- In CM-VM
- Others

### Combined vascular malformations*

- CM + VM: capillary-venous malformation
- CM + LM: capillary-lymphatic malformation
- CM + AV: capillary-arteriovenous malformation
- LM + VM: lymphatic-venous malformation
- CM + LM + VM: capillary-lymphatic-venous malformation
- CM + LM + AV: capillary-arteriovenous malformation
- CM + LM + VM + AV: capillary-lymphatic-venous-arteriovenous malformation
ISSVA Classification is the only complete system to determine vascular malformation / vascular Tumor

Based on the histologic diagnosis on the cellular level

Helps to achieve uniform reporting standards