Vascular malformations

DR RAVUL JINDAL MS FRCS
DIRECTOR VASCULAR SURGERY
FORTIS HOSPITAL
MOHALI
Basic principles

- Rare
- Difficult to understand and treat
- Very distressing for patient and physicians
- Multidisciplinary approach
Vascular Anomalies

- **Vascular tumors**
  - Hemangiomas - birthmarks (naevi), words for food ("strawberry" and "cherry") and drink ("port-wine")
  - Hemangioendothelioma
  - Angiosarcoma

- **Vascular malformations**
  - Arterial (AVM, AVF, coarctation, ectasia, aneurysm)
  - Capillary
  - **Venous**
  - Lymphatic
  - Combined

*International Society of Vascular Anomalies in 1996*
Vascular tumors

- Arise by endothelial hyperplasia
- Appears in the early neonatal period
- Distinctive growth cycle
  - Proliferation phase - early rapid growth
  - Involutional phase - slow regression
Vascular malformations (VM)

- Normal endothelial turnover
- Malformed vessel that results from developmental arrest during embryogenesis
- Presents at birth as an inborn vascular defect
- Continues to grow with body growth
# Hamburg classification of VM

<table>
<thead>
<tr>
<th>Type</th>
<th>Truncular</th>
<th>Extratruncular</th>
<th>Extratruncular – cavernous hemangiomas</th>
<th>Diffuse/Limited/localized</th>
<th>Deep/ Superficial</th>
<th>Diffuse/ Limited</th>
<th>Venous and arterial</th>
<th>Hemolymphatic</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Arterial-1%</strong></td>
<td></td>
<td>Truncular</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Venous-48%</strong></td>
<td></td>
<td>Extratruncular</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Arteriovenous-36%</strong></td>
<td>Truncular</td>
<td>Extratruncular</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Combined-15%</strong></td>
<td>Truncular</td>
<td>Extratruncular</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Venous malformation- Embryology

Arrested development in the syncytial stage/ undifferentiated stage of vein formation
Clinical features

- Present at birth; not always clinically evident

- VM grows proportionately to the child, expands slowly, and often enlarges during puberty

- Skin and subcutaneous tissues, muscle, abdominal viscera, and CNS

- Pain and stiffness in early morning
Bluish, soft, compressible and enlarges on dependency and Valsalva maneuver
Diffuse involvement
Venous malformation
Venous malformation
Head and Neck

- Unilateral
- Facial asymmetry
- Intraorbital
- Oral VM
- Pharyngeal, laryngeal, and deep cervical-oropharyngeal - OSA
Chest VM
Leg involvement

- Skin or can extend into muscles, joints, and bone
- Intraosseous VM - pathologic fracture
- Synovial lining of the knee
Limb involvement
KTS- Klippel-Trenaunay syndrome

- Sporadic
- obvious at birth
- Capillary-lymphatic-venous malformation (CLVM)
Investigations

- Coagulative profile
  - Extensive VM
  - Stagnation causes a localized intravascular coagulopathy

- Phleboliths - 2 years of age
Imaging

- Duplex
- MRI

- Misguided "AGA" approach ("always get an angiogram") still prevails
Treatment

• Indication
  • Appearance, pain, or functional problems

• Conservative
  • Elastic compression stockings
  • Low-dose Aspirin (75 mg every day)
    • Painful phlebothromboses

• Resection

• Sclerotherapy

• LASER
Neck cavernoma
Excision
Sclerotherapy

- **Small mucous or oromucosal VM**
  - 1% sodium tetradecyl sulphate

- **Large cutaneous or intramuscular VMs**
  - Absolute ethanol (100%)
  - Foam - STS/polidoconol
  - Local complications
  - Systemic complications - hemolysis and renal toxicity and cardiac arrest
24 years with right hip venous malformation
Contrast injection

MRI 2 years later
Left hand venous malformation
KTS post laser and foam
Result

- 75% to 85% of patients report good results, in terms of cosmetic improvement or relief of pain.
- Cure is rare because the defect affects all of the veins of the affected anatomic area.
- Recanalization can occur after sclerotherapy.
Right cheek lymphatic malformation
Right thigh AVM
Scalp AVM
CT
Post procedure
Post procedure
Conclusion

- Mainly a clinical diagnosis

- Can treat symptomatic patients with sclerotherapy /excision /LASER and embolisation with fairly good results
Questions ?