VASCULAR ANOMALIES OF THE FACE

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Vascular Anomalies are the abnormal formation or development of blood vessels affecting capillaries, arteries, veins and lymphatic channels.

Vascular anomalies are localized defects of vascular development.
Etiology
Vascular anomalies are histopathologically characterized by a focal increase in the number of vessels that are abnormally tortuous and enlarged\(^1\).

Classification
## ISSVA classification for vascular anomalies

(Associated with other anomalies)

<table>
<thead>
<tr>
<th>Vascular Tumors</th>
<th>Simple</th>
<th>Combined</th>
<th>Vascular Malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td>Capillary m. (CM)</td>
<td>CM+VM</td>
<td>capillary-venous m.</td>
</tr>
<tr>
<td></td>
<td>Lymphatic m. (LM)</td>
<td>CM+LM</td>
<td>capillary-lymphatic m.</td>
</tr>
<tr>
<td></td>
<td>Venous m. (VM)</td>
<td>CM+AVM</td>
<td>capillary-arteriovenous m.</td>
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<tr>
<td></td>
<td>Arteriovenous m. (AVM)</td>
<td>LM+VM</td>
<td>lymphatic-venous m.</td>
</tr>
<tr>
<td></td>
<td>Arteriovenous Fistula (AVF)</td>
<td>CM+LM+VM</td>
<td>capillary-lymphatic-venous m.</td>
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</tbody>
</table>

### Anomalies of major named vessels
- Affect
- lymphatics veines arteries
- origin
- course
- number
- length
- diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm)
- valves
- communication (AVF)
- persistence (of embryonal vessel)

### Associated with other anomalies
- Klippel-Trenaunay syndrome
- Parkes Weber syndrome
- Servelle-Martorell syndrome
- Sturge-Weber syndrome
- Maftucci syndrome
- Macrocephaly
- Microcephaly
- CLOVES syndrome
- Proteus syndrome
- Bannayan-Riley-Ruvalcaba syndrome
## ISSVA binary classification for vascular anomalies

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<thead>
<tr>
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<th>Vascular Malformations</th>
</tr>
</thead>
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<tr>
<td>Hemangioma</td>
<td>Slow Flow</td>
</tr>
<tr>
<td>Hemangioendothelioma</td>
<td>Capillary Lymphatic Venous</td>
</tr>
<tr>
<td>Angiosarcoma</td>
<td>Fast Flow</td>
</tr>
<tr>
<td></td>
<td>Arterial</td>
</tr>
<tr>
<td></td>
<td>Combined</td>
</tr>
</tbody>
</table>

FIGURE 11.1 Diagrams illustrating the channel morphology of the most common forms of vascular anomalies. (Reprinted, with permission, Burrows and Fellows, 1995) A. Infantile hemangioma consists of a solid cellular mass with organized, acinar pattern of arterial supply and drainage into dilated regional veins. B. Arteriovenous fistula is focal macroscopic connection between artery and vein. C. Arteriovenous malformation typically consists of a nidus or network of abnormal vascular channels with feeding arteries and draining veins. D. Venous malformation is a post-capillary lesion composed of abnormally shaped, dilated venous channels. Major conducting veins can be involved. E. Lymphatic malformation composed of fluid-filled spaces or channels lined with lymphatic endothelium.

Clinical Manifestation
Vascular Tumors
Infantile Hemangioma

Benign vascular neoplasms

Have a characteristic clinical course marked by

early proliferation and

followed by spontaneous involution.

**PROLIFERATION PHASE**

*Neonate*

Proliferation of primitive cells

**INVOLUTING PHASE**

1-5 years

Due to Apoptosis

Mast cells interact with macrophages and fibroblasts = Transgranulation

**INVOLUTED PHASE**

>7 years

Revascularization

Deposition of fat cells

**Third Trimester**
Cutaneous Hemangioma
small/large/extensive
Ulcerative/non-ulcerative

Deep Hemangioma

Involuting Hemangioma

Involuted Hemangioma

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Clinical Manifestation
Vascular Malformations
Capillary Malformations

- Caused by a defect of autonomic nervous system supplying capillaries
- The number of blood vessels are normal, but the diameter of the affected vessels is much larger.
- This enlargement results in increased blood flow.
- Since the vessels are close to the surface, this increased flow gives the skin its pink to purple appearance.
- The affected blood vessels will continue to enlarge and thicken with age, causing the color of the lesion to darken.
Venous Malformations

- Made up of **malformed veins**
- Vary in color from **blue to dark purple**, depending on how deep the malformation extends.
- Tend to swell with activity/exercise
- The mass is usually **soft and compressible** and then **refills when released**.
- There may be small hard masses palpable in the lesion, called **phleboliths**, which are small collections of calcium that have resulted from slow blood flow and blood clots.
Lymphatic Malformations

- Exact cause is unknown. Errors in the formation and development of lymphatics during fetal development.
- Made up of abnormal, dilated lymph channels that can be focal or diffuse.
- Increase in size with infection such as upper respiratory infections.
- Difficult to treat if they are diffuse (affecting more than one small area).
- Three types. Micro cystic, Macro cystic and Mixed
Arterio-Venous Malformations (AVM)

- Involve an abnormal connection between arteries and veins
- Consist of a blood vessel "nidus" (nest) through which arteries connect directly to veins, instead of through capillaries.
- Symptoms include throbbing pain and growth/thickness of the area involved.
- Palpation over lesion will reveal a pulsation or thrill.
- If bleeding occurs it can be quite brisk and may require medical attention.

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Combined Malformations
Sturge Weber Syndrome
Sturge-Weber syndrome consists of

Tortuous slow-flow vessels
    involving the conjunctiva, episclera, retina or choroid.

**Glaucoma** is the most common and serious ophthalmological complication; the prevalence is 60% (Sujansky and Conradi, 1995a).

Sudden corneal clouding is the pathognomonic sign of acute glaucoma; this is an emergency.
Diagnosis
Ultrasound...

Ultrasoundography of AV malformation of upper lip
Note the arterial flow, venous flow and nidus of capillaries
CT Scan

CT scan helps locate the position and extent of the lesion. They also help in identifying bony structures adjacent to the lesion.

Coronal CT Scan of A-V malformation of the cheek.
Arteriogram

Arteriogram is a CT scan with contrast that offers a clear view of the vessels in the vascular malformation.

Arteriogram of Lymphangioma of Cheek

Note the absence of feeder vessels.

Note the size of the cystic lesion.

If it is more than 2 cm it is a macrocystic lesion otherwise it is microcystic.

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Arteriogram of Venous malformation of cheek
Note the presence of feeder vessels and phleboliths
Arteriogram of Arterio-Venous malformation of cheek
Note the difference between right and left External Carotid Artery
MRI
The MRI helps the surgeon visualize the extent of the lesion in relation to surrounding soft tissue structures.

MRI Scan of AV malformation of upper lip
Note the clarity of the extent of the lesion.
Treatment
Treatment

Treatment for Vascular Malformations and Hemangiomas is usually as follows:

For Low Flow Superficial Lesions
Sclerotherapy followed by Conventional surgery

For High Flow Lesions
Subtraction angiography with embolization with gel foam or stents followed by Conventional surgery within 72 hours

In India Angiography is beyond the capacity of most patients. Therefore angiography is considered only if any great vessel is involved.
Cutaneous Hemangiomas

**Sclerotherapy**

- **Syrup or Tablet Propranolol:** 0.5 -1 mg/kg of body weight in two divided doses for 6 months under strict pediatric supervision
  
  *(Propranolol, β-blocker, vasoconstrictor, regulating angiogenic pathways inducing apoptosis of vascularized endothelial cells)*

- **Injection Triamcinolone (Kenocort):** One 20 mg /ml vial diluted in 2 ml saline and 1ml lignocaine injected intralesionally, once a month for six months.
  
  *(Triamcinolone, corticosteroid suppresses vasculogenic capability of multipotent stem cells)*

- **Contractubex (10% aqueous onion extract, 50 U heparin per gram of gel,1% allantoin) gel and olive oil:** massage on the lesion twice daily till the regression of the lesion.
All Vascular Malformations and Hemangiomas

**Bleomycin Treatment**

- **Pingyangmycin (Bleomycin A5):** 2-6 ml (0.5 - 4 mg/ml concentration) given intralesionally and repeated every 4 weeks for a maximum of 12 sessions. **OR**

- **Bleomycin:** 0.5 – 1.0mg/kg body weight up to a maximum of 6mg (0.5-105 mg/ml concentration) given intralesionally and repeated every 4 weeks for a maximum of 12 sessions.

- **Bleomycin** acts by producing a sclerosing effect due to its direct action on the endothelial cells of the lesion producing non-specific inflammatory reaction

- Can be given in Capillary, Venous, Arterio-venous and Lymphatic malformation and Hemangiomas.
Surgical Protocol

• **Key is Accessibility**
  
  Accessible = Surgery
  
  Inaccessible = Embolisation and surgery

• **Ligation** of all possible blood vessels in the vicinity of the lesion

• **Aim of surgery**
  
  • HARMONIC SCALPEL is used to radically excise all affected tissue as remnants of necrotic tissue can form a focus of a granuloma or further infection.
  
  • Reconstruct what ever possible
  
  • Post operative maintenance with steroidal injections intra-lesionally
• Cutting instrument that can cut and coagulate tissue simultaneously
• Can cut through thicker tissue and create less toxic surgical smoke than a Bovie
• Offers greater precision than a Bovie
• Cuts via vibration. Bovie cuts via an electrical current (and production of heat)

Therefore Harmonic Scalpel causes less lateral thermal damage
Treatment...

Ligating sutures

Capillary Hemangioma

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Treatment...

Hypertrophied Capillary Malformation
Treatment with full thickness skin graft harvested from right groin
Treatment...

Low Flow Venous Malformation

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Treatment...

High Flow Venous Malformation

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Treatment...

Lymphatic Malformation

Surgery is only performed as a cosmetic adjuvant to other therapies. Macrocystic lymphatic malformations are treated with drainage and ethanol injections as a sclerosing agent. Microcystic lymphatic malformations are treated with doxycycline injections as sclerosing agent.
High Flow A-V Malformation
Treatment...

High Flow A-V Malformation

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Treatment...

Sturge Weber Syndrome

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Treatment...

Sturge Weber Syndrome

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Complications

Hemangioma

• Very problematic, interfering with eating, breathing, seeing, hearing, and speaking.

Vascular malformations: capillary, venous and arterio-venous

• Patients with port-wine stains should be evaluated and monitored for a larger syndromic entity.

• Malformations that are part of the Klippel-Trenaunay-Weber syndrome can be located on the lungs, spleen, liver, bladder, or colon. Visceral involvement can often lead to substantial morbidity in the form of internal hemorrhage.
Complications…

Vascular malformations - Lymphatic malformations

• Diffuse cervicofacial disease can result in mandibulomaxillary hypertrophy because of direct invasion of the bone and growth of the malformation within the bone.

• Lymphangiomas often swell with the onset of general viral infection or remote bacterial infection. This typically resolves with the resolution of the infection.

• Lymphangiomas can become infected
Do not confuse a Vascular malformation with...
Slowly enlarging (~5mm per year), non-tender neck masses located just anterior to the sternocleidomastoid muscle at the level of the hyoid.

The mass may transmit the carotid pulse or demonstrate a bruit or thrill, which might confuse the clinician to think it is a vascular malformation.

As these tumors enlarge, progressive symptoms of dysphagia, odynophagia, hoarseness and other cranial nerve (IX-XII) deficits appear.

Carotid angiography is by far the most useful diagnostic test for paragangliomas.
Treatment…

Carotid Body Tumor
Vascular Malformations of the head and neck region are something that are treatable in most situations.

Care must be taken, however, to do a work up for the patient.

Most lesions that you will find in your practice will be low flow lesions.

To diagnose the flow of vascular malformation lesions requires nothing more than an ultra sound.

The key to treatment is Accessibility.