Congenital Vascular Malformations

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Introduction

- History and Classification
- Diagnosis
- Lymphatic Malformations
- Venous Malformations
- Capillary Malformations
- Arteriovenous Malformations
Among the most common congenital and neonatal abnormalities.

In the past sometimes confusing classifications were developed.

Understanding of the various biological characteristics of vascular tumors has been impeded.

Misconception that most of these lesions spontaneously disappear within the first years of life.

As a consequence congenital vascular malformations were often misdiagnosed and left untreated.
History

Virchow and his student Wegener, in 1880, separated all vascular tumors into angiomas and lymphangiomas characterized as "simplex", "cavernosum", and "racemosum".
Mulliken and Glowacki, in 1982, developed a biological classification encompassing physical findings, clinical behavior and cellular kinetics.

They distinguished hemangiomas from vascular malformations with two main characteristics distinguishing each.
Hemangiomas

- Usually are not present at birth, but become apparent within the first weeks of life.
- Show a rapid proliferation during the first two years of life, followed by a slow involution.
Hemangiomas

- True tumors, with proliferation of the vascular endothelium
Vascular Malformations

- Present at birth, but may not be clinically evident.
- Show proportionate growth in relation to the body volume and show no signs of spontaneous involution.
Vascular Malformations

- Normal endothelium
- Defect in vascular smooth muscle
- Progressive dilation of vascular channels
Classification

- Burrows, in conjunction with Mulliken, in 1983, further described malformations as either:
  - High flow - having a connection to the arterial or capillary system
  - Low flow - having a connection to the venous or lymphatic system.
Classification

- High-flow vascular anomalies, such as arteriovenous fistulas and arteriovenous malformations, are traditionally addressed by means of transarterial embolization.
- Low-flow malformations found to be solitary or combined in capillary, venous, or lymphatic vessels are successfully treated with sclerotherapy.
Classification

- 1988, 7th meeting of the International workshop on Vascular Malformations, Hamburg
- Classification differentiates between truncular and extratruncular malformations
<table>
<thead>
<tr>
<th>Type</th>
<th>Truncular forms</th>
<th>Extratruncular forms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Predominately arterial defects</td>
<td>Aplasia or obstruction Dilatation</td>
<td>Infiltrating or limited</td>
</tr>
<tr>
<td>Predominately venous defects</td>
<td>Aplasia or obstruction Dilatation</td>
<td>Infiltrating or limited</td>
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<tr>
<td>Predominately AV shunting defects</td>
<td>Deep AV fistula Superficial AV fistula</td>
<td>Infiltrating or limited</td>
</tr>
<tr>
<td>Predominately vascular defects</td>
<td>Arterial and venous Hemolymphatic</td>
<td>Infiltrating hemolymphatic Limited hemolymphatic</td>
</tr>
<tr>
<td>Predominately lymphatic defects</td>
<td>Aplasia or obstruction Dilatation</td>
<td>Infiltrating or limited</td>
</tr>
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Diagnosis

Noninvasive studies

- MRI
- Duplex ultrasound
- Whole body blood pool scintigraphy
- Transarterial lung perfusion scintigraphy
- Lymphoscintigraphy
- CT
Diagnosis

➢ Invasive studies
  – Selective and Superselective angiography
  – Direct puncture (percutaneous) arteriography
  – Standard phlebography
  – Direct puncture (percutaneous) phlebography
  – Direct puncture (percutaneous) lymphangiography
Diagnosis

- 3 non-invasive tests are sufficiently accurate and obviate the need for invasive studies
- Invasive procedures are usually reserved for treatment planning
MRI

- Imaging modality most commonly used
- Should include T1- and T2-weighted spin-echo imaging in multiple planes, fat-saturated T1-weighted imaging with the intravenous administration of a gadolinium-based contrast agent, and gradient-recalled echo (GRE) imaging.
- T2-weighted images are used mainly to evaluate the extent of the abnormality.
- GRE images are used to identify the hemodynamic nature of the condition (high- vs low-flow lesions); and contrast-enhanced images are used to determine the extent of the malformation and to distinguish low-flow vascular anomalies (venous malformation versus lymphatic malformation).
For any vascular anomaly, the basic approach is first, to evaluate fat-suppressed T2-weighted images to determine the extent of the anomaly, and second, to evaluate the GRE images to decide whether the anomaly is a high-flow lesion.

If the anomaly is a low-flow lesion, arteriovenous malformation, arteriovenous fistula, and hemangioma can be excluded from the differential diagnosis.
MRI

- Low-flow vascular anomalies (venous malformation, lymphatic malformation, capillary-lymphatic-venous malformation) can be further differentiated on the basis of their morphologic appearances and contrast-enhancement patterns.
If the anomaly has no contrast enhancement or a minimal degree of peripheral contrast enhancement (rings and arcs), lymphatic malformation should be considered foremost in the differential diagnosis.
If the anomaly has easily noticeable patchy areas of contrast enhancement, venous malformation should be suspected.
If the lesion is a high-flow anomaly, in hemangiomas, fast-flow vessels are usually at the periphery of the mass, and the mass usually enhances homogeneously.
A mass lesion is not expected in an arteriovenous malformation.

If there are any remaining questions, the high-flow nature of an arteriovenous malformation can be easily confirmed with Doppler examination, which reveals high-flow, low-resistance arteries and an arterialized waveform in the draining veins.
Treatment Indications

- Hemorrhage
- Risk of high-output heart failure
- Chronic venous hypertension
- Airway impingement
- Lesion threatens vital functions
Treatment Indications

- Disabling pain
- Functional disability
- Cosmetic deformity
- Recurrent infection
- Persistent lymph leakage
Low flow malformations

- Lymphatic Malformations
  - Microcystic
  - Macrocystic
- Venous Malformations
- Capillary Malformations
- Combined types
Lymphatic Malformations

- Microcystic (lymphangioma)
- Clusters of vesicles on the buccal mucosa, tongue, or conjunctiva.
- The vesicles can be clear, red or black as a result of microscopic bleeding.
Lymphatic Malformations

- Macrocystic (cystic hygroma)
- Almost a 50% association with chromosomal disorders such as Turner syndrome, trisomy 21, trisomy 18, and Noonan syndrome
- Often located below the level of the mylohyoid muscle
- Present as cervical cystic swelling, often with the overlying skin having a bluish hue
Lymphatic Malformations

- Since most lymphatic malformations are mixed-form malformations (macro- and microcystic), the most common therapeutic approach is sclerotherapy for the macrocystic portion of the malformation, then surgical excision of the remaining microcystic portion if needed.
- Aspiration or drainage results only in temporary shrinkage.
- Macrocytic lesions, if excised are ideally removed in one procedure, because repeated excisions are complicated by fibrosis and anatomic distortion.
- Microcystic lesions are often difficult to resect, because there are no distinct tissue planes between the malformed and normal structures.
Lymphatic Malformations

- Lee, 2005, reviewed 315 patients treated for LM.
- All head and neck LM were of the extratruncular form.
- Sclerotherapy with OK-432 showed 90% success rate with macrocystic LM, but 50% with microcystic type.
Lymphatic Malformations

- Jian, 2005, published a retrospective study to evaluate the results of Jian or Dingman glossectomy for lymphangiomatous macroglossia
Lymphatic Malformations

- Cosmesis and function improved after surgery in 7/7 patients. The tongue healed well, and the patients had no long-term complications.

- The authors conclude that although partial surgical excision, injection of sclerosing solutions, electrocoagulation, and radiation have been the chief modalities of treatment of diffuse lymphangioma of the tongue, surgical management is the most effective treatment.
FIGURE 2. Case 4: A 9-year-old boy with lymphangiomatous macroglossia managed by Jian glossectomy preoperative and postoperative frontal and lateral views.

A, Frontal view preoperatively.

B, Lateral view preoperatively.

C, Frontal view postoperatively.

D, Lateral view postoperatively.
Venous Malformations

- 2/3 of all vascular malformations.
- Are low-flow lesions.
- Present in a spectrum, ranging from an isolated skin varicosity or localized spongy mass to complex lesions infiltrating various tissue planes.
- May occur in the craniofacial skeleton, most commonly in the mandible.
Venous Malformations

Normal vein

Lumen

Multiple layers of smooth muscle cells

Endothelial cell

Venous malformation

Lumen

Endothelial cell

Single layer of smooth muscle cells
Venous Malformations

- The VM is a soft, compressible, nonpulsatile mass with rapid refilling.
- Expansion will occur on compression of the jugular vein or Valsalva's maneuver.
Venous Malformations

- Treatment algorithm proposed by Yao et al, 2001
MRI  US

Low-flow malformation

Symptomatic

Sclerotherapy

Follow-up with US

Asymptomatic

Observation

Surgical procedure

Physical therapy
Venous Malformations

- Treatment of choice is transcutaneous sclerotherapy
- 80% ethanol is the most commonly used agent
- According to literature complications occur in 10-15% of ethanol injections
Venous Malformations

- Complications of ethanol sclerotherapy include:
- Necrosis or ulceration of the skin
- Neuropathy
- Complications of systemic absorption, such as pulmonary vasoconstriction, or direct depressant activity on the myocardium
Venous Malformations

- Systemic absorption can be decreased by
- Selecting only lesions with minimal connection to the systemic circulation
- Using a double needle technique
Contrast medium is injected via the first catheter (top, arrow).

The lesion fills with contrast medium, the pressure in the lesion increases and the contrast medium flows along the path of least resistance and leaves the lesion via the second catheter (bottom, arrow).

During this procedure, it is easy to rule out any connection to the adjacent system.
Ethanol sclerotherapy as independent therapy applied to VM lesion at lower lip. A Clinical finding of rapidly expanding VM lesion at lower lip with recurrent bleeding and pain. B WBBPS finding of VM lesion in lower lip, more distinctive on the regional view (right-hand pair of photographs). C Duplex sonographic view of localized type of VM lesion within lower lip.
D,E MRI of extratruncular form of VM lesion extended throughout lower lip. F Angiographic view of ethanol sclerotherapy selectively given to nidus of VM lesion directly by percutaneous direct puncture technique with satisfactory control.
Venous Malformations

- Alternative sclerosing agents have been tested
- OK-432, derived from Streptococcus pyogenes
- Luzatto, in 2000, reported 100% success rate treating lymphangiomamas
- Giguere, in 2002, however, reported only 60% success
- No toxicity has been reported in the literature
Capillary Malformations

- Also known as “port wine stain”
- Light pink to dark purple patch
- Can be associated with hypertrophy of soft tissues or facial skeleton
Capillary Malformations

- Patients with capillary staining of the ophthalmic (V1) and maxillary (V2) dermatome may have Sturge-Weber syndrome.
- Vascular anomalies of the leptomeninges causing progressive degeneration and atrophy of the cerebral hemispheres.
Capillary Malformations

- Tunable flashlamp pulsed-dye laser (585-nm wavelength) is widely regarded as the optimum treatment
- Complications include hypo- or hyperpigmentation and scarring
Capillary Malformations

- Goldman, 1993
- 43 children with 49 separate port-wine stain vascular malformations were treated with pulsed dye laser at 585 nm. Overall, 16% of patients had more than 95% resolution of their port-wine stains after an average of 4.8 (1 to 11) treatments.
- Average improvement of 69% in those lesions not clearing completely
- Lesions in patients less than 4 years of age were almost twice as likely to clear than were those in older children (20% vs 12%), and in fewer treatments (3.8 vs 6.5).
- No episodes of scarring or persistent pigmentary changes in any of the patients.
Capillary Malformations

- Theories exist on reasons for persistence of lesions after laser thermolysis.
- In 2005, Sivarajan et al. investigated changes in capillary depth and diameter that occur with serial laser treatments.
- Their findings show that persistent vessels in capillary malformations after treatment are deeper and narrower than those in untreated lesions.
- The authors suggest that since depth and diameter are crucial to the most effective wavelength and pulse duration, respectively, of a therapeutic laser, adjusting these laser parameters for treating resistant lesions may be effective.
Arteriovenous Malformations

- Erythematous mass with a thrill or bruit
Arteriovenous Malformations
Arteriovenous Malformations

- There are four recognized stages of AVMs:
- A stage I lesion has a pinkish-bluish stain and warmth.
- In stage II, the lesion has pulsations, thrill, and bruit.
- In stage III, the patient has dystrophic skin changes, ulceration, bleeding, and pain.
- Finally, in stage IV, the patient has high-output cardiac failure.
Arteriovenous Malformations

- Extracranial arteriovenous malformations of the head and neck differ from AVM of the extremities.
- Decompensation failure resulting from AVM is very rare in the head and neck region.
- HNAVM communicate with the deep venous system less frequently because they usually occur in the more superficial tissue layers.
- The main symptoms of HNAVM are cosmetic disfigurements, deformities caused by the expansion of the lesions, and bleeding with pain or ulceration.
Arteriovenous Malformations

- Small, superficial arteriovenous malformations can be removed surgically.
- However, embolization has been the only feasible treatment option for most arteriovenous malformations.
- Embolization, which closes off the arterial feeders of the malformation, is generally effective in arteriovenous malformations to stabilize the malformation.
- In some patients, AVMs can be cured with repetitive embolizations. Most AVMs require several sessions, generally 2 months apart, with regular follow-ups.
- Also, with successful embolization, a surgical excision can become feasible in some AVMs.
Arteriovenous Malformations

- During the embolization procedure, the nidus needs to be embolized, but the large arterial feeders should not be embolized.
- Similarly, surgical ligation of the arterial feeders should not be performed. If the arterial feeders are embolized percutaneously or ligated surgically, the arteriovenous malformation nidus recruits new smaller arterial feeders, which then can not be accessed for nidus embolization and makes AVM management very difficult.
Arteriovenous Malformations

- Lee, 2004, retrospectively reviewed 66 patients treated for AVM with embolosclerotherapy and/or surgery.
- The authors report 100% success rate using preoperative embolism with NBCA glue plus surgery.
- They also report >90% success rate using embolosclerotherapy alone for surgically inaccessible (infiltrating) lesions.
Arteriovenous Malformations

- Han et al 2006 reviewed 20 patients over 7 years treated for AVM of the head and neck
- Ethanol sclerotherapy, surgical excision and embolization were used as treatments, either alone or in various combinations.
- Ethanol sclerotherapy had a success rate of 50.0% and a permanent complication rate of 8%.
- Surgical excision combined with embolization yielded 100% successful resolution of their HNAVM.
- 15% suffered from permanent complications including CN VII weakness.
- In total, 16/20 patients (80.0%) eventually achieved a ≥75% reduction in the size of their lesions.
Arteriovenous Malformations

Pt with HNAVM

MRI, Ultrasonography, Angiography, $^{99m}$Tc RBC scintigraphy

Percutaneous accessibility to AVM nidus

(+)

Sclerotherapy

Size decrease $\geq 50\%$ in 3-times sclerotherapy

(+) Follow-up

(-)

Embolization

Excision

Follow-up

Repeat (option)
Pearls

- If you suspect CVM, consult your friendly neighborhood interventional radiologist
- U/S and MRI are good initial tests
- Don’t rush into treatment if the patient is asymptomatic