

Vascular Anomalies

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Vascular Anomalies

Can be divided into two types:

- Tumours
- Malformations

Vascular Tumours

Can be divided into two groups:

- Congenital- present at birth
- Non-congenital- appear later

Vascular Tumours Classification

Mulliken 1982

Congenital vascular tumours include:

- Haemangioma
- Kaposiform Haemangioendothelioma (KHE)
- Tufted angioma
- Haemangiopericytoma

Vascular Tumours Classification

Mulliken 1982

Acquired vascular tumours include:

- Pyogenic granuloma
- Kaposi's Sarcoma
- Angiosarcoma
- Unilateral nevoid telangiectasias
- Angiolymphoid hyperplasia with eosinophilia
- Haemangioendotheliomas

Vascular Tumours

Haemangiomas:

- Proliferative vascular tumour
- Not present at birth
- Grow disproportionate to the child's growth
- Evolution → involution (when involuting develops areas of grey discoloration)
- Many sub-types: RICH, NICH, Segmental, etc

Rapidly Involuting Congenital Hemangioma (RICH)

- Vascular tumor
- Fully developed at birth
- Undergoes rapid involution and regression
- Rare entity
- Has positive glucose transporter (Glut-1)

Non-Involuting Congenital Hemangioma (NICH)

- Present before birth
- Grows in proportion with patient
- Does not involute
- If removed early (2-4 years) is similar to RICH
- Has negative glucose transporter (Glut-1)

Haemangiomas

- Can be life-threatening
- Treatment with oral steroids, supportive therapy, excision
- Managed by Pediatric Dermatologists, pediatric plastic surgeons
- 'Strawberry haemangioma' should not be used.

Kaposiform Haemangioendothelioma (KHE)

- Found in infants, less common in adults
- Close association with Kasabach-Merritt syndrome
- Tumour size may be large (>5cm)
- Localized:
 - Retroperitoneum 50%
 - Proximal thigh and groin 20%
 - Arms 20%
 - Other areas 10%

Kasabach-Merritt Syndrome

- Present from birth to first few weeks old
- Key Characteristics on skin:
 - Large
 - Swelling
 - Rapidly growing
 - Tender
 - Bruising reddish-purple soft-tissue
- Management
 - Prednisone
 - Interferon alpha
 - Vincristine

Tufted Angioma

- Tumour found presence:
 - At birth 14%
 - First year of life 49%
- Characteristics
 - Firm
 - Flat
 - Hyperhidrosis
 - Hard
 - Painful
- Associated with pregnancy and liver treatment

Haemangiopericytoma

- Found most often in the pelvis, proximal femur, vertebrae and humerus
- Affect patients ranging from 12 to 50
- Grow slowly and may be 20 years before diagnosis made
- Symptoms
 - Painless swelling/lump
 - Difficulties using limbs
 - Breathing difficulties
 - Coughing

PHACE Syndrome

- Posterior fossa brain malformations, large facial Hemangioma, Arterial anomalies, Cardiac anomalies and aortic coarctation, Eye abnormalities, and Sternal clefting and/or Supraumbilical raphe
- Rare, more common in females
- Present from birth to first few weeks of life
- Unilateral left-side most common, can be bilateral

Vascular Malformations

- Structural abnormality
- Errors of morphogenesis (4th-10th week)
- Present at birth
- Grow proportionately to the child
- Do not involute

Vascular Malformations can be a combination of:

- Capillary
- Venous
- Arterial
- lymphatic

Terms **NOT** to use to describe VM:

- Port-wine stain
- Cherry angioma
- Salmon patch
- Campbell de Morgan spots
- Cavernous haemangioma
- Hepatic/ vertebral/ intramuscular haemangioma
- Naevus flammeus
- Lymphangioma
- Cystic hygroma

Vascular Malformation **Mulliken Classification 1982**

- Capillary Malformation (CM)
- Venous Malformation (VM)
- Lymphatic Malformation (LM)
- Arteriovenous Malformation (AVM)
- Any combination of the above

Doppler Findings

Predominantly a collection of vessels

High flow lesions

- Prominent arterial feeders
- AVM
- AVF

Low flow lesions

- Capillary/ Venous/ lymphatic

AVM vs. NICH

AVM

- Mostly vessels
- High flow
- Arterial feeder
- Will not go away
- Surgery:
- Wide excision
- Deeper excision

NICH

- Soft tissue mass
- High flow
- Arterial feeder
- Will not go away
- Surgery:
- Narrow margins
- Superficial fat

Capillary Malformation (CM)

- Dilated mature capillaries
- Present at birth
- No involution
- Uni or bilateral
- Pink in infancy → purple with age
- Raised and nodular after puberty

Capillary Malformation (CM)

- PWS
- Phacomatosis pigmentovascularis

Sturge Weber Syndrome

- Facial CM
 - Trigeminal nerve distribution (V1 ± V2 + V3)
 - Unilateral but could be bilateral
- Glaucoma
- Convulsions
- Hemiparesis
- Mental retardation

Venous Malformations

- Bluish phlebectasias
- Overlying skin may be normal or CM
- STP may occur

Usually localized around

- Face
- Trunk, limbs
- Brain, spinal cord, lungs etc

Generalized:

- Blue Rubber Bleb Syndrome
- Glomovenous Malformation (Glomangioma)

Blue Rubber Bleb (BRB) Syndrome

- Inherited AD
- Generalised distribution
- Multiple venous malformations
- Large draining veins
- DD
 - venous compression syndromes esp in UL
 - Multiple Hereditary Glomangiomas

BRB Syndrome

- Multiple cutaneous and GI lesions
- 0.1 to 5 cm nodules
- Lesions bleed causing Fe def
- Spontaneous thrombosis
- GI complications eg bowel infarction
- Other organs involved:
 - > Brain, heart, lung, nasopharynx, etc

Maffucci Syndrome

- VM
- Enchondromas
- Bony abnormalities
 - Usually asymmetric
 - Cause secondary fractures

Glomovenous Malformation: (Glomangiomas)

- Arise from glomus cells
- Glomus cells are of smooth muscle origin
- Glomus bodies:
 - arteriovenous shunts present on acral structures
 - Neuromyoarterial receptors responsible for thermoregulation

- Glomus tumours are:
 - Usually solitary- nail bed
 - Painful
 - Arise later in life
 - Not inherited

- Generalised
- Not tender
- Present in childhood
- Strong family history

Lymphatic malformations

- Can be classified as:
 - Microcystic
 - Macrocystic

- Complications involves:
 - Bleeding
 - Infection
 - Lymphedema when extensive
 - May be associated with bony hypertrophy

- Old terminology examples:
 - Lymphangioma
 - Lymphangioma circumscriptum
 - Cystic hygroma

Capillary-lymphatic Malformation (CLM)

Includes:

- Hypertrophic naevus flammeus
- ‘verrucous lymphangioma’
- Haemangiomalymphangioma
- Angiokeratomas
- **CLM- Angiokeratoma of Fordyce**
- **CLM- Angiokeratoma Corporis Diffusum (Fabry’s Disease)**
- **CLM- Angiokeratoma of Mibelli**

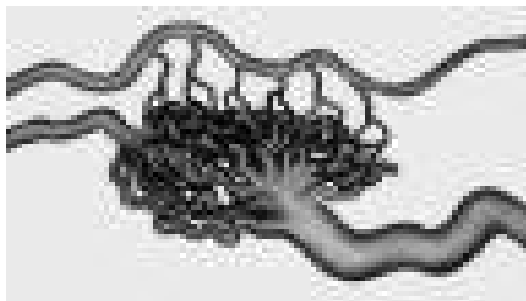
AVM Descriptions

- First appear as red or skin colored swelling
- Grow in proportion to the child’s growth
- At puberty may enlarge and bleed

AVM Classification

- Stage I: lesion has a pink-bluish stain and warmth. Doppler ultrasonography reveals arteriovenous shunting.
- Stage 2: the lesion has pulsations, thrill and bruit.
- Stage 3: the patient has dystrophic skin changes, ulceration, bleeding and pain.
- Stage 4: the patient has high-output cardiac failure.

AVM



AVMs

- May be confused with severe vv
- Chronic Aorto-caval fistulae
 - Venous dilatation in both lower limbs
 - Venous pulsation
 - 'Machinery' abdominal murmur
 - Abdominal pain, haematuria
- With macrofistulae may lead to
 - Limb hypertrophy
 - Tachycardia
 - Skin atrophy, ulceration

Complex Malformations

- Klippel Trenaunay Syndrome
 - C + V + L
- Parkes Weber Syndrome
 - C + V + A + L

KTS Identification

- CVL malformation
- Commonly presents as varicose veins
- Present at birth
- Usually unilateral
- Single limb or an arm and a leg on the same side
- Limb hypertrophy
- Subcutaneous fat is thickened
- May be painful
- Deep veins are normal
- Anterolateral vein of the thigh is usually involved

Proteus Syndrome

- Also known as Riley-Smith and Bannayan Syndromes
- Condition of abnormal growth
- Occurs bones, skin and head
- Symptoms includes:
 - Rough and raised skin
 - Increased size of an organ or bone

- Capillary malformations

Cobb Syndrome

- Also known as Cutaneomeningospinal angiomatosis
- Rare, non-inherited condition
- Presented at birth
- Neurologic complications develop in early adulthood

Treatment for Vascular Malformations

Alcohol Sclerotherapy/Embolization

- Traditional method for treatment of VM
- Absolute ethanol (95-98%) is an aggressive sclerosant, causes degeneration of proteins in body and rapid thrombosis
- Advantage: most effective especially for large VMs
- Disadvantage: painful procedure, serious side effects e.g. peripheral tissue damages, respiration difficulties, CNS depression, cardiac arrest, etc. (1-4)
- Requires the use of tourniquets and compression to minimize the passage of alcohol into the systemic circulation.
- Performed under general anaesthesia, with careful monitoring of cardiovascular intra-operatively (too much ethanol in circulation becomes toxic, hemolysis and cardiac arrest).
- Local complications: blistering, full-thickness cutaneous necrosis and damage to local nerves. If ethanol reached the capillary bed of any given tissues (e.g skin), it may cause significant soft tissue swelling and subsequently compartment syndrome (5).

Ultrasound Guided Sclerotherapy

- In foam format it has improved efficacy and is comparable with alcohol in potency. It is also safer than alcohol.
- UGS involves using duplex imaging to guide and monitor the intraluminal introduction and the subsequent flow of the sclerosant in the vascular space and the vasospastic reaction of the vascular lesion.
- Acts by removing the endothelial lining of the vessels and inducing fibrosis. The foam format has the advantage of being more efficient in displacing intra vascular blood and increasing the contact surface area of the endothelial layer with the sclerosant. The foam is produced by mixing the detergent sclerosant with air using a 3 way stopcock.
- Advantage: no need for monitoring (taking into account when the optimum volume of sclerosant is used), decreased side effects and complication rate. Also, ultrasound allows visualization so one can stop the injection at the appropriate time once optimal response seen, thereby increasing safety.
- Disadvantage: not suitable for very deep structures as ultrasound tissue penetration diminishes with depth and visualization becomes a problem.
- Suitable for treatment of low flow lesions e.g. VMs and lymphatic malformations.
- High flow lesions e.g. arteriovenous malformations are still probably best treated with embolization.

Surgery (6)

- Aim: to remove the whole lesion, prepare the lesion for other treatments or rectify whatever remains following involution.
- Depends on the child's age, weight and degree of impairment the VM is causing. E.g. VM near the eye is removed at an earlier time than a VM located at the back.
- Can be performed in phases to achieve a better outcome.
- Remains a controversial treatment option because of a high recurrence rate (7, 8) of 18% (9).

Fluoroscopic Ultrasound Guided Sclerotherapy (FUGS)

- Fluoroscopic monitoring remains useful in monitoring the draining of feeding vessels. For certain patients, both imaging modalities (fluoroscopy and ultrasound) may be used simultaneously (10).
- Procedure:
 - 1) Ultrasound guidance is used to cannulate patients.
 - 2) Omnipaque contrast medium is injected allowing visualization of the lesion and the normal draining veins.
 - 3) The detergent sclerosant (e.g. Sodium Tetradecyl Sulphate 3%) is prepared as foam using the Tessari technique (1:3 liquid to gas ratio).
 - 4) The Foam is introduced slowly into the lesion and appears radio-lucent on fluoroscopy. The injection is stopped when the draining veins appear to take up the foam.
 - 5) Compression is applied and maintained for 7 days post-operatively.

Treatment of Haemangiomas

Oral corticosteroids (11)

- Aim: slow the growth of haemangiomas.
- Example of treatment course: Prednisolone 2mg/kg per day for 2 weeks, 1mg/kg for a further 2 weeks, and 0.5mg/kg for the last 2 weeks. Repeat courses should be avoided.
- Since high doses of it are needed to be effective, the patient must be carefully monitored (6). In children, the child's blood pressure and sugar levels need to be carefully monitored as anti-hypertensives may need to be administered (renal department).
- Use of Interferon-alpha is controversial and mostly abandoned.
- When loose tissue remains, plastic surgery can be used to make the appearance look better.
- Upper respiratory tract lesions can be treated with laser therapy instead of surgery.

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