Vascular anomalies

Old classification (infantile hemangioma & vascular malformations)

Recent classification (tumors & malformations).

1_Vascular tumors :

Infantile hemangioma

*In***f***antile haemangiomas : ***female*** (f)> males
*Strawberry naevae
*Benign
*Can be localised or diffused
*Histologically share features of placental tissue
*Expression of GLUT-1
*PHACE association

Stages of infantile hemangioma:

Stage1:

- During the first 5-8 months of life
- Rapid proliferation phase
- Soft warm prominent Doppler signal
- Bright red appearance (strawberry naevus)

Stage 2:

- Prolonged phase lasts until 7-9 years
- The lesion becomes darker with a grey hue so they lose their colour
- Fine capillary telangiectasia

Stage3:

- Histologically the cellular parenchyma has been substituted with a fibro-fatty residue
- Characterised by the presence of a soft lump (visible if the lesion is superficial)

Management

Tx of infantile hemangioma

1- mostly expectant
Rarely we need to do (biopsy / CBC /US/MRI)
2-Active intervention necessary if:

- Large size or disfigurement
- Multiple lesions causing high output cardiac failure
- Obstruction of vital structures
- Persistent laceration

First line tx (propranolol: vasoconstriction/ dose daily)

- second line tx (steroids:intralesional/ dose every 4-6 weeks / rebound growth with systemic steroids)

Rebound growth can occur when systemic steroids are tapered too quickly or discontinued after a period of effective treatment. This phenomenon can be concerning, as the hemangioma may re-grow, sometimes to a degree similar to the original presentation

Embolization : high output cardiac failure & bleeding lesions .

Surgery :
*excision

*tracheostomy the hemangioma causes severe airway compromise that cannot be managed with medications or other less invasive treatments, a tracheostomy might be necessary to ensure the child can breathe adequately.

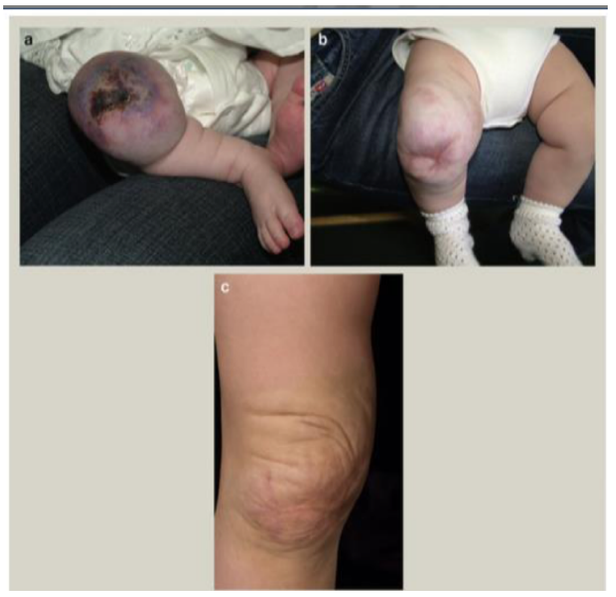
Pulsed dye laser
* no evidence that it alters natural hemangioma hx
* useful for surface telangiectasia
* for coagulation of ulcerated lesions but dressings are the principal form of wound care

Kaposiform haemangioendotheliomas
*aggressive / malignant
*clinical presentation: kasabach-Merritt phenomenon KMP
*TX: sirolimus for MTOR +ve tumors

Congenital haemangioma
*Have three subtypes

*Negative for GLUT1
Congenital hemangioma subtypes :
1-RICH rapidly involuting congenital hemangioma
Uncommon entities
*Fast involution and full regression by 1year of age
Large mass ? plaque like residuum ? atrophic patch

RICH vs infantile hemangioma	Faster involution	Firmer mass	With or without talengiectatic changes



2-Non-involuting congenital haemangiomas
*Mimic infantile haemangiomas &similar texture
Description of the mass:
Oval or round /flat shape or moderately bossed / with telangiectasia /may have halo
*don't exhibit further growth and do not regress
* tx is surgical excision

3-partially involuting congenital haemangiomas
*looks like NICH but regresses by 10 years age

Pyogenic granulomaPG
*lobular capillary haemangiomas
*rapid growth
*friable surface
*benign vascular tumor of skin & mucous membrane
* starts as small red papule growth over weeks and months stabilisation
* minor trauma causes : profuse bleeding and ulcers
(note * bleeding is difficult to control & often recurrent)

Vascular malformations
Classified according to vessel type :
1- Capillary

Capillary malformations (naevus flammeus, port-wine (stain, firemark
Definition: congenital, benign vascular malformations of the small vessels in the dermis
Epidemiology: may occur in association with a neurocutaneous disorder such as Sturge-Weber syndrome
-Clinical features: typically unilateral, blanchable, pink red patches that grow and become thicker and darker with age

Treatment: combination of supportive with involvement of clinical psychologist/ camouflage/ pulse dye laser
Surgery may be useful for reducing hypertrophied areas

Prognosis: benign skin lesion

2- Venous

Description of the mass: low-flow , blue , compressible soft tissue mass

Presentation:

- Disfigurement
- Pain
- Coagulopathy (D-dimer / fibrinogen)

5% genetic abnormalities Krit-1, TIE-2 & Glomulin , blue rubber bleb syndrome.

Mnemonic من سوق الجمعة :

اطو الكروت الزرقا اللامعة للحفلة

اطوِ من الطي يعني قريبة لمعنى tie /TIE-2

الكروت krit تعريب كلمة card

الزرقا blue rubber

اللامعة glomulin **glo**wing—<

Mx: compression garments/ NSAIDs/ scoerotherapy/ surgery

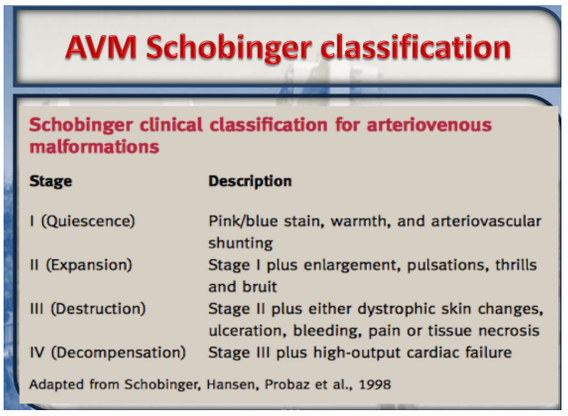
3- Lymphatic

Micro or macro cystic

Mx:

Sclerotherapy → injecting OK-432 sclerosing agent into the malformation directly
Surgery → for large or symptomatic and can lead to
1-seroma formation
2-infection

4- Arteriovenous malformation
*presentation varies according to the size and location
*characteristic nidus with arterial feeders and enlarged veins
* AVM schobinger classification



Mx:
Symptomatic stages (III&IV) → interventional rx, excisional surgery & reconstruction

Some lesions → repeated embolization (embolic agents like : ethanol , cyanoacrylate (glue), coils …)

5- Combined

*either isolated or associated with overgrowth disorders like(klippel-Trenunay / Proteus syndrome)
* heavy painful area &significant morbidity when involving the limb
*episodes of infection & wound breakdown

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