33- Vascular Lesions

Pre-Quiz:

1. **What is the difference between a vascular tumor and malformation, and what is an example of each?**
    - Vascular *tumors* (either benign *or malignant*) represent a ***proliferation* of *normal-appearing* cells or structures**
        - e.g. hemangiomas or pyogenic granuloma
    - Vascular *malformations* involve an ***error* in the *development* of blood vessels**
        - Tend to be *present at birth* and *persist* for a patient's lifetime
        - e.g. capillary malformations (port wine stains), lymphangiomas
2. **How do we categorize hemangiomas, and how do you counsel parents on what to expect?**
    - Two categories:
        - ***Congenital*** - *present* at birth
            - *rapidly* involuting congenital hemangiomas (RICH) or *noninvoluting* congenital hemangiomas (NICH)
        - ***Infantile*** - develop after birth, usually within the first few weeks of life
    - We expect them to **eventually go away**
        - They resolve at a rate of 10% per year, with 50% of lesions gone by age 5, 70% by age 7, 90% by 9 years old
        - *Median age of involution is 3 years old* and lesions are more persistent if they're present after 4 years of age
3. **What does PHACES syndrome stand for?**
    - **P**osterior fossae malformations - e.g. Dandy Walker malformations
    - **H**emangiomas - extensive on the face and often segmental
    - **A**rterial anomalies - e.g. aneurysms or anomalous branches of internal carotids or cerebral arteries
    - **C**ardiac anomalies - e.g. atrial or ventricular septal defects or coarctation of the aorta
    - **E**ye abnormalities - e.g. cataracts or retinal vascular changes
    - **S**ternal clefting or **S**upraumbilical raphe

New Vascular Reaction Pattern Order for Easier Memorization

1. Urticaria
2. Erythema multiforme – 2 forms (EM Major, EM minor)
3. The toxic erythema *group* with the ***three*** subclasses
    a. Drug - drug eruptions like SJS
    b. Bug - viral exanthems
    c. Toxin - toxin-mediated eruptions including staph scalded skin syndrome, Toxic shock syndrome, scarlet fever, and Kawasaki disease
4. The ***four*** *figurate* erythemas
    a. erythema annulare centrifigum
    b. erythema gyratum repens
    c. erythema migrans
    d. erythema marginatum
5. Vasculitis - inflammation of the blood vessel wall
6. Vasculopathy - vascular damage in the *absence of* vasculitis
7. Retiform purpura
8. Vascular growths - including neoplasms and vascular malformations.

Infantile Hemangiomas

Background:

- Extremely common, affect between **2-12%** of infants
- **Risk Factors**:
  - Female sex
  - Premature birth
  - Advanced maternal age
  - Placental abnormalities - e.g. placenta previa
  - Pre-eclampsia

Clinical Presentation:

- Up to 50% start with a **precursor lesion** such as a **red patch** or **telangiectasias**
- Typically **superficial** and have a **bright red** color, *but* can have **deeper components** that take on a **blue hue**
  - 25% of cases can be a mix of **superficial and deep components**
- **Grow *rapidly*** during the **first 5 months of life, plateau** between **9-12 months, slowly involute** thereafter

Diagnosis:

- Reasons for aggressive treatment/workup
  - Ulceration - more aggressive treatment needed
  - Threaten *function* by involving the periocular area, nose, ears, lips, or genitals
  - Extensive in the *beard* area - *60% risk* of airway involvement
  - Large/segmental in a *V1 nerve distribution* on the face - ↑ risk of PHACES syndrome
    - **Brain MRI** and **echocardiogram**
  - Large/segmental in the *lumbosacral area* - ↑ risk of LUMBAR/SACRAL syndrome

Treatment:

- **Topical timolol** or **Oral Propranolol:** rule out arterial and cardiac anomalies first if PHACES concern
- **Pediatric neurology/cardiology** + **ophthalmology** consults

PEARL: Neonates with a red patch in V1 need to be followed closely on a weekly basis, since *early* hemangiomas can look exactly like a port wine stain one day and then develop the classic elevation of hemangiomas within days to weeks

Nevus Flammeus (aka Port Wine Stain)

Background:

- Congenital *capillary* malformations that are present at birth
- Associated with Sturge-Weber Syndrome
  - Infants with an extensive port wine stain in a V1 nerve distribution on the forehead are at risk of Sturge-Weber syndrome

Clinical Presentation:

- Start as a **pink to red patch**
- **Well-demarcated**
- DO NOT rapidly proliferate and simply grow slowly *with* the patient
- DO NOT SELF RESOLVE and actually can become thickened and bumpy or papular later in life
- Sturge-Weber syndrome has a triad of a **port wine stain in a V1 distribution**, **leptomeningeal angiomatosis** (usually presents with **seizures)**, and **glaucoma**

Treatment:

- **Pediatric neurology** + **ophthalmology** + **radiology** consults

PEARL: Do NOT mix up the association between V1 *port wine stains* with *Sturge-Weber syndrome* and the V1 *hemangioma's* association with PHACES syndrome.

Nevus Simplex

Background:

- *the* most common vascular malformation affecting ~1/3 of newborns
- aka salmon patch, stork bite, or angel kiss

Clinical Presentation:

- Presents as a **pink, blanchable patch** that is often located in the midline of the *occiput* more so than the face or low back
- Compared to a port wine stain, a nevus simplex is **lighter in color**, is **less well-demarcated**, and usually resolves if it is on the face

Pyogenic Granuloma

Background:

- Typically affect children and young adults
- **Triggers:**
  - minor trauma in about 1/3 of cases
  - pregnancy
  - medications - e.g. systemic retinoids like isotretinoin and oral contraceptive pills

Clinical Presentation:

- Start as a **friable, red papule** that grows relatively quickly over the course of weeks to a few months
- Tend to appear on the **trauma-prone sites** of the gingiva, lips, fingers, or face
- Typically **solitary**, but there can be multiple lesions at times

PEARL: Use the full name pyogenic granuloma and do not call it a PG, because people often use PG to describe pyoderma gangrenosum, which is a completely different condition. Abbreviations are helpful in dermatology, but pyogenic granulomas and pyoderma gangrenosum are two conditions where you will probably want to avoid using the abbreviation PG.

Angiokeratomas

Background:

- Angio = superficial vascular nature, keratoma = hyperkeratotic look

Clinical Presentation:

- 5 types:
    - **Solitary or multiple angiokeratomas**
        - Tend to favor the **lower extremities**
    - **Angiokeratomas of Fordyce**
        - Classically appear on the **scrotum or vulva of older patients**
    - **Angiokeratoma corporis diffusum**
        - Often clustered in a **bathing suit distribution** and is associated with Fabry's disease
    - **Angiokeratoma circumscriptum**
        - Appears as **coalescing angiokeratomas** developing into a **plaque** in children
    - **Angiokeratoma of Mibelli**
        - Occurs in **teenagers** most often **on the hands and feet**

Histology:

- **Dilated vessels** in the **papillary dermis**
- **Acanthotic or thickened epidermis**

PEARL: At first glance, angiokeratomas basically look like a bloody seborrheic keratosis on path.

Tufted Angioma

- Classically **pink-red plaques on the neck of children less than 1 year of age**
- Can be associated with the **Kassabach-Merritt phenomenon (KMP)**
    - Refers to when platelets are trapped and destroyed in vascular lesions, leading to rapid lesion growth and coagulopathies

PEARL: KMP is classically seen in tufted angiomas or kaposiform hemangioendotheliomas (KHE). Remember that these two lesions **TA**KHE away platelets, with TAKHE spelled T-A-K-H-E and standing for **T**ufted **A**ngioma and Kaposiform HemangioEndothelioma.

Glomus Tumor

- Classically present as **solitary *painful* red papule on the finger**
- Can cause **significant nail dystrophy**

Angiolymphoid Hyperplasia with Eosinophilia (ALHE)

Background:

- Occur in **young to middle-aged adults**

Clinical Presentation:

- Presents as **grouped, pink to red-brown dome-shaped papules** *classically by the ear* but can be **anywhere on the head and neck**

Histology:

- Proliferation of **vessels in the dermis** with **large epithelioid endothelial cells**
- Background of **lymphocytic and eosinophilic inflammation**

Treatment:

- Variety of treatments:
    - **Excisional surgery** - most common
    - **Cryotherapy**
    - **Intralesional Kenalog**
    - **Imiquimod**

Kaposi's Sarcoma (KS)

Background:

- Low-grade vascular tumor

Clinical Presentation:

- 4 types:
    - **Classic KS**
        - Consists of **slow enlargement of macules into vascular plaques and nodules on the legs of older men** with a **Mediterranean** background
    - **African endemic KS**
        - Affects **young African males in endemic areas**
        - Can be **fatal**
    - **Iatrogenically immunocompromised KS**
        - Typically **skin-limited**
        - Occurs in **patients on immunosuppressive medications for organ transplants** or **autoimmune disease**
    - **AIDS-associated KS**
        - Caused by **HIV**

Histology:

- Stain for **Human Herpes Virus-8 (HHV-8)** on path to confirm diagnosis because it is present in *100%* of lesions

Treatment:

- Variety of treatments:
    - **Cryotherapy**
    - **Alitretinoin - topical retinoid**
    - **Systemic chemo** for progressive cases with internal organ involvement

Angiosarcoma

- High-grade malignant vascular tumor
- Classically presents in an **elderly Caucasian man** with a **bruise-like patch, plaque, or nodule on their face or scalp** that is **progressively enlarging**
- 5 year survival of <20%

PEARL: You have a patient with history of mastectomy and chronic lymphedema of their arm from the axillary lymph node dissection, and they start to develop purpuric papules and plaques of that limb. What do you they have? This vignette describes Stewart-Treves syndrome, which describes the development of angiosarcoma in an area of chronic lymphedema. It usually takes at least 4 years of problems with lymphedema before the angiosarcoma develops in the affected area.