10-Lichenoid Disorders

Lichen Planus

Background:

- 1% population
- Pt's usually >20 y/o w/ peak onset in **40-70s**
- Inflammatory disease that affects the **skin**, **mucous membranes**, hair and nails
- Unknown cause: pt may be predisposed to certain HLA subtypes + triggers (meds or virus) → alternate antigens on basal keratinocytes → targeted by T-Cells causing inflammation and lesions

Clinical Presentation:

- Remember the **6 Ps**!
    - **Pruritic**, **purple**, **polygonal**, **planar**, **papules**, and **plaques**
    - Skin manifestations: **anterior forearms** and **wrists**, hands, shins, neck, sacrum, genitals, and generalized
    - Look for **wickham striae** (fine grey-white dots or a net-like pattern overlying lesions)
    - Look for **koebnerization** = new lesions in the area of trauma (e.g. pt **scratching** → **new lesions in linear arrangement**)

PEARL: Most common location to see wickham striae is the oral mucosa. However, only 10% of patients w/ oral lichen planus go on to manifest the stereotypical skin findings.

- LP Variants: Location vs. Appearance vs. Cause
    - **Location: Genital LP, Mucosal LP, Nail LP, Inverse LP, Palmoplantar LP, Lichen Planopilaris**
        - *Genital*: 50% women & 25% men with cutaneous LP
        - *Mucosal*: 50% pts with cutaneous LP. Reticular wickham striae w/ asymptomatic lacy lines on the inside of the check. Can be caused by dental fillings, **amalgam** (mercury).
        - *Nail*: 10% of pts w/ cutaneous LP. Look for **lateral thinning, longitudinal ridges, pitting**, trachyonychia, dorsal pterygium
        - *Inverse*: **Intertriginous** areas of axilla, inguinal, and inframammary fold
        - *Palmoplantar*: pain ulcerations on the sole
        - *Lichen Planopilaris*: erythema and **perifollicular** scale around the **hair follicle** on the scalp and can **lead** to **scaring alopecia**.

    PEARL: LPP vs Discoid Lupus: LPP has more superficial inflammation at the infundibulum vs **D**iscoid Lupus that is **D**eeper at the isthmus

    PEARL: Clinical findings of **Graham-Little-Piccardi-Lasseur Syndrome**? Scarring alopecia of the scalp caused by LPP, non-scarring hair loss at the pubic and axillary lesions, follicular papules that appear as keratosis pilaris and classic LP lesions of the skin and the mucosa

    - **Appearance: Classic, Annular, Atrophic, Hypertrophic, Ulcerative, Linear, and Bullous, Pemphigoids, Pigmentosus**
        - *Classic*: 6 Ps!
        - *Acute*: classic LP w/ rapid onset of diffuse lesions that resolve more quickly over course of 3-9 months vs >1 year for classic LP
        - *Annular:* **ring-like** and typically affect the **penis** or **axilla**
        - *Atrophic:* **depressed** or hyperpigmented lesions on the **legs**
        - *Ulcerative:* affects the palmoplantar surfaces. Chronic ulcers are at > risk for SCC transformation
        - *Hypertrophic:* thick scaly **plaques** on the **shins** and dorsal **feet** that are often **symmetric** really **itchy** and can be confused with pickers nodules. SCC or KA develop in chronic LP.
        - *Bullous:* **blisters** that arise *within* **chronic** LP lesions
        - *Pemphigoides*: blisters arise mostly on *uninvolved skin*; 2/2 IgG auto-antibodies targeting BP antigen 2 of basement membrane (also targeted in bullous pemphigoid and herpes gestationis)
        - *Pigmentosus*: grey brown macules w/out prior erythema on the **sun exposed areas** of the **face** and **neck** w/ darker Fitzpatrick **skin types 3, 4.**
            - Typically older age of onset ~30-40 y/o (vs ashy dermatosis teens-20s)

    - **Cause: Actinic, Drug-induced**
        - *Actinic:* young **middle-eastern** adults and presents as **red/brown** plaques on the **sun-exposed** areas of the face, neck, and extremities
        - **Drug-induced**: 60 y/o w/ generalized lesions**, more eczematous, photo-distributed**, **NO** wickham striae, **spares** the **mouth** and **genitals**
            - **Delayed** rxn up to 1 year!
            - Look for **eosinophils** on path
            - **ACE-I, BB,** NSAIDs**,** gold**, HCTZ,** antimalarials**,** TNF-alpha inhibitors

Diagnosis:

- Biopsy

Histology:

- **Compact hyperkeratosis** w/out **parakeratosis**
- **Hypergranulosis** (opposite to psoriasis which typically has hypogranulosis)
- **Irregular acanthosis** w/ **sawtooth rete ridges**
- Dense **lichenoid lymphocytic** infiltrate leading to development of **civatte** bodies **(apoptotic keratinocytes)** or

Max-Joseph spaces **(separation between dermis and epidermis**)
- Eosinophils? Think drug-induced LP

PEARL: Think of classic LP as the strict parent that does not let parakeratosis or eosinophils around!

Treatment:

- **Classic LP: topical corticosteroids**
- **Oral or genital LP**: topical **calcineurin** inhibitors (pimecrolimus or tacrolimus)
- **Hypertrophic** lesions: **intralesional Kenalog**:
- **Generalized LP: Prednisone 20 mg multiple weeks** w/ **taper**, **Intramuscular kenolog, low dose methotrexate, metronidazole 500mg BID, phototherapy (UVB or PUVA)**
    - **Acitretin** or **cyclosporine** as **last resort**