8-Parapsoriasis

Background:

- Uncommon chronic idiopathic rash mimicking psoriasis

Epidemiology:

- Onset ~40 y/o. Males>female (M:F) 3:1

Clinical Presentation:

- **Small Plaque Parapsoriasis (SPP):**
  - **Scaly patches < 5 cm**
  - Wax and wane in course and then either resolve spontaneously or w/ treatment after several years
  - **"Bathing suit"** distribution
  - **NO association** w/ **cutaneous lymphoma**
  - Variant: *digitate dermatosis*: long finger-like patches along cleavage lines on the flanks

- **Large Plaque Parapsoriasis (LPP):**
  - **Scaly patches > 5cm**
  - **"Bathing suit"** distribution
  - **May progress to MF!**
    - **10-35% in 6-10 years**
  - Variant: *retiform parapsoriasis*. Wide-spread erythematous scaly patches in a net-like distribution.
    - Almost all cases progress to MF

Diagnosis:

- **Biopsy**: **interstitial infiltrate** of **CD4+ T-cells**

Histology:

- Both SPP and LPP have **parakeratosis**, corresponds to the scale seen clinically.
- Mild **acanthosis** and **spongiosis**.
- Mild **lymphocytic infiltrate** at the **DEJ** predominantly **CD4**+ and may have **T-cell clonality**, especially LPP

Treatment:

- **Topical corticosteroids** & various **phototherapies** like narrow band UVB

8-Pityriasis Rubra Pilaris

Epidemiology: acquired w/ bimodal onset in childhood/adolescence or in patients' 50's

Clinical Presentation:

- May begin with **scalp erythema** with scale→ **Red/brown follicular papules** with **central keratotic plug** having a "**nutmeg** grater" appearance affecting the **neck**, **head**, **trunk**, **extensor extremities**, and **dorsal fingers** → Eventually coalescing to **scaly plaques** w/ a **salmon to red orange color** that are **diffuse** and **symmetric** with **islands of sparing** (patches of normal skin within affected areas)
- Look for **keratoderma ("PRP sandals"**: thick hyperkeratosis of the skin of palms and soles
- Thickened yellow brittle nails, **NO PITTING** (vs. pitting seen in psoriasis)

PEARL: What are the 6 types of PRP?

- **Type 1) Classical Adult (55%):**
  - Rapid onset of clinical findings w/ good clinical prognosis.
    - 80% cases resolving over 3 years.
- **Type 2) Atypical Adult (5%)**
  - Chronic course of ichthyotic leg lesions, keratoderma, alopecia
- **Type 3) Classical Juvenile (10%)**
  - Clinical picture of classical adult but presenting in < 2 y/o
- **Type 4) Localized/Circumscribed Juvenile (25%)**
  - Symmetric keratotic follicular papules and erythematous plaques on extensor fingers, elbows, and knee
  - Most common type of PRP in children
- **Type 5) Atypical Juvenile (5%)**
  - Chronic follicular hyperkeratosis and erythema + *scleroderma-like* tightening of hands and feet
- **Type 6) HIV associated PRP (1%)**
  - Associated follicular spines, acne conglobata, hidradenitis suppurativa

Diagnosis:

- **Biopsy**

Histology:

- "**Checkerboard**" pattern: **alternating orthokeratosis** and **parakeratosis** w/ hyper and hypogranulosis
  - Looks like a "checkerboard" because you have orthokeratosis (without nuclei) sitting above purple hypergranulosis alternating with parakeratosis (has purple nuclei) sitting above hypogranulosis
- **Follicular plugging** (also seen in lichen sclerosis & discoid lupus)
- **Shoulder parakeratosis** (also seen in seb derm)
- **Irregular acanthosis**

Treatment:

- **Systemic Retinoids:** acitretin or **isotretinoin (1mg/kg per day)** may clear PRP in 6 months
- Methotrexate
- TNF-alpha
- Cyclosporine
- Azathioprine
- Phototherapy
- Note: topical corticosteroids are typically not that helpful