Invisible Dermatoses

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Pitted Keratolysis

- Discrete pits or craterlike lesions on the plantar surfaces
- Range from 1 to 7 mm in diameter and are similar in depth
- Some pits have a brownish color that may give the feet a dirty appearance
- Most cases are asymptomatic
- Hyperhidrosis is often noted on the feet
- Typically malodorous
Corynebacterium

- *Corynebacterium* genus
- Gram-positive, pleomorphic, aerobic rods
- Bacteria can hydrolyze keratin
- Hyperhydration greatly enhances growth of corynebacteria on the feet
  - Pitted keratolysis developed in 53% of 387 military volunteers whose feet remained wet for 3 or more days
Laboratory

- Wood’s Lamp shows characteristic coral red fluorescence
- Fluorescent examination of the patient's intertriginous areas may be helpful
  - Other corynebacteria-induced infections such as erythrasma and trichomycosis axillaris commonly coexist
Treatment

- **Hygiene**
  - Removal of the warm, moist conditions
  - Scrub their feet with an antibacterial soap.
  - Socks made of either cotton or absorbent synthetic material and change frequently

- **Topicals**
  - Aluminum chloride hexahydrate 20% (Drysol)
  - Erythromycin 2% solution
  - 1% clotrimazole cream
  - 2% miconazole nitrate cream
  - 1% clindamycin solution
  - Whitfield's ointment
  - 5% formalin solution
Tinea Versicolor

- Small and scaly white-to-pink-to-tan-to-dark spots
- Upper arms, chest and back, and may sometimes appear on the neck and face
- Prevents the skin from tanning normally
Treatment

- Topical or oral medications
- Uneven color of the skin may remain several months after the yeast has been eliminated
**Ichthyosis**

- **Xeroderma** *(xerosis)*
  - Mildest
  - Acquired, usually unassociated with systemic diseases
  - Lower legs of middle-aged or older patients
  - Exacerbated by cold weather and frequent baths

- **Inherited ichthyoses**
  - Excessive accumulation of scale on the skin surface, are classified according to clinical and genetic criteria
  - Consultation with a dermatologist is recommended to provide genetic counseling and guidance to treatments. Ichthyosis is a symptom in Refsum's syndrome (rare hereditary ataxia with polyneuritic changes and deafness caused by a defect in the enzyme phytanic acid hydroxylase) and in Sjögren-Larsson syndrome (hereditary mental deficiency and spastic paralysis); both syndromes are autosomal recessive.

- **Acquired ichthyosis**
  - Some systemic diseases (leprosy, hypothyroidism, lymphoma, AIDS)
  - Fine and localized to the trunk and legs or may be thick and widespread
Ichthyosis - Treatment

- Minimizing bathing is helpful
- Avoid Soaps and Hexachlorophene products
- Use emollient—preferably plain petrolatum, mineral oil, or lotions containing urea or hydroxy acids—should be applied twice daily, especially after bathing (for 10 min to hydrate the stratum corneum)
- Preparation containing 50% propylene glycol in water under occlusion (eg, thin plastic film or bags) every night after hydration of the skin is effective to remove the scale in ichthyosis vulgaris, X-linked ichthyosis, and lamellar ichthyosis,
  - Other useful agents include 5% or 6% salicylic acid gel, hydrophilic petrolatum and water (in equal parts), and cold cream and the hydroxy acids (eg, lactic, glycolic, and pyruvic acids) in various bases
- Most effective therapies for most ichthyoses are oral synthetic retinoids
  - Etretinate is effective in X-linked ichthyosis and epidermolytic hyperkeratosis
  - 0.1% tretinoin (vitamin A acid, retinoic acid) cream or oral for lamellar ichthyosis
Ichthyosis-Treatment for Epidermolytic Hyperkeratosis

- Epidermolytic hyperkeratosis (bullous congenital ichthyosiform erythroderma)
  - Long-term cloxacillin or erythromycin long as thick intertriginous scaling is present
  - Prevent superinfection with painful, foul-smelling pustules
  - Regular use of soaps containing chlorhexidine may also reduce the bacteria
Calcaneal Petechiae

- Self-limited, asymptomatic, trauma-induced darkening of the posterior or posterolateral aspect of the heel that occurs primarily in young adult athletes
  - First described in a group of basketball players in 1961
- Clinically insignificant but may be mistaken for malignant melanoma
- Similar lesion termed black palm (tache noir) has been described on the thenar eminence in weightlifters, gymnasts, golfers, tennis players, and mountain climbers
Black heel occurs in adolescents and young adults who participate in sports that involve frequent starts and stops, such as basketball, football, soccer, lacrosse, and racquet sports.

- Irregular dark lesion over the heel
- Usually is asymptomatic and does not inhibit the patient from performing routine daily activities

Examination reveals a blue-to-black macule or patch ranging in size from a few millimeters to several centimeters in diameter.

- The posterior and posterolateral heel are affected most commonly.
- On close inspection, multiple petechiae are centrally aggregated with a few scattered satellite patches.
- The dyschromia often is in a horizontal distribution; however, both circular and oval lesions may occur.
Macular Amyloidosis

- Pruritic eruption that is variable in severity often hyperpigmented
  - Small dusky-brown or grayish pigmented macules distributed symmetrically over the upper back and, in some patients, the arms
  - About 50% have reticulated or rippled pattern of pigmentation

- Nylon Towel Dermatitis
  - Constant friction and rubbing with a nylon brush or towel
Macular Amyloidosis-Histopathology

- Amyloid stains
  - Congo-red stain
  - Periodic acid-Schiff (PAS)
  - Methyl violet
  - Crystal violet
  - Cotton dyes (pagoda red, Sirius red)
  - Fluorescent dyes (thioflavin-T and Phorwhite BBU)

- Amyloid deposits are usually found within the dermal papillae
  - Globular, resembling colloid bodies, and may be in contact with basal cells at the DEJ
Macular Amyloidosis - Treatment

- **Control pruritus**
  - Sedating antihistamines
  - Topical dimethyl sulfoxide (DMSO)
  - Intrallesional steroids
  - Ultraviolet B (UVB)

- Laser vaporization, dermabrasion, and excision

- Electrodesiccation and curettage?
Urticaria (Hives)

- Lasts a few hours before fading without a trace
  - New areas may develop as old areas fade
  - Vary in size from as small as a pencil eraser to as large as a dinner plate and may join together to form larger swellings
  - Usually are itchy, but may also burn or sting
- 10-20 percent of the population will have at least one episode in their lifetime
  - Usually disappear quickly in a few days to a few weeks. Occasionally, a person will continue to have hives for many years.
- Difficulty breathing or swallowing you should go to the emergency room
Urticaria

- Biopsies are typically paucicellular
- Papillary dermal edema with scattered neutrophils and eosinophils
- Chronic or persistent lesions may have increased infiltrate
- Always rule out vasculitis
Brachioradial Pruritis

- Itch, burning and/or changed sensation arise in the areas of skin on either or both arms
  - Most commonly affected area is the mid-arm
  - Affected skin may appear entirely normal
  - Changes may arise from rubbing and scratching purpura and ecchymoses
Brachioradial Pruritis

- 22 patients with BRP-11 had cervical spine radiographs
  - The radiographs showed cervical spine disease that could be correlated with the location of pruritus in each of these 11 patients

**CONCLUSIONS**

- Patients with BRP may have underlying cervical spine pathology

Brachioradial Pruritis-Treatment

- Sun protection
- Cooling lotions (camphor and menthol)
- Cervical spine manipulation
- TENS
- Capsaicin cream-depletes nerve endings of their chemical transmitters
- Local anaesthetic creams
- Amitriptyline tablets
Argyria

- Gray to gray-black staining of skin and mucous membranes produced by silver deposition
- Early gray-brown staining of the gums develops, later progressing to involve the skin diffusely
- Slate-gray, metallic, or blue-gray color and may be clinically apparent after a few months to years
- Viscera tend to show a blue discoloration, including the spleen, liver, and gut
Argyria

- Long-term systemic treatment with silver salts containing drugs
  - Silver protein suspension for chronic gastritis or gastric ulcer, or as nose drops
  - Colloidal silver dietary supplements are marketed widely for cancer, AIDS, diabetes mellitus, and herpetic infections
  - Occupational disease in workers who prepare artificial pearls or who are employed in the cutting and polishing of silver

- Smallest amount of silver reported to produce generalized argyria in humans ranges from 4-5 g to 20-40 g.
  - Silver at 50-500 mg/kg body weight is the lethal toxic dose in humans
  - Normal human body contains about 1 mg silver
Argyria

- Small, round, brown-black granules appear singly or in clusters
  - Spare both the epidermis and its appendages, appearing in greatest numbers in the basement membrane zone surrounding sweat glands.
  - Favor the connective tissue sheaths around pilosebaceous structures and nerves
  - Predilection for elastic fibers and are best visualized as strikingly refractile with dark-field illumination
  - An increase in the amount of melanin in exposed skin also appears to occur
- Electron microscopy demonstrates electron-dense granules
- Neutron-activation analysis, atomic absorption spectrophotometry, or x-ray dispersive microanalysis can be used to confirm
- May decolorize the silver by placing histologic sections into 1% potassium ferricyanide in 20% sodium thiosulfate
Treatment

- 5% hydroquinone treatment
- Sunscreens and opaque cosmetics may be helpful
- Dermabrasion
- Chelation attempts unsuccessful
Morphea

- Usually asymptomatic and onset of lesions is insidious
- Arthralgias occasionally localized to an affected extremity
- Deep morphea may be associated with arthralgias, arthritis, myalgias, and carpal tunnel
- En coup de sabre lesions of linear morphea
  - Can present with seizures, headache, and visual changes
Localized vs. Systemic

- **Localized scleroderma**
  - Mainly the skin
  - May also affect muscles and bone, but it does not affect internal organs
  - Usually no progression to systemic
  - Two types of localized scleroderma
    - Morphea and linear
    - Inflammatory stage, followed by one or more slowly enlarging patches or plaques
    - Plaques are most commonly oval in shape and vary in size
      - Ivory/yellow center and are surrounded by a violet colored area.
  - Generalized morphea may involve almost the entire skin surface
Some cases need comparative biopsies of adjacent normal skin

- Atrophoderma of Pasini and Peirini

- Decreased thickness of dermis
Electron Microscopy

- 143 patients underwent axillary skin biopsies as part of evaluations for metabolic disease.
- Twenty-three (16%) had abnormalities
  - Mitochondrial (n = 12)
  - Lysosomal (n = 6)
  - Increased glycogen (n = 3)
  - Nonspecific cytoplasmic inclusions (n = 2)
  - Ceroid lipofuscinosis (n = 1)
  - Intradermal giant cells containing vacuoles and tubular inclusions (n = 1)
Electron Microscopy

- Muscle biopsies were performed in 13 of the 23 patients
  - Clinical or biochemical diagnosis was reached in 11 patients
    - Metachromatic leukodystrophy (n = 2)
    - Electron transport chain abnormalities (n = 2)
    - Glutaric aciduria type II (n = 1)
    - Unverricht disease (n = 1)
    - Lennox-Gastaut syndrome (n = 1)
    - Ketotic hypoglycemia of childhood (n = 1)
    - Probable Leigh disease (n = 1)
    - 5-methyl tetrahydrofolate homocystine methyltransferase deficiency (n = 1)
    - Pyruvate dehydrogenase deficiency (n = 1).

- Hum Pathol 2001;32:649-655
CADASIL

- Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy

- Disease of young adults and presents with migraines with or without an aura, mood disturbances, focal neurologic deficits, strokes, and dementia
  - Most patients will show symptoms by age 60 years
  - Recurrent subcortical ischemic events causing permanent deficits in as many as 2/3 of patients

- Mutations in the Notch 3 gene cause degeneration of vascular smooth muscle cells and multiple small infarcts in the white and deep gray matter of the brain

- Numerous areas, granular, electron-dense, osmiophilic material abutted vascular smooth muscle cells
CADASIL-MRI Findings

- Lesions usually symmetrically situated within the white matter and deep gray nuclei-periventricular white matter is preferentially involved
- Usually in the frontal lobe, temporal lobe, subinsular white matter, and internal and external capsules with relative sparing of the inferior frontal and occipital white matter in the early stages
- Brainstem affected in 45% of cases
Granular osmiophilic material abutting basement membrane of pericytes
Questions

- The advantage of a bad memory is that one enjoys several times the same good things for the first time.

-- Friedrich Nietzsche