

Dermatology Trivia

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Hidradenitis Suppurativa

- Chronic recurrent, debilitating disease predominantly affecting a young active population .
- Dysfunction of the folliculoinfundibular unit affecting the apocrine-bearing skin.

Hidradenitis Suppurativa

- **Characterized by recurrent abscess formation in areas of the skin that contains both terminal hair and apocrine glands**
- **Tender erythematous nodules that suppurate with scar formation and development of sinus tracts and scarring**
- **Post pubertal disease that is more common in women**
- **Prevalence remains controversial**



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Hidradenitis Suppurativa

- Etiology and pathophysiology of HS have yet to be fully elucidated
- Phenotype of the disease has different morphologic features covering a wide spectrum
- Has a negative impact physically, emotionally and psychologically on the patient

Hidradenitis Suppurativa

- Dysfunctional cutaneous immune response to commensal bacteria
- Painful inflammatory chronic condition
- Under-reported and misdiagnosed
- About a 1/3rd of patients have their first symptoms before the age of 18

Hidradenitis Suppurativa

- Deep painful abscesses and chronic draining sinus tracts are present in these regions:
 - Axillae
 - Anogenital
 - Inframmary
 - Inguinal

Hidradenitis Suppurativa

- Hallmark
 - Double comedone
 - Black head with 2 or more openings that communicate with the subdermis

Hidradenitis Suppurativa

- Diagnosis: Three criteria
 1. Typical lesions
 - ▶ Deep seated cystic lesions, abscesses, scarring, pseudo comedones
 2. Location
 - ≥ 1 of the areas HS:
 - ▶ Axillae, groin perineal region, buttocks, Inframammary, Intermammary
 3. Chronic nature

Hidradenitis Suppurativa

▶ **Systemic associations**

- ▶ Obesity (12%–18%)
- ▶ Metabolic syndrome
- ▶ Diabetes Mellitus (DM)
- ▶ Smoking
- ▶ Arthritis
- ▶ Spondyloarthropathy
- ▶ Hormone related disorders

Hidradenitis Suppurativa

- ▶ **Systemic associations**
 - ▶ Substance dependence
 - ▶ Depression
 - ▶ Inflammatory Bowel Disease—9 times more likely to develop HS than the general population

Hidradenitis Suppurativa

Systemic associations

- ▶ Follicular occlusion tetrad(HS,acne conglobata,dissecting cellulitis of the scalp and pilonidal cyst)

Hidradenitis Suppurativa

Systemic associations

- ▶ Pyoderma gangrenosum
 - ▶ PASH (pyoderma gangrenosum, acne, suppurative hidradenitis)
 - ▶ PAPASH (pyogenic arthritis, pyoderma gangrenosum, acne and suppurative hidradenitis)
 - ▶ PsAPASH (psoriatic arthritis, pyoderma gangrenosum, acne and suppurative hidradenitis)

Hidradenitis Suppurativa

▶ Scoring of Disease Severity

▶ Hurley Staging

- ▶ Stage I: Abscess formation, single or multiple without sinus tracts and cicatrization
- ▶ Stage II: Recurrent abscesses with tract formation and cicatrization, single or multiple, and widely separated lesions
- ▶ Stage III: Diffuse or near-diffuse involvement or multiple interconnected tracts and abscesses across the entire area

Hidradenitis Suppurativa

- ▶ HS is worsened by hormones particularly androgens
- ▶ 83% improvement was noted in 47 patients on a dairy free diet
- ▶ Avoidance of foods with high glycemic index and dairy might be beneficial.

Hidradenitis Suppurativa

- ▶ There is increasing evidence that the immune system plays a role
- ▶ Increased levels of cytokines such as IL1- β and TNF- α have been noted in tissue cultures
- ▶ Other cytokines thought to be upregulated include IL-10, IL-12, IL-17 and IL-23

Hidradenitis Suppurativa

- ▶ Systemic therapy is the mainstay of treatment
 - ▶ Antibiotics
 - ▶ Tetracycline, doxycycline, rifampin, clindamycin, dapsons
 - ▶ Retinoids
 - ▶ Acitretin
 - ▶ Isotretinoin

Hidradenitis Suppurativa

- ▶ Systemic therapy is the mainstay of treatment
 - ▶ Antiandrogens
 - ▶ Immunosuppresants
 - ▶ Tacrolimus
 - ▶ Cyclosporine

Hidradenitis Suppurativa

- ▶ Biologic agents for HS
 - ▶ Effective for moderate to severe HS
 - ▶ Adalimumab, anakinra, etanercept, infliximab, and ustekinumab
 - ▶ Adalimumab is a fully humanized monoclonal antibody that corresponds to the human immunoglobulin G1 and has heavy and light chain variable regions exhibiting specificity for human TNF α

Hidradenitis Suppurativa

▶ Biologic agents for HS

▶ Infliximab

- ▶ Chimeric antibody composed of both human and mouse proteins targeting TNF α

▶ Ustekinumab

- ▶ Human anti-p40 monoclonal antibody
 - ▶ P40 is a shared subunit of human interleukins (ILs)-12 and -23

▶ Anakinra is a recombinant IL-1 α receptor antagonist

Hidradenitis Suppurativa

- ▶ Biologic agents for HS
 - ▶ Strongest evidence with adalimumab and infliximab
 - ▶ May 2015 US FDA approved adalimumab for management of moderate to severe HS

Hidradenitis Suppurativa

- ▶ Topical therapy such as antibiotics and keratolytics combined with proper wound dressings are used as adjunctive treatment

Hidradenitis Suppurativa

- ▶ Laser and light therapy for HS
 - ▶ Laser hair reduction
 - ▶ Photodynamic therapy
 - ▶ CO₂ Excision and vaporization
- ▶ For advanced HS, surgical excision is the mainstay of treatment
- ▶ HS remains a widely unrecognized and difficult to treat condition resulting in diagnostic delay and patient dissatisfaction
- ▶ There is a renewed interest in understanding this devastating disease and current research is encouraging

Hidradenitis Suppurativa

Summary

- ▶ Hidradenitis suppurativa (HS) is an uncommon, but not rare inflammatory skin disease, it affects 98 per 100,000 people in the United States
- ▶ HS affects females more than males. African Americans and other ethnic populations are at higher risks
- ▶ One study showed highest incidence among 30-39 year old adults
- ▶ Clinical presentations include erythema followed by the presentation of deep-acne like cysts, folliculitis, boils mucopurulent discharge
- ▶ BMI of 40 and smoking are corelated with HS

Hidradenitis Suppurativa

- ▶ Patients with HS have increased risk of death from myocardial infarction, ischemic stroke, heart disease and other cardiovascular events
- ▶ Current treatment options include antimicrobials, immunosuppressants, anti-inflammatories and biologic therapy in severe cases
- ▶ In chronic cases, surgical excisions may be necessary, but recurrence is possible
- ▶ Intense pulsed light, neodymium-doped yttrium aluminum garnet laser and photodynamic therapy may benefit patients, but the clinical evidence for this practice is rated low.

Discoid lupus erythematosus

Discoid Lupus Erythematosus (DLE) Background

- ▶ Most common **chronic** cutaneous manifestation of SLE
 - ▶ 20% of SLE patients are affected
- ▶ Presents as hypo- or hyperpigmented patches or plaques, with erythema during active disease
 - ▶ May be variably atrophic or hyperkeratotic
- ▶ May occur as an isolated finding in the absence of SLE = *isolated DLE*



Source: Carol Soutor, Maria K. Hordinsky:
Clinical Dermatology
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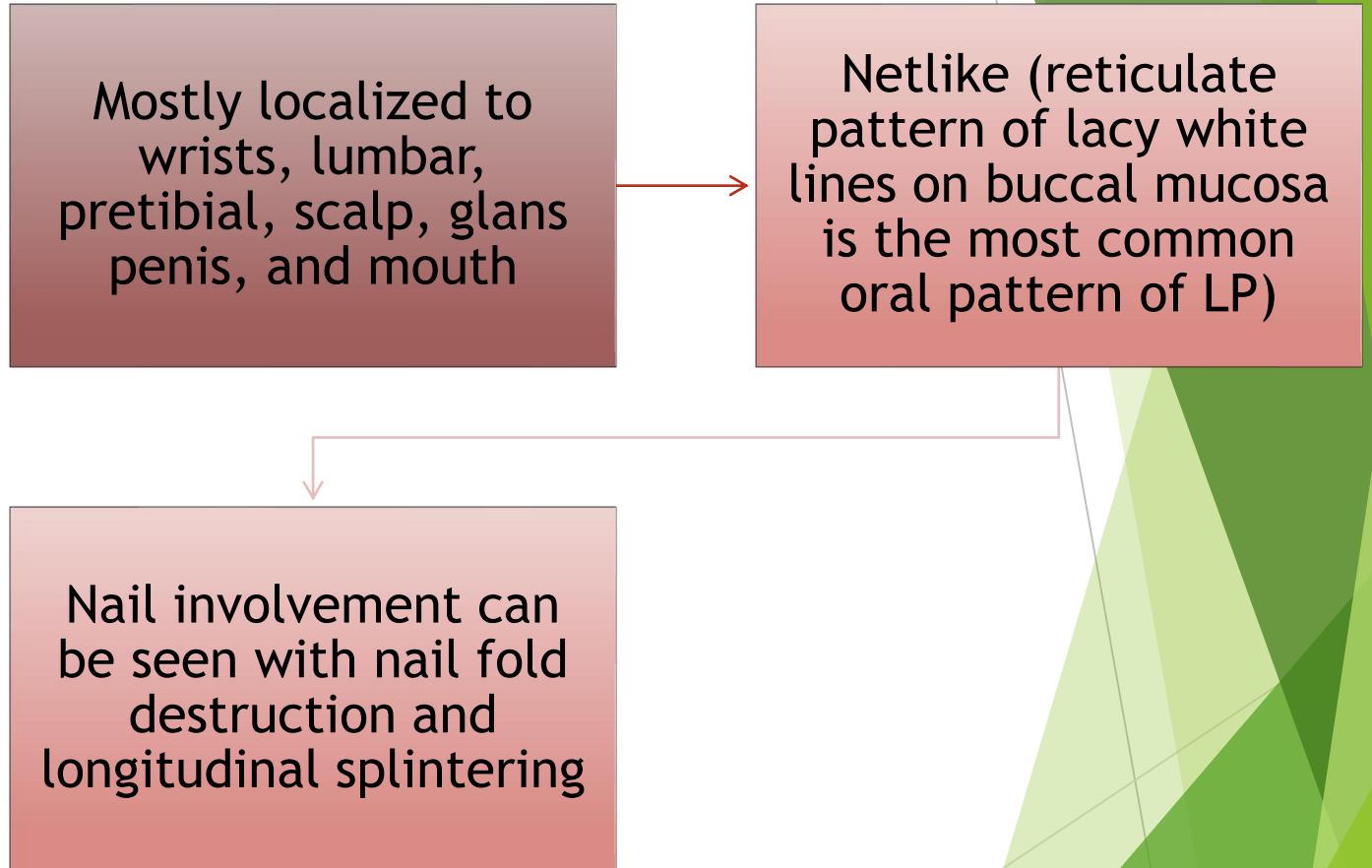
Lichen planus (LP)

- ▶ Acute or chronic inflammatory dermatosis
- ▶ Four P's - purple, polygonal, pruritic, papule
- ▶ Milky white reticulated patches in the mouth
- ▶ Idiopathic
- ▶ Drugs (gold, mercury)
- ▶ Infection (HCV)
- ▶ Flat topped sharply defined papules with white lines (Wickham's striae)

Mostly localized to wrists, lumbar, pretibial, scalp, glans penis, and mouth

Netlike (reticulate pattern of lacy white lines on buccal mucosa is the most common oral pattern of LP)

Nail involvement can be seen with nail fold destruction and longitudinal splintering



Due to cell-mediated autoimmune reaction



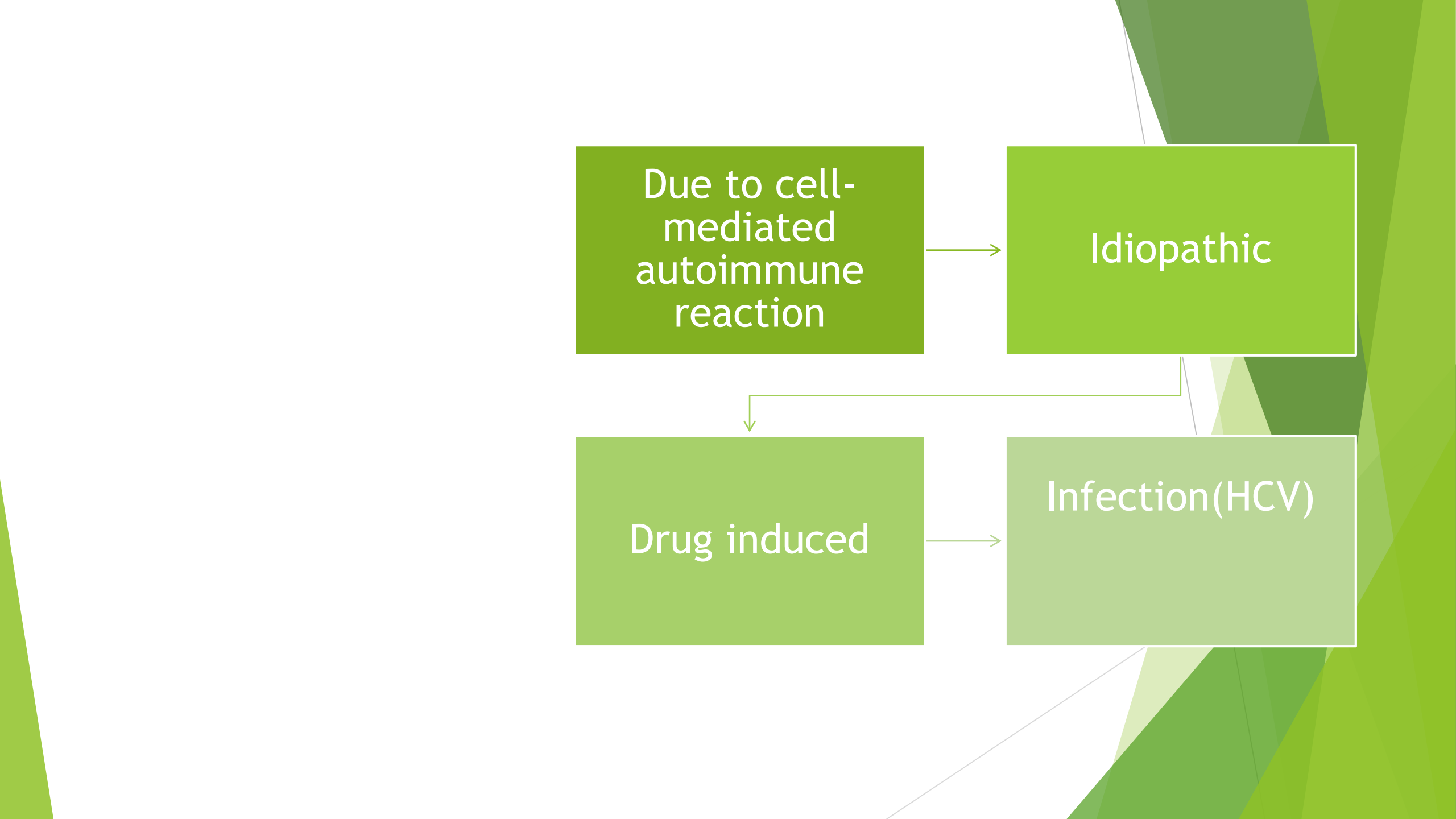
Idiopathic



Drug induced



Infection(HCV)





Lichen planopilaris



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Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology, in General Medicine, 8th Edition*: www.accessmedicine.com

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Vesiculo-bullous lichen planus. Vesicles and bullae with violaceous-erythematous papules and plaques on the foot.

Lichen planus

- ▶ Treatment
- ▶ Superpotent topical corticosteroids topical calcineurin inhibitors, intralesional and or systemic corticosteroids, phototherapy, methotrexate, acitretin, metronidazole

Lichen Planus



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Sweet Syndrome (Acute febrile neutrophilic dermatosis)

An inflammatory process expressed clinically by markedly edematous acuminate papules and edematous plaques situated mostly on the face, upper part of the trunk, and arms, especially the hands, and often accompanied by fever and leukocytosis.

- ▶ Described in 1964 by Sweet
- ▶ Four features
 - ▶ Fever
 - ▶ Leukocytosis
 - ▶ Acute tender lesions
 - ▶ Papillary dermal infiltrate of neutrophils
- ▶ Primarily affects adults
 - ▶ 86% of patients are women

- ▶ Four subtypes
 - ▶ Classic type (71%)
 - ▶ Associated with neoplasia (11%)
 - ▶ Associated with inflammatory disease
 - ▶ Associated with pregnancy (2%)
- ▶ SS is a reactive phenomenon and considered a cutaneous marker of systemic disease
 - ▶ AML
 - ▶ Streptococcal infection
 - ▶ Inflammatory bowel disease, solid tumors, pregnancy and other hematologic malignancy, infections
 - ▶ Medications such as granulocyte colony-stimulating factor, oral contraceptives, minocycline

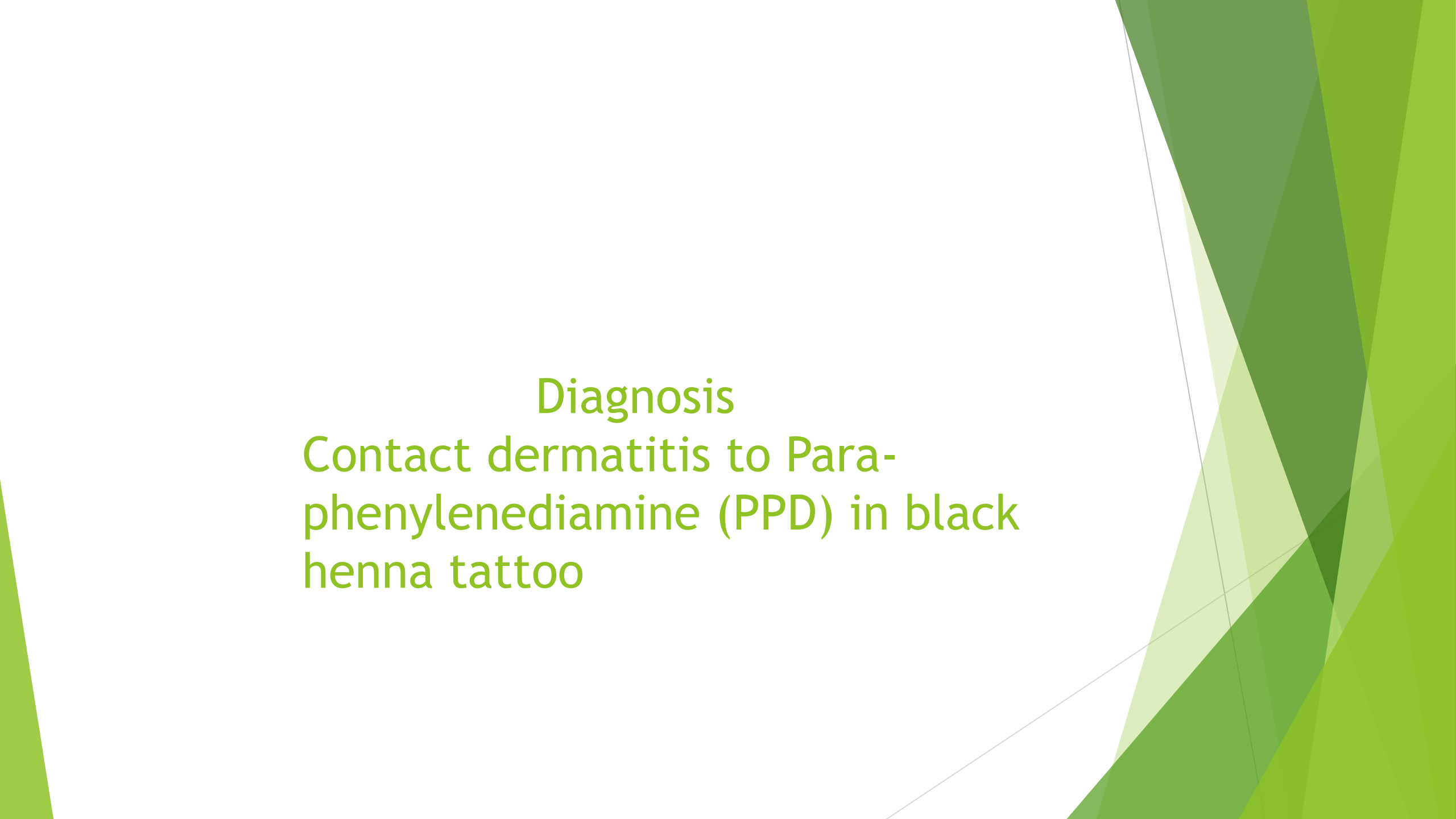
Clinical manifestations

- ▶ Acute, tender, erythematous, plaques, occasionally blisters with annular or arciform pattern occur on the head, neck, arms and legs
- ▶ The trunk is rarely involved
- ▶ Fever (50%)
- ▶ Arthritis, arthralgia or myalgias can be seen in up to 2/3rd of cases
- ▶ Conjunctivitis or episcleritis (30%)
- ▶ Oral apthae (13%)
- ▶ Cardiac, renal, hepatic and pulmonary involvement is rare

- ▶ Laboratory studies
 - ▶ Moderate neutrophilia (<50%)
 - ▶ Elevated ESR (>30mm/hr.)
 - ▶ Increased alkaline phosphatase
- ▶ The hallmark of Sweet syndrome is a nodular and diffuse dermal infiltrate of neutrophils with karyorrhexis and massive papillary dermal edema

Treatment

- ▶ Systemic corticosteroids -**Gold standard**
- ▶ Oral potassium iodide
- ▶ Colchicine
- ▶ Indomethacin
- ▶ Dapsone
- ▶ Doxycycline
- ▶ Cyclosporine

The background features abstract, overlapping geometric shapes in various shades of green, ranging from light lime to dark forest green. The shapes are primarily triangles and polygons, creating a dynamic, layered effect. The overall composition is clean and modern, with the text centered on a white background.

Diagnosis

Contact dermatitis to Para-phenylenediamine (PPD) in black henna tattoo

Allergic Contact Dermatitis to Henna Tattoo

- ▶ Allergic Contact Dermatitis (ACD) affects >14.5 million people in the US every year
- ▶ High economic burden
- ▶ Identification of the allergen combined with education can prevent progression of condition

Para-phenylenediamine (PPD)

- Contact allergen of the year 2006 by the American Contact Dermatitis Society (ACDS)
- PPD is a chemical substance commonly used in permanent hair dye
- Initially formulated for use in hair dye at the end of the 19th century

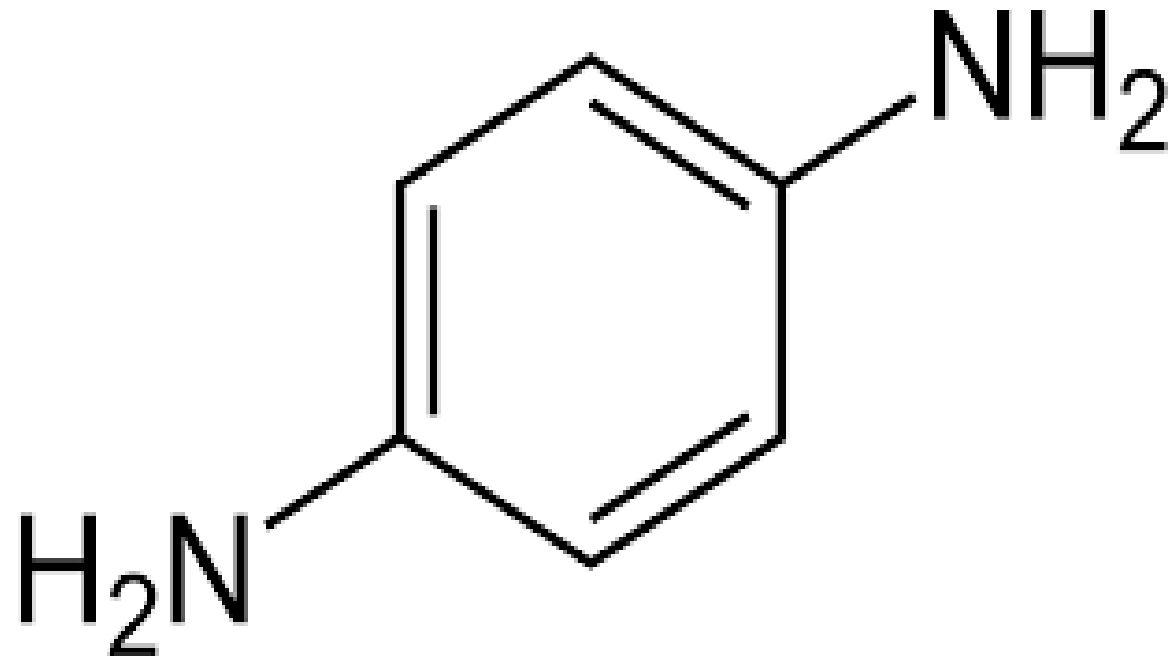
Para-phenylenediamine (PPD)

- ▶ PPD is also found in
 - ▶ Textile/fur dyes
 - ▶ Cosmetics
 - ▶ Temporary tattoos
 - ▶ Photographic developers
 - ▶ Printing inks
 - ▶ Black rubber mix
 - ▶ Oils
 - ▶ Gasoline

Para-phenylenediamine (PPD)

- ▶ Colorless substance that requires the oxidation process to become colored
 - ▶ The intermediate partially oxidized state causes sensitization in susceptible individuals

Para-phenylenediamine (PPD)



Paraphenylenediamine (PPD)

- ▶ Cutaneous reactions to PPD
 - ▶ Mild dermatitis: scalp, rim of ears, upper eyelids
 - ▶ Facial edema
 - ▶ Blistering edema
 - ▶ Rare cases of anaphylaxis

Para-phenylenediamine (PPD)

- ▶ Cross reactions
 - ▶ Azo and aniline dyes
 - ▶ Benzocaine
 - ▶ Procaine
 - ▶ Para-aminobenzoic acid (PABA)
 - ▶ Sulfonamides
 - ▶ Hydrochlorothiazide

Para-phenylenediamine (PPD)

- ▶ FDA prohibits the use of PPD on the skin
- ▶ Black henna tattoo is natural henna mixed with PPD
 - ▶ Can result in severe dermatitis, scarring and post-inflammatory pigment alteration
- ▶ Maximum permitted concentration in hair dye is 6%
- ▶ Levels of PPD in henna tattoos can be as high as 29.5%

Para-phenylenediamine (PPD)

- ▶ Patch testing
 - ▶ Critical in identifying the allergen
- ▶ Metallic and vegetable based hair dyes
- ▶ Para-toluenediamine sulfate (PTDS)
 - ▶ Tolerated by 50% of people allergic to PPD
- ▶ The Contact Allergen Management Program
 - ▶ Assists with identifying allergen free products

Erythema Elevatum Diutinum

- ▶ Erythema elevatum diutinum (EED) is a rare chronic leukocytoclastic vasculitis of unknown etiology.
- ▶ First described by Hutchinson (1888) and Bury (1889).
- ▶ The disease may occur in any age group but is more common in adults, particularly in the third, fourth, and fifth decades.
- ▶ Equal incidence in men and women.

- ▶ Cause is unknown but hypothesized to be an immune complex disease.
- ▶ Can be associated with inflammatory bowel disease, RA, SLE, IgA gammopathy, strep infection, multiple myeloma, myelodysplasia, celiac disease, HBV and HIV infection.
- ▶ Clinical features include symmetric, persistent, red-brown, red-purple or yellowish papules, nodules and plaques that favor the extensor surfaces of joints particularly the hands and knees.
- ▶ The mucous membranes and trunk are generally spared but the ears and face may be affected.

- ▶ Initially the lesions are soft but become fibrotic over time.
- ▶ Pain, aching, burning and hypo or hyperpigmentation are associated symptoms.
- ▶ EED is chronic condition lasting upto 35 years with periods of remission and exacerbation.
- ▶ EED may be present for many years before the diagnosis of hematological abnormalities becoming apparent in a patient.



<http://dermis.net>

- ▶ Histologically, acute lesions resemble Sweet's syndrome, showing papillary dermal edema, neutrophilic vasculitis with leukocytoclasia and fibrinoid change. Older lesions, on the other hand, reveal perivascular fibrosis and granulation tissue that clinically present as firm, dome-shaped nodules.
- ▶ In addition, these chronic lesions may show xanthomatization (extracellular cholesterosis) that clinically may give a yellowish tinge to the nodules.
- ▶ Depending on the degree of dermal edema and infiltrate, there may be a zone that is unaffected in the papillary dermis.

- ▶ Dapsone is considered the drug of choice in treating EED, primarily due to its rapid onset of action. However, lesions promptly recur following dapsone withdrawal.
- ▶ Other oral medications proven effective include niacinamide, colchicines, chloroquine, phenformin, clofazimine, and cyclophosphamide.
- ▶ Systemic corticosteroids are generally ineffective. One report of a patient with EED with IgA paraproteinemia and refractory to other modalities responded to intermittent plasma exchange.

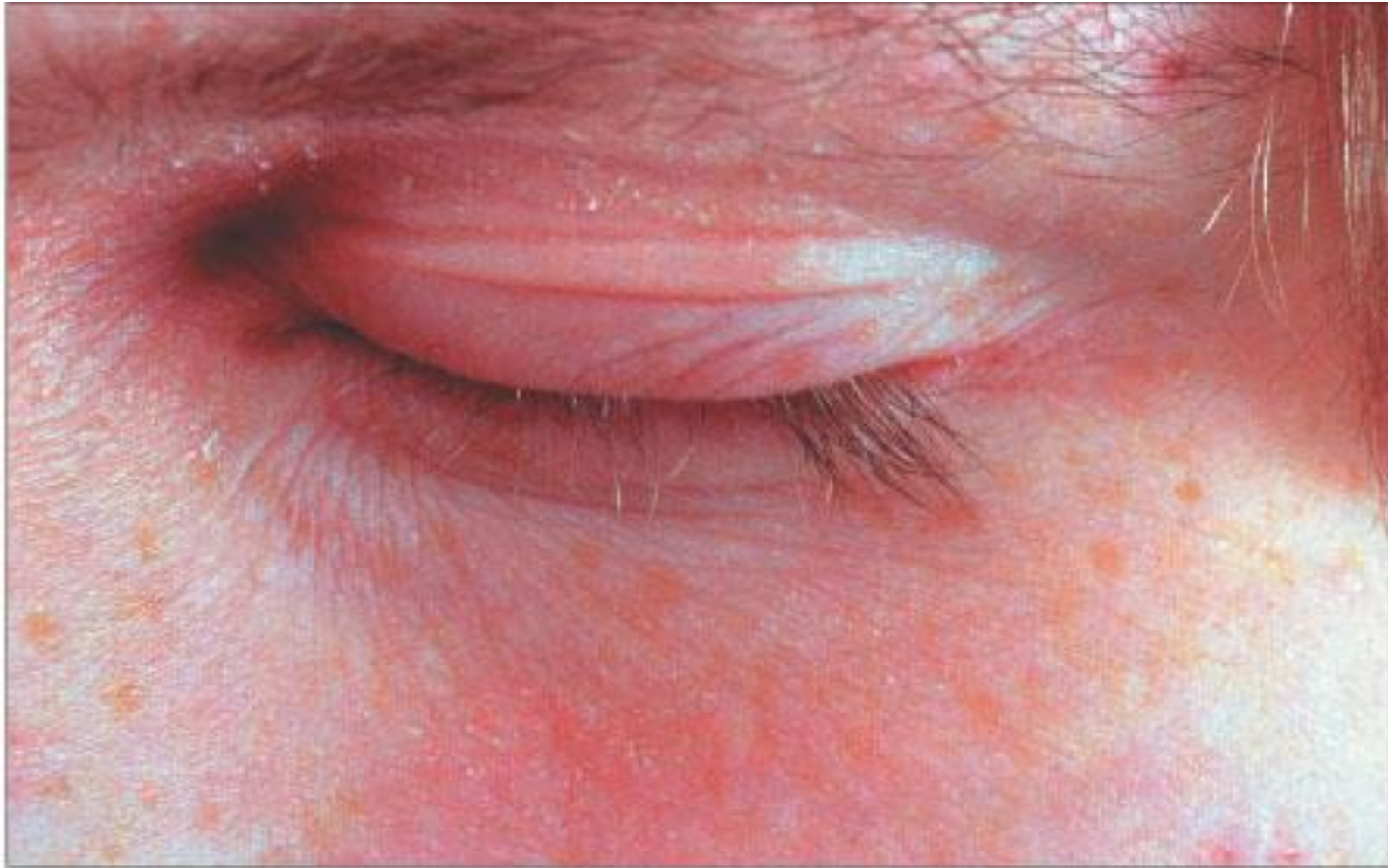
Pityriasis alba

Melasma

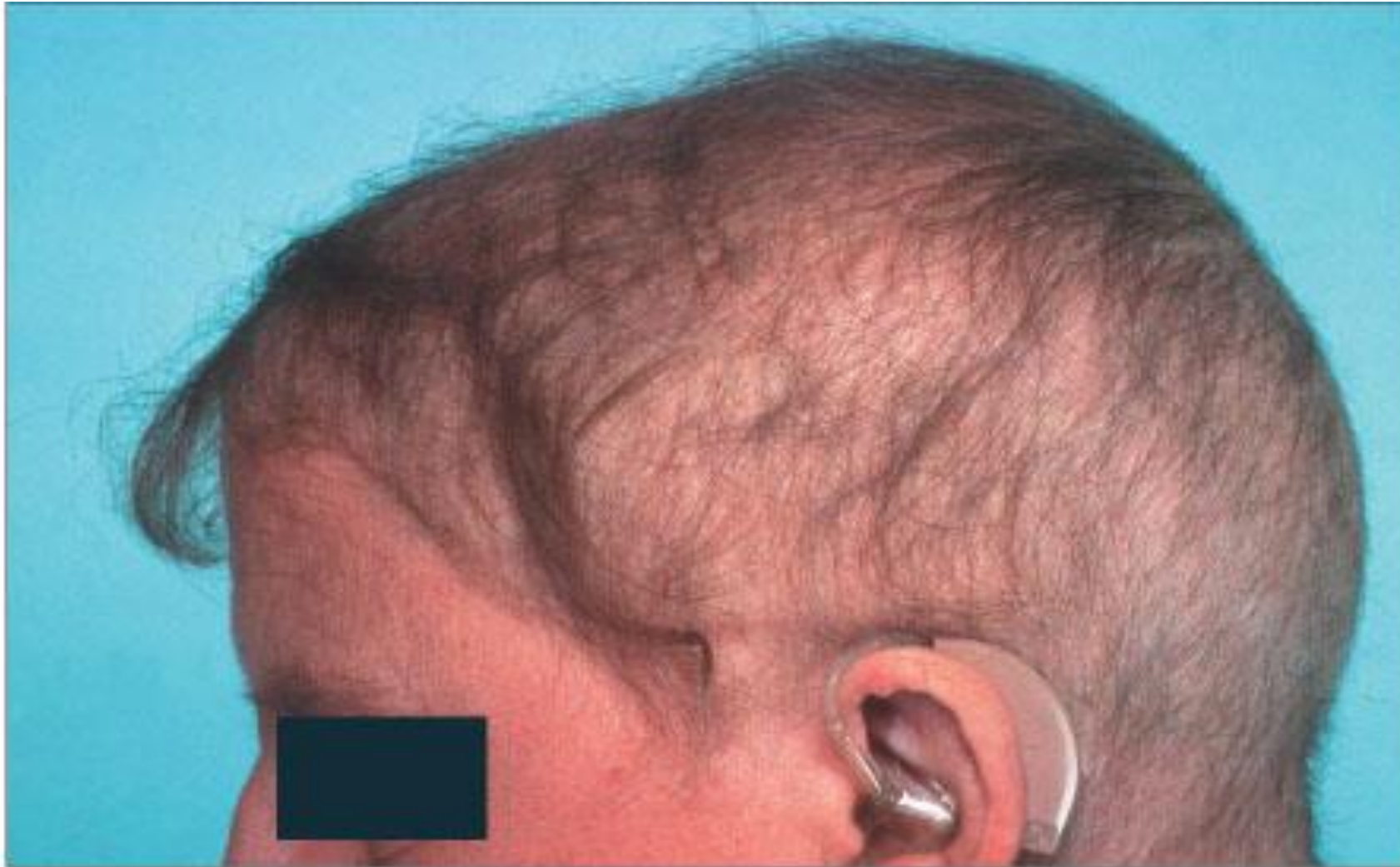


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Alopecia areata

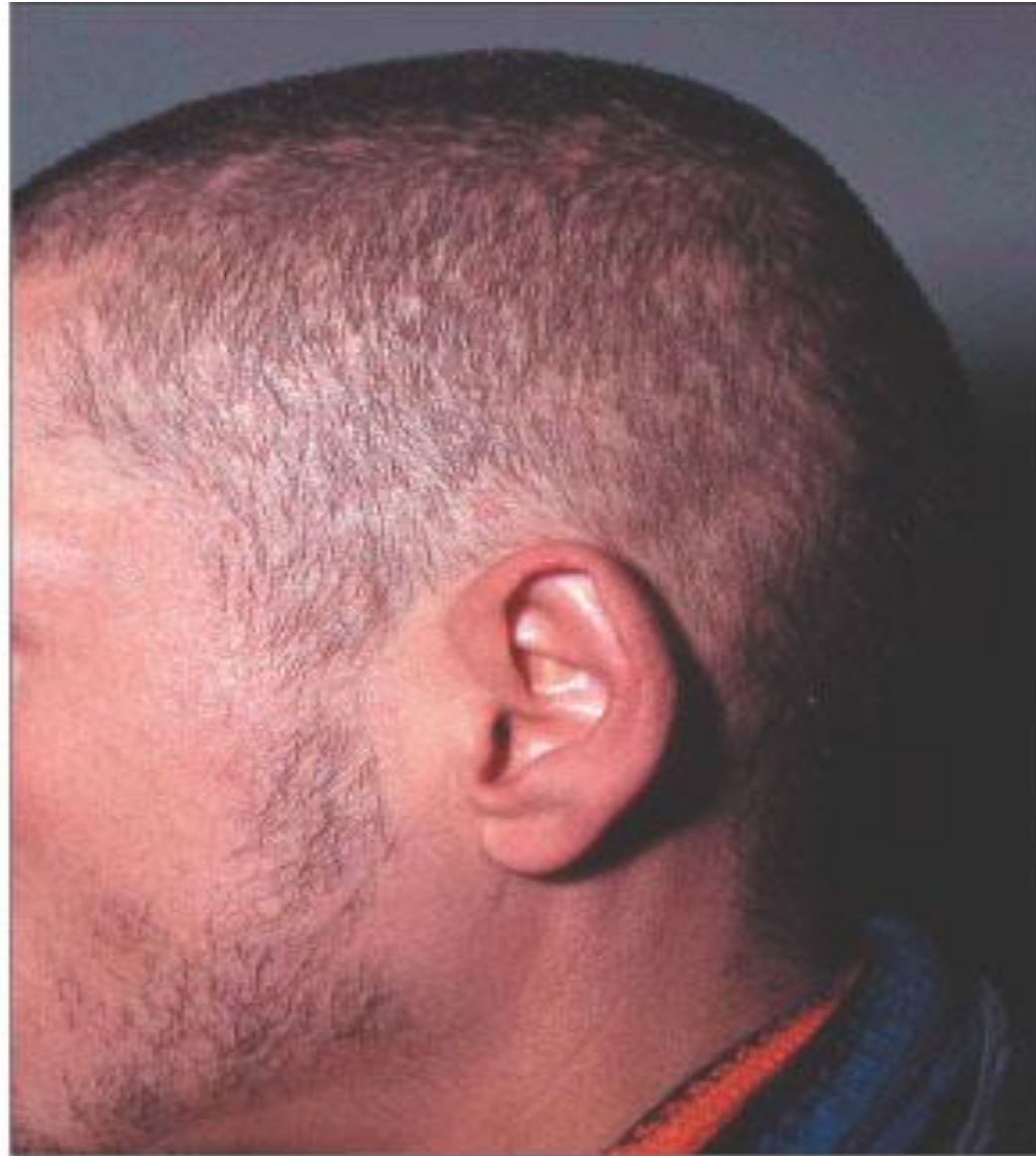


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Trichotillomania



Syphilitic alopecia



Thank you