Papulosquamous Diseases

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LECTURE

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30 March 2008

Papulosquamous Diseases

- a group of dermatoses with distinct morphologic features
- the primary lesion is most commonly a papule, usually erythematous, with a variable degree of scaling on the surface
- plaques form through the coalescing of primary lesions.

Papulosquamous Diseases

- Psoriasis (all forms)
  - gutate psoriasis
  - plaque psoriasis
  - pustular psoriasis
  - nails psoriasis
  - psoriatic arthritis
- Erythroderma
- Keratosis Follicularis (Darrier Disease)
- Parapsoriasis
- Pytiriasis Rosea

Papulosquamous Diseases

PSORIASIS

Psoriasis

- is a chronic inflammatory and proliferative disorder of the skin clinically manifested as well-circumscribed, erythematous papules and plaques covered with silvery scales typically located over the extensor surfaces and scalp
- course of the psoriasis is unpredictable
- immune system dysfunction in the background of a genetic predisposition is believed to be at the core of the disease process
- psoriasis is a very common disease and affects one to two per cent of the population in all geographic regions

Aetiology and Pathogenesis

- despite being the subject of intensive research over the years, the precise aetiology of psoriasis still remains unknown

2. Genetic factors
   - different groups of psoriatics population showed the following genes to be significantly associated with psoriasis: HLA Cw6, B13, B16, and B27

Aetiology and Pathogenesis

1. Provocating factors
   - **Trauma** - All types of trauma can lead to the development of plaque psoriasis (eg, physical, chemical, surgical, infective, and inflammatory). The development of psoriatic lesions at a site of injury is known as the ***Koebner phenomenon***
   - **Infection** – An acute eruption of guttate psoriasis may be provoked by streptococcal pharyngitis. ***HIV infection*** may be associated with increase in disease severity
   - **Drugs**. Lithium ,withdrawal of systemic corticosteroids, beta-blockers, antimalarials, and NSAIDs may cause flare of the disease

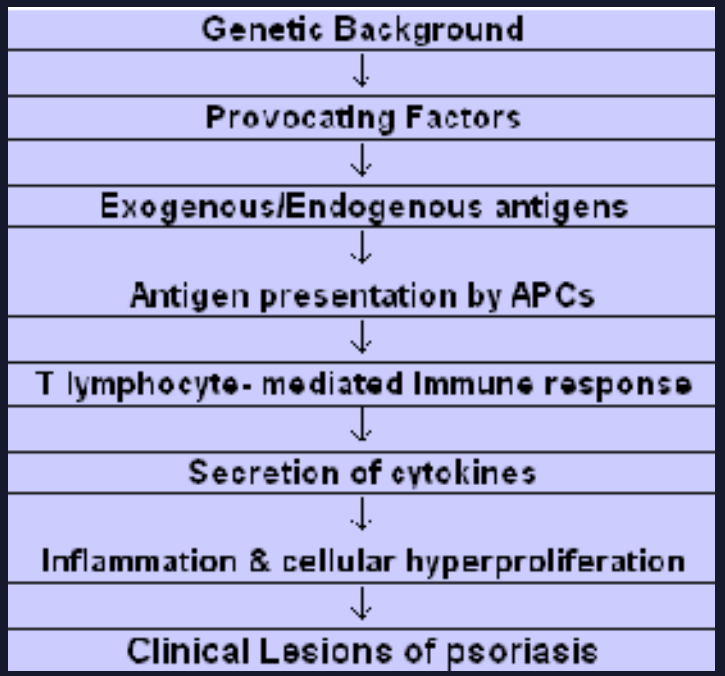
Aetiology and Pathogenesis

- **Sunlight** – although sunlight is generally considered to be beneficial for most of the patients, strong sunlight may worsen the disease in a small minority
- **Stress** - Many patients report an increase in the psoriasis severity with psychological stress
- **Smoking** - Cigarette smoking is associated with an increased risk of chronic plaque psoriasis
- **Alcohol** - Alcohol is considered a risk factor for psoriasis, particularly in young to middle-aged males
- **Endocrine** – the disease state may fluctuate with hormonal changes. Psoriasis may begin during puberty. Pregnancy may improve the disease. while a flare may occur during post-partum period

Aetiology and Pathogenesis

1. Role of immune response

   The epidermis and dermis of an active psoriatic plaque contain increased numbers of several different cells of the immune system, including activated T cells, activated antigen-presenting cells (APCs) (Langerhans cells, other dendritic cells and macrophages), neutrophils, and hyperproliferating keratinocytes



Pathology of Psoriasis

The pathology of psoriasis reflects the underlying immune-mediated inflammation and cellular hyperproliferation

- Hyperkeratosis with parakeratosis (presence of nucleated keratinocytes in the stratum corneum due lack of maturation of cells since rapid transit time do not permit normal maturation of cells)
- Reduced or absent granular layer
- Acanthosis with elongation of rete ridges and a corresponding upward elongation of dermal papillae
- Infiltrate: Mononuclear in dermis and polymorphs in the upper epidermis forming collections called ‘microabscess of Munro’
- Upper dermal vasculature shows dilatation

Clinical Features – History

- Family history is frequently elicited.
- First manifestation at any age but two peaks are observed: the first peak in the second decade, the second in the sixth decade
- Onset may follow trauma, infection, sunburn or psychological stress.
- Patients complain of prominent itchy, elevated red areas with increased scaling.
- Patients may recognize that new lesions appear at sites of injury to the skin (E*his isomorphic phenomenon (**Koebner reaction**) typically occurs 7-14 days after the skin has been injured and has been found in 40-80% of patients with psoriasis)*
- Joint pain, stiffness, and deformity are complaints in about10% of patients who have psoriatic arthritis.

Types of Presentations

- **Chronic plaque psoriasis**. The commonest type, presenting with typical plaques of psoriasis of the extensors and scalp.
- **Guttate psoriasis**. Is commoner in childhood. Acute eruption of drop-shaped lesions distributed widely over the body. Usually follows an upper respiratory infection.
- **Flexural psoriasis**: lesions are present over the flexors and intertriginous areas(axilla, groin, umbilical region, inframammary folds) the lesions may be moist and lack the typical scaling.
- **Pustular psoriasis**. may be **localized** or **generalized**. Localized pustular psoriasis usually presents with persistent pustular eruptions of the hands and feet.
  Generalized pustular psoriasis may occur as an explosive eruption of generalized pustules with systemic disturbances. This may follow withdrawal of systemic steroid therapy or application of irritants.

Types of Presentations

- **Arthropathic psoriasis** . Arthritis may accompany any variety of psoriasis in about ten per cent of patients . Psoriatic arthritis may take several forms. The commonest type is *asymmetrical oligoarthritis*, other types are: *symmetrical seronegative rheumatoid-like disease , distal interphalangeal* involvement(most characteristic, but relatively rare*), axial skeletal involvement*, and a destructive mutilating form *(arthritis mutilans*)
- **Erythrodermic psoriasis**. Psoriasis may present with erythroderma (exfoliative dermatitis). There is generalized inflammatory erythema with profuse scaling

Physical Exam

- The lesions are very **well marginated** with distinct border
- The lesions are **raised above the surface**
- The plaques usually have a diameter of one to several centimeters and have a **round or oval shape**.
- The plaques typically have a rich **red color** and may be surrounded by a pale halo (the ***halo of Woronoff***)
- The lesions are covered with a silvery white, mica-like, loosely adherent **scales** which, on removal may reveal punctate bleeding points (***Auspitz sign***)
- **Symmetry**: the lesions are symmetrically disposed on extensor surfaces of the body. Typical sites of affection are the elbows, knees, shin, knuckles, sacral areas and scalp.
- **Uniformity**: the psoriatic plaques tend to have the same features irrespective of site except for certain locations like the palms and soles, and the flexors.

Differential Diagnosis

•Seborrhoeic dermatitis •Lichen simplex •Pityriasis rubra pilaris •Leprosy •Candidiasis •Discoid lupus erythematosus •Cutaneous T-cell lymphoma	•Pityriasis rosea •Secondary syphilis •Lichen planus •Tinea corporis and cruris •Hyperkeratotic eczema of hands and feet •Parapsoriasis

Treatment

- Since psoriasis is a chronic disease, therapy is long-term. There are many treatment options available. Treatment regimens must be individualized according to age, sex, occupation, type and extent of disease and available resources
- Three main modalities of therapy are used: *topical, phototherapy* and *systemic agents*.
- These are used either alone or in combinations

Topical Therapy

- **Topical steroid** :Potent topical steroids, applied twice a day is effective in controlling limited plaque psoriasis. Intralesional injection of triamcinolone acetonide into isolated chronic plaques may also be used
- **Anthralin (Dithranol):** Derived from chrysarobin (active ingredient of Goa powder, derived from the bark of South American Araroba tree), it has antiproliferative and immunosuppressive action. It is best applied as short-contact therapy (washing off the medicine after a contact period of 10 to 30 minutes). Dithranol is irritant and can stain cloth , it causes a reversible brownish pigmentation of the treated skin
- **Vitamin D3 analogues: Calcitriol** and **Calcipotriol** are as effective as topical steroids and anthralin. Act by regulating keratinocyte proliferation and maturation. Side effects include irritation and hypercalcemia and kidney stones with high dose

Topical Therapy

- **Retinoids:** The synthetic retinoid **Tazarotene** when used topically, can regulate keratinocyte proliferation and maturation. Main side effect is irritation. Special precaution: women of child-bearing age.
- **Coal tar:** Tars have antiproliferative effect. May be combined with steoid therapy. Coal tar solution may be used for scalp psoriasis.
- **Salicylic acid:** As a two to ten per cent ointment, often combined with topical steroids, helps remove scales and also helps in penetration of steroids.

Phototherapy

- **UVB - Ultraviolet B (UVB) irradiation** utilizes ultraviolet radiation with wavelengths 290-320 nm (visible light range is 400-700 nm). It is used alone or combined with one or more topical treatments. The **Goeckerman regimen** uses coal tar followed by UVB exposure and the **Ingram method** is based on anthralin application following a tar bath and UVB treatment. UVB more commonly is now combined with topical corticosteroids, calcipotriene, tazarotene, or simply bland emollients. UVB phototherapy is extremely effective for treating moderate-to-severe plaque psoriasis. **Narrow-band UVB** (UVB of 311 nm wave length) is now increasingly used for its effectiveness and low potential for photodamage.

Phototherapy

- **PUVA photochemotherapy** - In this therapy (also known as PUVA), a photosensitizing drug methoxsalen (8-methoxypsoralens) is given orally, followed by ultraviolet A (UVA) irradiation to treat patients with more extensive disease. UVA irradiation utilizes light with wavelengths 320-400 nm. PUVA, decreases cellular proliferation by interfering with DNA synthesis, and also induces a localized immunosuppression by its action on T lymphocytes. Therapy usually is given 2-3 times per week on an outpatient basis, with maintenance treatments every 2-4 weeks until remission. Adverse effects of PUVA therapy include nausea, pruritus, and burning. Long-term complications include increased risks of photo damage and skin cancer.

Systemic Therapy

- **Methotrexate** is the most popular agent used. A weekly oral or intramuscular dose of 15 to 25 mg is used. Started with low dose and gradually increased to the desired dosage. 5mg 12 hourly X 3 doses per week is the usual oral regimen. CBC, platelets, LFT, urinalysis, and creatinine. must be normal before starting therapy. These tests are repeated at regular intervals to monitor toxicity. Main side effects are nausea, vomiting, hepatic dysfunction (even fibrosis on long term therapy), and microcytic anemia
- **Acitretin**: A synthetic vitamin A derivative. Is used in a dose of 10 to 20 mg/day gradually increased to the maximum dose of 50mg/day. Main side effects are : teratogenicity (the drug should not be used in women of child-bearing potential) , hypertriglyceridemia, xerosis of skin, chelitis and alopecia. It may be used in combination with phototherapy, methotrexate or cyclosporine

Systemic Therapy

- **Cyclosporine**: this drug selectively inhibits T-helper cell production of IL-2 and thus exerts an immunosuppressive effects. The usual dosage is 3 to 6mg/kg per day. A new microemulsion formulation may be used in a lower dosage. The commonest side effects are hypertension and nephrotoxicity
- **Mycophenolate mofetil**: still under evaluation, this newly introduced drug inhibits synthesis of the nucleotide guanosine. The recommended dosage is500mg four times a day with a maximum dose of 4 gm . side effects are: teratogenicity, neutropenia, GI symptoms, and opportunistic infections
- **Sulphasalazine** may be used to treat psoriasis either alone or combined with methotrexate, particularly in psoriatic arthropathy

Complications

- Complications are relatively uncommon
- Many of the complications (pustular psoriasis, erythroderma) are commonly due to inappropriate and aggressive therapy
- Psoriatic arthritis
- Pustular psoriasis
- Erythroderma and its metabolic complications
- Infection, particularly Staph. infections of the patches
- Eczematization due to topical agents
- Amyloidosis , rare sequel to arthropathic of pustular psoriasis
- Psychological consequences: depression, anxiety, lack of self-esteem
- Potential complications of systemic therapy should not be overlooked