DEFINITION

1. Psoriasis is a proliferative and inflammatory disease of the skin which affects people who are genetically predisposed. There is a tenfold increase in the speed of epidermal cell proliferation and nucleated cells are exfoliated before forming a horny layer. The characteristic lesions are chronic, sharply demarcated, dull red, scaly plaques, particularly on the extensor prominences and on the scalp. There is evidence that the clinically uninvolved skin of psoriatics is not normal. Psoriasis is enormously variable in its duration and extent and there are several morphological variants. The disease is very rarely fatal.

CLINICAL MANIFESTATIONS

2. Psoriasis affects about 2% of the world's population. It may arise at any age from early childhood onwards, with a tendency to arise at an earlier age in females or if there is a family history. It is generally chronic and persistent, without shortening life expectancy, so that its prevalence increases with age.

3. Its manifestations are extremely variable, with lesions which grow and regress. The condition may be acute, especially in children and adolescents, but more often it is chronic with exacerbation at varying intervals. Its course is unpredictable and its duration varies from a few weeks to a lifetime.

4. The most common location of psoriatic rash is on the scalp, which is affected in about 80% of cases. Otherwise, it tends to affect the extensor surfaces of the limbs, particularly the prominences, and the back.

5. There are several morphological variants, which may coexist:

    5.1. **Nummular psoriasis** (psoriasis vulgaris, common chronic stable plaque psoriasis). This is the commonest type. The lesions have a characteristic pattern. They are pink in colour and well defined with varying degrees of scaling and induration. The scales are usually silvery white and of variable thickness. On removal of the scales, small bleeding points are revealed in the underlying skin. This constitutes the Auspitz sign. There is considerable variation in the size and number of lesions. Demarcation is usually sharp, without gradual change to normal skin, but pale zones or haloes sometimes encircle the plaques. The outlines of large lesions are polycyclic, indicative of origin in several smaller units.

    5.2. **Guttate psoriasis.** This type occurs in children and young adults chiefly after acute streptococcal infections. The lesions are usually small and multiple and they are scattered over the trunk and limbs.

    5.3. **Elphantine psoriasis** (inveterate psoriasis). This term describes plaques with massively thickened horny layers.

5.4. **Rupioid psoriasis** (psoriasis ostracea). In this type, there is gross hyperkeratosis with limpet-like, cone shaped lesions which have horizontal rather than vertical lamellations.

5.5. **Unstable psoriasis.** This is a transition phase of chronic plaque psoriasis to a more extensive involvement, either erythrodermic or pustular psoriasis. It is regarded as a response to triggering factors in patients with a relatively low erythema threshold to other stimuli.

5.6. **Erythrodermic psoriasis.** Two forms exist:

5.6.1. The first form is a plaque psoriasis involving almost all the cutaneous surface. Chronic psoriasis may slowly transform into this type or it may occur as a result of sensitivity to treatment.

5.6.2. The second form is part of the spectrum of unstable psoriasis. It involves the whole skin and may occur acutely. It is commoner where there is arthropathy and can be precipitated by infections hypocalcaemia, or various topical therapies. There is severe systemic upset which may include hypo- or hyperthermia, electrolyte imbalance, dehydration, renal failure and high-output cardiac failure. The profuse scaling may lead to protein loss. There is an appreciable mortality.

5.7. **Pustular psoriasis.** Neutrophil accumulation in the epidermis occurs in all types of psoriasis but the term pustular psoriasis is used where macroscopic pustules appear. There are two main types of pustular psoriasis. In the **localised** form, the disease is chronic and confined to the hands and feet. The **generalised** form may involve the whole body and the course may be sub-acute, acute or even life-threatening.

5.7.1. **Localised pustular psoriasis**. The commonest variant is **chronic palmo-plantar pustulosis**, a predominantly adult condition with a female preponderance. Scaly, erythematous plaques studded with sterile pustules are found on the palms and/or soles. There is some evidence that this condition is a separate entity from nummular psoriasis, which coexists in less than 20% of cases. Also, there are genetic differences. Palmo-plantar pustular psoriasis may occur in an acute form.

5.7.2. **Acrodermatitis continua** is a rarer type of localised pustular psoriasis in which the lesions are located on the tips of the fingers and toes, sometimes with destruction of the nail plate and even osteolysis of the phalangeal tuft. It is extremely disabling.

5.7.3. **Generalised pustular psoriasis (GPP)**. This rare condition tends to occur in middle aged patients with common chronic psoriasis and may be precipitated by therapy, including treatment for the psoriasis. It is particularly seen in relation to corticosteroid therapy, when it can occur on withdrawal of either systemic steroids or potent topical steroids like clobetasol propionate. It may also arise from the use of other drugs including lithium, trazodone, salicylates, NSAID's, sulphonamides, progesterone, beta-blockers, amiodarone, anti-malarials and some anti-inflammatory drugs. Erythroderma occurs in about 60% of patients with GPP.

5.7.4. **Impetigo herpetiformis** is a form of GPP occurring in pregnancy. It has a predilection for flexural areas.

5.7.5. Infantile and juvenile pustular psoriasis are very rare.

6. Where psoriasis first presents in later life there may be a rapid spontaneous progression to the generalised pustular form. The condition may lead to exhaustion, toxicity or infection or there may be a remission within days or weeks.

7. The manifestations of psoriasis vary with its site. Scalp psoriasis has several patterns and may be associated with alopecia. A solitary patch may occur on the penis. Flexural psoriasis occurs in about 4% of patients. It may involve the axillae, groins, submammary region, retroauricular folds, perineum or vulva. In these sites scaling is not prominent and diagnosis may be difficult. Psoriasis on the palms and soles may be nummular or rupioid.

8. **Extracutaneous manifestations** of psoriasis can occur in several sites:

8.1. Psoriasis of the nails occurs in about 30% of psoriatic patients with skin lesions alone and in about 75% of cases where there is joint involvement. Both the nail bed and matrix are affected, resulting in pitting, discolouration, subungual hyperkeratosis and onycholysis. Splinter haemorrhage may occur. Psoriasis of the nails occurs equally in males and females and is commoner in older patients.

8.2. Mucosal lesions may occur in the pustular and exfoliative forms of the disease, usually in or around the mouth.

8.3. Ocular lesions may include conjunctivitis, iritis, blepharitis, and keratitis. These lesions tend to be active at the same time as those of the skin.

8.4. **Psoriatic arthritis** (psoriatic arthropathy, PsA) In about 8% of patients psoriasis is associated with peripheral and/or spinal arthropathy. This arthropathy is more common where the psoriasis is pustular or severe. The serological test for rheumatoid disease is negative.

8.4.1. The arthritis usually begins between the ages of 35 and 45, with a gradual onset in most cases but acute in about 30%. In most cases, it first affects the distal joints of the hands and feet and involvement of tendon sheaths may cause "sausage fingers". The large joints may also be affected and there is sacroiliac and/or spinal disease in 20-40% of patients.

8.4.2. Histologically, psoriatic arthritis resembles rheumatoid arthritis, but it tends to be less severe, with more frequent remissions and less joint destruction. Soft tissue nodules, vasculitis, pulmonary or renal involvement which may complicate RA are rare in PsA.

8.4.3. Gout is more common is psoriatics. Different investigators have found elevated uric acid levels in 30-50% of patients.

9. Psoriasis is a relatively common condition which may co-exist with other conditions including other skin disorders. There is an association with seborrhoeic dermatitis, lichen simplex and lichen planus. The exact significance of these associations is not known and they may well be chance findings.

10. Associations have also been reported with intestinal diseases including Crohn's disease, ulcerative colitis and malabsorption, and with occlusive vascular disease. Again the evidence is conflicting. Any association may not be with psoriasis per se but it may rather reflect some common genetic feature of the individual concerned.

11. The course and prognosis of psoriasis are unpredictable. Guttate forms carry the best chance of complete remission with no recurrence but for the most part psoriasis is a disease of relapse and remission.

12. A number of complications of psoriasis are recognised. They are uncommon and include secondary infection of lesions, a variable degree of itching and the exceedingly rare complications of renal or hepatic failure, atypical pulmonary fibrosis and amyloidosis.

13. Psoriatic patients who have been treated with high-dose PUVA or cyclosporin are at increased risk of developing squamous cell carcinomas. The same applies to those who have been treated with X-rays or arsenic, which are no longer used. In contrast, psoriatic patients who have not been so treated have a remarkably low incidence of sun-related cancers and solar keratosis.

AETIOLOGY

14. Psoriasis is a condition of unknown aetiology. However, both genetic and environmental factors are involved in the cause and course of the disease.

15. There is evidence from twin, family and population studies that psoriasis may be inherited. The exact mode of inheritance remains unclear, but it is probably multifactorial rather than due to a single gene. The HLA antigen system is linked to psoriasis. HLA-B13 and B17 are associated with psoriasis, but the strongest association is with HLA-Cw6. Individuals with this phenotype have more than ten times the normal risk of developing psoriasis. No other disease is known to have a primary link with HLA-C. On the other hand, in some cases of psoriasis there is no recognisable familial component and there is less than 100% concordance in monozygotic twins, indicating that there must be an environmental contribution.

16. Racial and ethnic variation in the prevalence of psoriasis is evidence of a major genetic factor in its aetiology. For example, compared with Western Europeans, it is low in oriental people, rare in pure native Americans and undetectable in a survey of 25,000 Latin American Indians. In Singapore, psoriasis is more common in Indians than in Chinese or Malays. It is more common in Kenyans and Ugandans than in West Africans. The prevalence in Norwegians is nearly 5%, but only about 0.6% in pure Norwegian Lapps, whose HLA patterns resemble those of Mongolians.

17. In genetically predisposed individuals, external factors may initiate psoriasis or exacerbate pre-existing disease. Such factors include the following:

    17.1. **Trauma**. The occurrence of psoriasis at the site of an injury is well recognised. The trauma may be physical, chemical, mechanical, electrical or surgical and psoriasis is produced in previously normal skin. This reaction, which is not exclusive to psoriasis, is called the Koebner phenomenon. The reaction occurs 7-14 days after the injury and its frequency is between 50 and 75%. A reverse Koebner reaction can also occur, with clearing of psoriasis following injury. These two phenomena are mutually exclusive.

    17.2. **Infection**

        17.2.1. Acute guttate psoriasis is known to be provoked by streptococcal infection, especially in the throat, and there is evidence that streptococcal infection may have a role in some cases of chronic plaque psoriasis. Psoriatic patients are not at increased risk for bacterial infections, although lymphangitis and lymphadenitis may occur with palmoplantar pustulosis.

        17.2.2. There is an association between severe psoriasis, psoriatic arthropathy and human immunodeficiency virus (HIV) infection. In individuals with HIV, psoriasis may flare severely or appear *de novo* in an explosive form. The prognosis of patients with AIDS is worsened in the presence of psoriasis.

17.3. **Endocrine factors**. The incidence of psoriasis peaks at puberty and the menopause. In 50% of pregnancies in patients with psoriasis, the disease worsens in the post-partum period. GPP may be provoked by pregnancy or high-dose oestrogen therapy and tends to exacerbate premenstrually.

17.4. **Sunlight**. The majority of psoriatics are improved by sunlight but in a small minority exposure to the sun triggers a Koebner reaction. Photosensitivity in psoriasis is generally associated with skin type 1, advanced age and female gender.

17.5. **Therapeutic Drugs**. Treatments for a variety of conditions may induce or exacerbate psoriasis. The substances most often implicated include lithium, the beta-adrenergic blockers and anti-malarials. Morphine, used therapeutically or otherwise, has also been implicated. Withdrawal of either systemic or the more potent topical corticosteroids may cause psoriasis to rebound.

17.6. **Smoking and alcohol**. There is no proof that alcohol consumption directly exacerbates psoriasis. Psoriatic patients who smoke are at increased risk of developing palmo-plantar pustulosis.

17.7. **Psychogenic factors**. Stress does not cause psoriasis but it may be associated with clinical deterioration of established psoriasis. Studies have been confounded by the fact that the disease itself is a potent stressor.

17.8. **Metabolic factors**. Renal dialysis and low calcium levels (e.g. following parathyroidectomy) can precipitate psoriasis.

CONCLUSION

18. **Psoriasis** is a disease of the skin which results from a genetic predisposition. It is probably a systemic disorder of the immune system capable of provoking activation and disordered growth of certain skin cells. Factors which may precipitate the onset of the condition, provoke an attack or worsen the condition in an individual who is already affected have been described above. In most cases, the onset or exacerbation of psoriasis occurs without apparent antecedent cause.

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