Generalised Pustular Psoriasis

Synonyms: acute generalised pustular psoriasis of von Zumbusch, pustular psoriasis von Zumbusch variant

This is a generalised eruptive form of psoriasis accompanied by fever and toxicity. Acute erythema is seen with a rapid spread of multiple sterile pustules over the body, concentrated in the flexures, genital regions and fingertips. It is a medical emergency that requires urgent hospital referral.

Generalised pustular psoriasis needs to be distinguished from a localised form of pustular psoriasis known as palmoplantar pustulosis (PPP).

Epidemiology

It is a rare condition: precise prevalence figures for the UK are not available. Prevalence is 7 per 1 million in Japan.[1] The average age of those affected is 50 but the range is wide.

Recognised triggers[2, 3]
- Systemic steroid withdrawal.
- Drugs: salicylates, iodine, lithium, phenylbutazone, oxypHENbutazone, trazodone, penicillin, hydroxychloroquine, calcipotriol, interferon alfa and recombinant interferon-beta injection.
- Topical medications causing irritation: coal tar, anthralin, steroids under occlusive dressing and zinc pyrithione in shampoos.
- Infections.
- Sunlight or phototherapy.
- Cholestatic jaundice.
- Hypocalcaemia.

No cause is identified in many patients.

Presentation[4, 5]
- The skin becomes very red and tender.
- Clusters of pustules appear within hours. Multiple, small (2-3 mm), yellowish pustules develop on an erythematous background and may spread rapidly to a generalised pattern. Over about 24 hours, the pustules coalesce, dry and desquamate in sheets. Further crops of pustules may appear over days to week, leaving the patient uncomfortable and exhausted.
- The flexural and anogenital regions are most commonly affected. Facial lesions are less common.
- Pustules may occur on the tongue and under the nails, causing dysphagia and loss of nails respectively.
- Patients are usually systemically unwell with symptoms including headache, fever, chills, arthralgia, malaise, anorexia and nausea. On examination, there is maybe a fast respiratory rate, tachycardia and pyrexia suggesting a toxic state. The tongue is dry and cracked.
- As the pustules remit, most systemic symptoms ebb but the patient can be left with erythroderma or with residual lesions of ordinary psoriasis.

While children and infants can develop a generalised pustular psoriasis of the adult form, a circinate or annular type is more usual, which tends to run a subacute/chronic course and to be more benign:
- There are recurrent episodes of annular or circinate erythematous plaques, with pustules on the periphery.[6]
- They are mostly over the trunk but also involve the extremities.
- They expand peripherally whilst healing in the centre.
- Systemic symptoms are mild or absent.
- Spontaneous remissions are frequent.

Differential diagnosis
- Drug eruptions
- Sepsis

Investigations
- FBC - typical raised white cell count with a marked rise in neutrophils and fall in lymphocytes.
- Elevated inflammatory markers.
- Hypoalbuminaemia.
- Abnormal LFTs. [7]
Abnormal renal function parameters due to acute kidney injury or tubular necrosis.

No microbial growth from cultures of pustules and blood.

**Management**[5, 8]

Admit to hospital urgently, as this can be a life-threatening condition:

- Intensive nursing and supportive therapy with attention to hydration, fluid balance, nutrition and temperature regulation.
- Bland topical compresses and saline or oatmeal baths assist in soothing and debriding affected areas.
- There is a very limited evidence base as regards the efficacy of different systemic therapies separately or in combination, partly due to the rarity of the disease. Treatment can be challenging, with problematic side-effects or lack of response. Indeed, case reports have implicated some of the agents used to treat generalised pustular psoriasis with aggravating/inducing the condition - for example:
  - Ciclosporin,[9]
  - Etanercept and infliximab.[10]

- Specific medications include:[8, 11]
  - Oral retinoids, usually acitretin.
  - Systemic corticosteroids,
  - Methotrexate.
  - Ciclosporin.
  - Colchicine.
  - Biological therapies - eg, infliximab, etanercept, ustekinumab, adalimumab and anakinra.

- Psoralen combined with ultraviolet A (UVA) treatment (PUVA) cannot normally be tolerated in the early stage, except perhaps in children who tend to experience a milder disease. Once the patient is stable, usually after several days of acitretin, PUVA may be commenced.

**Complications**[3, 12]

- Secondary bacterial infection of the skin may occur.
- Hair loss with telogen effluvium is possible.
- With subungual involvement, nails will be lost.
- Hypoaalbuminaemia may result from loss of plasma protein into tissues.
- Reduced circulating volume may cause renal tubular necrosis.
- Poor circulation and general toxicity may cause liver damage.
- Malabsorption and malnutrition.
- Death due to cardiac failure, sepsis or acute respiratory distress syndrome.

**Prognosis**

- Older patients with the von Zumbusch-type variant have a poorer prognosis.
- Children tend to have a good prognosis unless there is secondary infection.
- Prognosis is better where the pustular psoriasis was preceded by ordinary chronic psoriasis.

**Further reading & references**

- Generalised Pustular Psoriasis, von Zumbusch Type; DermIS (Dermatology Information System)
- The Psoriasis Association
- Acute forms of psoriasis; DermNet NZ
- Psoriasis: Signs and symptoms; American Academy of Dermatology
- Pustular Psoriasis; DermNet NZ

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