



seen. Linear IgA deposition is found in the epidermal basement membranes. C3 may also deposit in some cases.

### Laboratory findings, Diagnosis

Linear IgA deposition on the epidermal basement membranes can be identified by direct immunofluorescence (IF). Anti-epidermal basement membrane IgA autoantibodies may be detected in the patient's serum by indirect IF.

### Differential diagnosis

LAD should be differentiated from dermatitis herpetiformis. In LAD, ① there is a histopathological finding of linear patterns of in vivo IgA deposition, ② there are anti-basement membrane IgA auto-antibodies in serum in some cases, ③ there is no involvement of HLA-B8, DR3, or DQ2, ④ there is involvement of the mucosa, and ⑤ there is no sensitivity to gluten.

### Treatment

DDS is effective, as are oral steroids.

## Pustular diseases

### 1. Palmoplantar pustulosis (PPP) ★ ★

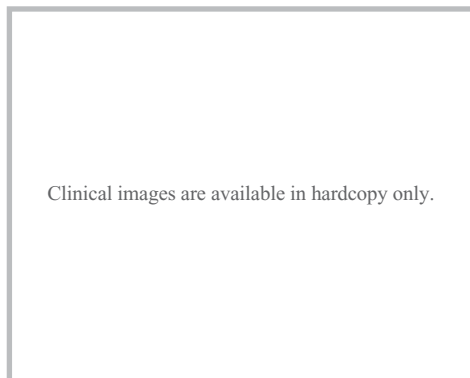
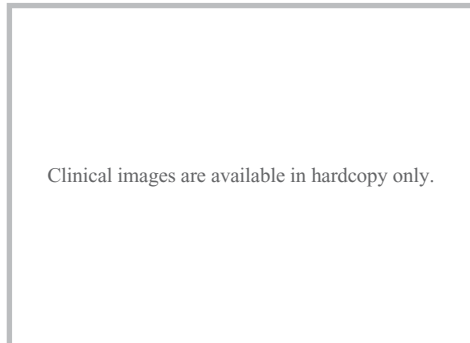
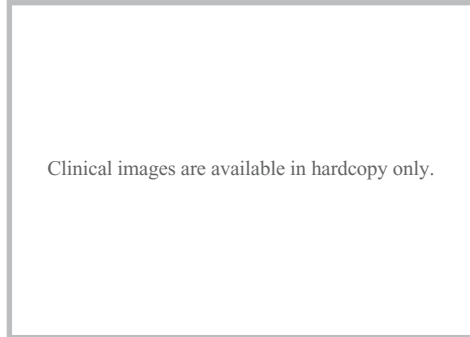
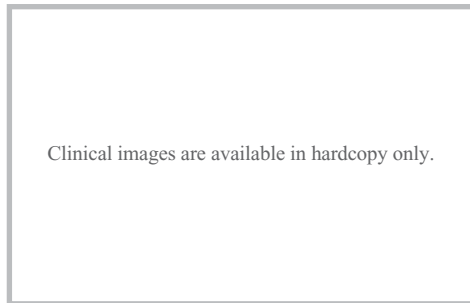
Synonym: Pustulosis palmaris et pustulosis

#### Outline

- Multiple sterile pustules form symmetrically on the palms and soles of the middle-aged and elderly, becoming chronic.
- Smoking, bacterial infection (tonsillitis), dental caries and dental metal allergy are associated with occurrence of PPP.
- Sternocostoclavicular ossification and pain may develop as complications.
- Topical steroid application, smoking cessation and tonsillectomy are the main treatments.

#### Clinical features

Multiple vesicles occur on the thenar and antithenar regions of the palms and arches of feet, and these become pustular. Erythema develops at the periphery of the lesions and fuses into plaques (**Fig. 14.35**). Itching may be present. Punctate depressions and thickening occur frequently in the nails. Pustules recur in 2- to 4-week cycles and progress chronically. They may appear on the knees, lower extremities and scalp. In 10% of palmoplantar pustulosis (PPP) cases, sternocostoclavicular ossification accompanied by chest pain develops as a complication.



**Fig. 14.35 Palmoplantar pustulosis (PPP).**  
Multiple pustules aggregate on the hands and feet.

### Pathogenesis

PPP patients are mainly female and tend to have smoked at least a pack a day for more than 20 years. If focal infections (e.g., tonsillitis, dental caries) are found and treated, PPP may improve. Cases induced by bacterial allergic reaction and metal allergy have been reported. It is hypothesized that PPP is a localized type of pustular psoriasis.

### Pathology

Sterile pustules in the epidermis contain neutrophils and degenerated epidermal cells. Mild cellular infiltration into the dermis is seen.

### Laboratory findings

To detect focal infections such as tonsillitis and dental caries, the white blood cell count, antistreptolysin O, CRP and sedimentation rate are examined by peripheral blood tests. In some cases, skin lesions are aggravated by a tonsil provocation test. For confirmation of dental metal allergy in the mouth, metal patch tests are conducted. Tests for detection of arthropathy and ossification (sternocostoclavicular) are also conducted.

### Differential diagnosis

The disease should be distinguished from tinea manus, pustular psoriasis, contact dermatitis, pompholyx and Reiter disease.

### Treatment

In most long-term smokers, the disorder disappears with cessation of smoking. Focal infectious diseases, if any, are treated. Prevention of tonsillitis and otological and dental treatments are important. Oral antibiotics are useful. Tonsillectomy may be performed. For skin lesions, topical steroids are the first line of treatment; additionally, vitamin D3 ointment and PUVA therapy are applied. Methotrexate, colchicin and etretinate are administered orally in severe cases.

## 2. Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) ★

### Clinical features

Subcorneal pustular dermatosis occurs infrequently but most commonly in women over age 40. Erythema and pustules appear in a ring-shaped or serpiginous pattern on the trunk and on sites exposed to friction. Pustules quickly dry, leaving crusts and scales (frilly scales) (Fig. 14.36). Itching or systemic symptoms are not present, and the mucosa is not involved. The disease becomes chronic with recurrent aggravation and remission.

### Pathogenesis, Pathology, Differential diagnosis

The etiology of subcorneal pustular dermatosis is unknown in

many cases. In some cases, IgA myeloma or ulcerative colitis exists as a complication. Sterile pustules mainly containing neutrophils are found histopathologically under the horny cell layer. Spongiform pustules, seen in pustular psoriasis, are not found.

Subcorneal pustular dermatosis is distinguished from intracellular IgA dermatosis by immunofluorescence. IgA class autoantibodies are not detected immunohistologically in the epidermal intercellular space.

### Treatment

DDS and oral steroids are effective. Etretinate (a vitamin A derivative), and PUVA are effective in some cases.

## 3. Eosinophilic pustular folliculitis ★

Synonyms: Ofuji's disease, Eosinophilic pustular dermatosis

### Outline

- Itching papules and pustules are produced. They aggregate, mainly in hair follicles.
- Male adults are the most commonly affected. Progress is chronic, with recurrences and remissions. The etiology is unknown.
- Multiple eosinophils are contained in the pustules.
- The disease may accompany HIV infection.
- Indomethacin is effective as a treatment.

### Clinical features, Pathogenesis

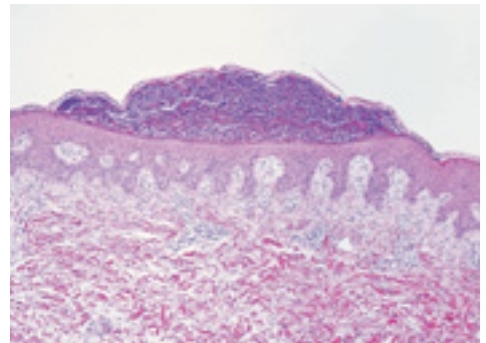
Eosinophilic pustular folliculitis occurs most frequently in men in their 20s or 30s. Sterile, follicular and itching papules or small pustules occur and aggregate to form erythematous plaques that spread centrifugally (**Fig. 14.37**). The face, upper body and extensor surface of the upper arms are the most severely affected regions; in some cases, however, the eruptions can occur in the hands and soles, where hair follicles do not exist. They leave abnormal pigmentation when they heal. Eosinophilic pustular folliculitis recurs with remissions. The disease may accompany a hematologic disorders or HIV infection. Most cases have been reported from Japan. The cause is unknown.

### Pathology, Laboratory findings, Differential diagnosis, Treatment

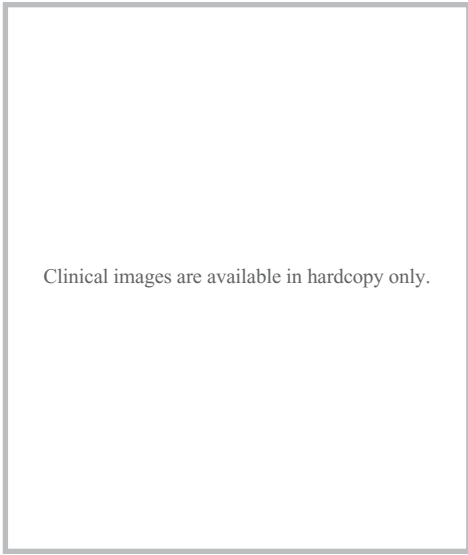
Eosinophils are found in large quantities in pustular components. Eosinophilic infiltration into the hair follicles and hair apparatuses results in destruction of hair follicles. Elevated levels of eosinophils are seen in the peripheral blood. Eosinophilic pustular folliculitis needs to be differentiated from tinea, candidiasis, folliculitis, acne, rosacea and contact dermatitis. When it occurs on the hands and soles, differentiation from pustulosis palmaris et plantaris is difficult. Indomethacin is very effective.

Clinical images are available in hardcopy only.

Clinical images are available in hardcopy only.



**Fig. 14.36** Subcorneal pustular dermatosis.



**Fig. 14.37 Eosinophilic pustular folliculitis.**  
Follicular papules and pustules aggregate, accompanied by itching.

#### 4. Acute generalized pustular bacterid

The appearance of acute sterile pustules on the trunk and extremities is followed by upper respiratory infection. Since the pathogenetic mechanisms have not been clarified, there is a controversy over whether acute generalized pustular bacterid is an independent clinical entity.

#### 5. Infantile acropustulosis

The extremities of infants are affected. Infantile acropustulosis is a recurrent pustular disease that causes multiple sterile pustules and vesicles with intense pustular psoriasis.

Refer to Chapter 15 for pustular psoriasis.