

PUSTULOSIS PALMARIS ET PLANTARIS

Prevalence, Clinical Observations and Prognosis

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Abstract. 85 outpatients with pustulosis palmaris et plantaris, 70 women and 15 men, have been studied from anamnestic and clinical aspects. 78 (92%) patients were followed-up after 5 years for a prognostic study. Patients with psoriasis present before the onset of the pustular eruption were excluded from the study. The prevalence in total populations of 12 669 persons in two geographical areas of Sweden is 0.05%. The incidence at the Outpatient Department of Dermatology in Gothenburg is 0.37%. In the outpatient group, 82% were women. The age of onset is mostly between 40 and 59 years of age. The lesions often started unilaterally on the palm or sole, and later spread to both hands and feet. The course is protracted in spite of traditional topical therapy, as 75% of the patients still had pustular lesions 5 years after their first consultation. Elimination of infectious foci played little part in producing a permanent cure. Every sixth patient had to be hospitalized for treatment, and 5% had to change occupation because of the persistent skin lesions.

Pustular eruptions of the palms and soles are often resistant to treatment and are highly incapacitating to the patient. Many disseminated dermatoses have these symptoms but in this study only those conditions will be dealt with where the skin lesions on the palms and soles are the only skin changes present. In this group of patients those with acrodermatitis continua (Hallopeau) can often be recognized because of its acral suppurative skin lesions and loss of nails, histopathologically characteristic spongiform pustules and the risk of a transition, sooner or later, to generalized pustular psoriasis.

After the exclusion of patients with acrodermatitis continua, the remaining cases with chronic palmar and plantar pustules have much in common: clinically, a close resemblance to each other; histopathologically, intraepidermal absces-

ses; cultures are negative for bacteria, virus or fungi, and the long-term prognosis for restitution is in many cases poor. Attempts to relate them to common dermatoses (i.e. psoriasis) or to delineate different syndromes among them have been done reflecting different opinions of etiology. Pustular psoriasis of the extremities was suggested by Barber (4) and Ingram (9, 10) who considered that psoriatics are more susceptible to this pustular reaction of the hands and feet. Pustular bacterid of Andrews (2) underlines the importance of focal sepsis, often in tonsils and teeth. Removal of the focus should be followed by a permanent cure of the skin disease. However, the often chronic nature of the disease has initiated diagnostic terms as "intractable pustular eruptions of the hands or feet" (5) and "relapsing pustular eruptions of the hands and feet" (3). Pustulosis palmaris et plantaris (PPP) (11) is a name which has the advantage of being only descriptive. It will also be used by us. It thus includes all chronic, sterile pustular eruptions of the hands and feet with the exclusion of acrodermatitis continua. A survey of suggested diagnostic terms is presented in Table I.

Some writers have published large series of PPP cases. Among the more recent reports may be mentioned those published by Everall (5), Ingram (10), Veltman & Schuermann (16) and Ashurst (3).

As long as the discussion of the etiology and treatment of PPP persists, however, we believe that further studies of clinical material may lead to a closer understanding of the disease and may possibly furnish clues concerning its pathogenesis. The prognosis for permanent cure is also insuf-

Table I. *Suggested diagnostic names in chronic, sterile palmo-plantar pustulosis*

Acrodermatitis (Dore, 1928)
Pustular psoriasis of the extremities (Barber, Ingram, 1930)
Pustular bacterid (Andrews, 1934)
Pustulosis palmo-plantaris (Bonnievie, 1939)
Acrodermatitis pustulosa perstans (Sachs et al., 1947)
Intractable pustular eruptions of the hands and feet (Everall, 1957)
Pustulosis palmaris et plantaris (Lever, 1961)
Relapsing pustular eruptions of the hands and feet (Ashurst, 1964)

ficiently investigated. The aim of the present investigation was to study anamnestic and laboratory data in a clinical material of PPP. Of particular interest was the prognosis for cure and the relation between PPP and psoriasis.

Definition of pustulosis palmaris et plantaris

PPP is defined as a chronic eruption of yellowish sterile pustules occurring especially on the thenar and hypothenar eminences, and the sole or sides of the heel. The skin of the affected areas is shiny and reddened. The pustules dry up to form brownish scabs, which are gradually exfoliated. Typical psoriatic lesions are absent.

MATERIAL

This study is based on outpatients with a diagnosis of PPP, treated at the Department of Dermatology in Gothenburg, Sweden, during 1964-65. The patients selected for this study were only those in which there were no questions as to the accuracy of the diagnosis. As we have a clinical impression that there exists an acute form of PPP, sometimes combined with pustules on the extremities and/or the trunk, which starts after an upper respiratory infection and usually disappears quickly, only patients with a duration of PPP for 4 months or more were accepted in this study. Patients who earlier or on their first visit to the hospital suffered from Reiter's disease or had patches of psoriasis (vulgaris, pustulosa) or impetigo herpetiformis on the rest of the body were not included in the study. After a survey of the diagnostic cards, 85 patients remained who did fulfil these criteria (70 women and 15 men).

Five years after their first visit to the Department the selected patients received by post questions contained in a prepared form schedule. Replies were received from 78 patients (64 women and 14 men). An analysis of their replies to the questionnaire and their clinical data is presented in the following.

Forty of the patients with PPP and forty healthy controls matched as to sex, age and time of examination underwent laboratory tests at the routine laboratory of the

hospital. The statistical tests used in the comparisons were the Sign-test and Student's *t*-test (level of significance 0.05).

RESULTS

A. Clinical and Anamnestic Data

Incidence. The incidence of PPP in relation to other skin diseases, calculated from the outpatients' registers at the Department of Dermatology in Gothenburg, was 0.22% for males, and 0.50% for females, in total 0.37%. For comparison, the incidence of psoriasis at the same Department is about 3% for women and 3% for men (6). Data from the literature are sparse. Ingram (9), however, reports that out of 25 000 consecutive patients with skin diseases seen in private practice, 6.5% had psoriasis and 0.53% had persistent pustular eruptions of the extremities.

Prevalence. The prevalence of PPP in total populations of 12 669 persons, 6 184 males and 6 485 females, in the County of Kristianstad, Sweden, was 0.050% (8). This figure can be compared with the calculated prevalence of psoriasis in Sweden, 1.9% (7). Agrup (1) in a survey of hand dermatoses in 107 206 persons in south Sweden, found 27 cases of PPP in 1 659 persons who reported that they suffered from skin diseases on their hands and accepted a free examination (prevalence 0.027%). In the Faroe Islands the prevalence is 0.01% (13).

Sex distribution of PPP in the present series was 82% women and 18% men (females/males = 4.6). The prevalence was 0% among 6 184 males and 0.092% among 6 485 females in a population study (8). Ashurst (3) in a clinical study of 43 cases, found females to be affected nearly eight times more frequently than males. Agrup (1) reports the female/male ratio to be 1.2 in her field study of hand dermatoses. In psoriasis, the sex prevalence in Sweden is 2.3% in males and 1.5% in females (7).

Age at onset. The age at onset of 78 patients with PPP was: 20-29 years, 10; 30-39 years, 10; 40-49 years, 18; 50-59 years, 27; 60-69 years, 8; 70-79 years, 5. Ingram (10) reports the average age of onset to be 40-50 years, and in Ashurst's series (3) it was 31-50 years of age. In psoriasis, the onset shows a maximum for men at 10-24 and for women 10-19 years of age (6).

Duration of the skin lesions at the first time

of investigation. 4-6 months, 12; 7-24 months, 33; 2-4 years, 16; 5-9 years, 14; 10-14 years, 2; more than 15 years, 1 person.

Localization at onset. The initial localization of PPP at onset is shown in Table II. Hellgren (6) in a study of 194 psoriatics found the primary lesion to be localized to the hands in 6.2%. In no case did psoriasis start on the feet.

Maximum distribution of lesions in 78 patients with PPP. Palms only, 9; soles only, 21; palms and soles, 48 persons. The right and left sides of extremities are affected with the same frequency.

Heredity

1. *Of palmoplantar pustulosis.* Two patients (2.5%) declared that a father and a brother seemed to suffer from the same skin disease as they did. However, no further data about these two men have been available and a definite diagnosis of PPP is impossible.

2. *Psoriasis.* Among the close blood relatives (parents, brothers and sisters, children) of 6 patients (7.7%) there was a history of psoriasis, verified by doctors. In psoriasis, every third patient can be expected to have a relative with psoriasis (6).

3. *Allergic diseases.* The patients were asked if any of their blood relatives suffered from asthma, urticaria, eczema or hay fever. There were no significant differences between the PPP patients and a group of matched healthy controls.

Other factors

At the onset of PPP 9 of 78 persons (11.5%) were aware of upper respiratory infections, infections of the teeth or urinary organs, and 7 (9%) stated that the eruption followed a skin injury or local infection. None had been vaccinated or exposed to intense sunlight the month before the outbreak of skin lesions. Three women declared that the onset was within 3 months of parturition.

The season of onset was for 28% of the patients spring (March, April, May), 24% summer (June, July, August), 33% autumn (September, October, November) and 15% winter (December, January, February). 20% of the patients were regularly better during summer and 10% during winter, while 6% regularly got worse during sum-

Table II. *Localization at onset of pustulosis palmaris et plantaris in 78 patients*

	Palm(s)	Sole(s)	Palm and sole
Unilateral	22	25	
Bilateral	12	10	9
	34	35	9

Table III. *Focal infection in 26 investigated patients with pustulosis palmaris et plantaris*

	Patients no.	Cured	Still affected
Patients with focal infection			
Focus removed	9	3	6
Focus not removed	2	1	1
Patients with no focal infection	15	4	11

mer and 10% in winter. 60% reported that the season of the year did not influence the severity of the skin lesions. 27 patients had travelled in countries around the Mediterranean but only 7 of them could enjoy a temporary remission in the warm climate.

Soap irritated the lesions in 10% of the patients. Menstruation had an adverse effect in 6 out of 30 women (20%) in the fertile ages. Six patients (13%) had noted that acute infections were followed by exacerbations of the skin lesions.

Therapy and prognosis for restitution

The instituted treatment consisted of potent corticosteroids in ointments (mostly applied under occlusive dressings) and intralesional injections, tar, Bucky radiation therapy, and systemic short-term administration of antibiotics or sulphonamides. In one-third of the patients an extensive physical, laboratory and radiological investigation was performed in the search of an asymptomatic infectious focus (Table III). In about every second of these thoroughly investigated middle-aged or old patients a suspect focus of infection could be demonstrated, mostly in or around the teeth. The proper treatment of these foci coincided only in a few patients with a permanent cure of the skin lesions. The cured patients did not differ significantly from the rest of the patients in any way. The result is in accordance with the opinion of many dermatologists nowadays, that in PPP re-

removal of an infectious focus plays little or no part in producing a permanent cure (5, 10, 12). This opinion is, however, not generally accepted (16).

The 5-year follow-up showed 59 patients (75%) still to have active pustular lesions of the hands and feet. 35 of them had permanent lesions, and in 24 cases the lesions were episodic. Three-fourths of our 78 cases with PPP were thus still sick 5 years after their first visit to the Department. This figure can be expected to rise as some of them who were free of symptoms at the time of the follow-up may only have a temporary remission. In fact, remissions lasting 5 years have been reported (5).

29 patients (36%), 10 men and 19 women, had been unable to work on one or several (8 patients) occasions because of the skin lesions. Four patients (5%), 2 men and 2 women, had been forced to change occupation because of the socially crippling palmo-plantar lesions. Thirteen patients (17%), 2 men and 11 women, had been hospitalized with extensive, painful and resistant lesions, or because of secondary bacterial contamination with lymphangitis and fever.

None had developed psoriatic lesions on the rest of the body during the 5 years following the first visit to the department.

B. Laboratory Tests

The following laboratory tests were performed in some 40 patients with PPP and in 40 matched controls: determinations of the haemoglobin level, red and white blood counts, neutrophilic, eosinophilic and basophilic leucocytes, monocytes, lymphocytes and erythrocyte sedimentation rate. All tests showed normal values in comparison with matched controls. According to Piérard (14) and Veltman & Schuermann (16) there is often a leucocytosis during exacerbations of PPP but we found no significant difference between the patients and the healthy controls.

Antistreptolysin titres (AST) were elevated in 6 of 28 patients, antistaphylolysin titres (ASTA) in 1 of 28.

In roentgenographic examinations pathological infectious lesions in the pulmonaris were found in 1 of 31 patients, inflammation in the accessory nasal sinuses in 2 of 26 patients and abscesses at the teeth roots in 8 of 26 patients.

COMMENT

As can be seen from the prevalence figures, PPP is not a common disease in the population. Consultations with patients having PPP are, however, not uncommon to dermatologists (and often patients consult several) owing to its poor prognosis for restitution. Traditional therapy has an unsatisfactory effect on the disease. This is illustrated by the fact that 75% of the patients still have their skin lesions 5 years after their first visit to the Department of Dermatology. Complete blood analysis and radiological investigations sometimes demonstrate a focus of infection, but its elimination does not apparently lead to a quicker cure of the skin lesions than if these investigations and treatments not had been done. This is in accordance with Everall's (5) results in 70 patients with PPP. Of course this does not exclude the possibility that an infectious focus in the body can initiate a pustular, sterile skin eruption but when the dermatosis has, for some reason or other, manifested itself, then the eradication of the infectious focus will not bring a cure of the dermatosis.

The persistence of the skin lesions and their unfortunate localization will also lead to social complications. Every third patient has sometime had to be on the sick-list, and every sixth was hospitalized for treatment. 5% of the patients with PPP had to change occupation because of wet and dirty or restaurant or service jobs.

These discouraging results will probably not improve until more is known about the etiology of the syndrome.

Patients with PPP differ from psoriatics in some respects in this study. They have a lower frequency of psoriasis among their blood relatives, reversed male/female ratio, considerably higher age of onset and as a rule they have no seasonal variations typical of psoriasis. Histopathologically, the spongiform pustules of pustular psoriasis are not seen in PPP (11, 14, 15). None of the patients with PPP in this study had developed psoriasis within 5 years of their first consultation, but the time of observation is comparatively short.

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