

terone and estradiol did not worsen in the postpartum phase.

Our patient developed PAN during pregnancy, but in a cutaneous form. This is a distinct clinical entity (5) usually limited to the medium-size arteries in the deep dermis and panniculus. It is accompanied (2) by pain, myalgia and peripheral neuritis of the lower extremities. Compared with the systemic form it has a benign course and this may explain the maternal survival in the 3 cases of cutaneous PAN.

In our case, the infant survived, as in 6 of the 7 cases where pregnancy could proceed to full term. The only infant death (9) was related to placental arteritis and not to infant vasculitis. Thus the pathological process in pregnant women with PAN is apparently not transmitted to the fetus *in utero*.

Conversely, of the 28 reported cases of PAN in infancy (6) only 3 began shortly after the birth (10 days, 25 days (10), 6 weeks (7) with symptoms since the birth), which may suggest an *in utero* involvement. In these cases no maternal illness or complication of pregnancy was reported. Furthermore, of the 28 cases, only one maternal connective tissue disease (rheumatoid arthritis) was reported (1).

The fetus resistance to PAN in pregnancy may be explained by the fact that the fetus cannot synthesize antibodies until after 4 weeks of *ex utero* life (8). Further follow-up of the infants was not reported. In our case no sign of vasculitis could be detected in the child throughout a 4-year period of regular follow-up.

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Palmoplantar Pustulosis: A Neurocutaneous Disease?

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Abstract. A case of palmoplantar pustulosis in a 74-year-old woman is presented. The clinical appearance suggests a neurocutaneous pathogenesis.

Key words: Palmoplantar pustulosis; Neurocutaneous disease

Palmoplantar pustulosis (PPP) is a chronic and persistent eruption of yellowish sterile pustules occurring predominantly and most often symmetrically on the thenar or hypothenar eminence and the soles or sides of the heel. Occasionally the pustules are more diffusely scattered over the palms or soles (1).

The aetiology is still undetermined (1, 4). The present case suggests that PPP is a neurocutaneous disease.

CASE REPORT

A 74-year-old woman was referred with PPP. A brother had psoriasis. The right hand had been amputated at the age of 4 because of complications after an antebrachium fracture.

Since 1978 the patient had been treated for PPP with local steroids, salicylic ointment, Bucky and K_2MnO_4 baths without effect. Examination revealed a crusting, hyperkeratotic element localized to the hypothenar of the left hand (Fig. 1). On the right antebrachium, crusting and hyperkeratotic elements were found at the distal end (Fig. 2). The soles showed numerous yellow and brown pustules and crusting. Fig. 3 shows the therapeutic situation after 5 weeks of treatment with 25 mg aromatic retinoid (Ro 10-9359).



Fig. 1. Hyperkeratotic elements on the hypothenar of the left hand.

DISCUSSION

The present case suggests that PPP is a neurocutaneous disease, since the patient had a crusting, hyperkeratotic eruption on the amputated arm.

The symmetry of the disease is distinctive (4). A neurocutaneous pathogenesis could explain the eruption seen in this case and the symmetry generally observed.

Abnormal function of the granulocytes has been reported in PPP as well as in other chronic diseases (2, 3). Recently, increased prevalence of antithyroid antibodies and thyroid diseases in PPP has been described (5). Tetracycline treatment improved the lesions of PPP in double-blind cross-over investigations (7, 9).

One could speculate that in some cases infection could provoke an automimmune response where the autoimmune reaction is directed partly against

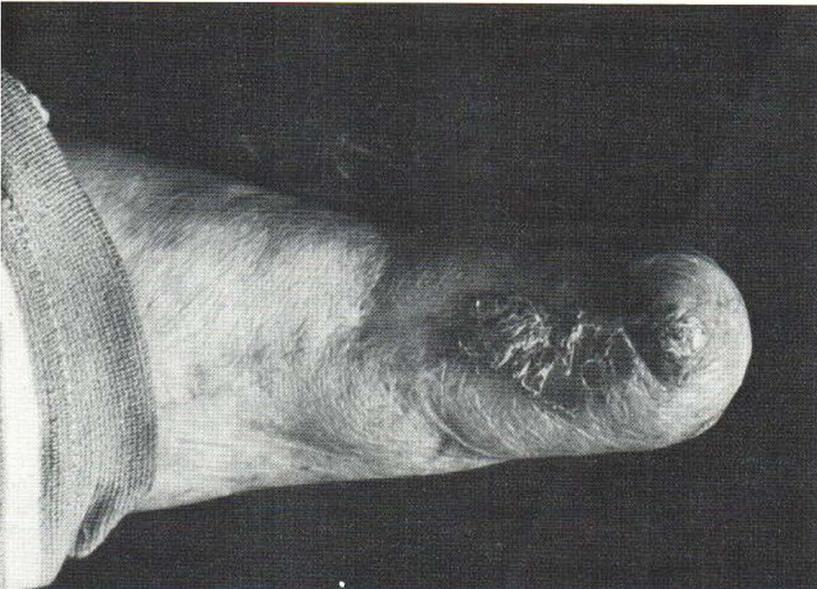


Fig. 2. Hyperkeratotic elements on the amputated anterior aspect of the right antrachium.

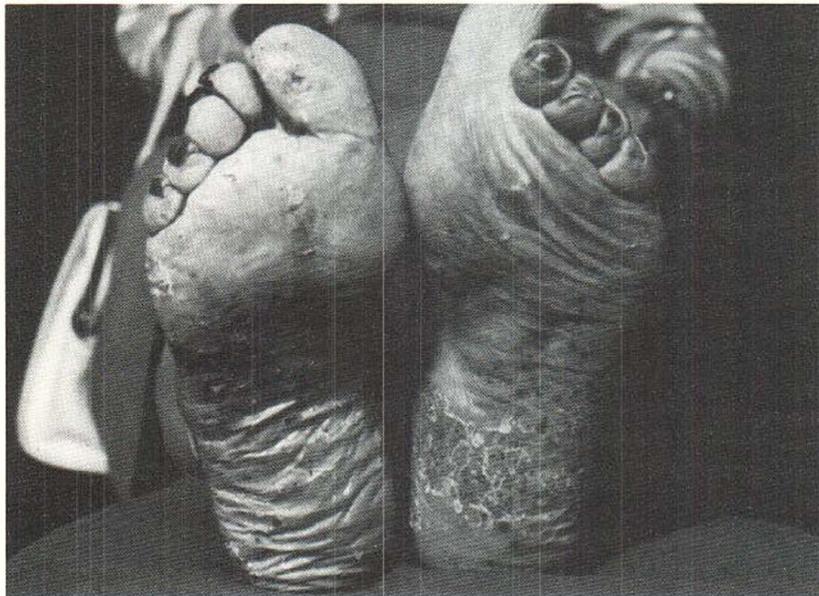


Fig. 3. Soles after 5 weeks of treatment with Tigason® (Ro 10-9359).

neurocutaneous elements. The intra-epidermal abscesses might be the result of trans-epidermal elimination of immune deposits from the dermis (8). A neurocutaneous pathogenesis would support the "lower epidermis" theory concerning the morphogenesis of PPP (i.e. the first phase in characterized by spongiosis and vesicle formation in the lower layers of epidermis) (7).

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Tyrosinase-negative Albinism with Congenital Malformations and Squamous Cell Carcinoma of the Genitalia

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Abstract. A 41-year-old tyrosinase-negative male albino presented with congenital bilateral inguinal hernias, club feet and squamous cell carcinoma of the glans penis. Such associations do not appear to have been reported previously.