

## Acral Pseudolymphomatous Angiokeratoma of Children: a Case with a Lesion on the Wrist

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Sir,

We report on a rare case of acral pseudolymphomatous angiokeratoma in children (APACHE) (1), presenting as a lesion on the wrist. This site of the lesion has not been described in the nine cases of APACHE reported previously (1–4).

### CASE REPORT

A 2-year-old Japanese girl had a past history of eczema on the whole body which had been well controlled with mild corticosteroid ointment. She developed an asymptomatic papular lesion on her right wrist, and visited our outpatient clinic. The lesion was composed of linearly arranged, deep red-coloured, papules varying in size from 1 mm to 4 mm, and coalesced, on the dorsal site of the right wrist (Fig. 1). There were no similar lesions on other sites of the body. Under a provisional clinical diagnosis of epidermal nevus, the papular lesion was followed-up for 4 years, without therapy, and showed almost no change in clinical appearance. The lesion was surgically removed.

Histologically, the epidermis was flattened in most parts of the lesion, covering a well circumscribed, dense cell infiltrate of massive mononuclear cells, confined to the superficial through mid-dermis (Fig. 2A). The epidermis showed hydropic degeneration of the basal cell layer with the cell infiltration (Fig. 2B), and focal



Fig. 1. Clinical feature of the lesion, which is composed of linearly arranged, deep red-coloured, papules on the dorsal site of the right wrist.

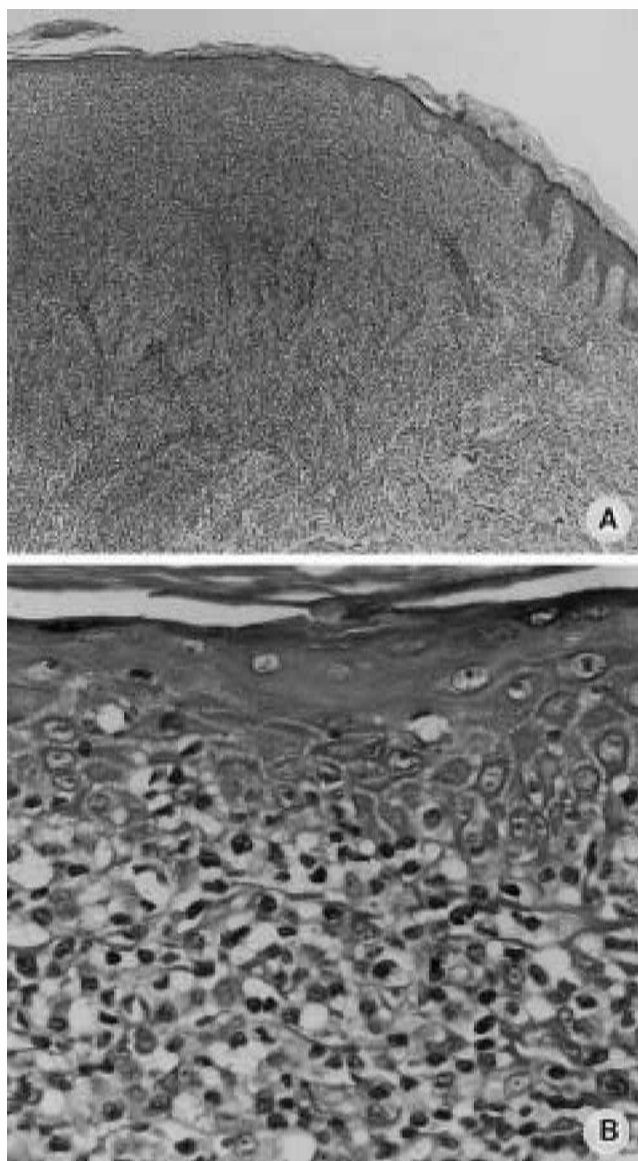


Fig. 2. Histological findings of the lesion (H&E). (A) Well circumscribed, dense cell infiltrate beneath a flattened epidermis (× 30). (B) Hydropic degeneration of the basal cell layer with mononuclear cell infiltration, and cell infiltration in the dermis composed mainly of small lymphocytes with a considerable number of plasma cells (× 300).

hyperkeratosis with parakeratosis. The infiltrating cells were mainly small lymphocytes, with a considerable number of plasma cells (Fig. 2B). No atypical cells were found within the cell infiltrate, and conspicuous, dilated capillaries without thickening of the walls were seen.

Immunohistochemically, there was an admixture of

focal B (CD20+) and dispersed T (CD45RO+) lymphocytes. CDw75 antibody, which stains germinal centre B lymphocytes, was negative. Ki-67 was negative. Kappa chain-positive lymphocytes and lambda chain-positive lymphocytes were observed almost equally. The lesion has shown no recurrence for at least 3 years after the surgical removal.

## DISCUSSION

APACHE has been reported to arise mostly in childhood, and to present as unilateral eruptions of multiple angiomatic papules, mostly on acral sites of the hands and feet (1–4). Histologically, APACHE has been documented to correspond to pseudolymphoma (1–4). Immunohistochemical examination of infiltrating lymphocytes did not reveal any monoclonal proliferation of T or B cell subsets nor aberrant expression of surface markers (1–4). The present case is regarded as belonging to the category of APACHE in both histological and clinical features, except for the site of the lesion.

As regards the term “APACHE”, the disorder has not shown any histological features of angiokeratoma (1–4), with the exception of one case of a lichenoid reaction (2). It arises not just in childhood (1–4) but

also in adulthood (3), and is seen not just on the digits (1–4) but also on the back (3), and, as in the present case, on the wrist. The term “APACHE” does not therefore seem suitable for the condition. Recently, the terms “small lymphoid papules of the extremities” (3) and “small papular pseudolymphoma” (3) have been put forward for the condition. A kind of mechanical stimulation might be a factor inducing the condition, as suggested by the acral localization of lesions in most cases of this skin disorder.

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## Prurigo Pigmentosa in Association with *Helicobacter pylori* Infection in a Caucasian Turkish Woman

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Sir,

Prurigo pigmentosa (PP) is a rare inflammatory dermatosis of unknown origin characterized by intensely pruritic papular eruption which resolves leaving gross reticulate hyperpigmentation. The majority of cases are young Japanese women (1–4). To date, only 17 cases have been reported outside Japan (2, 5, 6). This is a report on a case of PP associated with gastritis due to *Helicobacter pylori* in a young Caucasian Turkish woman.

## CASE REPORT

An 18-year-old female presented with a 1-year history of intensely itching, recurrent papular eruption leaving gross reticular pigmentation on her back. The condition had not responded to previous treatments with antihistamines, antidepressants, sulfamethaxazole/trimethoprim, fluconazole, ciprofloxacin or topical cortico-

steroids. She had had no past or current medical problem except mild dyspepsia. On examination, there were erythematous papules, some of which were excoriated and exulcerated, on the background of hyperpigmented keratotic plaques distributed in reticular pattern covering the middle portion of the back and scapulas symmetrically.

A punch biopsy from a papule revealed orthokeratosis, spongiosis, intraepidermal vesicles with accumulation of neutrophils and eosinophils, focal vacuolar basal layer degeneration, perivascular lymphohistiocytic infiltrate, melanophages and upper dermal oedema. Direct immunofluorescence was negative for IgA, IgG, IgM and C3 deposits. With a diagnosis of PP, the patient was started on doxycycline, 200 mg daily. However, on the third day of therapy, she experienced a phototoxic reaction, which resolved with topical steroids, and her gastric complaints became more severe. Doxycycline was therefore discontinued. Laboratory