Acrodermatitis continua of Hallopeau (ACH) is a rare, chronic disease characterized by acropustular eruptions predominantly involving the distal phalanges of the hands and feet with marked involvement of the nail bed. The sterile pustules may coalesce to form groups of lesions, which, over time, may spread proximally to involve the dorsal side of the hands, forearms and feet. Pustulation of the nail bed and nail matrix are often associated with onychodystrophy and even onychomadesis of the involved digits. Atrophic skin changes, onychodystrophy and osteolysis are frequently present, causing painful and disabling lesions (1).

CASE REPORT

A 9-year-old girl (body mass index (BMI) 18.9) presented to our department with a 3-year history of ACH, with recurrent flares of painful pustular scaly lesions on the distal portion of the fingers and toes, with associated severe onychodystrophy (palmoplantar pustular psoriasis area and severity index (PP-PASI) score 62) (Fig. 1A). No other lesions were noted over the patient’s body surface area. Her past medical history was otherwise unremarkable, with no thyroid dysfunction and no family history of psoriasis. Clinical and microbiological examination did not show any bacterial or fungal infection. Histological features of psoriasis were found in a biopsy specimen from skin lesion. X-ray examination revealed no osteolytic changes of the bones and no joint deformities. Based on the clinical and histopathological findings, ACH was confirmed.

Previous treatments with potent topical corticosteroids, cyclosporine (3 mg/kg/day for 6 months) and etanercept (0.4/kg/twice a week for 3 months) had all been unsuccessful. Because of the lack of efficacy of previous therapies and severe disability, we decided to use the human anti-tumour necrosis factor alpha (anti-TNFα) monoclonal antibody adalimumab. After receiving approval from the local ethics committee, adalimumab was introduced at a first dose of 80 mg, 40 mg after 1 week and then 40 mg every other week. After the first 4 weeks of treatment, a substantial improvement in the lesions was noted, without formation of pustules, leaving only mild onychodystrophy (PPPASI score 6) (Fig. 1B). At 8 weeks of treatment there was complete resolution of the clinical aspects. The effects of adalimumab persist after 12 months of therapy, with no adverse events, relapse or need for additional treatment. The young patient currently remains under treatment and follow-up.

DISCUSSION

ACH is generally considered as a rare variant of pustular psoriasis, though some authors classify it as a separate entity. Histology shows neutrophils filling the subcorneal cavity and aggregations of leukocytes between the epidermal cells forming spongiform pustules (of Kogoj), with a lymphohistiocytic infiltrate and focal oedema of the superficial dermis (2). The disease runs a chronic, relapsing course with slow proximal progression and may evolve into generalized pustular psoriasis. It is more common in middle-aged females and often begins after localized trauma (3), but is rare in children. The disease
Short communication

is difficult to treat, with limited success reported with numerous agents, including topical treatment such as corticosteroids, tar, dithranol, fluorouracil, calcipotriol and calcineurin inhibitors (4), phototherapy (5), cyclosporine, methotrexate combined with oral propylthiouracil (6), retinoids (7), dapsone (8), thalidomide in association with ultraviolet (UV) (9) and tetracyclines (10). Recently, there have been reports of effective treatment with the biologic agents infliximab (11) and etanercept (12). Sopkovich et al. (13) described successful treatment of ACH with adalimumab. In childhood the treatment can be a challenge because many therapeutic options have drawbacks or are not approved. We report here the case of a 9-year-old female patient who was treated successfully with adalimumab.

Adalimumab is a recombinant, fully human, anti-TNF-α monoclonal IgG1 antibody that acts primarily by binding both soluble and cell-bound forms of TNF-α and neutralizing the activity of cytokine (14). TNF-α blockers have shown efficacy in refractory ACH, supporting the relationship of TNF-α to this kind of disorder (15). Since our case is a paediatric patient, double-blind, placebo-controlled trials would be desirable to ascertain the safety and efficacy of this treatment.

REFERENCES