

Pityriasis Rubra Pilaris Successfully Treated with Infliximab

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Sir,

Pityriasis rubra pilaris (PRP) is an uncommon, idiopathic, papulosquamous disorder that often progresses to erythroderma and causes disabling keratoderma. The disease is classified into 5 groups on the basis of clinical appearance, behaviour and prognosis, as proposed by Griffiths in 1980 (1). Recently, a sixth group has been proposed in order to acknowledge the HIV-associated type of PRP (2).

No single therapy is universally effective, and some cases are resistant to multiple therapies. We describe here a case of recalcitrant type I, adult-onset PRP that was treated successfully with infliximab.

CASE REPORT

A 63-year-old woman presented with a 2-month history of well-defined, erythematous scaling patches involving her scalp and trunk. A biopsy from a lesion showed a spongiotic dermatitis. Treatment with topical steroids was initiated. Her medical history was remarkable for hypertension, diabetes mellitus, hypercholesterolaemia, hyperuricaemia and anxiety. Her medications included irbesartan, metformin, fenofibrate, allopurinol and amitriptyline hydrochloride. She had no family or personal history of psoriasis.

Despite the topical treatment the lesions progressed in a cephalo-caudal distribution. Therapy with oral cyclosporine, 5 mg/kg/day, was started. Two biopsy specimens revealed hyperkeratosis with alternating

orthokeratosis and parakeratosis, moderate acanthosis, a normal granular layer and a mild perivascular chronic inflammatory cell infiltrate consistent with PRP.

Based on the clinical and pathological findings, the patient was diagnosed as having a PRP, adult type.

Despite treatment with cyclosporine the lesions progressed. Examination revealed erythroderma with several 1–1.5 cm “islands of sparing”. Yellow palmo-plantar keratoderma with fissuring was noted (Fig. 1A). Treatment was switched to acitretin, 50 mg/day. After 5 months of acitretin monotherapy, the skin eruption failed to improve. A trial of intravenous infliximab was initiated. Full blood count, electrolytes, liver function tests, urine microscopy, viral hepatitis and HIV serologies, PPD and chest X-ray were all normal or negative prior to initiation of therapy.

Three infusions of infliximab (weeks 0, 2 and 6) were administered at a dose of 5 mg/kg over a 2-h period. The dose of acitretin was tapered and stopped after 2 weeks. The patient improved dramatically within 6 weeks of the first infusion, with resolution of erythroderma and pruritus (Fig. 1B).

Given the response to infliximab, the patient was given 3 further infusions at 8-week intervals with no other concomitant treatment. No side-effects were observed.

DISCUSSION

Therapy for PRP is based largely on anecdotal reports owing to the idiopathic nature of the disorder,



Fig. 1. (A) Erythroderma with fingertip-sized islands of sparing on the chest and abdomen and yellow palmar keratoderma with fissuring. (B) Erythema and scaling subsided following 3 infusions of infliximab.

the rarity of cases and the lack of controlled trials. In addition, some cases of PRP resolve spontaneously without therapy; therefore evaluation of the success of any therapy is controversial. Several therapeutic approaches for treating PRP have been published (3). In 1985, Fox et al. found that more than 20 different agents have been used to treat the disorder (4). Systemic retinoids are commonly used as a first-line therapy and methotrexate has traditionally been a good second-line treatment. Other therapies include immunosuppressive medications, such as azathioprine or cyclosporin A, stanozolol, phototherapy, extracorporeal photochemotherapy, fumaric acid esters and calcipotriol.

Infliximab is a neutralizing, chimeric, monoclonal antibody that binds to the pro-inflammatory cytokine tumour necrosis factor (TNF)- α . Initially developed for the treatment of Crohn's disease, this medication has been reported to be effective in treating psoriasis (5) and a number of inflammatory dermatoses, including hidradenitis suppurativa or pyoderma gangrenosum (6, 7). There is a clinical and therapeutic response overlap between psoriasis and PRP. As infliximab is efficacious in treating psoriasis, it could be also useful in PRP.

Only a few cases of the use of anti-TNF- α immunotherapy as treatment for PRP have been published (7–10). The first report (8) describes a patient with PRP and polyarthritis treated with infusions of the p55 TNF- α receptor immunoadhesin, with no response of the skin condition, but with remission of the arthritis. Three subsequent reports have demonstrated the efficacy of infliximab in the treatment of 6 patients with adult-type PRP (7, 9, 10). No significant adverse effects have been noted in the literature.

In our patient it is highly likely that infliximab was responsible for the clinical response. Failure of the cyclosporine and acitretin therapies to control the disease, and the temporal improvement related to the infliximab infusions, suggest an active role of the drug. The patient experienced a significant improvement after the third infusion at week 6, which was infliximab monotherapy, as acitretin had been stopped 4 weeks previously. The

therapy was well-tolerated in our patient, with no side-effects.

PRP may pose a therapeutic challenge. In view of the response in the case described here and the previous successful reports (7, 9, 10), infliximab could be included as an alternative agent for treating adult-onset PRP. TNF- α is a pro-inflammatory cytokine that plays an immunomodulatory role in a variety of systemic and dermatological diseases. Given the response to anti-human TNF monoclonal antibodies, TNF- α may play a pathophysiological role in PRP.

Conflicts of interest: None identified.

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