Erythrodermic pityriasis rubra pilaris managed at home: intensive community care followed by ustekinumab

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A 74-year-old woman presented with a rash on her right upper chest, which then progressed to become generalized over 2 months. There was a prominent follicular component to the rash, islands of sparing, and thick, orange hyperkeratosis of the palms and soles (Fig. 1). The features were those of classic erythrodermic pityriasis rubra pilaris (PRP). Histological examination of a skin biopsy confirmed the clinical diagnosis.

Following the onset of erythroderma, the patient developed bilateral pedal oedema, had poor urine output, and felt shivery and unwell. Hospital admission for ongoing management was recommended, but declined by the patient, who opted for home treatment supported by community nurses and carers. A dermatology specialist nurse visited weekly to monitor progress, while carers visited three times daily for emollient application. Additionally, the patient self-applied emollients between these multiple home visits. Approximately 5–6 kg of emollient was used each week. District nurses visited weekly to perform venepuncture and for weight, urine output and observation monitoring. Subsequently, Telehealth was introduced, a self-monitoring system with which the patient takes their own physical observations (weight, pulse, blood pressure), which are entered into a monitor and analysed remotely by an experienced community nurse. The patient remained haemodynamically stable throughout the course of her treatment.

Cotton leggings, a long-sleeved cotton top and cotton gloves were provided. Increased oral fluid intake and a high protein diet were advised, with good compliance. By week six, the patient had stabilized, but the erythroderma persisted. The shivering had stopped, urine output was normal, and the patient was able to independently apply the emollients regularly, thus the home visits by nurses and carers were withdrawn.

The erythroderma remained persistent, with > 90% of the skin affected. There was no discernible improvement with methotrexate (highest dose 17.5 mg weekly) over 3 months. Subsequent treatment was started with acitretin 20 mg daily, but this was stopped after 3 months because of adverse effects and lack of efficacy. The patient’s Dermatology Life Quality Index (DLQI) at this stage was 21.

Ustekinumab was commenced, and administered at 45 mg (for patients with bodyweight < 100 kg) in weeks 0, 4 and 12. Initially, there was no response, but after 6 weeks, a small but significant improvement was noted. The erythroderma cleared slowly over the subsequent 6 weeks. The rate of improvement increased after the second dose of ustekinumab, with almost complete clearance by the third dose at week 12.

PRP is a rare papulosquamous keratotic dermatosis of unknown aetiology. The management of PRP is always challenging.1 In severe disease, patients may become very unwell with erythroderma that is unresponsive to systemic agents. The evidence base for systemic agents is thin, owing to the lack of clinical trials1 in this rare condition. Dermatology textbooks suggest retinoid or methotrexate as first-line therapy, tumour necrosis factor-α inhibitors or ustekinumab as second-line, and fumaric acid or intravenous immunoglobulin as third-line.2 There is growing evidence from case reports of a good response of PRP to ustekinumab therapy. However, case reports and small case series are likely to be the only evidence for ustekinumab therapy in PRP.
The rationale for using ustekinumab as a treatment for PRP is that it works in psoriasis, a condition with similar clinical and histological features. Nevertheless, the exact therapeutic mechanism of ustekinumab in PRP is unclear. PRP is thought to be a T cell-mediated autoimmune condition. Ustekinumab is an anti-p40 monoclonal antibody that blocks interleukin (IL)-12/IL-23, thus blocking the activation of T cells.

One of the complications of PRP is erythroderma, an extensive exfoliative dermatitis that is potentially life-threatening. By convention, hospital admission is recommended for erythrodermic PRP to allow intensive nursing care, administration of nutritional supplementation, frequent application of emollients, and appropriate monitoring of routine observations and fluid balance. However, the intense pressure on acute hospital beds creates a suboptimal environment for such care. The exfoliative erythrodermic phase of PRP often lasts for weeks or even months, making admission into an acute medical bed a last resort, rather than the preferred course of action. Following widespread closure of dermatology beds across the NHS in England and Wales and high rates of acquired infection, inpatient care could potentially make the situation worse for patients with PRP in an environment uniquely unsuited to the care of patients with erythroderma. Although earlier controlled trials have shown improved outcome in chronic conditions with home-based care, a recent study raised concerns about reduced quality of care in the community. Management of erythroderma at home was considered for this patient only because of her reluctance to be admitted to hospital. This case illustrates that in the context of a supportive family and a compliant, stable patient, erythrodermic PRP can do well without the need for inpatient admission, with treatment and regular monitoring provided by an experienced community-care team led by an experienced dermatology nurse.

In conclusion, we report a patient with classic erythrodermic PRP managed in the community. Systemic therapy progressed through methotrexate and acitretin, before ustekinumab was prescribed to good effect.

References

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CPD questions

Learning objective
To demonstrate up-to-date knowledge of the management of refractory pityriasis rubra pilaris.

Question 1
Erythrodermic pityriasis rubra pilaris (PRP) is usually difficult to treat. Which one of these treatments has been effective in treating refractory cases of erythrodermic PRP?

a) Methotrexate.
b) Acitretin.
c) Omalizumab.
d) Ustekinumab.
e) Hydroxychloroquine.

Question 2
What is the mechanism of action of ustekinumab?

a) Anti-p40 monoclonal antibody which blocks interleukin (IL)-12 and IL-23.
b) Anti-tumour necrosis factor (TNF)-α.
c) Anti-CD11a monoclonal antibody.
d) Anti-CD20 monoclonal antibody.
e) Anti-IgE monoclonal antibody.

Instructions for answering questions

This learning activity is freely available online at http://www.wileyhealthlearning.com/ced

Users are encouraged to

- Read the article in print or online, paying particular attention to the learning points and any author conflict of interest disclosures
- Reflect on the article
- Register or login online at http://www.wileyhealthlearning.com/ced and answer the CPD questions
- Complete the required evaluation component of the activity

Once the test is passed, you will receive a certificate and the learning activity can be added to your RCP CPD diary as a self-certified entry.

This activity will be available for CPD credit for 2 years following its publication date. At that time, it will be reviewed and potentially updated and extended for an additional period.