Narrowband UVB phototherapy in erythropoietic protoporphyria: case series

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Sir, Erythropoietic protoporphyria (EPP) is an inherited disorder of the haem metabolic pathway. It causes disabling cutaneous photosensitivity and, sometimes liver disease due to the accumulation of protoporphyrin in blood, liver and other tissues.¹

Holme SA et al reported the median age at onset and diagnosis in patients with EPP in the UK as 1 and 12 years respectively, with diagnosis being delayed until the age of 20 years or more in 34% of their patients.² However, late presentation of EPP in adults has been reported.³

The mechanism of cutaneous photosensitivity is not entirely known. The primary event appears to be endothelial damage caused by the photodynamic reaction of the accumulating
protoporphyrin. Mast cell and complement activation may also be responsible for the clinical manifestations in EPP.\textsuperscript{4}

Behavioural measures, wearing protective clothing and opaque sun blocks are useful in preventing photosensitivity but often unacceptable in this young group of patients. A systematic review of treatment options in EPP did not detect an efficacy of beta-carotene, vitamin C and N-acetyl cysteine in well-designed studies.\textsuperscript{5}

Narrowband ultraviolet B phototherapy (NBUVB) is widely used as a prophylactic treatment for many photodermatoses, especially for polymorphic light eruption (PLE). A two to four fold increase in minimal erythema dose (MED) to relevant visible wavebands was demonstrated after NBUVB in five patients with EPP. Patients also reported a favourable response after treatment with NBUVB with sunlight exposure tolerance increasing up to 2 hours.\textsuperscript{6}

We would like to share our experience of treating EPP patients with NBUVB over the last 20 years. We did a retrospective analysis of case notes of all EPP patients who received NBUVB. The regimen followed for NBUVB treatment was similar to that for PLE with starting dose of 50\% to 70\% of MED and 20\% increments, and then reduced to 10\% increments, with usually up to 15 treatments.

Twelve patients (9 females) were treated with a total of 80 annual courses of narrowband UVB phototherapy (Philips TL-01 lamp) between 1991 and 2011. Mean age of patients was 28 years and the majority was of skin phototype I. The mean age of onset of the disease was 2.4 years except for one patient who had late onset of EPP at the age of 44 years. The mean duration to diagnosis from the onset of disease was 3.7 years. The diagnosis was established based on clinical history, signs, qualitative plasma porphyrin scan and quantitative erythrocyte porphyrins. Ten who were phototested had abnormal monochromator response to relevant visible wavebands. None of them had abnormal response to any UVB waveband. MED to NBUVB at 24 hours was normal in all of them.

The mean total treatments per course ranged from 8 to 15 and mean final dose per course was between 0.19 and 0.92 J/cm\textsuperscript{2}. Seven patients reported good benefit, 4 patients reported no benefit and no information on efficacy was available for 1 patient. Two patients reported an increase in duration of tolerance to sunlight up to 1.5 hours after NBUVB compared to few minutes prior to treatment. Three patients commented on a better tolerance of sunlight abroad. Three patients commented on the duration of efficacy and felt that it lasted for 6-9 months.

The treatment was well tolerated with well - demarcated erythema as a side effect only in 2 patients. EPP like symptoms occurred in 2 patients but was due to day light exposure early in the course of treatment. Both were able to continue with treatment after a short interval. Two received their NBUVB phototherapy at home: one received 3 courses and the other had 4 courses. Number of treatments ranged from 15 to 20 per course. Both of them reported good benefit and tolerated the treatment well without any EPP like symptoms.

NBUVB phototherapy was beneficial and well tolerated in our group of patients with EPP. Home phototherapy was also found to be effective and well tolerated in our patients.

The main mechanism of induced tolerance causing the therapeutic benefit could be due to physical photoadaptation. It is possible that effects of NBUVB on cutaneous immune function, such as effects on mast cells, could also contribute to the therapeutic benefit.
An ideal study would be a randomised controlled trial which due to the low prevalence of EPP would have to involve more than one centre. Home NBUVB phototherapy could be offered in such a study to allow recruitment of patients over a wide geographical area.

References:
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