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Topic: Inflammatory skin diseases

Evaluation of systemic and biologic therapies in adult erythrodermic pityriasis rubra pilaris: a 10-year single-centre study in Russia

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Introduction

Pityriasis rubra pilaris (PRP) is a rare papulosquamous disorder frequently characterized by erythroderma. Due to limited data on the etiopathogenesis of PRP, treatment of this disease is challenging. Systemic retinoids are regarded as the first-line therapy, but they are not always sufficiently effective. The similarity between PRP and psoriasis pathogenic mechanisms has led to the consideration of genetically engineered biological agents (GEBAs) as an adjunctive treatment for PRP.

Materials and Methods

The study was conducted at the Department of Dermatology and Venereology, Russia, from May 2014 to May 2024. It included 21 patients over the age of 18 with the erythrodermic form of PRP. Patients were selected from archive records based on the following criteria: age 18-80 years and presence of the erythrodermic form of PRP (Psoriasis Area and Severity Index (PASI) ≥20). The Dermatology Life Quality Index (DLQI) was evaluated at baseline and monitored monthly during the treatment period. Twenty-one consecutive adults with erythrodermic PRP were treated with systemic therapies and followed prospectively during routine clinical care. The treatment distribution was as follows: 38.1% (n=8) received acitretin, 14.3% (n=3) isotretinoin, 38.1% (n=8) methotrexate, 28.6% (n=6) prednisolone, 4.8% (n=1) dexamethasone, 19% (n=4) netakimab, 4.8% (n=1) ustekinumab, and 4.8% (n=1) phototherapy (PUVA, UVB-311nm). The average PASI score before treatment was 28 ± 8.6, and the average DLQI was 21 ± 2.1.

Results

Following acitretin therapy, patients experienced significant regression of lesions, with a corresponding decrease in PASI to an average of 5 ± 3.2 points (achieving PASI75) and in DLQI to an average of 3.7 ± 1.9 points, indicating a positive therapeutic effect. One patient (4.8%) underwent phototherapy (PUVA, UVB-311nm) as monotherapy without achieving a clinical effect. However, when phototherapy was combined with other treatments, it demonstrated stabilization of the skin process and improved disease dynamics. Specifically, combination therapy with acitretin and phototherapy (RePUVA) achieved mean scores of 7.2 ± 4.6 (PASI), representing a decrease in PASI by ≥75%, and 5 ± 1 (DLQI), demonstrating enhanced efficacy. The effectiveness of methotrexate and phototherapy as

monotherapy for PRP remains controversial. In cases of relapse or resistance to initial therapy, GEBAs were used and found to be at least as effective as acitretin. Remission (achieving PASI 90) following successful treatment with netakimab and ustekinumab was maintained for up to 18 months. Follow-up continues to assess the long-term outcomes of biological therapy for PRP.

Conclusions

The data confirm the rationale for the first-line use of systemic retinoids in PRP. In this small cohort, acitretin appeared to provide greater overall clinical improvement than isotretinoin. Genetically engineered biological therapy may be considered an effective adjunct for severe and treatment-resistant PRP, contributing to sustained remission and improved quality of life. However, the interpretation of these findings is limited by the small sample size, single-centre retrospective design, absence of a control group, and the purely descriptive nature of the analysis.