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Title:

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Date:

2018-08-01

Citation:

Davenport, R., Le, S. T., Gin, A., Goh, M. S. Y. & Foley, P. (2018). Resolving classic pityriasis rubra pilaris, mimicker of erythema gyratum repens. *Australasian Journal of Dermatology*, 59 (3), pp.232-233. <https://doi.org/10.1111/ajd.12752>.

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<https://hdl.handle.net/11343/293930>

Resolving Classic Pityriasis Rubra Pilaris – mimicker of Erythema Gydatum Repens

Running Heading: A mimic of Erythema Gydatum Repens

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KEYWORDS:

Classic adult type Pityriasis rubra pilaris

Type I Pityriasis rubra pilaris

Erythema gyratum repens

Secukinumab

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This is the author manuscript accepted for publication and has undergone full peer review but has not been through the copyediting, typesetting, pagination and proofreading process, which may lead to differences between this version and the [Version of Record](#). Please cite this article as [doi: 10.1111/ajd.12752](https://doi.org/10.1111/ajd.12752)

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Article type : Case Letter

Resolving Classic Pityriasis Rubra Pilaris – mimicker of Erythema Gydatum Repens

A 58-year-old male presented with type I classic pityriasis rubra pilaris (PRP), characterised by a non-pruritic erythematous squamous eruption progressing in a cephalo-caudal fashion over 4 weeks to an erythroderma affecting over 95% body surface area with small islands of sparing. There was thick yellow palmoplantar keratoderma and nail dystrophy. Skin histology showed hyperkeratosis, parakeratosis, acanthosis and focal hypergranulosis, consistent with the diagnosis. Our patient does not have a past history of malignancy, and remains well, with negative age-appropriate cancer screening.

Initial treatment involved topical betamethasone dipropionate 0.05% ointment and acitretin 25mg daily. After 6 months, methotrexate 10mg weekly was added due to lack of response. This was discontinued after 3 weeks because the liver alanine aminotransferase (ALT) rose from a baseline of 24 to 169 U/L (normal range 5-40 U/L). There was no response to 6 weeks of narrowband UVB (3 times per week) or infliximab 5mg/kg (weeks 0, 2, 6, 14). Fifteen months after presentation he was commenced on secukinumab 300mg/month. He remained on acitretin between 25-50 mg/day throughout. Gradual improvement was noted in the severity and extent of erythema and scale, and the thickness of the keratoderma. Secukinumab was ceased after 9 months and he continued to improve thereafter. As the pityriasis rubra pilaris resolved, annular concentric rings of erythema and inward trailing scale were noted on the trunk and proximal limbs (Fig. 1a-b), reminiscent of erythema gyratum repens (EGR). Unlike true EGR, which is often malignancy-associated and pruritic, these figurate lesions were asymptomatic and did not migrate rapidly from day to day. Five

months on, the EGR-like eruption persists and he continues on acitretin at a reduced dose of 10mg daily.

Pityriasis rubra pilaris and erythema gyratum repens are two rare but clinically distinct dermatological entities.¹ PRP is an idiopathic papulosquamous condition that presents with follicular keratosis and erythema, whereas EGR is a figurate eruption, consisting of mobile serpiginous bands of erythema, and is often associated with internal malignancy.

We wish to highlight that pityriasis rubra pilaris presenting in the classical form, can assume an EGR-like appearance upon its resolution. To date, this unusual observation has been described in seven patients.¹⁻⁵

The differential diagnoses of pityriasis rubra pilaris include paraneoplastic erythroderma and cutaneous T-cell lymphoma. The question remains whether PRP simulating EGR necessitates the investigation for an underlying malignancy. Like other authors, we believe that in patients such as ours, the EGR-like appearance may simply be a stage in the resolution of classic PRP rather than an ominous sign heralding internal malignancy.² In all previously reported cases, screening and follow-up, which spanned 11 months to 7 years, failed to identify malignancy.¹⁻⁵ We report this case to remind clinical dermatologists of this rare but distinct phenomenon of classic PRP resolving with an EGR-like appearance.

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Figure Legends:

- Figure 1. (a, b) Figurate erythema with “wood – grain” appearance on trunk, axilla and upper thigh.

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