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Single Case

A Case of Recalcitrant Circinate Generalized (Lapière) Psoriasis Successfully Treated with Risankizumab

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Keywords
Risankizumab · Lapiere's psoriasis · Circinate psoriasis

Abstract
Circinate (Lapière) psoriasis represents a rare variety of generalized subacute pustular psoriasis clinically characterized by rapid onset of annular circinate lesions with micro-pustules at the borders without classic plaque psoriasis manifestations. Most reported cases have been described in childhood with a relative benign course, fast and long-term remission after treatment. However, the recalcitrant course may result in an important negative impact on patients' quality of life. Many systemic treatments have been reported for the management of moderate to severe forms, with variable clinical outcomes. However, data about the use of biologics in this rare psoriasis subtype are still lacking. Herein, we report the first case of circinate psoriasis unresponsive to methotrexate and almost all classes of biologics approved for psoriasis (anti-tumour necrosis factor (TNF), anti-interleukin (IL)-12/23, and anti-IL-17) which was successfully treated with risankizumab (anti-IL-23).

Introduction

Circinate psoriasis, also known as erythema annulare centrifugum type or Lapière's psoriasis, represents a rare variety of generalized pustular psoriasis, usually appearing with a subacute-chronic course with less systemic manifestations than plaque psoriasis [1].

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Clinically, it is characterized by rapid onset of annular circinate lesions with micro-pustules at the borders. Typical psoriasis lesions may be missing and the diagnosis is based on clinical course and histological findings [2]. Although most reported cases have been described in childhood, with a relative benign course, fast and long-term remission after treatment [3], it may be severally recalcitrant [2, 3]. To date, many topical and systemic treatments have been reported, with variable clinical outcomes. However, data on the use of biologic treatments in the management of this rare subtype of psoriasis are still lacking. Herein, we report the first case of a severe circinate psoriasis unresponsive to topical, traditional, and most biologic systemic treatments successfully treated with risankizumab.

Case Presentation

A 38-year-old male patient referred at our outpatient clinic on April 2019, with an history of 3 years of erythematous annular circinate elements of the trunk. Dermatological examinations revealed micro-pustules at the borders of the lesions, which were disseminated at trunk and arms. The patient reported an erroneous previous diagnosis of erythema annulare which was unsuccessfully treated with topical steroids. However, due to the borders' pustulation, which was not clearly diagnostic for erythema annulare, a skin biopsy from the pustular edge on an annular lesion was performed. Histological examinations revealed acanthosis, hyperkeratosis, and spongiform pustules in the upper epidermis; these findings resulted diagnostic for pustular psoriasis. Thus, histological and clinical findings were consistent for circinate psoriasis. No other comorbidities were reported in his medical history. Laboratory examinations were all within normal ranges except for erythrocyte sedimentation rate (ESR) (43 mm/h – normal value: <15 mm/h). Hence, methotrexate 10 mg weekly was started, without showing any clinical improvement after 12 weeks. After methotrexate withdrawal, due to the severity of skin manifestations and the strong itchy symptoms adalimumab was prescribed. Despite an initial clinical improvement, after 3 months of therapy, the patient experienced a recurrence of skin manifestations and symptoms. Thus, different biologic classes were consecutively prescribed including anti-interleukin (IL)-12/23 (ustekinumab) and anti-IL-17 (ixekizumab) both suspended for inefficacy. Particularly, after 3 months of ustekinumab treatment the patient showed a complete clinical remission, which was maintained up to 8 months, when patient reported a severe flare-up. Consecutively, standard dose of ixekizumab was started, with only partial improvements after 5 months. Given the worsening of manifestations and itching, which resulted in an important negative impact on patient's quality of life (psoriasis-area-severity-index [PASI]: 18.5; body-surface-area [BSA]: 19%; dermatologic-life-quality-index [DLQI]: 24), ixekizumab was suspended (Fig. 1a–c). Furthermore, the patient asked for a therapy with a larger period of administration; risankizumab treatment was prescribed. Standard risankizumab dose led to achieve significant clinical improvements after 12 weeks (Fig. 1d–f) (PASI: 1.5; BSA: 4.5%), with a complete resolution of itching and an important improvement on DLQI, which decreased from 24 to 5. These clinical outcomes were still maintained at last follow-up visit (week24).

Discussion

Pustular psoriasis represents a group of psoriasis subtypes characterized by infiltration of neutrophil granulocytes in the epidermis clinically resulting in sterile pustules [3]. Most frequently described subtypes of pustular psoriasis include (i) generalized pustular psoriasis (described by L. Von Zumbusch); (ii) acrodermatitis continua of Hallopeau; (iii) palmoplantar

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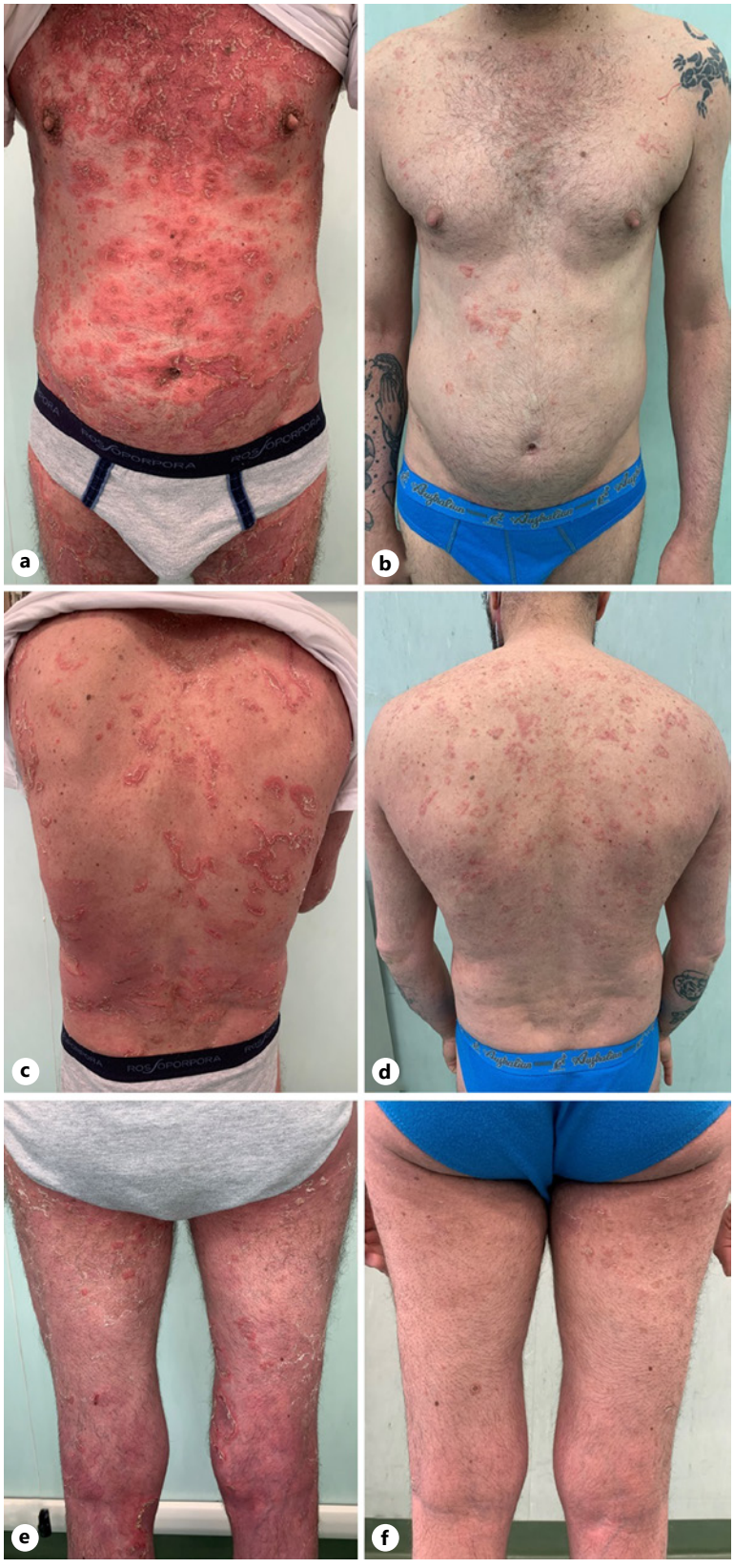

Fig. 1. Clinical manifestations of severe circinate psoriasis at baseline (**a–c**) and after 12 weeks of treatment with risankizumab (**d–f**).

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pustulosis (the most common, firstly described by Barber) [3, 4]. However, other less frequent forms of pustular psoriasis have been described, such as circinate or Lapiere's type [5]. Circinate psoriasis, rare variety of generalized subacute pustular psoriasis, has been more frequently described in childhood with a benign chronic recurrent course [5]. Typical psoriasis findings may be lacking, while annular recurring and moderately pruritic eruption with superficial pustules of the borders are usually described. Due to the lacking of specificity of manifestations, several conditions should be considered as clinical differential diagnosis. These include granuloma annulare, tinea corporis, erythema annulare centrifugum, nummular atopic dermatitis, and other pustular conditions (i.e., acute generalized exanthematous pustulosis) [1, 6]. However, differential diagnosis may be difficult, needing a biopsy and histological examination to reach a diagnosis. Fortunately, in most reported cases, annular pustular psoriasis showed a relatively good prognosis, with rapid resolution after treatments. Different treatments have been reported in the management of this subtype of psoriasis, with variable clinical outcomes. In mild forms, topical corticosteroids have been used with a complete remission of manifestations. As regards moderate-to-severe forms, many systemic treatments have been used, including methotrexate, dapsone, etretinate, leading important improvements on clinical manifestations [1, 7, 8]. However, spontaneous resolution and long-term remission were mostly reported in children than in adults. Indeed, adult pustular psoriasis still represents a challenge in dermatology, showing a chronic remitting course and a more frequent unresponsive rate than classic plaque psoriasis. The exact pathogenesis of this rare subtype of psoriasis is still unproven; however, it has been widely considered as a subtype of pustular psoriasis; hence, it may share some common cytokine pathways with its pathogenesis. Indeed, it has been shown that these subtypes of psoriasis have different pathogenic mechanisms than the classic plaque type, with an important role played by IL-36 signalling alteration, and IL-17A pathway [9]. Indeed, while in plaque-type psoriasis the role of TNF, IL-17, and IL-23 has been well documented in both preclinical and clinical studies, and indirectly confirmed by the effectiveness of new biologic therapies, pustular psoriasis showed a peculiar cytokine profile [10–12]. Particularly, an unopposed IL-36 signalling may promote TCR-driven proliferation of CD4+ T cells and cause higher IL-17A production [9]. Moreover, a recent study evaluating skin cytokine levels in pustular psoriasis skin found that IL-23 and IL-17 expressions were higher in the epidermis and perivascular dermal area of pustular psoriasis patients, suggesting that the expression of IL-23 might play a role in this subtype of psoriasis [13]. As regards the use of biologics in pustular psoriasis, anti-tumour necrosis factor (TNF) resulted the most effective biologic treatment, while anti-IL-12/23 and anti-IL-17A may be considered in refractory pustular psoriasis [8]. However, to date there is still no experience in the use of biologics in both child and adult annular pustular psoriasis. In addition, the effectiveness of the new molecules may be variable according to different factors, as also demonstrated by the appearance of paradoxical reactions with other chronic inflammatory skin diseases following the use of biotechnological drugs [11].

Herein, we reported the successful use of risankizumab, an anti-IL-23 recently approved for moderate to severe psoriasis [14], in a recalcitrant circinate generalized (Lapière) psoriasis. Particularly, our patient showed primary or secondary inefficacy to almost all available biologic classes (anti-TNF [adalimumab], anti-IL-12/23 [ustekinumab], and anti-IL-17 [(ixekizumab]), while remitted during treatment with risankizumab, one of the newest anti-IL-23 approved for moderate to severe psoriasis. Indeed, to date there are no official guidelines or recommendations indicating how to choose the best biologic treatment in this rare subtype of psoriasis. Our report showed risankizumab as a potential safe and effective option in the treatment of this rare subtype of pustular psoriasis, as well in multifailure patients. However, more studies are needed to better understand the potential role of anti-IL-23 in the

management of rarer forms of pustular psoriasis, with studies focusing on pustular psoriasis outcomes during risankizumab treatment.

Statement of Ethics

All procedures adopted in the present study were in respect to the ethical standards in the World Medical Association Declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. Ethical approval was not required for this study in accordance with local/national guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Angelo Ruggiero, Wanda Lauro, Gabriella Fabbrocini, Chiara Miano, Alessia Villani, and Claudio Marasca made substantial contributions to the conception or design of the work, or acquisition, analysis, and interpretation of data for the work; drafting and revising the work critically for important intellectual content; gave the final approval of the version to be published; and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Data Availability Statement

All data generated or analysed during this study are included in this article. Further enquiries can be directed to the corresponding author.

References

1 Liao PB, Rubinson R, Howard R, Sanchez G, Frieden IJ. Annular pustular psoriasis: most common form of pustular psoriasis in children: report of three cases and review of the literature. Pediatr Dermatol. 2002 Jan–Feb;19(1):19–25.
2 Resneck JS, Cram DL. Erythema annulare-like pustular psoriasis. Arch Dermatol. 1973;108(5):687–8.
3 Navarini AA, Burden AD, Capon F, Mrowietz U, Puig L, Köks S, et al. European consensus statement on phenotypes of pustular psoriasis. J Eur Acad Dermatol Venereol. 2017 Nov;31(11):1792–9.
4 Von Zumbusch L. Psoriasis und pustuloses Exanthem. Arch Dermatol Syphilol. 1910;99:335–46.
5 Filippeschi C, Battini ML, Tiradritti L, Mastrolorenzo A, Staderini C, Rapaccini AL, et al. A case of recurrent circinate erythematous psoriasis (Lapière). G Ital Dermatol Venereol. 1990 Aug;125(7–8):341–3. (Italian).
6 Karamfilov T, Wollina U. Juvenile generalized pustular psoriasis. Acta Derm Venereol. 1998;78:220.
7 Rosinska D, Wolska H, Jablonska S, Konca I. Etretinate in severe psoriasis of children. Pediatr Dermatol. 1988;5:266–72.

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DOI: 10.1159/000523756

8 Staughton R. Infantile generalized pustular psoriasis responding to dapsone. Proc R Soc Med. 1977;70:286–7.
9 Wang WM, Jin HZ. Biologics in the treatment of pustular psoriasis. Expert Opin Drug Saf. 2020;19(8):969–80.
10 Megna M, Ruggiero A, Camela E, Fabbrocini G, Marasca C. A case of erythrodermic psoriasis successfully treated with guselkumab. Dermatol Ther. 2020;33(2):e13238.
11 Marasca C, Megna M, Balato A, Balato N, Napolitano M, Fabbrocini G. Secukinumab and hidradenitis suppurativa: friends or foes? JAAD Case Rep. 2019 Jan 30;5(2):184–7.
12 Ruggiero A, Fabbrocini G, Cinelli E, Megna M. Guselkumab and risankizumab for psoriasis: a 44-week indirect real-life comparison. J Am Acad Dermatol. 2021 Mar 21;85(4):1028–30.
13 Song HS, Kim SJ, Park TI, Jang YH, Lee ES. Immunohistochemical Comparison of IL-36 and the IL-23/Th17 axis of generalized pustular psoriasis and acute generalized exanthematous pustulosis. Ann Dermatol. 2016;28(4):451–6.
14 Megna M, Fabbrocini G, Cinelli E, Camela E, Ruggiero A. Guselkumab in moderate to severe psoriasis in routine clinical care: an Italian 44-week real-life experience. J Dermatolog Treat. 2020;33(2):1074–8.