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CASE REPORT

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Complete response with guselkumab in a patient with double presentation of recalcitrant hyperkeratotic psoriasis over 18 months: a case report

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ABSTRACT

Background: Psoriasis is a chronic inflammatory skin disease with many clinical presentations. The most common variety is chronic plaque psoriasis. Rare chronic plaque psoriasis subtypes include annular, lichenified, and hyperkeratotic. Hyperkeratotic forms include ostraceous, rupioid, and elephantine. Some authors ignore the distinction between the types of hyperkeratotic psoriasis. Nevertheless, biological therapies have not been extensively researched in such rare cases of psoriasis. We present a case of intractable hyperkeratotic psoriasis that was successfully treated with guselkumab.

Case presentation: According to the physical examination of our 45-year-old male patient, the abdomen had confluent plaques and thick sero-exudative crusts. Erythematous, crusted, limpet-like lesions covered the back. The scales passed the Grattage test. The biopsy showed psoriasis features. After two months of guselkumab therapy, lesions were PASI 100. After a year of 8-week guselkumab medication, the patient's reaction was unchanged. Accordingly, guselkumab was given every 10 weeks for six months. The patient was disease-free except for a few minor lesions on the back and abdomen that disappeared following topical steroid cream.

Conclusions: Our patient had elephantine and rupioid psoriasis on the abdomen and back, respectively. This twofold presentation in the same patient supports the idea that hyperkeratotic psoriasis subtypes are interchangeable. The natural evolution of hyperkeratotic psoriasis lesions may explain this appearance, which has never been recorded. In this case report, guselkumab was highly effective at treating severe hyperkeratotic psoriasis in a real-world clinical environment for 1.5 years.

KEYWORDS

Psoriasis; Guselkumab; Hyperkeratotic psoriasis; Elephantine psoriasis; Rupioid psoriasis

ARTICLE HISTORY

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Introduction

Psoriasis is a common chronic inflammatory skin disease that can manifest clinically in a variety of ways. The most prevalent subtype of psoriasis, chronic plaque psoriasis, is characterized by well-defined, erythematous plaques with an overlying layer of silvery-laminated scales. A few rare subtypes of chronic plaque psoriasis include annular, lichenified, and hyperkeratotic forms. Rarely reported in the medical literature, the hyperkeratotic form is further classified as ostraceous, rupioid, and elephantine. Some authors do not differentiate between hyperkeratotic forms of psoriasis [1]. Due to the dense scales observed in these cases, systemic treatment in the form of oral cyclosporine or methotrexate injections was primarily used to manage the reported cases [1-5]. With infliximab, a case of ostraceous psoriasis with psoriatic arthritis has been resolved [6]. Moreover, a patient with generalized ostraceous psoriasis was cured by adalimumab [7]. In such uncommon cases of psoriasis, however, biological treatments have not been thoroughly studied. We report a case of recalcitrant hyperkeratotic psoriasis that was effectively treated with guselkumab with a consistent response over an extended period of time.

Case Presentation

A 45-year-old man presented to our clinic with a 9-month history of disseminated squamous plaques with dense scales on the trunk and extremities [psoriasis area severity index (PASI): 30]. The patient did not report any personal or familial psoriasis history. He also denied taking any medications recently. According to the physical examination, confluent plaques and thick sero-exudative crusts were present on the abdomen (Figure 1a). The back displayed distinctly demarcated, erythematous, well-defined lesions resembling limpets covered with scales and crust (Figure 1c). The scales exhibited a positive Auspitz sign. Nails, hair, face, palms, and mucous membranes were unaffected. HIV, venereal disease research laboratory (VDRL), and QuantiFERON-TB gold tests were negative. The biopsy revealed hyperkeratosis, acanthosis, papillomatosis, and dilated dermal papillae with perivascular lymphocytic infiltrate and exocytosis of neutrophils in the stratum corneum, all of which are consistent with psoriasis. Before presenting to our clinic, he had received topical steroid treatment without improvement. Our initial treatment regimen included 20 mg of methotrexate per week, 1 mg of folic acid per day, and calcipotriene/betamethasone dipropionate (0.005%/0.064%) cream as a topical. After two months of methotrexate treatment compliance, the lesions did not improve, so the patient requested additional intervention due to cosmetic

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disfigurement and diminished quality of life. Therefore, the patient continued to receive methotrexate for an additional two months, while guselkumab (100 mg subcutaneous injection) was administered initially at weeks 0 and 4, and then every eight weeks. Lesions exhibited a complete response (PASI 100) within two months of biological therapy (Figures 1b and 1d). After a year of continuous treatment with guselkumab every 8 weeks as a monotherapy, the patient's response remained unchanged. After that, guselkumab was administered every 10 weeks for a total of six months (Figures 2a and 2b). Nonetheless, the patient remained disease-free, except for a few tiny lesions on the back and abdomen, which vanished after applying a topical steroid cream.

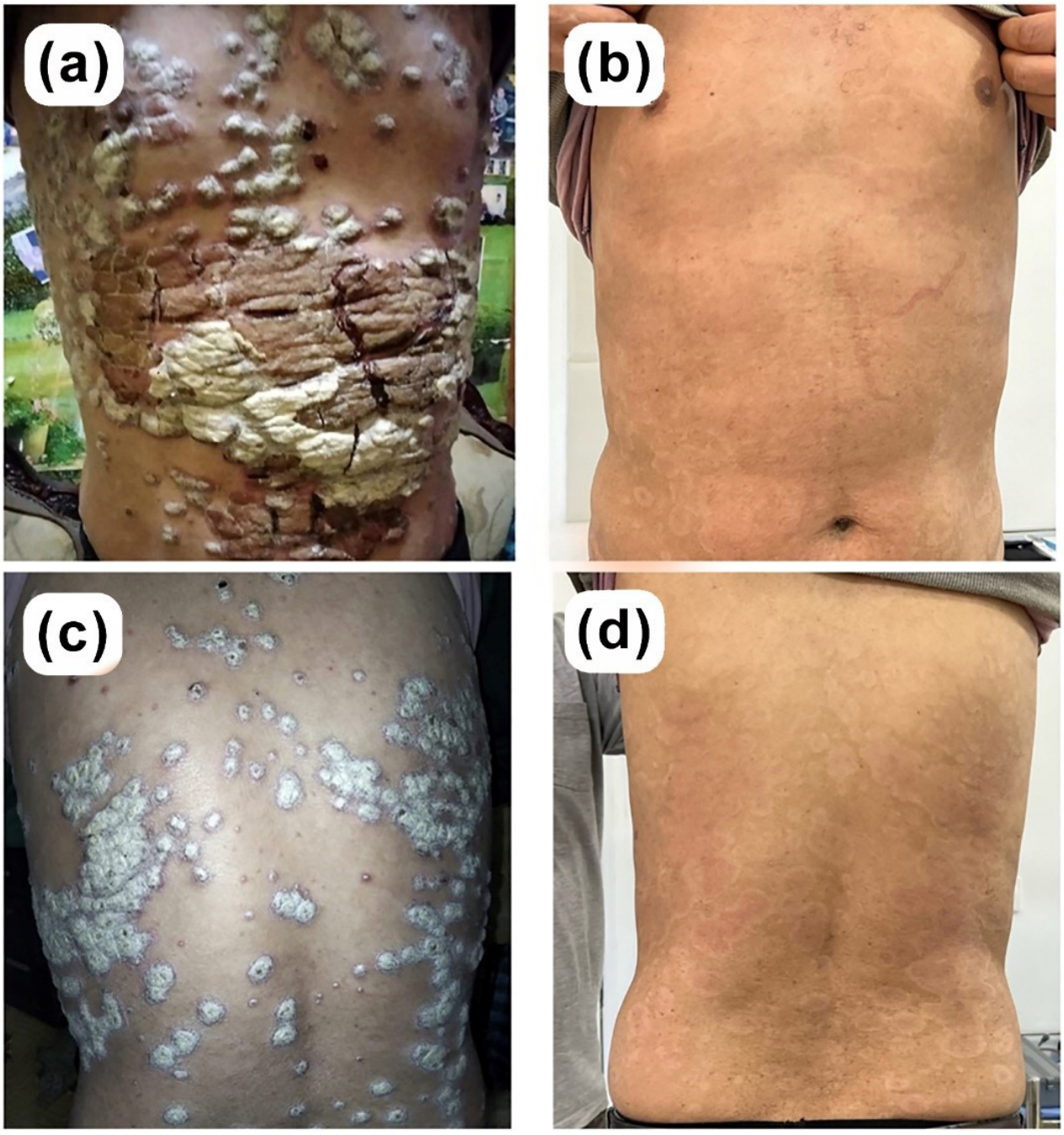

Figure 1. (a) A 45-year-old man with large, thick, flat, long-standing plaques over the abdomen. (b) After two months of guselkumab injections (initially at weeks 0 and 4, but then every 8 weeks), the abdomen, and (c) The back demonstrates sharply demarcated, erythematous, well-defined limpet-like plaques covered with scales and crust. (d) back, showing complete remission with post-inflammatory hypopigmentation.

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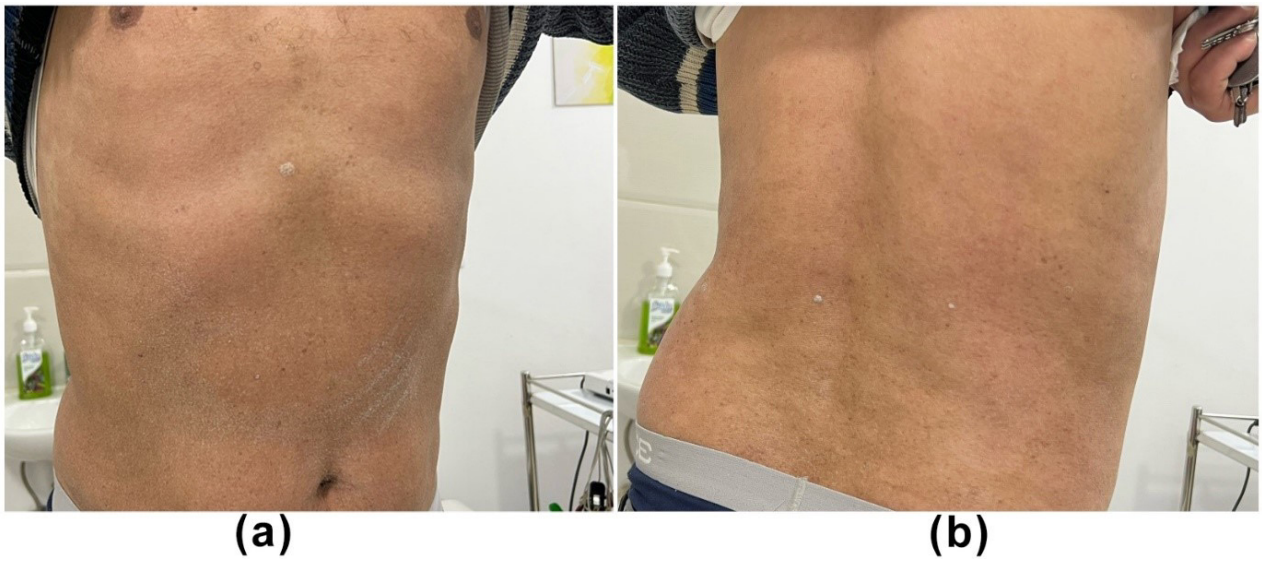

Figure 2. (a) Follow-up of the lesions in the abdomen and (b) back, after 18 months of treatment with guselkumab showing a consistent response.

Discussion and Conclusions

Rarely reported in the literature, the hyperkeratotic form of psoriasis is classified as ostraceous, rupioid, and elephantine. Ostraceous psoriasis is characterized by lesions with firmly adhered dense scales, varying colors, and surfaces resembling an oyster shell. Hyperkeratotic, concentric, circular, and cone-shaped lesions that resemble limpets are what distinguish the rupioid form. Elephantine psoriasis lesions have massive, thick, flat, persistent plaques [8]. Our patient presented with elephantine and rupioid psoriasis on the abdomen and back, respectively. This double presentation in the same patient supports the hypothesis that the terms for the various forms of hyperkeratotic psoriasis are interchangeable. This presentation has never been documented before; however, it is possible that it could be partially explained by the natural progression of hyperkeratotic psoriasis lesions from one stage to another. Because the lesions in the abdomen emerged first, then the lesions in the back.

The discovery of the IL-23/IL-17 axis has resulted in a significant increase in our understanding of the pathogenic immune events present in psoriasis and a paradigm shift in the treatment of this condition. The development of IL-17 inhibitors demonstrated that blocking the pathway of this cytokine was associated with high levels of efficacy and rapid onset of action in moderate-to-severe psoriasis, as well as superior clinical responses compared to TNF-inhibitors and ustekinumab, a nonselective IL-23 inhibitor. However, side effects such as neutropenia, candidiasis, and Crohn's disease exacerbations have been linked to these agents, highlighting the need for novel therapeutic approaches. Recently, selective IL-23 inhibitors such as guselkumab, tildrakizumab, and risankizumab have emerged, exhibiting a profile that is highly effective, long-lasting, and safe. Guselkumab is a human IgG1 monoclonal antibody (mAb) that binds to the p19 subunit of IL-23. It is the first drug of its kind to be approved by the US Food and Drug Administration (FDA) and the European Medicines Agency (EMA) for the treatment of moderate-to-severe plaque psoriasis in adult patients. In the treatment of moderate-to-severe plaque psoriasis, the superiority of guselkumab over other biologics is also evident in the less frequent dosing regimen of this selective IL-23 inhibitor (guselkumab: initially at weeks 0 and 4, but then every 8 weeks). From the patient's perspective, characteristics such as less frequent drug administration or the option for self-administration contribute to improved adherence and, consequently, improved clinical outcomes [9]. Furthermore, this infrequent administration is advantageous in nations with limited resources.

Multiple studies have demonstrated the long-term efficacy and safety of the guselkumab treatment. Patients in randomized controlled trials (RCTs) have typically received fewer prior therapies and have fewer comorbidities than patients in the real world. Therefore, real-world data from post-marketing studies and international or local registries are of great interest to the scientific community, as they provide crucial information on patients who are typically excluded from RCTs [10]. In the current case report, guselkumab was shown to be exceedingly effective for the treatment of severe hyperkeratotic psoriasis in a real-world clinical setting over a period of 1.5 years.

Availability of data and materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Disclosure statement

No potential conflict of interest was reported by the authors.

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