OXFORD

Oxford Medical Case Reports, 2024, **11**, 515–517
https://doi.org/10.1093/omcr/omae138
Case Report

Rupioid psoriasis, a unique presentation treated with Ustekinumab

Sdrah Diab[1], Moatasem Hussein Al-janabi[2,*], Ibrahim Diab[3], Fouz Hassan[4,‡]

[1]Department of Dermatology and sexually transmitted disease, Tishreen University Hospital, Lattakia, Syria
[2]Department of Pathology, Cancer Research Center, Tishreen University Hospital, Lattakia, Syria
[3]Department of Pediatrics, Tishreen University Hospital, Lattakia, Syria
[4]Department of Dermatology, Tishreen University Hospital, Lattakia, Syria

*Corresponding author. Department of Pathology, Cancer Research Center, Tishreen University Hospital, Lattakia, Syria. E-mail: dr.3esami2022@gmail.com
‡Vice President of Syrian Society of Dermatology, Assistant secretary of Pan Arab League of Dermatology

Abstract

Rupioid psoriasis is a rare subtype of psoriasis characterized by distinctive lesions resembling oyster shells, known as rupioid lesions. This subtype is particularly uncommon in the pediatric population and is often associated with poor treatment compliance. Ustekinumab, an IgG monoclonal antibody, targets IL-12 and IL-23, reducing the release of proinflammatory cytokines TNFα, IL-2, and IL-17α, which play vital roles in psoriasis pathophysiology. Approved for pediatric patients aged six years and older, ustekinumab provides a therapeutic option for moderate to severe psoriasis. We present the case of a 10-year-old girl diagnosed with psoriasis vulgaris at age two. She presented with rupioid lesions following a urinary tract infection that had been treated with oral cefixime (200 mg). After conducting appropriate tests, ustekinumab (45 mg subcutaneously) was administered, leading to significant improvements in the thickness of the lesions and overall appearance. This case demonstrates ustekinumab's efficacy in treating this challenging form of psoriasis.

Keywords: Rupioid psoriasis; Pediatric; Ustekinumab; monoclonal antibody; psoriasis treatment; childhood psoriasis; inflammatory cytokines

Introduction

Psoriasis is a common, chronic, inflammatory skin disease with an immunological and genetic basis. It can manifest at any age but is rare in children under ten. This condition significantly impacts the quality of life and morbidity of patients [1]. While there are various types of psoriasis, some forms are particularly unusual, such as ostraceous, rupioid, and elephantine psoriasis [2]. The rupioid variety is characterized by hyperkeratotic, well-demarcated, limpet-like, cone-shaped, circular plaques. Due to its distinct appearance, rupioid lesions can also be indicative of other conditions, including disseminated histoplasmosis, reactive arthritis, secondary syphilis, photosensitive skin lesions associated with aminoaciduria, and keratotic scabies, necessitating comprehensive diagnostic approaches such as biopsies and blood tests [3]. Rupioid psoriasis (RP) may be resistant to topical treatments and is often associated with complications like psoriatic arthritis [2–4]. This paper presents a case of prolonged rupioid psoriasis in a child, which showed partial improvement following treatment of an associated infection and significant improvement with the administration of Ustekinumab.

Case presentation

A 10-year-old girl, diagnosed with psoriasis vulgaris at age two with no remission, presented to our outpatient clinic with oyster-like, long-standing, pruritic lesions that had worsened during a recent urinary tract infection. She presented with rupioid lesions that were widespread, sharply demarcated, erythematous, thick, hyperkeratotic plaques with firmly adherent scales and hemorrhagic crusts, resembling oyster shells, and covered her trunk, scalp, face, and limbs, surrounded by discrete pustules and hyperkeratotic papules (Fig. 1).

The recent exacerbation coincided with dysuria, turbid and foul-smelling urine, and mild fever. Laboratory tests confirmed an elevated white blood cell count with a left shift, elevated C-reactive protein, and the presence of bacteria and white blood cells in the urine. A urine culture identified *Escherichia coli* as sensitive to cefixime. Based on the symptoms and test results, she was diagnosed with a urinary tract infection (UTI).

To exclude other potential causes of the rupioid lesions, comprehensive assessments were performed, including a chest X-ray, complete blood count, metabolic panel, tuberculosis screening, hepatitis tests, HIV (human immunodeficiency virus) screening, and syphilis serology (*Treponema Pallidum* Hemagglutination Assay). All returned normal results and were negative for hepatitis, tuberculosis, TPHA, and HIV.

The skin biopsy showed psoriasiform acanthosis, parakeratosis with notable Munro microabscesses, thin suprapapillary plates, an absent granular layer, and prominent papillary dermal edema (Fig. 2), leading to a diagnosis of rupioid psoriasis.

Received: January 20, 2024. **Revised:** July 26, 2024. **Accepted:** September 6, 2024

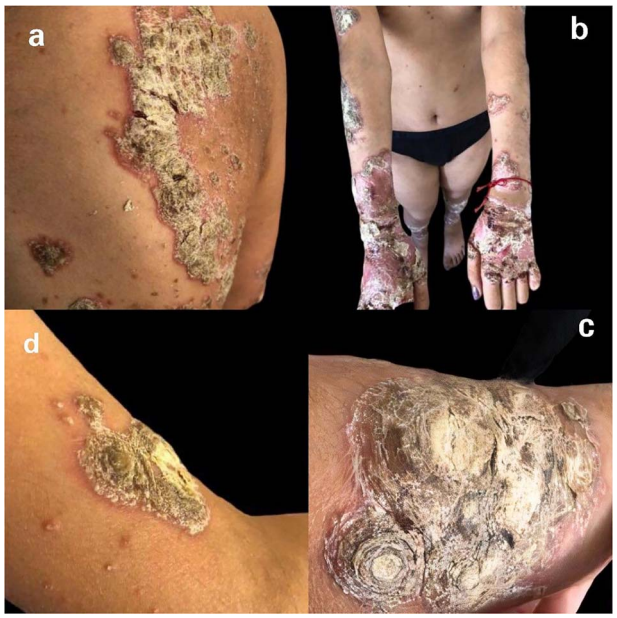

Figure 1. Clinical presentation of rupioid psoriasis in a 10-year-old patient. Panels [a and b] show erythematous, scaly plaques spread across the trunk and extremities. Panels [c and d] feature well-demarcated plaques covered with thick, oyster shell-like scales, illustrating the distinctive morphology of rupioid psoriasis.

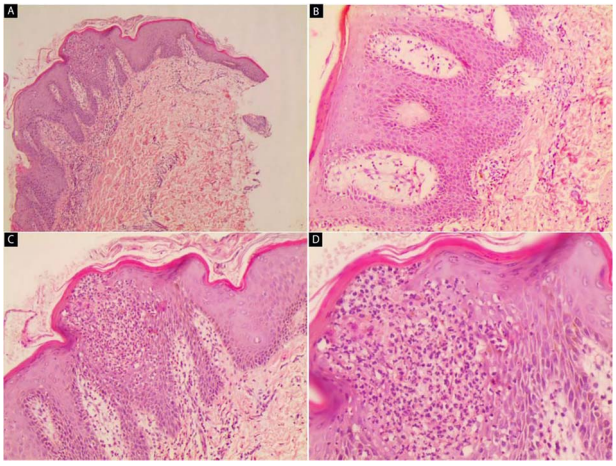

Figure 2. H&E-stained sections of the psoriatic lesions. (a and B) low power magnification (40x and 100x, respectively) displays regular acanthosis with elongated rete ridges, parakeratosis, and perivascular lymphocytic infiltrate in the upper dermis, typical features of psoriasis. (C and D) high power magnification (100x and 200x, respectively) shows a marked subcorneal pustule, providing insight into the acute inflammatory aspect of the disease.

The UTI was treated with cefixime 200 milligrams, resulting in an improvement of the skin lesions by the end of the antibiotic course. Ustekinumab was administered after the resolution of the UTI at a dose of 45 mg subcutaneously, based on her weight of 27 kilograms, a Psoriasis Area Severity Index (PASI) score of 17.9, and a Body Surface Area (BSA) involvement of 20%.

Three weeks post-administration, a follow-up revealed a significant improvement, with a PASI of 50% (Fig. 3). No adverse effects from the medication were noted. The patient will continue receiving ustekinumab doses every three months, with long-term follow-up planned to assess full efficacy after the third dose and subsequent treatments.

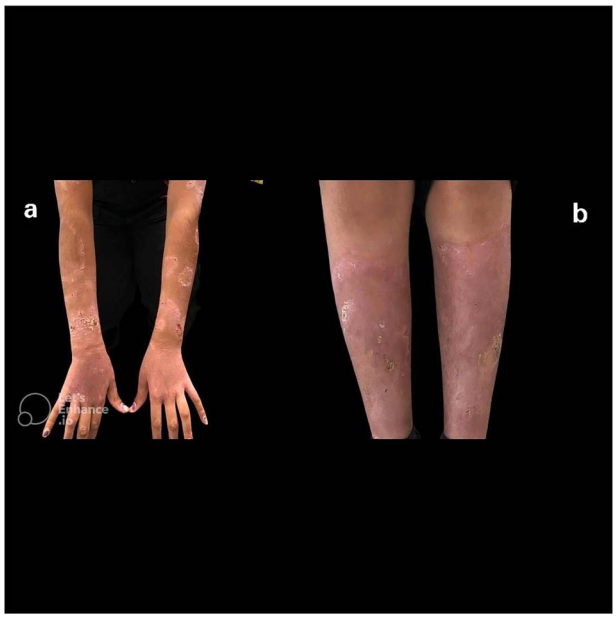

Figure 3. Clinical improvement in the rupioid psoriasis lesions after the first dose of Ustekinumab, captured just before the administration of the second dose, scheduled four weeks after the first. Panels [a and b] document the notable reduction in erythema and scaling, demonstrating the therapeutic response to the biological treatment after three weeks.

Discussion

The term 'rupioid' originates from the Greek word *rhupos*, meaning filth or dirt, and was first described in 1948 by Polish dermatologist Marian Grzybowski [3]. Rupioid psoriasis (RP) features well-demarcated conic plaques with thick, dark, lamellate, and adherent crusts that resemble limpet or oyster shells. Histologically, this 'dirty' appearance reflects serosanguineous exudate and thickened skin [3]. As reviewed in the dermatological literature, RP is a morphologic subclass of plaque psoriasis characterized by these distinctive hyperkeratotic lesions. According to recent studies, psoriasis with thick plaques tends to show a male predominance and has a higher incidence of psoriatic arthritis and nail disease; additionally, a greater body surface area is affected compared to patients with thin plaque type [3]. Although psoriasis is rare in children under ten years old, it represents about 4% of pediatric skin diseases, with comorbidities being notably more common in pediatric patients compared to their peers [1, 5].

Our patient's lesions appeared at age two, and symptoms of psoriatic arthritis started in the current period. Extensive literature indicates that upper respiratory tract infections can precipitate the onset and flare-up of psoriasis, with streptococcal species often acting as superantigens that trigger Th17 cell activation, crucial in the pathogenesis of psoriasis [6]. However, Ramirez-Bosca et al. examined blood samples from psoriasis patients and discovered increased DNA translocation caused mainly by intestinal microbes, including *E. coli*, Shigella fresneli, and *Enterococcus faecalis*. Bacterial DNA translocation is accompanied by a higher level of inflammatory response [7]. Lipopolysaccharide (LPS), an endotoxin of *E. coli*, is thought to work as a potent inflammagen and contributes to the inflammatory response in psoriatic patients [8]. Alongside the antimicrobial effect of Cefixime, it has an anti-inflammatory role, as it caused a decrease in CRP, TNF-α, CD8, and IL-6 levels and an increase in CD3+, CD4+, and

CD4+/CD8+ levels in children with UTI treated with cefixime [9]. Interestingly, our patient had a urinary tract infection caused by E.coli, which was thought to trigger the RP flare-up. The partial improvement of cutaneous lesions following treatment for the infection with cefixime lends support to this hypothesis.

A comprehensive review of the literature reveals that RP flare-ups can also be triggered by factors such as beta-blockers, lithium, synthetic antimalarials, and HIV [10]. Treatment of RP poses significant challenges due to poor compliance, and the challenging nature of topical agent penetration highlighting the necessity to identify treatment regimens that patients can easily adhere to, and emphasizing the importance of regular follow-up sessions to enhance compliance. Other treatments options for rupioid psoriasis reported in the literature include methotrexate, which is used for its immunosuppressive effect; cyclosporine, which inhibits T-cell activation and is beneficial for rapid control of psoriasis; intralesional steroids that directly reduce lesion inflammation; and adalimumab, another biologic that targets TNFα, a potent inflammatory mediator in psoriasis [4, 10]. We decided to use Ustekinumab because of the high BSA score, thick scales, and more suitable of the patient.

Ustekinumab is an IgG1κ monoclonal antibody that specifically binds the p40 subunit of cytokines IL-12 and IL-23, thereby inhibiting IL-12-induced activation of Th1 cells and reducing the secretion of proinflammatory cytokines such as TNFα and IL-2. It also decreases IL-23-induced activation of Th17 cells and reduces the secretion of the proinflammatory cytokine IL-17α [11]. These cytokines have effects on several cells like keratinocytes and epithelial tissue. This communication between the epidermis and the immune system causes keratinocyte hyperplasia seen in psoriasis [12].

Ustekinumab was approved for use in the pediatric population aged ≥6 years for the treatment of moderate-to-severe psoriasis, Ustekinumab offers a favorable dosing schedule of a subcutaneous injection every 12 weeks after an initial dose four weeks post the first administration [10].

Conclusion

Identifying rupioid psoriasis is crucial due to its implications for patient compliance, treatment complexity, and its association with psoriatic arthritis. This form of psoriasis often presents challenges in treatment plans, necessitating the selection of regimens that are both effective and manageable for patients. Ustekinumab, approved for children with moderate-to-severe psoriasis, has shown promising results in treating this demanding variant. Its efficacy highlights the importance of tailored therapeutic approaches in pediatric dermatology, especially for cases resistant to conventional treatments.

Conflict of interest

The authors declare no conflicts of interest.

Funding

There was no funding for this publication.

Ethical approval

No ethical approval was required for this publication.

Consent

Informed and written consent from the patient was taken prior to publication. Written consent for the publication of patient photographs and medical information was obtained.

Guarantor

Fouz Hassan.

References

1. Kharibam P, Laishram S, Choudhary N. *et al.* Concomitant interstitial lung disease with psoriasis. *Curēus* 2022;**14**:e26979. https://doi.org/10.7759/cureus.26979.
2. Antonio JR, de Lucca LCP, Borim MP. *et al.* Case for diagnosis. *An Bras Dermatol.* 2014;**89**:1003–4. https://doi.org/10.1590/abd1806-4841.20143156.
3. Chung HJ, Marley-Kemp D, Keller M. Rupioid psoriasis and other skin diseases with rupioid manifestations. *Cutis* 2014;**94**:119–21.
4. Feldman SR, Brown KA, Heald P. "Coral reef" psoriasis: a marker of resistance to topical treatment. *J Dermatol Treat* 2008;**19**:257–8. https://doi.org/10.1080/09546630802032611.
5. Thatiparthi A, Martin A, Liu J. *et al.* Biologic treatment algorithms for moderate-to-severe psoriasis with comorbid conditions and special populations: a review. *Am J Clin Dermatol* 2021;**22**:425–42. https://doi.org/10.1007/s40257-021-00603-w.
6. Rowawi R, Dwipangestu G, Suwarsa O. *et al.* A successful treatment of Ostraceous psoriasis associated with psoriatic arthritis in children: a case report. *Psoriasis: Targets Ther* 2020;**Volume 10**:61–6. https://doi.org/10.2147/PTT.S285832.
7. Ramirez-Bosca A, Navarro-Lopez V, Martinez-Andres A. *et al.* Identification of bacterial DNA in the peripheral blood of patients with active psoriasis. *JAMA Dermatol* 2015;**151**:670–1. https://doi.org/10.1001/jamadermatol.2014.5585.
8. Liu S, He M, Jiang J. *et al.* Triggers for the onset and recurrence of psoriasis: a review and update. *Cell Commun Signal* 2024;**22**:108. https://doi.org/10.1186/s12964-023-01381-0.
9. Han J, Luo D, Han X. Effects of Cefixime on immune functions and inflammatory factors in children with urinary tract infection and targeted nursing strategies. *Arch Esp Urol* 2023;**76**:313–8. https://doi.org/10.56434/j.arch.esp.urol.20237605.36. PMID: 37545149.
10. Cavazos A, Benjanuwattra J, Morataya C. *et al.* Rupioid psoriasis presents with extensive hyperkeratotic plaques. *Case Reports* 2023;**16**:e252500–0. https://doi.org/10.1136/bcr-2022-252500.
11. Necas M, Vasků V. Ustekinumab in the treatment of severe rupioid psoriasis: a case report. *Acta Dermatovenerol Alp Pannonica Adriat* 2010;**19**:23–7.
12. Griffiths CEM, Strober BE, van de Kerkhof P. *et al.* Comparison of ustekinumab and etanercept for moderate-to-severe psoriasis. *N Engl J Med* 2010;**362**:118–28. Available from. https://doi.org/10.1056/NEJMoa0810652 https://www.ncbi.nlm.nih.gov/pubmed/20071701.