



A rare entity: Keratoderma blenorrhagicum in a patient with Reiter's syndrome

Nadir bir antite: Keratoderma blenorajikumlu bir Reiter sendromu

● Şirin Yaşar, ● Dua Cebeci, ● Zeynep Altan, ● Pembegül Güneş*, ● Sema Aytekin

Istanbul Haydarpaşa Numune Training and Research Hospital, Clinic of Dermatology; *Clinic of Pathology, Istanbul, Turkey

Abstract

Reiter's syndrome is a systemic disorder, originally occurs as a triad of arthritis, urethritis and conjunctivitis. Cutaneous finding consist of a palmoplantar keratoderma, circinate balanitis or vulvitis, psoriasis-like skin lesions, and buccal ulcerations.

We present a case of Reiter's syndrome in a 25-year-old woman who developed the typical skin lesions - keratoderma blenorrhagicum and circinate vulvitis.

Keywords: Reiter's syndrome, keratoderma blenorrhagicum, circinate vulvitis

Öz

Reiter sendromu, artrit, üretrit ve konjunktivit üçlüsünden oluşan sistemik bir hastalıktır. Deri bulgusu olarak palmoplantar keratoderma, sirsine balanit ya da vulvit, psoriasis benzeri deri lezyonları ve bukkal mukozada ülserasyonlar izlenir. Burada 25 yaşında, deride keratoderma blenorajikum ve sirsine vulvit ile karakterize Reiter sendromlu bir olgu sunulmaktadır.

Anahtar Kelimeler: Reiter sendromu, keratoderma blenorajikum, sirsine vulvit

Introduction

Reiter's syndrome (RS) is a systemic disorder, occurring as a triad of arthritis, urethritis and conjunctivitis. The term RS is usually used as a synonym for reactive arthritis (ReA) and ReA is used instead of RS. The most common skin findings are palmoplantar keratoderma, circinate balanitis or vulvitis, psoriasis-like skin lesions, and buccal ulcerations^{1,2}. The symptoms arise few weeks following urogenital tract infections (e.g. Chlamydia trachomatis and Ureaplasma urealyticum) or gastrointestinal tract infections (e.g. Yersinia and Shigella infections). The pathogenesis of ReA is unknown but it has been defined that people with a particular genetic type called human leukocyte antigen (HLA)-B27 have an increased risk of developing the syndrome (about 80% of people with ReA carry this gene). Skin and mucosal involvement is observed in about 10% of cases. We present a case of ReA in a 25-year-

old woman with keratoderma blenorrhagicum of genitalia and oral aphthous ulceration.

Case Report

A 25-year-old female was admitted to the gynecology clinic with the complaints of painful genital ulcers, dysuria, and abdominal and joint pain. The patient had no history of sexually-transmitted disease. Her complaint of arthralgia started two months after urethritis treatment. Two weeks after the onset of acute arthritis and arthralgia, the patient developed red patches on the palms and soles, which quickly evolved to circinate plaques. The patient had fever, fatigue and malaise. Dermatologic examination revealed erythematous, confluent, desquamative papulonodules and pustules on the palms, feet, fingers and toes, and diffuse yellowish hyperkeratotic plaques on the soles (Figure 1a-c).

Address for Correspondence/Yazışma Adresi: Şirin Yaşar MD, İstanbul Haydarpaşa Numune Training and Research Hospital, Clinic of Dermatology, İstanbul, Turkey
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ORCID ID: orcid.org/0000-0002-4110-1874

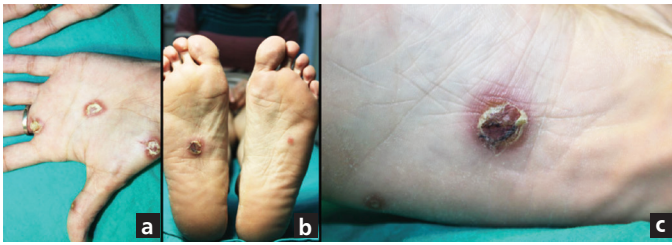


Figure 1. a-c) Diffuse yellowish hyperkeratotic plaques on the palms and soles



Figure 2. a-c) Aphthous ulcers were detected on lips and circinate vulvitis occurred on vulvae

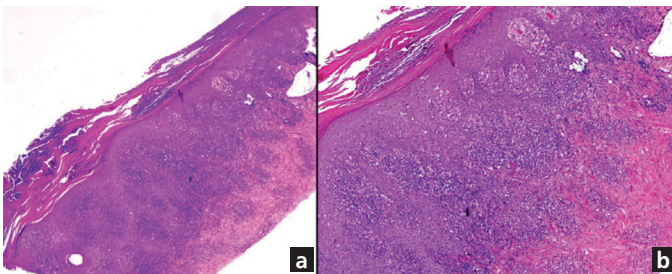


Figure 3. a, b) Skin biopsy showed psoriasiform changes- hyper- and parakeratosis, acanthosis, elongation of the rete ridges in the epidermis and mixed inflammatory infiltrate in the upper dermis

An ovoid major aphthous ulcer with circumscribed margins, covered with a whitish membrane and an aphthous ulcer with gray base were detected on the soft tissues of the lip. In the genital mucosa, multiple ulcers, features of painful, isolated or tending to merge circinate, elongated vaginal mucosa-located in the right posterior of the labium majus were detected (Figure 2a-c). Urinalysis was normal and urine, stool and urethral swab cultures were negative. Routine hemogram, rheumatoid factor (RF), and erythrocyte sedimentation rate were within the normal limits. Venereal disease research laboratory and human immunodeficiency virus (HIV) (ELISA) test were negative. Antistreptolysin O titer was found to be 325 (0-200 IU/mL). The C-reactive protein level was within the normal range (<0.8 mg/dL). HLA-B27 haplotype was found to be positive. X-ray of the hand and feet showed normal radiological findings. Histopathological investigation showed psoriasiform changes, hyperkeratosis and parakeratosis, acanthosis, elongation of the rete ridges in the epidermis and mixed inflammatory infiltrate in the upper dermis (Figure 3a, b). The diagnosis of RS was made according to the typical history, clinical symptoms and histological findings. In this context, the described skin lesions were interpreted as keratoderma blenorrhagicum. The patient underwent topical and systemic therapy with antibiotics [Ciprofloxacin 750 mg 2x1 and Naproxen 2x1-non-steroidal anti-inflammatory drugs (NSAIDs)]. The hyperkeratotic papules and plaques were gradually resolved and desquamated. Complete clinical resolution of the skin lesions was observed in one month with treatment. Her joint symptoms subsided

in 1 week and skin lesions in 4 weeks. She was followed up for 3 months after remission and remained asymptomatic. Informed consent for publication of this report was obtained from the patient.

Discussion

During the First World War, Hans Reiter described RS but, it has been recognized since 1818³. In 1818, Brodie published a case of ReA after venereal infection, Fiessinger and Leroy reported four cases of urethritis, conjunctivitis and arthritis of the peripheral and spinal joints following an outbreak of bacillary dysentery in France and since then, the term "Fiessinger-Leroy syndrome" is used in France⁴.

ReA is characterized by the triad of urethritis, arthritis and conjunctivitis after a urogenital or gastrointestinal infection. Although our patient had also urethritis and arthritis, conjunctivitis was not detected. ReA is classified as a seronegative spondyloarthropathy, because the patients are negative for RF. The positivity of HLA-B27 haplotype is an important risk contributing to the development of ReA and 80% of patient have positivity of HLA-B27. It has been hypothesized that HLA-B27 is responsible for an autoimmune cross-reaction via sharing some molecular characteristics with bacterial epitopes⁵.

The following complaints and findings were investigated to diagnose ReA;

- Acute onset ReA, with arthritis, malaise, fatigue, and fever.
- Arthritis: asymmetrical oligoarthritis of the lower extremities is the most common symptom, dactylitis, enthesopathy, sacroileitis.
- Genitourinary tract: non-gonococcal urethritis associated with dysuria, urgency, and urethral discharge, meatal edema and erythema, prostatitis, vulvovaginitis, circinate balanitis.
- Eye: conjunctivitis, photophobia, burning of eyes, cataract, keratitis, anterior uveitis may be seen.
- Gastrointestinal tract: diarrhea, abdominal pain.
- Skin-nails: keratoderma blenorrhagicum, onychodystrophy and severe psoriasiform dermatitis commonly involve the flexures, scalp, palms, and soles in HIV-positive patients.

Cutaneous involvement includes psoriasiform lesions (keratoderma blenorrhagicum) (in about 10%), painful and erosive lesions on the tips of the fingers and toes, nail dystrophy (20-30%) and erythroderma¹. Painful, aphthous ulcerations occur in oral and pharyngeal mucosae. Our patient had malaise, fever, fatigue, abdominal pain and arthralgia. Antibiotic treatment was given two months ago for urethritis. Acute arthralgia was developed two weeks ago and then painful, aphthous oral ulcer, circinate vulvitis and psoriasiform hyperkeratotic eruption (keratoderma blenorrhagicum) occurred on her hands and feet.

Psoriasiform epidermal hyperplasia and neutrophilic microabscesses in the stratum corneum are the most significant findings. Skin biopsy obtained from the psoriasiform lesion showed psoriasiform changes hyperkeratosis and parakeratosis, acanthosis, elongation of the rete ridges in the epidermis and mixed inflammatory infiltrate in the upper dermis^{2,3}.

Non-steroidal anti-inflammatory drugs and sulfasalazine are most commonly used in the treatment of ReA. If the NSAIDs are not helpful, steroids are administered⁶. Today, infliximab, etanercept, adalimumab, golimumab, and certolizumab, are shown to achieve partial or full remission of symptoms⁷. Non-biologic disease-modifying anti-rheumatic drugs, such as methotrexate and sulfasalazine, are

effective in resistant cases⁸. The case we presented shows the typical localization, morphologic features and clinical course of keratoderma blennorrhagicum as a characteristic cutaneous manifestation of ReA. The foremost finding was genital ulceration in ReA which can be included among disorders causing genital ulceration and must be kept in the differential diagnosis of genital ulceration.

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Informed Consent: Informed consent was taken.

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Authorship Contributions

Surgical and Medical Practices: Ş.Y., Concept: Ş.Y., D.C., Design: D.C., Z.A., Data Collection or Processing: Ş.Y., D.C., Analysis or Interpretation: Ş.Y., D.C., S.A., P.G., Literatür Arama: Ş.Y., D.C., Literature Search: Ş.Y., D.C., Writing: Ş.Y., D.C., Z.A., S.A., P.G.

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References

1. Uystepruyst P, Tellier E, Dardenne B, Darimont M: Reiter's syndrome: apropos of a case Acta Urol Belg 1998;66:29-33.
2. Reveille JD: HLA-B27 and the Seronegative Spondyloarthropathies. Am J Med Sci 1998;316:239-49.
3. Graham RM: Reiter's Disease. Ed: Champion RH, Burton JL, Burns DA, eds. Rook/Wilkinson/Ebling text book of Dermatology. 6th ed. London (UK). Blackwell Science 1998: 2759-71.
4. Hodgetts TJ: Reiter's disease: an historical review of a soldiers' disease. J R Army Med Corps 1990;136:170-2.
5. Eapen Br: A new insight into the pathogenesis of Reiter's syndrome using bioinformatics tools. Int J Dermatol 2003;42:242-3.
6. Palazzi C, Olivieri I, D'Amico E, Pennese E, Petricca A: Management of reactive arthritis. Expert Opin Pharmacother 2004;5:61-70.
7. Ahogo KC, Bataille M, Lasek A, Wantz M, Lebas D, Modiano P: A case of reactive arthritis (oculo-urethro-synovial syndrome) treated with infliximab Ann Dermatol Venereol 2017;144:804-5.
8. Kanwar AJ, Mahajan R: Reactive arthritis in India: a dermatologists' perspective. J Cutan Med Surg 2013;17:180-8.