

## Atypical Reiter's Syndrome

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**Genital ulceration is a rare manifestation of Reiter's syndrome (RS). We report a case of a young Asian male with atypical Reiter's syndrome who presented with painful and malodorous perianal and genital ulcerations, low back pain, and psoriasiform skin lesions. A review of the literature revealed only 4 reported cases of Reiter's syndrome and genital ulceration, all in females. To our knowledge perianal and genital ulceration in a male patient with RS has not been described before.**

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Classic Reiter's syndrome (RS) is characterized by aseptic inflammatory arthritis, urethritis or cervicitis and conjunctivitis. Cutaneous manifestations consist of a palmoplantar keratoderma, circinate balanitis or vulvitis, psoriasis-like skin lesions, and buccal ulcerations. The symptoms occur few weeks following infectious disease of the urogenital tract (e.g. *Chlamydia trachomatis* and *ureaplasma urealyticum*) or the gastrointestinal tract (e.g. yersinia and shigella)<sup>1-3</sup>. These inflammatory reactions occur in individuals with a genetic predisposition and are triggered by bacterial antigens that interact mostly with human leukocyte antigen (HLA)-B27 positive. Sixty to 80% of patients with RS are HLA-positive<sup>4</sup>. There are no well recognized diagnostic criteria for RS and the diagnosis is usually based on medical history and clinical findings. Histopathology, positive HLA-B27, and positive results of infectious agents are supportive of the diagnosis. The presentation of RS may be atypical or incomplete as in our case.

The aim of this report is to describe an atypical case of RS in a young male with genital ulceration. A review of the literature found only four reported cases of RS and genital ulceration, all in females<sup>5-8</sup>. To our knowledge, this may be the first case of RS with genital ulceration in a male patient to be described in literature.

### THE CASE

A twenty-nine years old Asian male presented with five months history of painful perianal and genital ulcers with bleeding and malodorous discharge. Prior to his presentation, he was treated with several systemic antibiotics (nature unknown), for it was presumed to be an infection but no benefit was obtained. The patient had lost 6 kgs over the previous five months. His history was significant for redness and soreness of the eyes, an ulcer of the mucosa of the upper lip, swelling of the nail folds, dysuria, easy fatigability, and low back pain. He had no fever and no abdominal pain. The patient denied any significant past medical history including any of sexually transmitted diseases or diarrheal illness.

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Physical examination revealed perianal linear shallow ulcerations (see Figure 1) and scattered shallow ulcerations over the groin (see Figures 2 and 3). The edges of the ulcers were flat with normal surrounding skin. Several erosions over the penis and the scrotum were noted (see Figure 3). There were no other skin or mucosal lesions and no enlarged lymph nodes. Ophthalmologic examination revealed signs of dry eye syndrome.



**Figure 1: Shallow, Linear Perianal Ulcerations**



**Figure 2: Scattered Shallow Ulcers in the Groin**



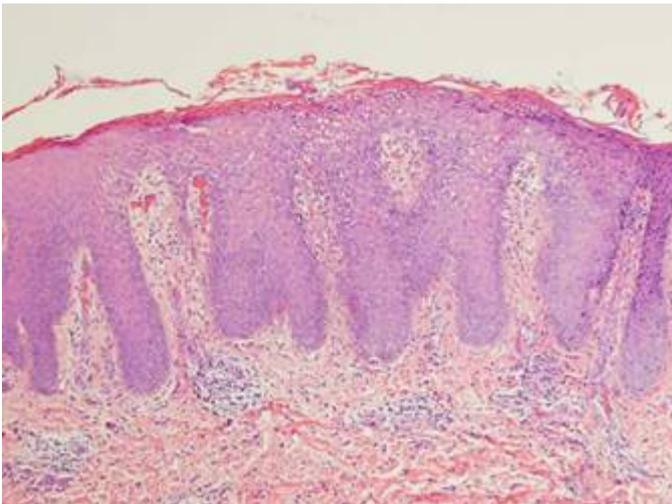
**Figure 3: Shallow Ulcers on Penis, Scrotum and Groin**

Laboratory studies revealed erythrocyte sedimentation rate of 37 mm per hour (0-20 mm/h); C-reactive protein 6.3 mg per liter (0-3 mg/L); total serum protein 87 gram per liter (64-82 g/L); and serum globulin 44 gram per liter (15-30 g/L). The results of routine chemistry profile and white blood cell count with differential were normal. Radiography of sacroiliac joints disclosed right sacroiliitis (see Figure 4).

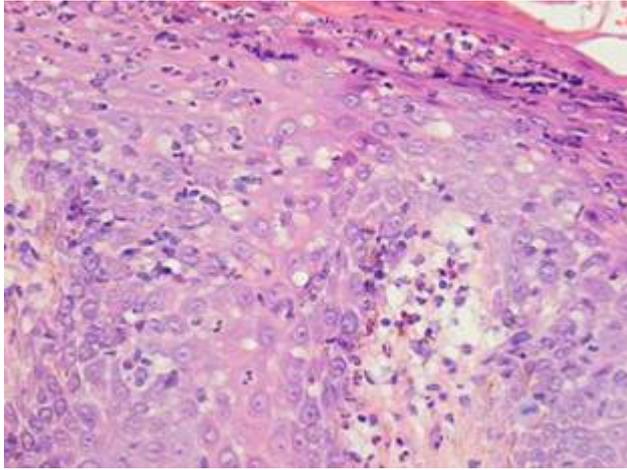


**Figure 4: Radiogram Showing Right Sacroiliitis**

The patient tested negative for hepatitis serology, HLA-B27, urethral culture for Chlamydia, stool microscopy and culture, and human immunodeficiency virus (HIV) enzyme-linked immunosorbent assay. He was non-reactive to the Venereal Disease Research Laboratory test (VDRL). Tissue cultures from all lesions produced no growth. A histopathologic examination of the biopsy specimen from the edge of one of the groin ulcers demonstrated hyperkeratosis, parakeratosis and psoriasiform epidermal hyperplasia with focal spongiosis and thinning of suprapapillary plates (see Figure 5). Within the stratum corneum, there were aggregates of neutrophils which were also present singly and scattered in the stratum spinosum (see Figure 6).



**Figure 5: Hyperkeratosis, Parakeratosis and Psoriasiform Hyperplasia of the Epidermis and Perivascular Mononuclear Cell Infiltrate**



**Figure 6: Neutrophilic Collections in the Stratum Corneum, with Neutrophils Scattered in the Epidermis**

Occasional lymphoid cells were seen in the lower epidermis. Numerous neutrophils were seen around capillaries in the papillary dermis; deeper to this, there was a perivascular inflammatory infiltrate rich in lymphocytes with few neutrophils. Fungal stains were negative and Donovan bodies were not seen. Based on clinical and histopathological findings, a diagnosis of Reiter's syndrome was made. The patient was treated with Neotigason 50 mg daily, potassium permanganate soaks, Voltaren® 50 mg daily and lubricant liquid for the eyes. Significant improvement with complete clearance of the skin lesions occurred within the first three months. Unfortunately, he was lost for follow-up.

## **DISCUSSION**

RS was first described by Hans Reiter during the first World War though it had been recognized since 1818<sup>9</sup>. It is a genetically determined disease in association with HLA-B27. Classically, RS is characterized by the triad of urethritis or cervicitis, conjunctivitis, and arthritis. Additionally mucocutaneous findings such as circinate balanitis in the uncircumcised penis and crusted erosions on the circumcised penis and on the scrotum may be observed<sup>1,2</sup>. Keratoderma blennorrhagicum and oral ulcers may be seen<sup>3</sup>. Although it is a disease of young males with HLA-B27, other HLA types that are linked with HLA-B27 and other age groups are not excluded<sup>3,10</sup>. Caucasians are most commonly affected, probably because of more prevalent HLA-B27 in this population group<sup>11</sup>. It occurs in response to enteric infections (such as salmonella, shigella, yersinia, campylobacter) or to urethral infection from *Chlamydia trachomatis*<sup>3</sup>. Severe cases of RS can occur as a late manifestation of HIV<sup>12</sup>. Other factors can induce the disease such as immunotherapy with BCG and alpha interferon, or following hepatitis B vaccination<sup>13-16</sup>.

The diagnosis in our patient rests on a constellation of signs and symptoms that are suggestive of RS. These include redness and soreness of the eyes, mouth ulcers, swelling of nail folds, dysuria, and sacroiliitis. More significant are the histopathological findings of psoriasiform epidermal hyperplasia and neutrophilic microabscesses in the stratum corneum. His response to Neotigason supports the diagnosis of Reiter's syndrome. Other diagnoses considered were psoriasis and Behçet's disease, the former for the histopathological findings and the latter for the mucosal and genital ulcers. RS fits best albeit the presentation is atypical or unusual in that there were no circinate balanitis, keratoderma blennorrhagicum or HLA-B27 positivity,

though the absence of any of these does not negate the diagnosis. The other interesting finding is the genital ulceration in this patient. Few cases of genital ulceration in females with RS have been reported but none in male patients<sup>5-8</sup>. This patient lacked overt urethral discharge, but dysuria in patients with RS can be the only manifestation of urethritis especially as this patient received several antibiotics prior to his presentation, which might have suppressed overt urethral discharge and contributed to the negative urethral and stool cultures.

## CONCLUSION

**We present this case as an atypical RS because of the discrepancies between it and classic RS. Foremost finding was genital ulceration, which has not been previously reported in male patients. RS can be included among disorders causing genital ulceration and must be kept in the differential diagnosis of genital ulceration.**

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