Ulcerative vulvitis in Reiter's syndrome

A case report

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SUMMARY In a case of acute Reiter's syndrome with severe vulvitis the diagnosis was based on the presence of a vaginal discharge and dysuria, arthritis, conjunctivitis, buccal ulceration, keratoderma blenorrhagica, and HLA B27 tissue-typing antigen. The vulval lesions were similar in appearance to those of circinate vulvitis. The acute histological change were confined to shallow ulceration with an inflammatory infiltration of the subjacent dermis. Coincidental lichen sclerosus et atrophicus was present, which could have been masked by the acute lesions.

Introduction

Reiter's syndrome is uncommon in women,¹ and vulval lesions have been reported previously in only three cases.^{2,3} In two of these cases shallow circinate erosions were the major feature.³ Our patient had similar lesions, and we report for the first time the histological changes as well as the clinical course.

Case report

A 28-year-old unmarried Caucasian woman presented with an acute asymmetrical inflammatory arthropathy affecting the right knee, both ankles, and the right shoulder. One month before its onset she had experienced explosive watery diarrhoea lasting three days. This was followed three weeks later by a vaginal discharge, dysuria, frequency of micturition, mild conjunctivitis, and painless buccal ulceration. She denied any previous urogenital symptoms.

CLINICAL FEATURES

She was febrile (temperature 39.7°C) and effusions were present in the right knee and ankle. There was a pronounced vulvitis consisting of a widespread area of erythema around the introitus and extending to the perianal region. The vulva was covered by many scattered, well-defined, round shallow ulcers, and an underlying atrophy was suspected. A thick white vaginal discharge was present, but the cervix appeared healthy. There was no urethral discharge.

Based on these appearances, and supported by a consultant dermatologist's opinion, a combined diagnosis of circinate vulvitis and suspected lichen sclerosus et atrophicus was made. There were no other skin lesions, and general examination including sigmoidoscopy was otherwise normal.

INVESTIGATIONS

Microscopy of vaginal smears and culture indicated the presence of candidosis. Urethral, vaginal, cervical, and rectal cultures for *Neisseria gonorrhoeae*, *Trichomonas vaginalis*, and *Chlamydia trachomatis* gave negative results. Stool cultures for *Salmonella* spp, *Shigella* spp, *Yersinia* spp, and *Campylobacter* spp also gave negative results. *Klebsiella oxytoca* was present in the first stool specimen but not on three following occasions. A mid-stream urine specimen contained $0.4 \times 10^9/1$ white blood cells, but culture for eubacteria gave a negative result. Synovial fluid aspirated from the right knee contained $9.4 \times 10^9/1$ white blood cells (90% mature polymorphonuclear cells and 10% small mononuclear cells with active chromatin), but culture for bacteria, including gonococci, gave negative results.

Paired sera for Widal testing, complement fixation tests for psittacosis, mycoplasmas, and herpes simplex virus, and microimmunofluorescence for chlamydia all gave negative results.

The haemoglobin concentration was 10.3 g/dl, white cell count $7.8 \times 10^9/1$ and erythrocyte sedimentation rate (Westergren) 120 mm in the first hour. Tissue typing showed that HLA B27 was present.

Fluorescent antibody tests for antinuclear factor and antibodies to mitochondria, gastric parietal cells,

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thyroid colloid, and thyroid epithelium gave negative results. Rheumatoid factor was absent. Protein electrophoresis showed an acute-phase reaction with raised alpha₂ globulin and lowered albumin. The Venereal Diseases Research Laboratory (VDRL) and *Treponema pallidum* haemagglutination (TPHA) tests gave negative results.

X-ray films of chest, knees, ankles, and feet showed no abnormalities, as did a rectal biopsy. Vulval biopsy showed changes consistent with lichen sclerosus et atrophicus (fig 1). Additional focal areas of ulceration were present with an inflammatory infiltration of the subjacent dermis (fig 2).

The patient's regular boyfriend (and only recent sexual contact) was examined. Neither *N gonorrhoeae* nor *C trachomatis* was isolated from the urethra.

TREATMENT AND COURSE OF ILLNESS

The patient was initially treated with rest in bed and given piroxicam, naproxen, and intra-articular steroid injections. Her fever subsided after 48 hours,

FIG 1 Vulval biopsy showing hyperkeratotic, non-ulcerated epidermis and papillary dermis composed of oedematous homogenised connective tissue consistent with lichen sclerosus et atrophicus. $\times 100$.

FIG 2 Vulval biopsy showing focal ulceration of the epidermis with acute inflammation of the immediately subjacent dermis. $\times 40$.

and the arthritis gradually disappeared over the next six weeks. On the advice of a consultant venereologist a two-week course of erythromycin and nystatin pessaries was given once all the specimens for culture had been collected. The vulval lesions, which were treated with salt baths and local miconazole nitrate 2% and hydrocortisone 1% cream, had healed two months later.

Two weeks after presentation she developed a few brown papules on the sole of the left foot, the appearance being consistent with keratoderma blennorrhagica; these disappeared spontaneously three weeks later.

Three months after her first presentation all symptoms and signs of Reiter's syndrome had resolved; the vulval atrophy did not progress. The erythrocyte sedimentation rate fell to 20 mm in the first hour and the haemoglobin concentration rose to 12.0 g/dl. All treatment was stopped. At five months she had developed atrophic lesions of the skin, which were typical of lichen sclerosus et atrophicus, over the left shoulder and reticulate lesions on the buccal

mucosa. She refused biopsy of these lesions. At one year the lesions had progressed no further, and there has been no recurrence of the Reiter's syndrome.

Discussion

This woman's presentation was typical of post-dysenteric Reiter's syndrome. She had at least five of the features of the clinical hexad⁴ (when this is modified for her sex)—namely arthritis, conjunctivitis, mucosal lesions, keratoderma blennorrhagica, and circinate vulvitis. The diagnosis was also supported by the presence of HLA B27. Although urethritis was not proved, a history of dysuria and white cells in the urine does suggest that it may have been present. Urethritis is not, however, essential to the diagnosis of Reiter's syndrome, as Paronen's⁵ large study found it to be absent in 23% of cases; indeed cystitis has been proposed as the female counterpart for urethritis.⁶

The candidosis was probably coincidental, as in one of the cases reported by Thambar.³ Unfortunately, we were unable to determine the precipitating organism for the Reiter's syndrome in this case.

The histological changes of ulceration are the first to be recorded. No other acute features were present; in particular no psoriatic changes could be found, but this has to be confirmed by future cases. A biopsy is indicated because underlying disease, which could otherwise be masked, may be missed.

The lichen sclerosus et atrophicus was probably a coincidental but important finding. An increased

incidence of organ-specific antibodies (absent in this case) has been noted in this condition,^{7,8} and vulval lesions may carry an increased risk of neoplastic change.⁹ Follow-up of such patients is advised¹⁰; it might have been overlooked in this case if the lichen sclerosus et atrophicus had not been confirmed by the biopsy.

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