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Painful Joints With Scaly Rash - A Case Report

Atiya Mahboob, Zafar Iqbal, Farrukh Iqbal, Shandana Tarique
Department of Medicine, Shaikh Zayed Medical Complex, Lahore.

SUMMARY

We present an uncommon case of Reiter's syndrome. The patient presented with pain in joints and scaly skin lesions. Presence of skin lesions helped in reaching the diagnosis.

INTRODUCTION

Reiter's syndrome is characterized by the presence of non-suppurative polyarthritis of more than one-month duration followed closely by a lower urogenital or enteric infection particularly in young men carrying HLA B-27 antigen. Inflammatory eye disease and mucocutaneous manifestations are common.

CASE REPORT

A 56 years old man was admitted in medical ward through Dermatology Out Patient Department, at Sheikh Zayed Hospital in February 2000. He presented with complaints of pain in the neck, right knee and left ankle joint which was accompanied by skin lesions on various parts of body for last two months. He also complained of an increase in frequency of micturition and watering from eyes. Pain in joints was continuous, aggravated by movement and relieved partially by simple analgesics. There was no history of trauma to limbs. Initially he could perform his routine work but due to increasing in intensity of pain his activity was gradually limited. These joints became red and swollen. At the same time he developed skin lesions on medial side of feet, front of legs, right side of lower abdomen including external genitalia and right arm. These lesions were multiple, itchy, red, raised, varying from half to one centimeter in size, round to oval in shape with watery discharge and crust formation. The patient developed polyuria with an urge to micturate after every 1-2 hours. Urine was pale in color and was about half a cup in quantity. At the end of stream he experienced extreme burning sensation in urethra. There was no history of passage of pus, blood, gravel particles or froth in urine. He had no complaint of increased intake of water or abdominal pain. He also complained of bilateral ocular watery discharge that remained through out the day and got worsened on exposure to light. He had no pain, purulent discharge, redness or gritty sensation in his eyes. His vision was not affected. He had low-grade intermittent fever during this period, chills accompanied it but there was no evening rise or night sweats.

On systemic review, patient gave history of cough with occasional white expectoration. There was no history of shortness of breath, wheeze, palpitation or chest pain. He had no history of nausea, vomiting, heartburn, altered bowel habits, weakness of any part of body, loss of sensation, photosensitivity, oral ulceration or bleeding from any part of the body. He had occasional mild pain in joints of lower limbs for the last 14 years. There was no past history of diarrhea, any major urinary complaint, polyuria, polydypsia or surgical operation. Personal and family history was unremarkable.

At the time of examination, patient was conscious and fully oriented in time and space. He was unable to sit or stand due to pain in his various joints. He was pale and having temperature of 100°F. Examination of locomotor system revealed swelling of left ankle and right knee joints. These were red, tender and mildly warm. Active and passive movements were limited. No deformity of feet, toes, hands or fingers were noted. Movements of neck in all directions were limited. Tenderness

was present in lower cervical region. Rest of spine was normal. Dermatological examination showed multiple round to oval papulonodular lesions of half to one centimeter in size on feet, shins, lower abdomen and right arm. These were red, crusted having collarette of scales at periphery surrounded by erythematous halo (Fig. 1). Crusted vesiculopustular lesions were present on scrotum, shaft of penis and around external urethra coalescing to form polycylic and circinate pattern. Palatal mucosa was red in color with no ulceration. Dorsum of tongue was normal. There was bilateral pterygium formation and excessive watery discharge in both eyes. Early lenticular changes were noted on slit lamp examination. No signs of uveitis were present. Fundi were normal. Rest of the systemic examination was insignificant.

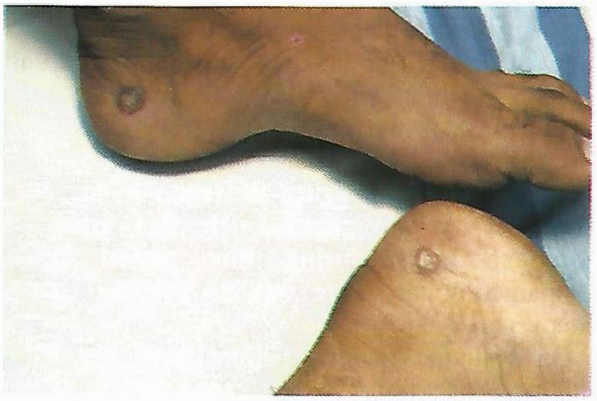

Fig. 1: Typical psoriasiform lesion on medial sides of feet.

A provisional diagnosis of Reiter's syndrome was made. His haemoglobin was 9.7 gm/dl with normal peripheral picture. Total count was $6.6x10^2/mm^3$ with 82% polymorphs. Platelets were $432,000/mm^2$. His ESR was 120mm in first hour. Urine examination showed 20-25 pus cells per high field. His renal function tests, serum electrolytes, uric acid, CPK, rheumatoid factor, antinuclear antibodies, anti-double stranded antibodies, VDRL and HIV were negative. Stool complete examination was normal. Repeated urine and blood cultures showed no growth. Biopsy of skin lesion showed psoriatic pattern with subcorneal pustule formation.

X-ray of left foot and ankle joint revealed presence of spurs at insertion of plantar fascia and tendoachillis (Fig. 2). X-ray spine showed syndesmophyte formation and squaring of vertebral bodies.

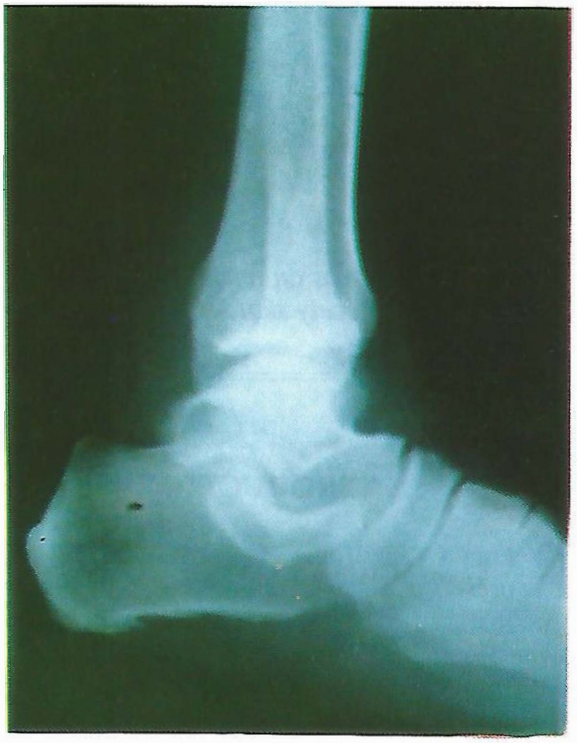

Fig. 2: X-ray foot showing presence of spurs at insertion of plantar fascia and tendoachillis.

Patient was given analgesics, steroids preparation for skin lesions and oral tetracycline. Initially he responded to treatment but after two weeks his condition became static and after 4 weeks injection methotrexate was added for his joint complaint.

DISCUSSION

Reiter's syndrome is a genetically determined and often protracted host response to specific antecedent enteric or genitourinary infection. The earlier description of characteristic triad of arthritis, conjunctivitis and uretheritis following enteric or venereal infections was given by Stoll in 1776[1] and by Brodie in 1818[2]. Feissinger and Leroy and Reiter independently described Reiter's syndrome after World War I.

Reiter's syndrome is a member of a family of diseases termed the sero-negative spondyloarthropathies that exhibit familial clustering; an inherited predisposition associated with the presence of certain major

histocompatibility gene complex (MHC) class I, *e.g.* HLA-B27, varying degrees of arthrocutaneus and ocular manifestations[3]. In addition to psoriasis and Reiter's syndrome, which have striking cutaneous involvement, the spondyloarthropathy group includes other entities, e.g., ankylosing spondylitis, enteric arthritis, Bechet's syndrome and Whipple's disease. The diagnosis of Reiter's syndrome can be simple or exceedingly difficult.

The classic presentation is a young male, after having a nonspecific enteric or urogenital infection develops fever, conjunctivits, urethritis, arthritis and extensive arthropathy. However in a patient with arthritis, ocular involvement and scaly cutaneous lesions following diseases should be differentiated.

Psoriasis

It is a chronic, recurrent inflammatory disorder of unknown etiology. Typical lesion is well demarcated erythematous papule covered by white silvery scales, mainly on the extensor surfaces of elbows, knees and sacral area. Scalp, palm, soles and nails are also affected. With sudden withdrawal of high doses of oral steroids, infections, pregnancy, hypocalcaemia, these lesions become studded with sterile pustules. Such pustules may be present on palms, soles, and trunk or under surface of nails. The condition may become generalized. Tongue, buccal mucosa and even the esophagus may be involved. There may be circinate or erosive lesions on the tongue forming geographical tongue. Conjunctivitis, keratitis, iridocyclitis, uveitis all may occur especially in pustular psoriasis. Prevalence of psoriasis is 1.5-3.5% in Western Europe. Estimates of the prevalence of arthritis in psoriatic patients vary from 6-34%. Female and male ratio is 1.3:1. In 65% of cases skin lesions precede arthritis while in 16% skin and joints are affected simultaneously[4]. Following types of joint involvement is seen in psoriasis:

1. Predominantly peripheral mono or asymmetrical oligoarthritis.
2. Classic form with predominantly distal interphalangeal arthritis.
3. Predominantly symmetrical, rheumatoid-like polyarthritis. Rheumatoid arthritis (RA) factor is negative.
4. Arthritis mutilans, relatively uncommon, severely deformed arthritis mainly affecting fingers and toes. Gross osteolysis may cause digital foreshortening and ankylosis.
5. Predominantly axial arthritis. Pseudo spondylitis and/or sacral ileitis, with or without peripheral arthropathy.

The diagnosis is confirmed by histopathology of skin lesion, revealing hyperkeratosis, parakeratosis, absent or reduced granular layer, acanthosis, subcorneal munro micro - abscesses, elongated retepegs, papillomatosis and thinning of epidermis overlying papillae.

2. Lupus Erythmatosus (LE)

Joint, cutaneous psoriasiform lesions, eye and mucosal involvement is seen in following types of LE.

a. Sub acute cutaneous LE
b. Chronic LE or DLE
c. Systemic LE
d. Antinuclear antibody (ANA) negative SLE

a. Sub acute cutaneous LE

This subset comprises about 10% of patient with L.E who have annular polycyclic (one-third) or non-scarring papulosquamous (two third). Lesions are usually above the waist, particularly around the neck, on the back, front of trunk and on the outer aspects of arms. The borders may show vesiculation and crusting. Follicular plugging and hyperkeratosis are less prominent and the lesions resolve leaving grey white hypopigmentation and telangiectasis Fifteen percent of patients have psoriasiform lesions or erythroderma has been noted. About half of the patients fulfill the criteria for SLE of American Rheumatism Association (ARA) with arthritis. The most frequent features, i.e. fever, malaise and CNS involvement occur but renal disease is mild and infrequent. ANA is found in 60%, anti Ro in 80% and Anti- La in 40% of patients. In 88% of patients with papulosquamous LE, direct immuno-fluoresence is positive[5]. Mucous membrane of nose shows ulceration. There may be perforation of nasal cartilage leading to depressed bridge of nose. Oral mucosa may show small erythematous purpuric areas. Superficial painful erosions, white patches resembling oral thrush or even infarction of the

tongue may occur. In females erythema of vulva and perianal area occurs. Vulval and vaginal ulceration may be present. Nail folds may show vascular necrosis and ragged cuticles and dilated blood vessels. Nail plate changes include pitting, ridging, splinter haemorrhages, onycholysis, striate leuconychia and red lunulae. Nail changes occur in 25% of patients. Recurrent Osler's nodes and clubbing may also be seen. Gangrene of tips of fingers may develop insidiously.

b. DLE[6]

The patient usually present with a rash. There is history of Raynaud's phenomenon or chilblain. Joints pain occurs in 25% of patients. Lesions are present on any part of the body but mainly the exposed parts. Typical lesion is well-demarcated plaque hyperpigmented at the periphery, hypo-pigmentation, atrophy and adherent scales in the centre. When these scales are removed, follicular plugging is seen. Sometimes this scaling is so prominent that it is easily mistaken for psoriasis. In 24% of patients mucous membrane are involved. Ulceration of nasal mucosa occurs in 9% of patient. On buccal mucosa, lichen planus like plaques are present. Superficial erythematous erosions can be seen on buccal mucosa, tongue and palate. Oral lesions may resemble leukoplakia. Erythematous lesion occur on vulva and around the anus. In eyes, velvety edema and marked redness of conjuntiva may occur. Eyelids are red, slightly infiltrated and always scaly. The horny spike of follicular plugging is seen on removal of scale. The plaques may be associated with conjuntival scarring and symblepharon. Superficial punctate keratopathy and stomal keratitis have been reported. Nail may show subungual hyperkeatosis, red blue discoloration, longitudinal striae and crumbling of nail.

c. ANA-negative SLE

Patient has similarities with patients suffering from SCLE. In about 5-10% of patients with SLE, ANA cannot be demonstrated by using standard substrates. Clinically the patients have a non-scarring malar flush, oral ulceration and photosensitivity with papulosquamous or annular lesions on face, trunk and arms[7]. Arthritis, serositis, renal disease and haemotological involvement is also present. Involvement of joints varies from 25% in DLE to 90% of patients of SLE. A rheumatoid like deformity is present in 25% of cases with marked soft tissue swelling especially of dorsa of fingers, hands and wrists. Radiological changes are uncommon. RA factor is positive in 40% of cases. Migratory polyarthritis, sacroiliitis may be present. Salmonella infection occurs in patients in SLE and may be associated with septic arthritis.

Histopathology of skin lesion shows:

i. Liquefactive degeneration of basal cell layer.
ii. Degenerative changes in the connective tissue, consisting of hyalinization, edema and fibrinoid changes.
iii. A patchy dermal lymphocytic infiltrate with a few plasma cells and histiocytes particularly around the appendages, which may be atrophic. Other changes include hyperkeratosis without parakeratosis, keratotic plugging of the hair follicles and glandular orifices, atrophy or acanthosis of prickle cell layer.

Lupus vulgaris (L.V)

In some cases lupus vulgaris develops at the site of primary inoculation, in others it occurs by direct extension from underlying infected glands or joints or by lymphatic spread from mucous membrane of nose or throat. Rarely multiple foci arise by haematogenous dissemination particularly following measles. The earliest lesion is a tiny, reddish brown, flat plaque of soft almost gelatinous consistency. The lesion gradually becomes brown, raised, more infiltrated and by peripheral extension gyrate or discoid in shape. In plaque form scaling occurs and leads to a psoriasiform appearance especially on legs and arms[8]. Plaque covers a large part of body. Irregular scarring leave islands of activity. The edge becomes thickened and hyperkeratotic. Nasal, buccal or conjuctival mucosa is involved in many cases and must be examined carefully. One percent of patients with tuberculosis have skeletal involvement. Joint involvement in children occurs in the primary stage of the disease. In adults it is secondary to pulmonary or renal disease. It is due to haematogenous spread to subchondral bone or the spinal intervertebral discs.

There is usually monoarticular arthritis affecting the hip or knee in 30% or the sacroiliac or other joints in 20%. In 50% of patients there is spinal involvement. There is insidious onset of pain, swelling and restriction of movement associated with general symptoms of malaise anorexia and night sweats. Histopathological examination of skin lesion shows intradermal focus of caseous necrosis surrounded by well-formed epithelioid granulomas containing Langhan's giant cells.

Leprosy (B.B)

Multiple cutaneous macules, papules, plaques, or annular shape - plaque with a "punch-out" appearance are characteristics. These are dry, hypoasthetic with loss of hair and sweat. One or more nerves are likely to be thickened and non-functioning. The lesions in type 1 reactions become erythematous, edematous, and scaly[9], may desquamate or rarely ulcerate. Nerves become tender with loss of sensory and motor functions. Blindness due to leprosy occurs in at least 25% of patients. Lagophthalmos results from parasis of orbicularis occuli. Damage to ophthalmic branch of trigeminal nerves causes anesthesia of cornea and conjunctiva, leaves it at risk of minor trauma and ulceration. Iritis, iridocyclitis, uveitis occurs in type 2 reaction. Patient has pain in most of the joints in type 1 reaction but if he also develops simultaneous type 2 reaction, frank arthritis affecting many joints develop. Histopathology of skin lesion shows diffuse epithelioid cell granuloma in the dermis. There is narrow clear papillary zone called grenz zone.

Secondary Syphilis

Secondary syphilis is initiated by the appearance of the macular rash about 8 weeks after the exposure. The initial lesions are macular or maculopapular. Characteristically the lesions are non-itchy, coppery-red in colour and symmetrically distributed. The typical papule is firm, round to oval and shiny covered with thin layer of scales forming papulosquamous syphilide[10]. Psoriasiform papules of palms and soles are especially common in black people. On macerated skin surfaces and mucous membrane, eroded, weeping papules with a tendency to hypertrophy appear. On the genital area, besides small-eroded exudating papules, large eruption of hyperkeratotic, coalescing papules-condylomata lata may be seen. On the mucous membrane the basic papular eruptions are less distinctive, but they tend to be symmetrically distributed. As the surface epithelium dies, it turns grey and forms round or oval mucous patches on the palate, inner aspects of lips and cheeks. Ulceration is not common. Sharply defined, round or oval lesions devoid of dead epithelium may appear on the tongue and may be associated with flattened papillae. Non-specific symptoms are low grade fever, headache, pain in joints and muscles which occur in early stages of the disease. A generalized microlymphadenitis is more pathognomonic. Iridocyclitis, nephritis, hepatitis and splenomegaly are not rare. Histopathology of papule shows endothelial swelling of vessels of the dermis with a sleeve-like infiltrate mostly of plasma cell.

Therefore, whenever we observe a patient with joint pain accompanied by scaly skin lesions and mucosal involvement, the possibility of Reiter's syndrome should be kept in mind.

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The Authors:

Atiya Mahboob,
Assistant Professor,
Department of Dermatology
Division of Medicine,
Shaikh Zayed Medical Complex,
Lahore.

Zafar Iqbal,
Professor,
Department of Medicine,
Shaikh Zayed Medical Complex,
Lahore.

Farrukh Iqbal,
Associate Professor,
Department of Medicine,
Shaikh Zayed Medical Complex,
Lahore.

Shandana Tarique
Medical Officer,
Department of Medicine,
Shaikh Zayed Medical Complex,
Lahore.

Address for Correspondence:

Atiya Mahboob,
Assistant Professor,
Department of Dermatology
Division of Medicine,
Shaikh Zayed Medical Complex,
Lahore.