CASE REPORT

PSORIATIC ARTHRITIS MUTILANS IN A BLACK NIGERIAN PATIENT: A CASE REPORT

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ABSTRACT

Background: Psoriatic Arthritis Mutilans (PAM) is a rare destructive form of arthritis, especially in blacks and its diagnosis and treatment still remains a challenge.**Case Report**: A 55-year-old house wife with 30-years history of psoriatic skin lesions, developed swellings and pain of the small joints of the hands and feet, wrists, elbows, shoulders and knees associated with low back pain and alternating buttock pain shortly after onset of the rashes. She had dystrophic, pitting, yellowish nail changes with sub-ungual hyperkeratosis. The joint swellings rapidly progress into shortening of the digits of her hands and feet, with resorption of the interphalangeal joints and subluxation of the metarcarpophalangeal joints (MCPJs). Her Body Mass Index at presentation was 19.8kg/m^2, she had subluxation of the MCPJs with shortening and telescoping of the 3rd, 4th and 5th digits of both hands and bilateral knee swelling with ankylosis. Her rheumatoid factor was negative, CRP was 36mg/dl, white cell count of 3.0×10^9 cells/L with predominant lymphocytosis (63%). Her serum urea, creatinine and uric acid were normal. Radiographs of the hands and feet showed 'pencil in cup' appearance with marked periosteal reaction and osteolysis, complete joint erosion and subluxation. A diagnosis of PAM was made using **C**l**AS**sification for **P**soriatic **AR**thritis (CASPAR) criteria. She was placed on methotrexate 10mg weekly and topical steroids with short course of naproxen and was advised for total knee replacement and biologic agents for her treatment. **Conclusion**: PAM still remains a challenge in terms of early detection of the characteristic phenotype and treatment.

Keywords: Neonatal thrombocytopenic purpura, Differential diagnoses, Haematological profile

INTRODUCTION

Psoriatic Arthritis (PsA) is an irreversible, progressive, inflammatory condition associated with bone damage, joint pain, stiffness, swelling and extra-articular manifestation such as enthesitis and dactylitis in patients with psoriasis. [1]Moll and Wright[2] defined PsA as an inflammatory arthritis associated with psoriasis in the absence of rheumatoid factor (RF) and classified it into five different types, based on the patterns of joint involvement: (*i*) polyarticular; (*ii*) distal interphalangeal (DIP); (*iii*) spondyloarthropathy; (*iv*) oligoarticular; and (*v*) arthritis mutilans; which is described as the most severe form of PsA. Psoriatic arthritis mutilans (PAM) is rare with an estimated prevalence among patients with psoriasis of 0.6% to 21%.[3] A review by Haddad A and Chandran V [3] highlighted that, research on arthritis mutilans is hampered by lack of accepted case definition and therefore difficulties in getting adequate epidemiologic, clinical and radiographic features as well as clues to pathogenesis. Psoriatic arthritis is rare in black Africans, especially arthritis mutilans.[4-6] In a study involving 384 patients with PsA conducted over seven years in South Africa, they reported no single case of black individual with PsA. [4] We report a case of 55 year old with Psoriatic Arthritis Mutilans (PAM) referred to our clinic from the general outpatient department of University of Maiduguri Teaching Hospital.

CASE REPORT

A 55-year-old housewife with a background history of pruritic silvery scaled skin lesions involving the scalp, elbows, knees and feet for 30-years (figure 1) was referred to the Rheumatology clinic of University of Maiduguri Teaching Hospital on account of hand deformities. She developed progressive swellings and pain of the small joints of the hands and feet, wrists, elbows, shoulders and knees few months after onset of the rashes. There was dystrophy, pitting, sub-ungual hyperkeratosis and yellowish discoloration of her nails. She reported having severe dandruff and scaring alopecia.

Within seven months after noticing the swelling, she had progressive shortening of the digits of her hands and feet, with resorption of the distal and proximal phalanges, subluxation of the metarcarpophalangeal joints with ulnar deviation (figure 2). In addition, she had low back and alternating buttock pain with early morning stiffness that is relieved by activity and taking non-steroidal anti-inflammatory drugs (NSAIDs). Her knees became increasingly painful with marked swelling and inability to walk or squat properly rendering her incapacitated (functional class IV [7]). She reported having dry eyes and mouth but no nodules, eye redness, photophobia or dactylitis. She is para 7+0, three years postmenopausal with no hypertension or diabetes mellitus and had no family history of psoriasis. Her Psoriasis Area and Severity Index (PASI) score at presentation was 5.8.

On further examination, her weight was 52kg, height 1.62m (Body Mass Index, 19.8kg/m^2), blood pressure of 90/60mmHg, temperature of 36.8^{o}C with silvery scale hyper- and hypo-pigmented skin lesions on the knees and feet and dandruff on the scalp (figure 1). She had tenderness and swelling of both wrists, ulnar deviation and subluxation of the MCPJs with shortening and telescoping of the 3rd, 4th and 5th digits of both hands (figure 2). Bilateral knee swelling with marked flexion deformity and reduced range of motion. She had no sacroiliac joint tenderness and other systemic examination were normal.

Investigation revealed a negative rheumatoid factor, CRP of 36mg/dl and ESR of 52 mm in the first one hour. Full blood count showed pack cell volume of 40%, white cell count of 3.0×10^{9} cells/L with predominant lymphocytosis (63%) and platelet of 340×10^{9} cells/L. She had urea of 2.5 mmol/L, creatinine of 48 µmol/L and uric acid of 187 µmol/l. Liver enzymes were normal. Radiographs of the hands and feet showed 'pencil in cup' appearance with marked periosteal reaction and osteolysis, complete joint erosion and subluxation (figure 3). A diagnosis of PAM was made using CASPAR criteria based on current history of psoriasis, nail changes, negative rheumatoid factor and radiographic features. She was placed on methotrexate 10mg weekly and topical steroids with short course of naproxen and was advised for total knee replacement.

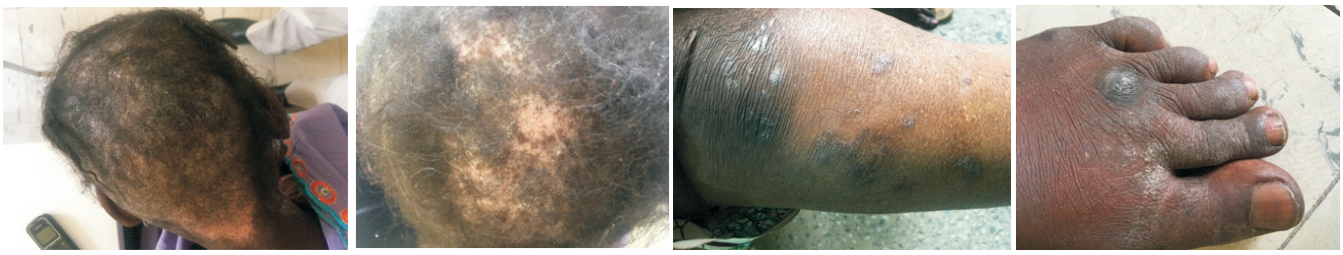

Figure 1: Psoriatic skin lesions in the index patient

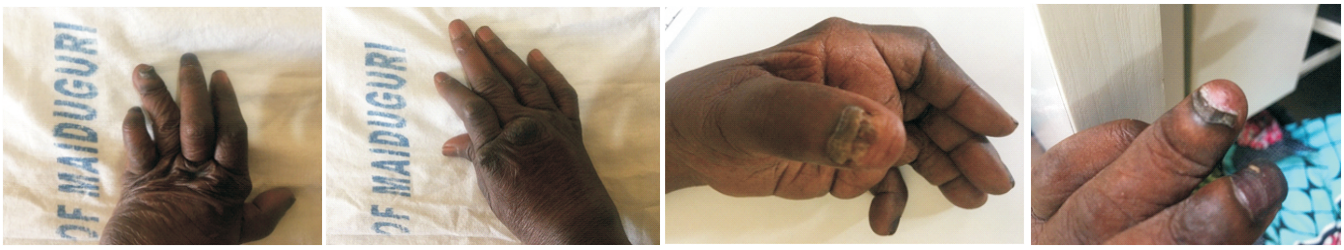

Figure 2: Hand and Nail deformities of the index patient.

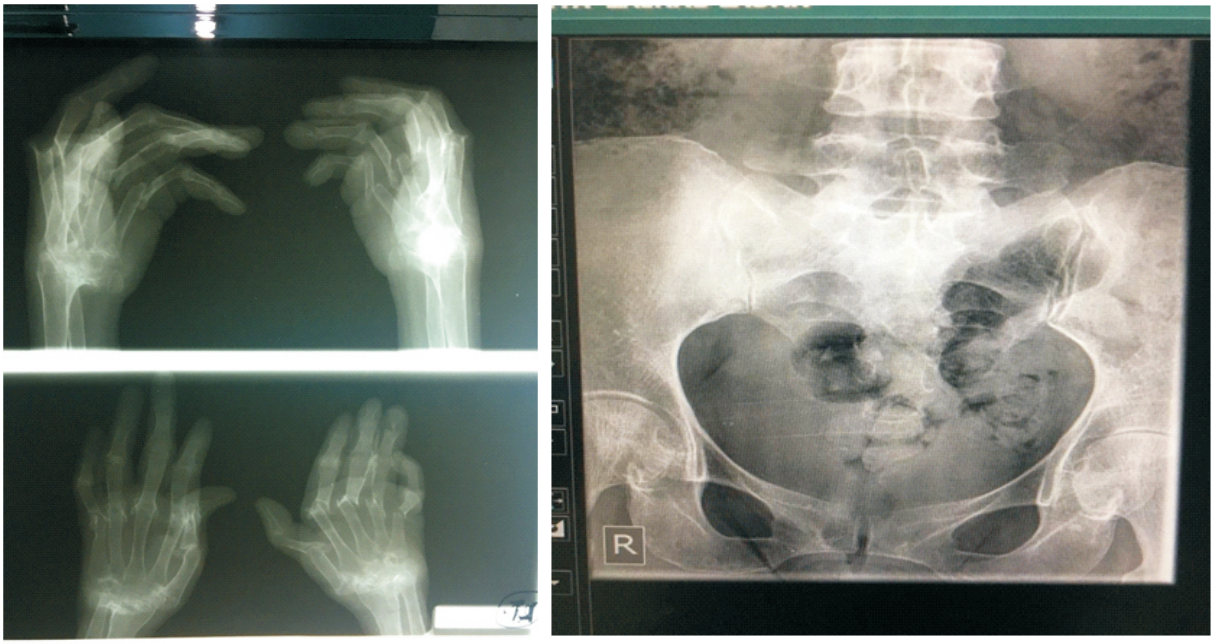

Figure 3: (a) Plain radiographs of the hands of the index patient showing marked destruction and osteolysis of the joints with "pencil in cup" deformities and (b) grade III and IV sacroilitis and pseudo-widening of the Sacroilicac joint (black arrow)

DISCUSSION

Psoriatic arthritis mutilans (PAM) is rare. The estimated prevalence among patients with psoriasis is between 0.6 to 21%, [3, 8] this wide range is due to lack of agreed clinical or radiographic definition of this sub-phenotype. There are few reports of PAM as part of patients with psoriasis in black Africans. [4-6] A case definition of PAM proposed by Moll and Wright [2] requires an arthritis manifesting as digital telescoping from severe osteolysis (opera glass finger), often associated with sacroilitis and classic radiographs showing whittling and cupping of phalanges, metacarpals or metatarsals, giving the classic appearance of pencil-in-cup deformity. In addition to the aforementioned features, the **G**roup for **R**esearch and **A**ssessment of **P**soriasis and **P**soriatic **A**rthritis (GRAPPA) in 2012[9]believed that the definition of arthritis mutilans should involve peripheral joints, especially of the hands and feet, but not axial joints. Both radiographic and clinical features were important, but radiographic features were believed to be more sensitive and a single joint involvement can also be used in diagnosis. [9]

Our patient has most of these features in addition to the skin lesions,however, the rapid nature of her joint destruction shortly after the onset of the skin lesions indicates a more aggressive course of her illness. The GRAPPA[9] initiative does not attached significance to the rapidity of bone destruction.

In a retrospective cohort study conducted by Jadon et al[10] comparing patients with PAM (n=36) and those without PAM (n=483), they reported PAM patients being younger in age, having more nail disease, marked impairment of functional status and more likely to have radiographic axial disease. A similar report from a multicenter study involving 360 patients indicates that PAM patients had longer disease duration, worse functional capacity, and more DIP joint involvement [11]. The index patient developed skin lesions at a younger age of 25 and PAM at age 26-years, had nail lesions and had a poor functional class too.

Intriguely, our patient did not report dactylitis, currently or in the past. In the Nordic countries, dactylitis was seen in 64% of patients with PAM, with 61% of them having dactylitis in the same finger or toe that had arthritis mutilans. [12]

The negative rheumatoid factor seen in this patient is consistent with the **C**l**AS**sification for **P**soriatic **AR**thritis (CASPAR) criteria requirement,[13] however, we were not able to perform an anti-citrullinated protein antibodies (ACPA) serology of the patient; Jadon et al [10] found no difference in ACPA positivity between patients with PAM and those without PAM, although some studies revealed thatACPA is seen in up to 10% of cases. [14]

Studies have shown that PAM patients have higher risk of having radiographic axial disease/sacroiliitis (OR 2.31/2.99) than non-PAM patients,[10] similarly, our patient had back pain with alternating buttock pain. Her plain radiograph revealed grade III and IV sacroilitis on the right and left respectively with classical pseudo-widening (Figure 3)

She was placed on methotrexate because of its affordability, availability and following the recommendations by the **Eu**ropean **L**eague **A**gainst **R**heumatism (EULAR) [15] updated in 2015 and was advice on total knee replacement therapy. Her symptoms have subsided but she however, needs biologics and/or other newer agents for her debilitating disease but the cost is prohibitive especially for individuals in low resource settings. [15] PAM, though rare, still remains a challenge in terms of early detection of the characteristic phenotype and management leading to significant impacts on the functional status of the patient.

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