CASE REPORT

A Psoriatic Arthritis Patient with Multiple Rare Complications – case report and review of literature.

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Abstract

A 28 year old male patient of psoriasis was admitted in a tertiary care hospital of Bangladesh with typical skin lesions for 18 years, nail changes for 16 years, arthritis for 12 years and eye changes for 3 years. In addition he had other rare extra articular complications like severe aortic stenosis, moderate aortic regurgitation and mild mitral regurgitation. He had a positive family history of psoriasis. HLAB27 is positive and X-ray of hands and feet showed classical findings of psoriatic arthritis. We presented the case to show the early age of onset, severity of the disease with rapid progression and multiple extra articular complications.

Key words
Psoriatic arthritis, psoriasis, extraarticular.

Introduction

Psoriatic arthritis is an autoimmune disease with known human leukocyte antigen (HLA)–associated risk factors. Psoriatic arthritis affects the ligaments, tendons, fascia, and joints, and it occasionally develops in the absence of detectable psoriasis. Psoriatic arthritis may occur at higher frequencies when skin involvement is more severe, especially when pustular psoriasis is present.[1] The National Psoriasis Foundation estimates that 10% to 30% of people with psoriasis also have psoriatic arthritis. Psoriatic arthritis can develop at any time, but for most people it appears approximately 10years after the onset of psoriasis. While psoriatic arthritis is most common in adults between the ages of 30 and 50 yrs, it can develop in anyone, including children. Psoriatic arthritis only affects people who have psoriasis. We presented a patient of psoriasis who has developed arthritis at a very early age. The extra articular complications are vivid and reached the severity of the disease within a short period.

Case Report

A 28year old male patient of psoriasis was admitted in a tertiary hospital of Bangladesh with the complaints of peeling of skin from the palm and sole for 18 years, ridging and destruction of nails for 16years, progressive joint deformity of small joints of hands and feet with gradual resorption of terminal phalanges resulting in shortening of fingers for 12 years.

He developed visual disturbances in the form of redness, irritation and watering from both eyes especially in bright light for 3 years. He had a family history of psoriasis. Her mother and two younger sisters are suffering from psoriasis.

On examination the patient was anxious looking with congested eyes & corneal haziness. Hand examination revealed, flexor deformity and tenderness of both DIP & PIP joints, resorption of the terminal phalanges resulting in shortening of fingers, swelling of the MCP joints of both hands [Fig-1].

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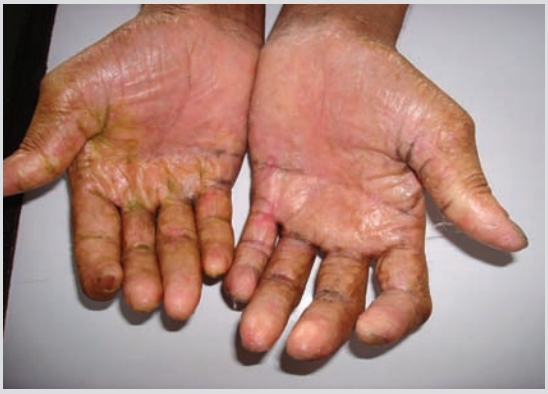

Figure 1: Psoriatic hand showing flexion deformity of DIP joints, resorption of terminal phalanx, extensive skin scaling and fissuring.

There is tenderness and flexion deformity of DIP joints of the feet. Examination of the eyes showed grossly reduced visual acuity of both eyes with poor fundal glow and bilat- eral bullous keratopathy. There was corneal haziness in the left eye [Fig-2].

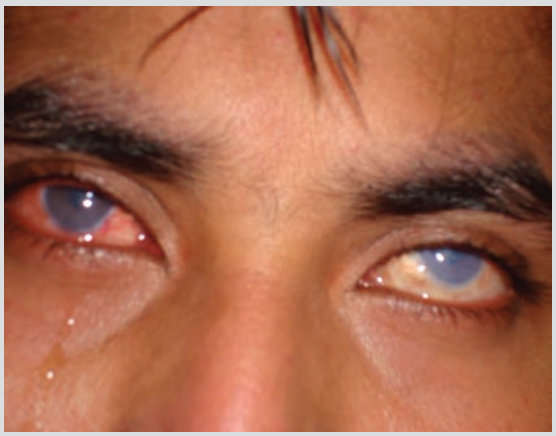

Figure 2: Eye changes showing corneal haziness, scleritis.

Examination of the skin revealed extensive palmo-planter erythematous plaque with scaling & fissuring [Fig-3], erythematous plaque with silvery scales around the umbilicus, positive Auspitz sign. There were transverse ridging, nail pitting, onycholysis and onychodystrophy of the nails of hands and feet. On examination of cardiovascular system there was both systolic and diastolic murmur in aortic area. Examination of other system was not significant.

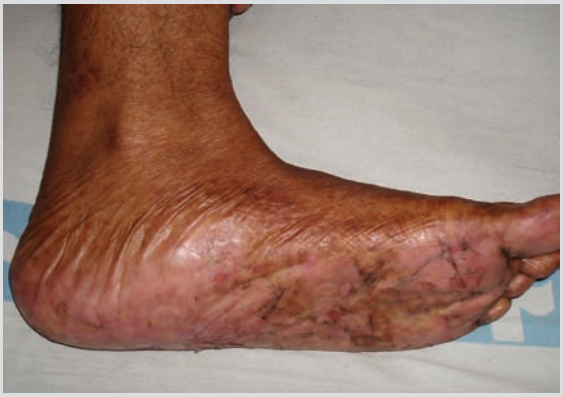

Figure-3: Erythematous plaques with scaling and fissuring of skin.

Laboratory investigations revealed high ESR with negative RF, positive HLA-B27.

X-ray of hands and feet showed joint space reduction in DIP joints; cyst formation and resorption of terminal phalanges; destruction of some DIP joints of hand; “Pencil in cup” appearance of the 1st, 4th & 5th DIP joints of hand. PIP and MCP joints are normal [Fig-4]. Echocardiography showed severe calcific aortic stenosis, moderate aortic regurgitation and mild mitral regurgitation.

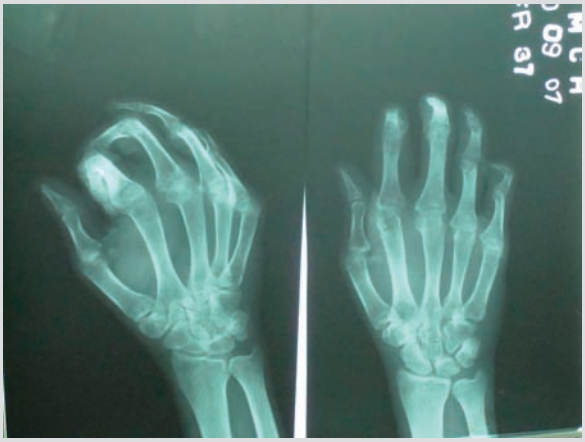

Figure-4: X-ray hand showing resorption of terminal phalanges, joint space reduction of DIP joints, cyst formation.

Discussion and review of literature

Psoriatic arthritis (also arthropathic psoriasis or psoriatic arthropathy) is a type of inflammatory arthritis that affects around 5-7% of people suffering from the chronic skin condition psoriasis.[2] In 1964, the American Rheumatism Association listed psoriatic arthritis as a clinical entity. However, diagnostic criteria have not been agreed upon, and several proposed definitions have stressed separate features of this multifaceted disease.

The high frequency of distal joint involvement in psoriatic arthritis compared with rheumatoid arthritis (RA) and arthritis mutilans (as an unusual but characteristic mani festation) has received special attention. The great variety of clinical manifestations was framed in the definition suggested by Moll and Wright in 1973, i.e., "An inflammatory arthritis associated with psoriasis, usually with negative sheep cell agglutination (SCA) test, i.e., rheumatoid factor."[3]

While many people consider psoriasis as a skin disease, its primary cause is a malfunctioning immune system. And it doesn't just affect the skin. Many of its worst effects can come from psoriatic arthritis, a swelling of the joints that develops in some people with psoriasis. Psoriatic arthritis causes symptoms like other types of arthritis -- stiff, painful and swollen joints -- and it can be serious. Untreated, psoriatic arthritis can cause bone loss and deformation of the joints. The course of psoriatic arthritis is usually characterized by flares and remissions. Experts don’t agree on how common psoriatic arthritis is. The National Psoriasis Foundation estimates that 10% to 30% of people with psoriasis also have psoriatic arthritis.[3]

While psoriatic arthritis is most common in adults between the ages of 30 and 50yrs, it can develop in anyone, including children.4 Psoriatic arthritis only affects people who have psoriasis. Even so, diagnosis can be difficult. The symptoms of psoriatic arthritis often appear years after the first signs of psoriasis on the skin. But sometimes the arthritis develops before the patient have any lesions. About 80% of those affected develop psoriatic arthritis after the onset of psoriasis, but in about 20% the arthritis occurs before the onset of psoriasis.[5]

Psoriatic arthritis is more common in white persons than in persons of other races. Men and women are affected equally; however, if the subsets of psoriatic arthritis are considered, male predominance occurs in the spondylitic form, whereas female predominance occurs in the rheumatoid form.[6]

Approximately 40% of patients with psoriasis or psoriatic arthritis have a family history of these disorders in first-degree relatives. The exact mechanism of the association between HLA and psoriatic arthritis is not clear. Psoriatic arthritis is associated with an increased frequency of HLA-B7 and HLA-B27 and a lower frequency of HLA-DR7 and HLA-Cw7.[7]

There are five different kinds of psoriatic arthritis. Asymmetric oligoarthritis makes up about 70% of all cases of psoriatic arthritis. It often involves one or a few joints, like the knee, hip, or fingers. Although it's frequently mild, it can sometimes be debilitating. The inflamed joints may be red and hands and feet may be swollen. Symmetric polyarthritis is the second most common form of psoriatic arthritis. It often causes symptoms in the same joints on both sides of the body. Symptoms are similar to rheumatoid arthritis, and symmetric

arthritis can cause permanent damage. Distal interphalangeal predominant (DIP), a less common form of psoriatic arthritis, affects the joints close to the fingernails and toenails. The nails are often affected too. Spondylitis can make movement painful, especially in the neck and back. It can also cause inflammation of the spinal column. Arthritis mutilans is a rare and often debilitating and destructive form of psoriatic arthritis.[8]

Arthritis generally is not considered to correlate strongly to any particular type of psoriasis or to the severity of the skin disease.[9] However, in one study, arthritis was noted more frequently in patients with severe skin disease, whereas in another, pustular psoriasis was associated with more severe psoriatic arthritis. In patients presenting with an undefined seronegative polyarthritis, looking for psoriasis in hidden sites such as the scalp, perineum, intergluteal cleft, and umbilicus is extremely important. A diagnosis of psoriatic arthritis may be missed because of an inadequate physical examination.[10] Nails are involved in 80% of patients with psoriatic arthritis but in only 20% of patients with uncomplicated psoriasis. Onycholysis, transverse ridging, and uniform nail pitting are 3 features of nail involvement that should be noted. A direct correlation exists between the number of pits and the diagnostic significance. When skin and joint disease begin simultaneously, nail involvement is frequently present at the onset.[11] The nail plate is deeply pitted, probably due to defects in nail growth caused by psoriasis, has a yellow to yellow-pink discoloration, probably due to psoriatic involvement of the nail bed. White areas appear under the nail plate. These are air bubbles marking spots where the nail plate is becoming detached from the nail bed (onycholysis). There may be reddened skin around the nail. The nail plate crumbles in yellowish patches (onychodystrophy), probably due to psoriatic involvement in the nail matrix. The nail is entirely lost due to psoriatic involvement of the nail matrix and nail bed.[12] Extra-articular features are observed less frequently in patients with psoriatic arthritis than in those with RA. Ocular involvement may occur in 30% of patients with psoriatic arthritis, including conjunctivitis in 20% and acute anterior uveitis in 7%. Scleritis and keratoconjunctivitis sicca are rare.[13]

Several reports have demonstrated an association between psoriasis and cardiovascular diseases such as hypertension, valvular disease and arrhythmia. However, the data is scarce. Inflammation of the aortic valve root, which may lead to insufficiency, has been described in patients with psoriatic arthritis and is similar to that observed more frequently in persons with ankylosing spondylities.[14]

No specific diagnostic tests are available for psoriatic arthritis. Diagnosis of the disease is made based on clinical and radiologic criteria in a patient with psoriasis. The most characteristic laboratory abnormalities in patients with psoriatic arthritis are elevations of the erythrocyte sedimentation rate (ESR) and C-reactive protein level. Patients with psoriatic arthritis are typically seronegative for RF, although RF is detected in 5-9% of patients. Synovial fluid is inflammatory, with cell counts ranging from 5000-15,000/mL and with more than 50% of cells being polymorphonuclear leukocytes. Within the synovium, the infiltrate consists predominantly of T lymphocytes. Synovial fluid complement levels are either within reference ranges or increased, and glucose levels are within reference ranges.

Radiological features have helped to distinpsoriatic arthritis from other causes of polyarthritis. Erosions will usually begin at the peripheral articular surfaces and extend centrally. Fluffy bone periostitis and erosions at the interphalangeal joints can create an appearance which resembles "mouse ears". As previously mentioned; there are five different patterns of psoriatic arthritis. Classic psoriatic arthritis or DIP arthritis, involves the distal digits with associated fingernail pathology. Resorption of the distal phlangeal tufts can often be seen; this is a process called acroosteolysis. Arthritis mutilans is a very destructive form of psoriatic arthritis with significant periarticular bone resorption. The erosions can cause a "pencil in cup" deformity where one articular surface is eroded creating a pointed appearance; the articulating bone can be concave, resembling an upside down cup. Symmetric polyarthropathy can strongly resemble rheumatoid arthritis with erosions and ankylosis of the interphalangeal joints. Asymmetrical oligoarthritis has the appearance of soft tissue swelling of a single phalanx known as a "sausage digit". The fifth and last pattern exhibits findings similar to ankylosing spondylitis with spinal sendesmophytes which occur in an asymmetric distribution. This is associated with sacroilitis, which will usually present in an asymmetric and unilateral pattern.[15] Recent studies have indicated that MRI may be a sensitive method for demonstrating the typical enthesopathic pathology of psoriatic arthritis, particularly in the hands and feet.

Moll and Wright's description of PsA suggested that the disease was less severe than that seen in RA.[16] However, over the past two decades it has become clear that PsA is much more aggressive than previously thought. About 20% of the patients develop a very destructive disabling form of arthritis. Over time there is clinically active arthritis such that by the time patients have been followed for more than 10 years, 55% have five or more deformed joints.[17]

Conclusion

We presented the case as the patient presented in an atypical manner with early onset of arthritis (within two years of psoriasis) in an aggressive manner and rapid development of extra articular features. PsA is included in the seronegative spodyloarthropathy group which has a less severe disease progression. But psoriatic arthritis patients with positive family history and with skin lesion starting at childhood may show a more aggressive course resulting in early morbidity and disability.

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