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Case Report

SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis); A Case of Backache in an Adolescent in Pakistan

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ABSTRACT

SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis) syndrome is a group of rare auto-inflammatory disorders with a wide range of dermatological and osteoarticular manifestations. Different nomenclatures, like Chronic Recurrent Multifocal Osteoarthritis (CRMO) and Chronic Non-Bacterial Osteomyelitis (CNO), have been used in the past for the same disease, but SAPHO Syndrome has been the accepted terminology since 1990. An adolescent, 17 years of age, with a six-month history of inflammatory backache and fatigue, reported to a rheumatology clinic. His health assessment questionnaire- disability index came out to be two. Acne on physical examination raised inflammatory markers, and characteristic findings of SAPHO syndrome on bone scan suggest the disease. The patient was treated with an excellent response to bisphosphonates. The report highlights the importance of the evaluation of inflammatory backache in adolescents. SAPHO syndrome is an underdiagnosed disease. Therefore, more awareness of this disease among healthcare physicians is required.

Keywords: SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis), Chronic recurrent multifocal osteoarthritis (CRMO), Chronic non-bacterial osteomyelitis (CNO).

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INTRODUCTION

SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis) Syndrome is a rare autoinflammatory disorder with less than 1200 cases reported worldwide. Its incidence is thought to range from 1 in 10,000 among the white population, while in the Japanese population, it is 0.00144 in 100,000 individuals.[1] Different nomenclature has been used in the past for the same disease, but now SAPHO syndrome is the accepted terminology proposed by Chamot in 1988. [2]

Different factors like genetics, auto-inflammation, infections (Propionibacterium acnes, Staphylococcus aureus) and immune dysregulation are thought to play a role in the pathogenesis of this disease. However, exact etiopathogenesis and its different phenotypes are different. Cytokines, including IL1, IL8, IL17, IL18 and anti-TNF alpha, are thought to be involved in causing different osteoarticular and dermatological manifestations of the disease.[2]

Osteoarticular and dermatological manifestations are the hallmarks of the disease, with joint pains, bone pains and bone swellings. Dermatological manifestations include palmoplantar pustulosis, severe facial acne, and hidradenitis suppurative. Other systemic manifestations include inflammatory bowel disease, fatigue, fever, uveitis and sciatica. A bone scan is diagnostic for the disease with characteristic Bull's Head signs of the anterior chest, osteitis and hyperostosis of the spine, jaw and peripheral bones. X-rays, CT scans, and MRIs may also show osteolytic lesions and bone and soft tissue oedema at later stages.[3]

We here report a case of an adolescent with acne and characteristic findings of SAPHO syndrome on a bone scan.

CASE REPORT

An adolescent, 17 years of age, was in the usual state of health six months ago when he developed off-and-on backache, which was insidious in onset, gradually progressive, worse in the morning, aggravated by rest and partially relieved by physical activity. The patient could not sit for 30 minutes during his study period. Backache was also associated with fatigue for the past four months, which was insidious, generalized, and gradually progressive. The patient had no temporal relationship with physical activity or rest and felt lethargic most of the day. The patient reported these symptoms to a local doctor, where an X-ray of the lumbosacral spine appeared normal. He was advised of NSAIDs and muscle relaxants, but when his

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symptoms did not improve, he was referred to a Rheumatologist on 21st Nov 2021. Systemic review was positive for acne for the past five years, and he had no history of oral ulcers, weight loss, cough, chest pain, or any gastrointestinal or genitou-rinary symptoms.

Physical examination revealed an adolescent with a lean build. The patient was vitally stable and had severe acne on his face. He had no pallor, jaundice, oedema or palpable lymph nodes. The joint and musculoskeletal examination was unremarkable with non-tender sacroiliac joints, while Schober's and FABER's were negative. The rest of the systemic examination was unremarkable.

The patient underwent an extensive laboratory and radiological workup. His age-adjusted serum erythrocyte sedimentation rate was raised (15mm at 1st hour), along with raised C-reactive protein levels (25mg/L), both markers of inflammation. His complete blood count, liver function and renal function tests were normal. Human Leukocyte Antigen (HLA B-27) was negative, and urinalysis was unremarkable, with serum electrolytes in the normal range.

His X-ray of the Sacroiliac (SI) Joints showed no pathology. His MRI SI Joint also came out to be normal. His bone scan was requested, which showed characteristic Bull's Head signs of the anterior chest, osteitis of the pubis and hyperostosis of the distal tibia (Figure). His Health Assessment Questionnaire- Disability Index came out to be two.

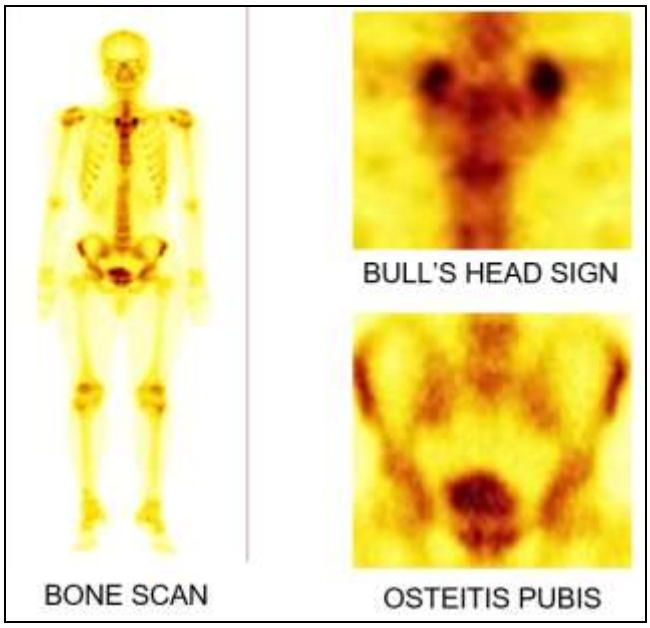

Figure: Bone Scan of the Patient with Characteristic findings of SAPHO Syndrome.

It was planned that the patient would be started on bisphosphonates, for which a pre-treatment work-up was done, which included serum calcium and vitamin D levels and a dental check-up. The patient was started on bisphosphonates (Tablet Alendronate 70mg) once weekly along with NSAIDS 12 hourly and calcium carbonate with cholecalciferol. A dermatological consultation was also made for acne, and the patient was started on Isotretinoin 20mg once daily and Tretinoin cream once daily.

At four weekly follow-ups, the patient showed marked symptomatic improvement in his osteoarticular and dermatological manifestations, significantly reducing HAQ-DI scores. He is being followed up every month.

DISCUSSION

SAPHO syndrome has many overlapping features with other auto-inflammatory disorders like psoriasis, ankylosing spondylitis and inflammatory bowel disease. It is usually a diagnosis of exclusion, with less than twelve hundred cases reported worldwide. However, it is still considered an underdiagnosed entity, and increased awareness among healthcare professionals is required for its early identification. The peak incidence among the Asian population is at a relatively younger age, ranging from the fifteenth month of age to 70 years of age.[3]

Being a rare disease, the management of SAPHO Syndrome is based on case reports and observational studies. Several studies have shown that the efficacy of bisphosphonates is equivalent to anti-TNF alpha.[4] These results are promising as patients on bisphosphonates do not require life-long anti-TNF alpha treatment, thus reducing the cost and side effects associated with anti-TNF alpha drugs.[5,6] As SAPHO Syndrome is an underdiagnosed entity, more awareness is required among healthcare professionals regarding its presentation and diagnostic evaluation.[7] Bone scans can pick up disease early; therefore, their judicious use in difficult-to-diagnose cases is recommended. More research and clinical trials regarding its effectiveness as a therapeutic option are also required.

Conflict of Interest: None.

Authors Contribution:

Following authors have made substantial contributions to the manuscript as under:

ARAD & MA & AK& MZH: Conception, data acquisition, drafting the manuscript, critical review, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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