Case Report

Undifferentiated Spondyloarthritis Progressing to Ankylosing Spondylitis

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ABSTRACT

Undifferentiated spondyloarthropathy (USpA) is one distinct form of spondyloarthropathy in the clinical spectrum of spondyloarthritides. The frequency and clinical spectrum of USpA have been ignored in previous epidemiologic and clinical studies. A generally accepted nosologic concept and definition of USpA may be needed to overcome this issue. With the help of the modified Amor criteria or European Spondyloarthropathy Study Group (ESSG) criteria, the real prevalence may be better defined in the future and may also lead to early recognition of such patients in clinical practice. Hereby, we present a case of a 52 year old female patient presenting with low back pain was initially diagnosed as a case of undifferentiated spondyloarthropathy which later on progressed to ankylosing spondylitis.

Keywords : Undifferentiated spondyloarthropathy, Ankylosing spondylitis

Introduction :

Undifferentiated spondyloarthropathy (UspA) is the most common subtype of the spondyloarthritides with a prevalence between 0.7% and 2.0%[1]. The term Usp A refers to patients with clinical and roentgenographic features suggestive of spondyloarthropathies but not fulfilling the diagnostic or classification criteria for any of the currently established disease categories. It is very difficult to differentiate the various forms in their early stages due to their overlapping clinical features. These are asymmetric lower limb arthritis, enthesopathy, unilateral or bilateral sacroiliitis, rheumatoid factor negativity, extra articular features like uveitis, familial aggregation and association with HLA B27[2]. There are no pathognomic clinical features for USpA, and the absence of diagnostic tests for any of the spondyloarthritides requires diagnosis to be made on a combination of history, clinical examination and supportive laboratory tests. USpA may represent an early phase or incomplete form of ankylosing spondylitis or another form of spondyloarthropathy. With the help of newer biological agents (TNF α blockers), the progression of the disease can be halt at its initial stage.

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Case History :

A 52year old female patient presented with low back pain since last 9 years. Pain was associated with early morning stiffness for about 30 minutes duration that improves with activity and associated with nocturnal exacerbation. Pain was gradually progressive so much that it was affecting her daily living activities and later on she needed support for walking. Her complaints responded to nonsteroidal anti-inflammatory drugs (NSAIDS), and her condition improved with exercise. Initially pain was persistent for 3 years then was subsided and she had intermittent exacerbations. Initially there was no history of other joint involvement but later she developed intermittent peripheral arthritis in the form of swelling over right ankle joint and pain in left shoulder joint.

There was no history suggestive of dactylitis, scaly skin rash, malar rash, abdominal pain, tenesmus, diarrhoea associated with mucous and blood. Also there was no history of fever, recurrent oral ulcers or genital ulcer or history suggestive of genitourinary tract infection or redness of eyes or increased lacrimation. There was no positive family history.

For above complaints, patient first consulted orthopedician and her MRI Lumbosacral spine was done s/o L5-S1 disc protrusion. She took treatment from orthopedician for about 2 years but had no relief.

Patient was then referred to rheumatologist in 2010 and on examination he was found to have tenderness at left gluteal region and ischial bursa, straight leg test was 75^{o} (bilateral) and Lasegue test was

negative. Modified Schober's test was negative, chest expansion was normal and there was no sacroiliac joint tenderness.

Her investigations revealed CBC : Hb-10 gm%, TLC 8100/mm^3, negative rheumatoid factor, Erythrocyte sedimentation rate - 44 mm/hr, C-reactive protein - 22 mg/L, urine test and rest routine blood investigations were normal. X-ray lumbar spine, pelvis & hip were normal. HLA B27 was positive. MRI lumbosacral spine (2010) was s/o L5-S1 disc protrusion. Tc bone scan (2010) was s/o bilateral sacroiliitis.

Diagnosisof **Undifferentiated Spondyloarthritis** was made as per modified Amor criteria[3].

Treatment : Patient was given NSAIDS, DMARDs (Sulphasalazine), Bisphosphonates & Physiotherapy. Patient had relief of her symptoms & was on regular follow up with rheumatologist since then. During last 7 years she had intermittent back pain, alternate buttock pain and peripheral arthritis which were controlled by NSAIDS. In March 2017 she had exacerbation of her disease in the form of severe back pain and pain in both gluteal region. On examination she was found to have bilateral sacroiliac joint tenderness, antalgic gait and decreased range of movements at both hip joint and also early parkinsonism features in the form of bradykinesia and rigidity. Because of parkinsonism, patient had minimal activity which might be the cause of exacerbation of her disease.

MRI bilateral sacroiliac joint with hip joint and spinogram (2017) was done s/o active marrow oedema along the bilateral sacroiliac joints with cortical irregularity and few erosions s/o active sacroiliitis, hip joint was normal and whole spine screening was s/o L5-S1 central extrusion with no spinal canal stenosis.

Patient was then given intra-articular steroid injection (depotmethylprednisolone 80 mg) in both sacroiliac joints under CT control with good response in one SIJ and was also started on anti-parkinsonian drug. Patient's Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) was calculated as 6.7 and after treatment it dropped down to 3.0.Now, patient has pain in left hip region only & able to walk on her own & on examination, Patrick's test (FABER) was positive on left side with painful left hip rotation.

Diagnosis : Patient was later diagnosed as a case of ankylosing spondylitis (as per modified New York criteria).[4]

Discussion : Undifferentiated SpA is a syndrome with features similar to other SpA, but affected patients do not fulfill criteria for any specific SpA. According to various cohort studies, they may represent either an incomplete form of ankylosing spondylitis or other forms of reactive arthritis or IBD associated SpA. In various cohort analysis it was found that about 30-40% proceed to develop AS in about 10 years and some 50-60% remain USpA, 10-15% go into spontaneous remission and about 1-2% manifest as psoriatic arthritis.[2,5,6] However, subsequent data suggest that these patients may represent a distinct disease entity on the basis of demographic and clinical criteria. Although no specific criteria are identified, modified Amor criteria / ESSG criteria can be helpful in confirming a clinical diagnosis of USpA.[3,7]

Figure 1 : **MRI with T1 sequence of sacroiliac joints - shows cortical irregularity and few erosions along the bilateral sacroiliac joints**

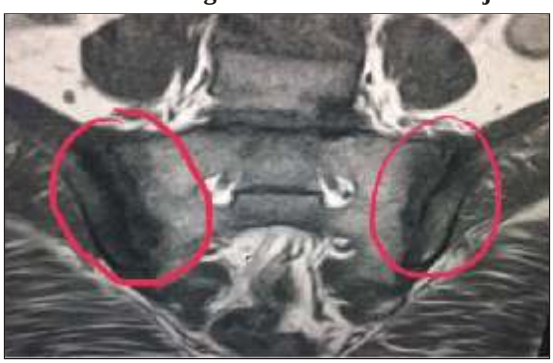
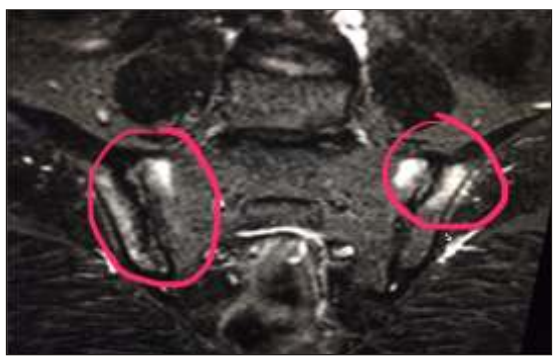


Figure 2 : **MRI with short tau inversion recovery (STIR) sequence showing bone marrow oedema s/o active sacroiliitis**



Prognostically they are better than other forms of SpAs. Most patients maintain good function without progressive disease or clinically significant radiographic changes. Treatment with NSAIDS is effective. Salazopyrin has been found to be effective in a large multicentre trial. Use of TNF α blockers have been advocated in severe forms of USpA.[1,2] Anti-TNF therapy may have the potential to benefit these patients symptomatically but also prevent progression to AS.

Conclusions : As the Amor criteria / ESSG classification criteria for SpA are becoming more widely used in clinical practice, it is important that clinicians become more familiar with the concept of USpA. Can we alter the natural course of the disease? With accurate patient selection and the availability of newer biological agents that are effective in suppressing spinal and peripheral joint inflammation, the door is open for significant improvements in the early treatment of disease, and even perhaps for strategies to halt the progression of disabling SpA.[1]

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Table 1 : **Modified Amor criteria for USpA**

Inclusion criteria Score	Exclusion criteria
Inflammatory back pain 1	Diagnosis of specific spondyloarthropathy
Unilateral buttock pain 1	Sacroiliitis on radiograph ≥ grade 2
Alternate buttock pain 2	Precipitating genitourinary or GI infection
Ethesitis 2	Psoriasis
Asymmetric peripheral arthritis 2	Keratoderma blenorrhagicum
Dactylitis (sausage digit) 2	IBD (crohn disease or ulcerative colitis)
Acute anterior uveitis 2	Positive antinuclear antibody titre> 1: 80
HLA B27 positive or family history of SpA 2	
Good response to NSAID 2	

A score of 6 or more is diagnostic of SpA

Table 2 : **Modified New York Criteria for AS (1984)**

Modified New York Criteria for AS (1984)
A) Clinical criteria :
1. Low back pain and stiffness (> 3 months, improved by exercise & not relieved by rest).
2. Limitation of lumbar spine motion in both the sagittal and frontal planes.
3. Limitation of chest expansion relative to normal values correlated for age and sex.
B) Radiological criteria :
Sacroiliitis grade ≥ 2 bilaterally or grade 3-4 unilaterally
Definite AS is present if the radiological criteria is present with at least one clinical criteria.
Probable AS if
- 3 clinical criteria or
- Radiological criteria present but no signs or symptoms satisfying clinical criteria