CASE STUDY

Ankylosing spondylitis with systemic lupus erythematosus: A rare co-existence

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Abstract

Systemic lupus erythematosus (SLE) and ankylosing spondylitis (AS) are two distinct immunological disorders with different etiopathogenic profiles and their co-existence is very rare. The present study discusses two cases of co-existing AS with SLE. The first case, which fulfilled the criteria for lupus, had HLA-B27 arthritis with predominant peripheral involvement, bilateral hip disease associated with non-scarring alopecia, recurrent oral ulcers and positive immunological markers. The second case was of a 37-year-old male with lupus (predominantly musculoskeletal and cutaneous) who presented with inflammatory low backache. The patient had high titers of ANA and anti-dsDNA positivity and was on methotrexate treatment. On evaluation, he was found to have HLA B27 positivity and bilateral active sacroiliitis. The distinguishing feature of both the cases was that the time frame of presentation of the disorders was almost same, while second case had a significant time frame between the two disorders.

Keywords: ankylosing spondylitis, systemic lupus erythematosus, HLA-B27, arthritis, sacroiliitis

Case 1

A 64-year-old female presented to rheumatology out-patient department with complaints of bilateral knee joint pain (right>left) and right ankle joint pain. Pain was inflammatory in nature and was associated with swelling and morning stiffness. There was no history of low backache or skin, eye or bowel involvement. Further investigations revealed the presence of bilateral hip pain associated with difficulty in walking. She underwent bilateral hip arthroplasty around 8 years back, secondary to hip arthritis. After hip replacement, she had on and off episodes of bilateral knee joint pain, right ankle joint pain and heel pain, which got aggravated in the last 6 months with no relief on NSAIDs. Her history revealed the occurrence of erythematous lesions over the scalp, followed by patchy hair loss with absence of regrowth of hair during the same time period. She also had recurrent oral ulcers over the tongue and buccal mucosa. There was no history suggestive of any sicca symptoms or Raynaud's phenomena. On physical examination, non-scarring patchy alopecia was present with no active lesions (Fig. 1). Oral ulcerations were present over the lateral border of the tongue and buccal mucosa. Systemic examination was suggestive of synovitis of bilateral knee and right ankle joint. Her hemogram, liver and kidney function tests, urine routine microscopy and protein creatinine ratio were normal. Whereas, ESR and CRP were elevated and HLA-B27 was positive. X-ray of pelvis revealed normal sacroiliac joints. ANA was positive with 3+ intensity at a titer of 1:320 with homogenous pattern. ANA profile was positive for nucleosome and the anti-dsDNA titers were significantly elevated (88.6 IU/ml). Serum complement levels (C3) were low. MRI of sacroiliac joint and skin biopsy were suggested, but the patient was not willing for further investigations. Based on the available clinical investigations, the diagnosis was concluded as spondyloarthritis with mucocutaneous lupus. She was managed with hydroxychloroquine, tapering dose of low-dose steroids and methotrexate. To the best of our knowledge, this is one of the few case reports on the co-existence of SLE and spondyloarthritis with similar period of presentation.

Case 2

In 2011, a 37-year-old male presented with inflammatory pain in small joints of both hands along with excessive hair fall and recurrent oral ulcerations. His investigations revealed positive ANA (1 in 2560 with 4+ intensity and homogenous pattern) and anti-Smith antibodies. Rheumatoid factor and anti-CCP levels were negative. Complement levels were low and urine protein to creatinine ratio was normal. The patient was started with methotrexate and low-dose steroids (which were tapered

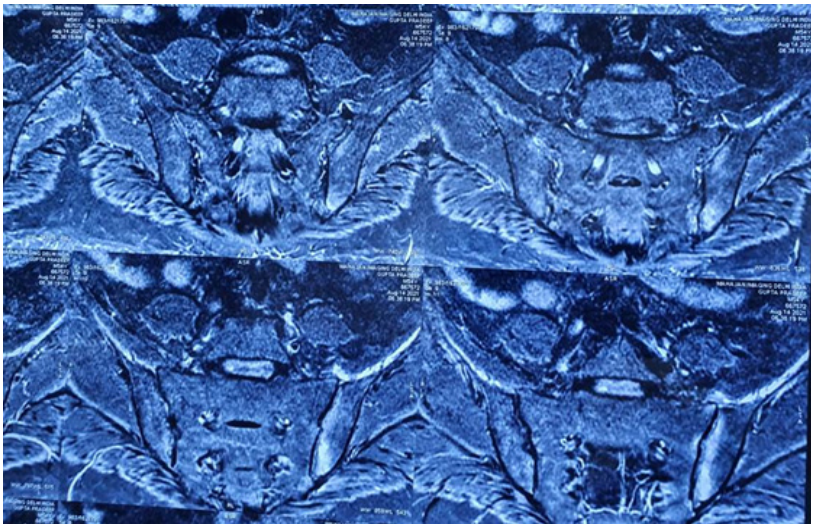
and stopped). However, he stopped the medications by himself after 3 years. The lupus flare recurred after 5 years of initial presentation, which was managed with tapering dose of steroids and methotrexate. During the recent presentation, he had inflammatory back pain and right knee joint pain associated with swelling for 2-3 months. Further evaluation revealed raised ESR and CRP levels. Whereas, hemogram, and liver and kidney function tests were normal. The PCR assay for HLA-B27 was positive. MRI of sacroiliac joint was suggestive of active left sacroiliitis and subacute to chronic right sacroiliitis (Fig. 2). The patient was diagnosed as ankylosing spondylitis with SLE and was advised to undergo NSAID therapy for 4 weeks. Biological therapy was also planned if there was no response.

Discussion

Literature review shows that there are only very few cases on co-existence of AS and lupus. The first case reported had non-scaring alopecia, recurrent oral ulcerations, ANA and anti ds-DNA positivity, and low complement levels. On the other hand, the patient fulfilled the Assessment of SpondyloArthritis international Society (ASAS) classification criteria for peripheral SpA such as HLA B-27 positivity, and the presence of arthritis and enthesitis. Most of the reported cases including the present case was noted in females. In

Fig. 1: Discoid lupus lesions and scarring alopecia

Fig. 2: MRI imaging showing bilateral sacroiliitis



most of the cases, the presence of SLE was noted before the occurrence of spondyloarthritis, whereas in the currently reported first case, both the disorders manifested at the same period of time. Clinical findings favoring the diagnosis of spondyloarthritis were pain with inflammatory nature, response to NSAIDS, high inflammatory markers, and MRI evidence of bone marrow edema in bilateral sacroiliac joint.

One contradictory point was the presence of inflammatory back pain, even in SLE patients with lupus. But the presence of associated peripheral joint pain, HLA-B27 positivity and sacroiliitis in MRI made the diagnosis in favor of spondyloarthritis, as the co-existence of these two disorders is very rare. In 1982, Nashel *et al.* have reported the first case of lupus with SpA in a 43-year-old male patient.[1] Recently in 2020, Nesilhan *et al.* have studied 281 SLE patients in whom inflammatory back pain was present in 16.3% and sacroiliitis in 7.8%. Only one patient was found to have HLA-B27 positivity.[2] De Smet *et al.* described sacroiliitis in 9 patients with active SLE with elevated radionucleotide uptake ratios in sacroiliac joints and further reported that uptake ratios returned to normal in patients who achieved remission.[3] Vivas *et al.* in 1985 reported sacroilitis in 16 male patients with SLE and HLA-B27 negativity was noted in all of the subjects.[4] In 2019, Enginar reported a case of spondyloarthritis with discoid lupus, with manifestation of both disorders at the same time of presentation.[5] Another case was reported by Tarhan in 2014, where a patient of SLE (biopsy-proven lupus nephritis) presented with spondyloarthritis manifestations 8 years after diagnosis of SLE.[7] Here the time frame may be comparable to the currently documented second case, but the patient was a young male who was diagnosed initially with lupus and diagnosed with spondyloarthritis after 10 years.

Studies have suggested that the combination of HLA-B27 with HLA-A1 and HLA-DR2, or with HLA-A1 and HLA-DR3 is very rare.[1]This finding suggests that apart from HLA association, some other linked pathogenesis exists between these two disorders. Many studies have recently suggested the importance of IL17/23 axis in ankylosing spondylosis pathogenesis. Even though SLE is mainly mediated by interferon pathways, recent studies have shown elevated IL-17 levels in SLE.[7] IL-17 can directly stimulate the B cells to produce autoantibodies and is responsible for autoimmune disease process.[8] Further, some other studies are suggesting that there is recruitment of IL-17 producing T helper cells in kidney, skin and CNS in SLE patients.[6] These findings indicate that the IL-17 can act as a common gateway for both disorders and further studies are needed to confirm this entity.

Conclusion

The present study highlights the need to consider the probability of co-existence of AS and SLE. The study also emphasizes the time frame of two different disease presentations, one case with two disorders at the same time frame and another case with a significant time gap between two disorders, which can evoke a therapeutic challenge for a treating rheumatologist.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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