Case Report

Co-existence of Systemic Lupus Erythematosus and Ankylosing Spondylitis - A Case Report

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Abstract

Co-existence of systemic lupus erythematosus and ankylosing spondylitis is not common. In this report, we present a 24-year-old woman with both the entities diagnosed in the course of time. Initially she presented with malar rash, photosensitivity, oral ulcer and polyarthritis. Her ANA and anti-Smith antibody were positive, but ant-ds DNA was negative. With these features, she was diagnosed as a case of systemic lupus erythematosus. Few months later, she was diagnosed with ankylosing spondylitis on the basis of inflammatory back pain and bilateral sacroilitis.

Key words: Systemic lupus erythematosus, ankylosing spondylitis, sacroilitis

INTRODUCTION

Systemic lupus erthematosus (SLE) is a systemic auto-immune disease characterized by diverse multisystem involvement with symptoms ranging from mild skin involvement to life threatening organ disease.[1,2] Ankylosing spondylitis (AS) is a chronic inflammatory disease which belongs to a group of heterogeneous conditions known as spondyloarthritis. Sacroiliitis is the hallmark of AS, especially in earlier disease stages.[3] Sacroilitis may be an infrequent manifestation of SLE itself[4,18] or may rarely represent as coexistence of SLE with AS[5].However, peculiarities of sacroilitis in SLE are absence of typical back pain and HLA B27.[4] So far, only 9 cases of coexistence of SLE and AS have been reported.[5-13] In this report, we describe another case of such coexistence.

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CASE SUMMARY

A 24-year-old woman was diagnosed with SLE on November 2016 on the basis of malar rash, oral ulcer, photosensitivity, polyarthritis, positive ANA and anti-Smith antibody. She was doing well with hydroxychloroquine and tapering doses of corticosteroid. Four months later, she experienced one episode of loss of consciousness,several occasions of vomiting along with 3 episodes of seizures, and at that time it was labeled as CNS involvement of SLE. She was then discharged with hydroxychloroquine, sodium valproate and prednisolone 7.5 mg daily. Two months later, she developed inflammatory back pain and severe pain in both hips (anterior). She did not have other features of SLE during this visit. Her SELENA SLEDAI score during admission was 2. Her hemoglobin was 7.1 gm/dl, reticulocyte count, bilirubin were normal,Coombs' test was negative,CRP-14.0 mg/dl, x ray SI joints showed sacroilitis grade II on right and grade III on left side with joint space reduction and periarticular sclerosis in both hip joints (image 1). MRI of both hip and sacroiliac joints revealed mild flattening of heads of femur bilaterally, T2WI fat suppression and STIR- mixed signal changes seen in femoral head of both sides with reduction of joint space of both hip joints (image 2). Joint effusion was noted in both hip joints. T1WI hypo, T2WI and STIR hyper intense signal changes were seen within subchondral marrow of both (more on left) sacroiliac joints with destruction of overlying articular cartilage resulting reduction of joint spaces (image 3). Ankylosing spondylitis was diagnosed on the basis of inflammatory back pain and bilateral sacroilitis. So, the final diagnosis of systemic lupus erythematosus, ankylosing spondylitis and bilateral avascular necrosis of femoral head was made.The cause of AVN was thought to be long term steroid therapy. Patient was then managed with NSAIDs and adjuvant analgesics and orthopedic consult was done for AVN.

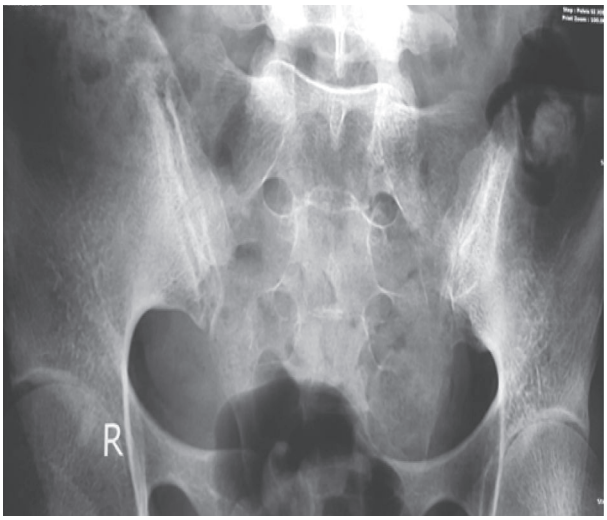


Figure 1: X ray pelvis showing bilateral sacroilitis, joint space reduction in both hip joints

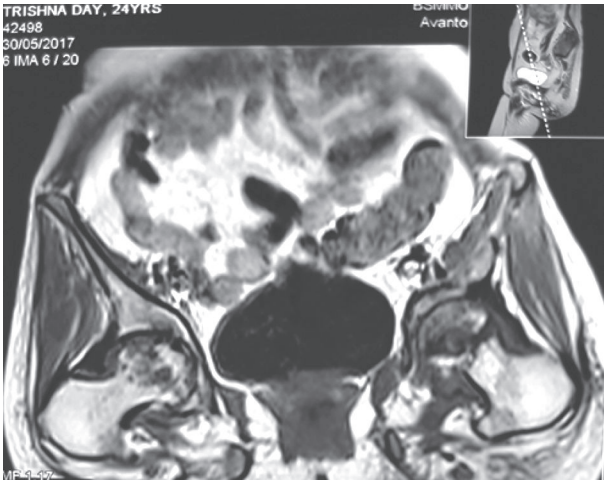
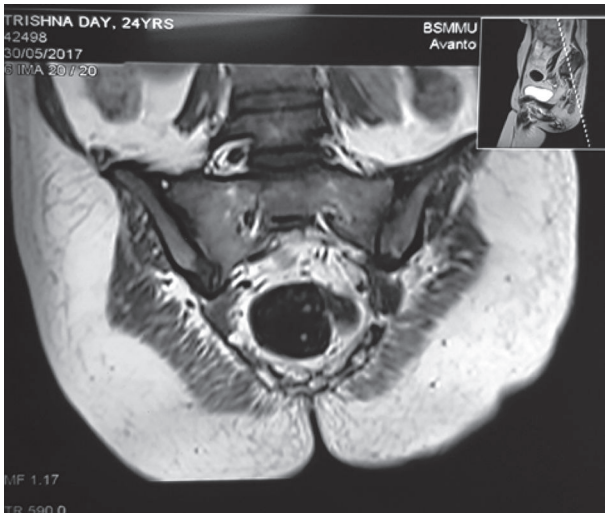


Figure 2: MRI of both hip and sacroiliac joints revealed mild flattening of heads of both femur, T2WI fat suppression and STIR- mixed signal changes seen in femoral head of both sides with reduction of joint space of both hip joints



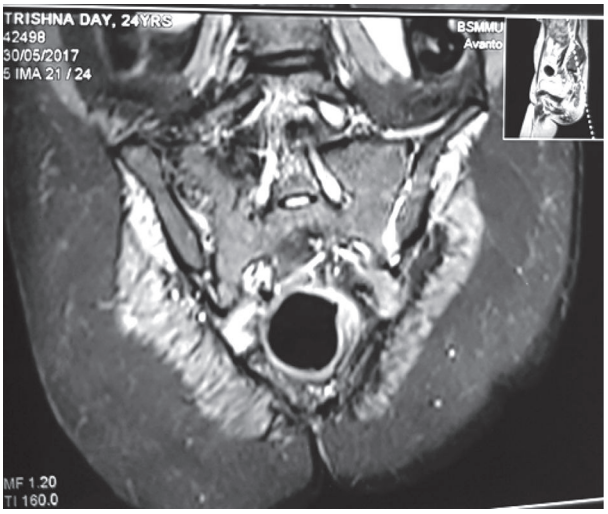


Figure 3: T1WI hypo (3a), T2WI and STIR (3b) hyper intense signal changes are seen within subchondral marrow of both (more on left) sacroiliac joints with destruction of overlying articular cartilage resulting reduction of joint spaces

DISCUSSION

SLE is predominant in women of reproductive age group with the F:M ratio of 8-15:1[14], whereas AS has F:M ratio of 1:1.2.[15] The coexistence of these two entities is very uncommon with only 9 case reports published in the literature. In our case, SLE preceded the features of AS by seven months. She fulfilled 1997 ACR criteriafor SLE with presence of malar rash, photosensitivity, oral ulcer, polyarthritis, positive ANA, anti-Smith antibody and lupus anticoagulant. Six months later, she developed inflammatory back pain with bilateral sacroilitis confirmed by MRI. Thus additional diagnosis of AS was made as per modified New York criteria.[16] Among the 9 cases mentioned above, 6 were female and 3 were male. ANA was positive in all the cases and anti-ds DNA in 8. In our case, ANA and anti- Smith antibodies were strongly positive, and anti-ds DNA was negative. The symptoms of SLE may or may not precede those of AS. In 3 cases including ours, symptoms of SLE preceded AS by few months to years (duration ranging from 4 months to 8 years).[5,8] However, in 5 reported cases, symptoms of AS preceded those of SLE by several years, duration ranging from 4 years to 17 years.[7,9,10,12,13]

Though sacroilitis may be a rare manifestation of SLE, its presence along with inflammatory back pain may be suggestive of an additional diagnosis. In a report by Nassonova et al, it was shown that 22 out of 43 male patients with SLE had radiological sacroilitis and 7 had ankylosis of sacroiliac joint. However, none had inflammatory back pain or positive HLA B27, and NSAIDs were not effective treatment for back pain.[4] In our case, patient presented with inflammatory back pain which improved with NSAIDs treatment.Chandrashekhar et al discussed a case of 21-year-old female with SLE, dermatomyositis overlap with inflammatory low back pain and symptomatic bilateral sacroilitis. HLA B27 was negative. The severity and duration of back pain correlated with high disease activity of SLE.[17] However, in our case, inflammatory back pain started when SLE was not active and patient's back pain responded to NSAIDs.

Similar to our case, Tarhan F et al has recently published one report of 55-year-old woman with SLE and subsequently developing inflammatory back pain, bilateral sacroilitis in MRI and positive HLA B27.[5] MRI has been used as a diagnostic tool for sacroilitis by our team and Tarhan F et al. Similar case has been discussed by Kucuk A et al, where sacroilitis was diagnosed by CT scan.[6] He reported a case of 33-year-old female with SLE and sacroilitis in CT, and gave history of inflammatory neck pain and back pain on query. Later she was found to be HLA B27 positive.

In previously reported cases, HLA B27 was positive in[8,5, 6, 8-13] whereas it was negative in 1 case[7] which is similar to ours. However the presence of other HLA genotypes was not searched in the previously mentioned case report[7] and our case.

CONCLUSIONS

Coexistence of SLE and AS is a rare event. Though sacroilitis may be rarely present in SLE,if present with inflammatory back pain in the background of low disease activity of SLE, meticulous search for coexisting AS should also be done.

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