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Case Report

Coexistence of Systemic Lupus Erythematosus and Ankylosing Spondylitis: Another Case Report

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Abstract

Coexistence of ankylosing spondylitis (AS) and systemic lupus erythematosus (SLE) is very rare. Untill now only 8–10 cases has been reported in English literature. Here, in this study, we will discuss another case with coexistence of these two diseases and review the previously reported cases. 35-year-female patient with previously diagnosed AS with typical inflammatory lower back pain and HLA b27 positivity, MRI confirmed the presence of B/L active sacroilitis 10 years back now came with locomotor, skin, hematopoitic system involvement, with typical autoimmune disorder facies, showing strong positivity for ANA BLOT study and specific pattern on ANA by IFA, hypocomplementimia thus diagnosed as SLE in our hospital. Including our case most of the cases of coexistence of AS and SLE are females and SLE precedes occurrence of AS.

Key words: ANA BLOT, Ankylosing spondylitis, Connective tissue disorder, HLA b27, Systemic lupus erythematosus

INTRODUCTION

Ankylosing spondylitis (AS) is a chronic inflammatory disease of axial skeleton which manifests as inflammatory back pain, progressive stiffness of spine, assymetrical peripheral oligoarthritis, and specific organ involvement such as anterior uveitis. AS is more prevalent in males. Systemic lupus erythomatosis, on the other hand, is complex rheumatologic disease involving skin, joints, serous membranes, kidneys, lungs, and other organs of the body. The most characteristic features include rash, sores in mouth, and musculoskeletal manifestations such as arthritis and currently diagnosis is based on ACR criteria. Systemic lupus erythematosus (SLE) classified as systemic autoimmune disorder since autoantibodies presence against antinuclear antibody (ANA), double stranded DNA. These two autoimmune rheumatologic diseases which have different etiopathogenesis and diverse clinical and genetic characteristics features are rarely seen together. Here, we report another case with coexistence of AS and SLE.[1-5]

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CASE REPORT

A 35-year-old female patient admitted to our hospital with h/o inflammatory back pain, and severe morning joint stiffness for 1–2 h was diagnosed as AS on basis of X-ray and magnetic resonance imaging showing b/l sacroilitis responding well to non-steroidal anti-inflammatory drugs. Now she came with c/o generalized weakness, fatigue, alopecia since 5–6 months, oral ulcers, severe joint pain since 1 month. On examination, she was afebrile, vitally stable, showing generalized lymphadenopathy. Her laboratory test results were as follows: Raised erythrocyte sedimentation rate 55 mm/h, C-reactive protein 107, rheumatoid factor negative, white blood cell count 2.7/μL (4.60–10.2), hemoglobin 8.7 g/dL (12.2–18.1), ANA test positive with titer of 1/2304 and homogenous pattern on ANA BY IFA, anti-double-stranded DNA (Crithidia test) was positive (2+), complement 3 (C3): 60 mg/dL (83–193) and complement 4 (C4): 6 mg/dL (15–57). Urine routine showing 1+protein with non nephrotic range proteinuria (247 mg/24 h). Liver and renal function tests, serum protein and creatinine phosphokinase levels, and thyroid function tests were within the normal limits. Montoux taest was positive. Hence, lymph node biopsy was done to rule out disseminated kochs shown reactive follicular hyperplasia and was negative for AFB stain, culture, and gene expert. Above symptoms along with anti-ds DNA and ANA blott strong

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Table 1: EULAR/ACR clinical domains and criteria for SLE

Domain	Criteria	Points
Constitutional	Fever	2
Hematologic	Leukopenia	3
	Thrombocytopenia	4
	Autoimmune hemolysis	4
Neuropsychiatric	Delirium	2
	Psychosis	3
	Seizure	5
Mucocutaneous	Non-scarring alopecia	2
	Oral ulcers	2
	Subacute cutaneous or discoid lupus	4
	Acute cutaneous lupus	6
Serosal	Pleural or pericardial effusion	5
	Acute pericarditis	6
Musculoskeletal	Joint involvement	6
Renal	Proteinuria >0.5 g/24 h	4
	Kidney biopsy class II or V lupus nephritis	8
	Kidney biopsy class III or IV lupus nephritis	10

SLE: Systemic lupus erythematosus

Table 2: EULAR/ACR immunologic domains and criteria for SLE

Domain	Criteria	Points
Antiphospholipid antibodies	Anti-cardiolipin antibodies or Anti-β2GP1 antibodies or lupus anticoagulant	2
Complement proteins	Low C3 or low C4	3
	Low C3 and low C4	4
SLE-specific antibodies	Anti-dsDNA antibody or anti-smith antibody	6

SLE: Systemic lupus erythematosus

positivity, with low C3, C4 complement levels, with raised titers on ANA by IFA (>1:2308) with homogenous pattern on IFA confirmed the diagnosis of SLE. Since there was only joint, skin, serous membrane involvement, the initial treatment included hydroxychloroquine (200 mg/day), and moderate to low doses of methylprednisolone (10 mg/day) as she does not responded that well to treatment dose of methylprednisolone increased (40 mg/day) and added mycofenolet mofetil (MMF) orally (360 mg twice a day) with oral proton pump inhibitor (40 mg/day) [Tables 1 and 2].

DISCUSSION

Coexistence of AS and SLE is very rare and to the best of our knowledge, there are only 8–10 reported cases in English literature. AS is frequently seen in male patients and SLE is seen mainly in female patient. Coexistence of both is seen in female patients and majority of patients complaints are related to AS. Diagnosis of AS was before diagnosis of SLE. Our patient is female her complaints started with AS and later developed SLE. There was combination of AS findings like b/l sacroilitis, HLA b27 positivity with SLE findings such as hematological pancytopenia, renal involvement with 1+ protein in urine, hypocomplementemia, positive autoantibodies for ANA, anti dsDNA. The coexistence of these two diseases with different genetic backgrounds in the same patient is much lower than expected based upon their prevalence in the general population. It has been suggested that the combination of HLA-B27 with HLA-A1 and HLA-DR2 is very rare. The rare combinations of the susceptibility genes of AS and SLE were speculated to explain the rarity of the coexistence of these two diseases. Diagnosis of SLE made on basis of ACR criteria as follows:[6-11]

Treatment for SLE

EULAR recommendations

For the treatment of SLE in 2008 and updated them in 2019. EULAR recommends that treatment in SLE aim at remission, or at low disease activity in all organ systems if remission cannot be achieved. Specific medication recommendations include the following:

1. Hydroxychloroquine is recommended for all patients with SLE
2. Glucocorticoids can provide rapid symptom relief, but the medium- to long-term aim should be to minimize the daily dose to ≤7.5 mg/day prednisone equivalent or to discontinue them
3. Subsequent initiation of immunosuppressive drugs facilitates more rapid tapering of glucocorticoids and may prevent disease flares. The choice of agent depends on prevailing disease manifestation(s), patient age and childbearing potential, safety concerns, and cost.

Regarding Immunosuppressive Drugs:

1. Consider methotrexate and azathioprine in patients with poor symptom control with glucocorticoids and hydroxychloroquine, or when hydroxychloroquine alone is unlikely to be sufficient
2. MMF is a potent immunosuppressant with efficacy in renal and non-renal SLE (but not in neuropsychiatric lupus), but its teratogenic potential and higher cost limit its recommendation in women of reproductive age with non-renal manifestations
3. Cyclophosphamide can be considered in organ-threatening disease (especially renal, cardiopulmonary, or neuropsychiatric) and as rescue therapy in patients with non–major organ manifestations refractory to other agents. Due to its gonadotoxic effects, it should be used with caution in women and men of fertile age; concomitant use of gonadotropin-releasing hormone analogues is recommended in premenopausal patients

Regarding biologic agents for SLE are as follows:

4. Consider belimumab in patients with extrarenal disease inadequately controlled by first-line treatments.

CONCLUSION

In conclusion, the coexistence of SLE and AS is very rare. Including the present case, there are only nine to eleven reported cases. Most of the cases are females. The present case is also a 35 yr female with known case of AS with HLA B27 positivity and B/L sacroillitis recently diagnosed with SLE with help of clinical symptoms, ANA BLOT study , ACR /EULAR criterias ,and other relevant blood investigations and treated accordingly.

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