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A CASE OF SYSTEMIC LUPUS ERYTHEMATOSUS WITH SJÖGREN'S SYNDROME OVERLAP

General Medicine

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ABSTRACT

Systemic lupus erythematosus (SLE) and Sjögren's syndrome (SS) are chronic autoimmune diseases that can coexist, leading to a complex clinical presentation known as SLE-SS overlap syndrome. This review aims to elucidate the unique clinical characteristics, diagnostic challenges, and management strategies associated with SLE-SS overlap. Studies have shown that around 14-19% of patients with SLE also meet the criteria for SS.(1) Diagnostic criteria for SLE-SS overlap involve meeting established guidelines for both SLE and SS, incorporating clinical findings and serological markers, salivary gland biopsy and imaging studies may also aid in confirming SS.(2) Management of SLE-SS overlap requires a multidisciplinary approach to address the diverse manifestations of both diseases. Treatment regimens typically include immunosuppressive agents,Biologic therapies, Supportive care to manage sicca symptoms improving patients' quality of life.(3) The prognosis of SLE-SS overlap varies, with some patients experiencing a more severe disease course due to the additive effects of both conditions. Ongoing research is necessary to better understand the pathophysiological mechanisms underlying the overlap and to develop targeted therapies.(4)

KEYWORDS

INTRODUCTION:
Systemic Lupus Erythematosus (SLE) and Sjögren's Syndrome (SS) are both systemic autoimmune diseases that share several clinical and immunological features. SLE is characterized by widespread inflammation and tissue damage in multiple organs, including the skin, joints, kidneys, and central nervous system. Sjögren's Syndrome primarily affects the exocrine glands, leading to dry mouth and dry eyes, but can also involve other organs and systems. The overlap of these two conditions, referred to as SLE/SS overlap syndrome, presents a unique clinical challenge and requires careful diagnostic and therapeutic considerations.(5)

Patients with SLE/SS overlap syndrome exhibit clinical features of both diseases, which may include polyarthritis, renal involvement, and characteristic autoantibodies such as anti-Ro/SSA and anti-La/SSB, commonly associated with SS, as well as anti-dsDNA and anti-Sm, typically seen in SLE. The coexistence of these autoantibodies can complicate the clinical picture and influence disease prognosis and management strategies.(6)

Recent studies have highlighted the importance of recognizing and diagnosing SLE/SS overlap syndrome early in its course to tailor appropriate treatment regimens and improve patient outcomes. The pathophysiological mechanisms underlying this overlap syndrome remain a subject of ongoing research, with a focus on genetic predisposition, environmental triggers, and immune dysregulation. Understanding these mechanisms is crucial for developing targeted therapies and improving the quality of life for affected individuals.(7)

In this review, we aim to provide a comprehensive overview of SLE/SS overlap syndrome, discussing its epidemiology, clinical manifestations, immunological profile, diagnostic criteria, and current therapeutic approaches. We will also explore the latest advancements in research that shed light on the complex interplay between SLE and SS, offering insights into potential future directions for clinical practice and research.(8)

Case Presentation :
A 58 year old female came to svp hospital with Chief complaint a low grade fever on and off since 4-5 years resolved after taking medications.

C/o mouth ulcer at soft palate, which was painful, recurrent and resolved with medication but not associated with bleeding or discharge since 4 years.

C/o rashes over sun exposed areas of skin like bilateral hands and legs which was reddish brown in colour, itchy, round shape , thick and scaly with h/o on and off since 3 years.

C/o falling of hair which gradually became thinner, broken,non scarring and patchy at temporal and parietal scalp region since 3 years.

C/o skin hyperpigmentation (bluish to black colour) over bilateral legs and foot which was non itchy, non ulcerative, painless since 2 years.

C/o dryness of mouth, crackles lips and tongue a/w difficulty in swallowing and speaking, loss of taste sensation since 2 years

C/o dryness of eyes , redness of upper eye lids a/w difficulty in near vision ,blurring of objects since 2 years

C/o of breathlessness on exertion since last one year.

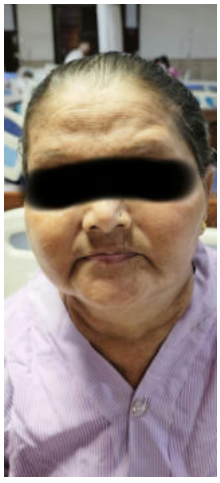
Past History :
K/C/O HTN since 2011

no history of diabetes mellitus , trauma, surgery, thyroid disease, surgery, trauma, infection

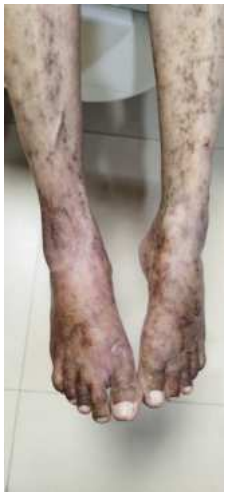
Examination :
hyperpigmentation + over B/L legs and foot
Ulcer + over left foot
Skin thickening + over fingers & cheeks
Alopecia +

RS, CVS and CNS systemic examinations were normal.

1).



2).



Investigation :

DATE	22/07/2020	20/07/2020	23/09/2021	16/02/2022	07/3/2023	15/6/2023	30/06/2023	16/9/223
Hb	12.4	11.9	6.39	9.6	11	10	9.5	11.7
WBC	3660	3200	3250	3800	2850	3290	3100	7.08
PLT	242	246	36K	245	228	186	172	217
N/L/E	8/14/2	84/10/2	75/21/1	62/35/2	68/26/2	68/24/2	60/25/3	87/8/1
MCV/RDW		90.2/15.2	88/13.4	90.9/13.3	90/14	95/13	89/12.7	98.7/15.7
HCT	38.3	36	21.2					35
UREA						47		
CREAT				1.18	0.93	0.94		0.79
Na+								
K+								
ALT/AST		13/-		21/-	18/11	12/-		20/23
ALP						54		
TB/DB						0.4/0.2		
TP/ALB								
CRP					7.1	6.5		1.32
CPK				21.35				
ESR				132	103	72		21

22/07/2020
URINE R/M :
blood +
protein -trace
rbc 4-5

Usg Abdomen :
liver-fatty changes

2D ECHO
EF -70%
reduced lv diastolic compliance
Rest normal

TSH - 4.8 (normal)
Hiv, Hbsag, Hcv -Negative
CT brain & MRI cervical spine- normal

14/2/2022
SPEP -serum protein electrophoresis-

total protein -6.92
albumin-3.10(decrease)
globulin-3.82(increase)
a/g ratio-0.81(decrease)
alpha 1 globulin -0.66(increased)
alpha 2 globulin-1.22(increased)
beta globulin-0.93
gama globulin -1
m-band absent

16/2/2023
RA factor - > 80 (<10)

12/3/2023
P-ANCA AND C-ANCA -Negative

ANA / IF :
speckled pattern ++++, 1:100 & 1:200
ANTI SM, SSA, SSB AB+

2019 Eular / Acr Criteria :
SCORE : 29 (>10 POSITIVE FOR SLE)

1.CONSTITUTIONAL :
Fever - 2
2. HEMATOLOGICAL :
Leucopenia - 3
Thrombocytopenia - 4
3. MUSCULOCUTANEOUS :
Nonscarring alopecia - 2
Oral ulcer - 2
Discoid lupus lesion- 4
4. MUSCULOSKELETAL :
Joint involvement - 6
5. IMMUNOLOGICAL :
Anti SM AB : 6

Treatment :
Patient was treated with following management measurements.

General Measures:
Hydration: Encourage frequent sips of water and use of saliva substitutes for dry mouth.

Ocular Lubrication: Artificial tears for dry eyes.

Pharmacologic Treatment:
Hydroxychloroquine
Corticosteroids
Methotrexate
Azathioprine
Rituximab
NSAIDs
Pilocarpine

Patient had been regularly monitored for disease activity through clinical and laboratory evaluations and explained about potential drug side effects and complications. Patient was also advised for regular ophthalmologic and dental check-ups.

DISCUSSION :
The coexistence of Systemic Lupus Erythematosus (SLE) and Sjögren's Syndrome (SS) in patients presents a multifaceted clinical entity that warrants careful consideration. The overlap of these two autoimmune conditions complicates diagnosis, influences disease progression, and impacts therapeutic strategies. Our review highlights several key aspects that need to be addressed for a better understanding and management of SLE/SS overlap syndrome.

The diagnosis of SLE/SS overlap syndrome is particularly challenging due to the overlapping clinical features and the presence of shared autoantibodies. The simultaneous presentation of symptoms such as arthritis, renal involvement, and sicca symptoms can obscure the differentiation between SLE and SS. The use of comprehensive diagnostic criteria that incorporate clinical manifestations and immunological profiles is essential for accurate diagnosis. The American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) criteria for SLE and SS, respectively, provide a framework, but there is a need for specific guidelines for overlap syndromes.(9)

Patients with SLE/SS overlap syndrome often exhibit a unique immunological profile characterized by the presence of both SLE-associated autoantibodies (e.g., anti-dsDNA, anti-Sm) and SS-associated autoantibodies (e.g., anti-Ro/SSA, anti-La/SSB). This dual expression not only complicates the clinical picture but also suggests a common pathogenic mechanism that links these two conditions.(10)

The management of SLE/SS overlap syndrome requires a tailored approach that addresses the specific manifestations of both conditions.

Immunosuppressive therapies, including corticosteroids, hydroxychloroquine, and biologics such as rituximab, have shown efficacy in treating SLE and SS individually. However, the optimal therapeutic regimen for patients with overlap syndrome remains to be determined. Clinical trials specifically designed for this patient population are needed to establish evidence-based treatment protocols.(11)

Advancements in the understanding of the pathophysiology of SLE/SS overlap syndrome are crucial for developing targeted therapies. Research into the genetic predispositions, environmental factors, and immune system dysregulations that contribute to this overlap can reveal new therapeutic targets. Additionally, the development of biomarkers for early detection and monitoring of disease activity can improve patient management. Collaborative efforts among researchers, clinicians, and patient advocacy groups are essential to drive progress in this field.(12)

CONCLUSION :

SLE/SS overlap syndrome represents a complex interplay between two systemic autoimmune diseases, presenting unique diagnostic and therapeutic challenges. This review underscores the need for further research to elucidate the underlying mechanisms, refine diagnostic criteria, and develop tailored treatment strategies. Enhancing our understanding of this overlap syndrome will ultimately improve patient outcomes and quality of life.

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