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CASE REPORT

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A rare case report of coexisting rheumatic heart disease and systemic lupus erythematous

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Abstract

This case report discusses the rare coexistence of Systemic Lupus Erythematosus (SLE) and Rheumatic Heart Disease (RHD) in a 46-year-old female patient, challenging the conventional understanding of their distinct presentations. The patient exhibited migratory joint pains, palpitations, and shortness of breath. Diagnostic investigations confirmed SLE based on EULAR/ACR criteria, with positive anti-nuclear and anti-dsDNA antibodies. Concurrently, transthoracic echocardiography revealed severe mitral stenosis and regurgitation, leading to the diagnosis of RHD. The patient underwent successful open-heart surgery with mitral valve replacement. The discussion explores the rarity of this coexistence, emphasizing the need for cautious consideration and further research into potential immunological overlaps between SLE and RHD. The report concludes with a call for comprehensive studies to enhance our understanding of the pathophysiology connecting these two conditions.

KEYWORDS

coexistance, rheumatic heart disease, systemic lupus erythematosus

1 | INTRODUCTION

Systemic lupus erythematous (SLE), a complex systemic autoimmune condition,[1] and rheumatic heart disease (RHD), a sequela of inadequately treated streptococcal infections involving cross-reactivity of antibodies against M protein affecting heart layers and valves.[2] Both SLE and RHD involve complicated immunological mechanisms. Understanding the potential association between these two conditions holds substantial clinical relevance. It is quite rare to see both of these major diseases present together and manifest with common symptoms at the same time.[3] There are only two similar cases reported till now on this subject; both had the initial presentation of acute rheumatic fever (ARF), followed by RHD, and later developed SLE.[4,5] Though most patients with RHD lack a classical history of ARF, the initial attack is indolent or subclinical.[6] Here, we report an extremely rare case of SLE coexisting with RHD without a typical natural course of ARF.

2 | CASE REPORT

2.1 | Patient history and clinical examination

A 46-year-old female patient presented with multiple joint pains all over the body for 3 years, palpitations, and

shortness of breath (SOB) for 2 years. Joint pain started with small joints in the hands and feet, then progressed to other large joints of the body. It was migratory in nature, present throughout the day, and not associated with swelling, joint deformity, fever, rashes, or skin changes. Palpitation was episodic, irregular in rhythm, and occurred on exertion and relieved on rest. It was associated with sweating and chest pain. SOB was insidious and gradually progressed, initially from no symptoms at ordinary activity (class 1 NYHA) to causing symptoms in less than ordinary physical activities (class 3 NYHA). It was associated with nighttime awakenings. The patient has no documented history of diabetes or hypertension. There is no history of fever, cough, night sweats, anorexia, vomiting, or pain in the abdomen.

She denies a history of syncopal attacks, weakness of limbs, decreased urine output, hematuria, hoarseness of voice, dysphagia, or recent drug intake.

On examination, she had a regular pulse of 72 beats per minute. Blood pressure was 100/70 mm of mercury, respiratory rate was 19 breaths per minute, and on cardiovascular examination, first and second heart sounds were heard with no murmur. The tender joint count was 12, and the swollen joint count was 0.

2.2 | Diagnostic investigations and treatment

Investigations showed positive anti-nuclear antibody, showing an end point titer of 1:40, positive anti-dsDNA antibody, and positive anti-histone antibody. RA factor, HLA-B27, and anti-CCP antibody tests were done to rule out rheumatoid arthritis, and they yielded negative results. Notable Laboratory findings are enlisted in Table 1. She was started on lifelong hydroxychloroquine and a low-dose steroid for 2 weeks, which was tapered over the next week.

Transthoracic echocardiography (TTE) revealed a thickened mitral valve, sub-valvular deformity present, both commissures fused, calcification present, severe mitral stenosis (MV area = 1.3 cm^2), a mean diastolic gradient of 9 mmHg, moderate mitral regurgitation (Figure 1,Figure 2), moderate tricuspid regurgitation (gradient = 50 mmHg), moderate pulmonary artery hypertension (PASP) of 60 mmHg, and a dilated left atrium with no clots and normal LV function (LVEF = 65%). (Figure 3).

After the diagnosis of RHD, the patient underwent open heart surgery with a midline sternotomy incision, followed by left atrial appendage ligation, mitral valve replacement, and tricuspid valve ring repair. A prosthetic mechanical mitral valve was used, and the patient was discharged with stable hemodynamic status. Lifelong warfarin therapy was initiated with regular monitoring of PT-INR.

TABLE 1 Significant investigations findings.

Tests	Results
1.Antinuclear antibody	Positive, titer is 1:40
2.Anti-dsDNA antibody	Positive
3.Anti-histone antibody	Positive
4.RA factor	Negative
5.HLA -B27	Negative
6.Anti-CCP antibody	Negative

3 | DISCUSSION

The coexistence of RHD and SLE in a single patient is a rare event, and only two case reports have been reported till now regarding it.[4,5]

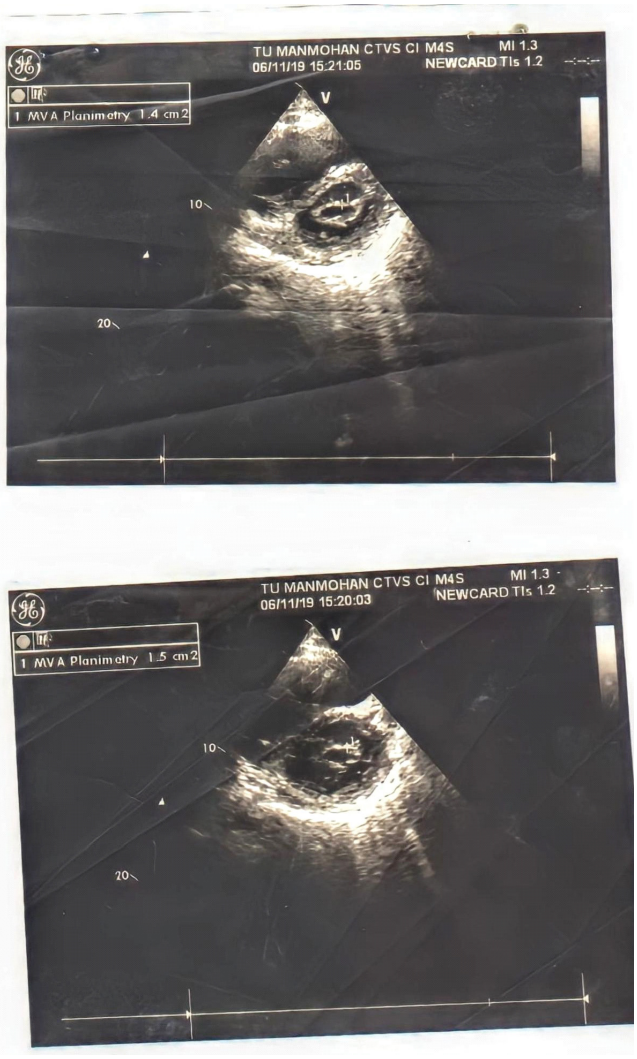
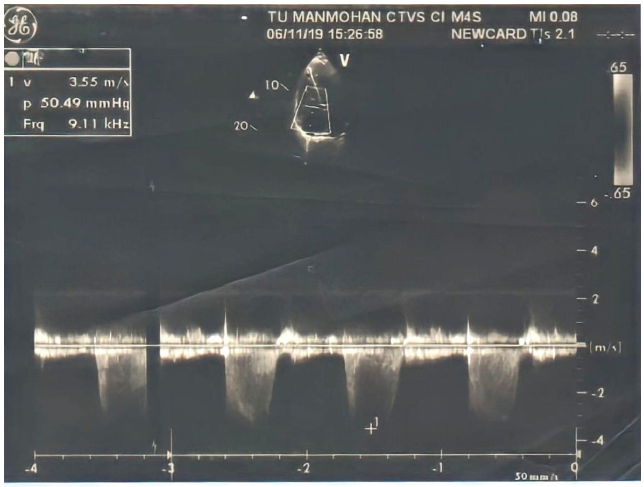


FIGURE 1 Transthoracic Echocardiographic (2D) showing stenosed Mitral valve.



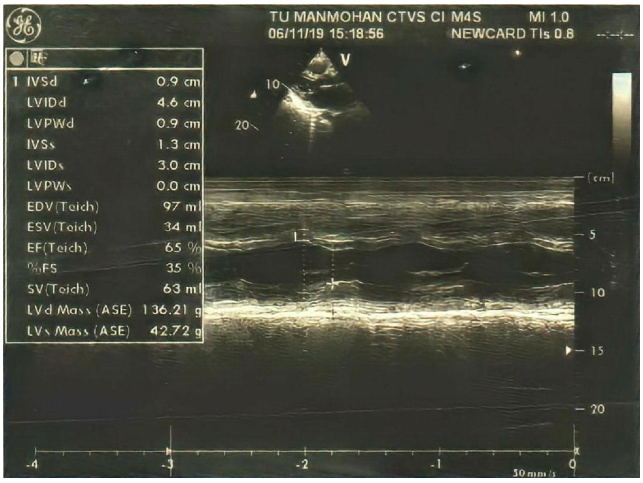


FIGURE 3 Transthoracic Echocardiographic (2D, M-mode) showing left ventricular status.

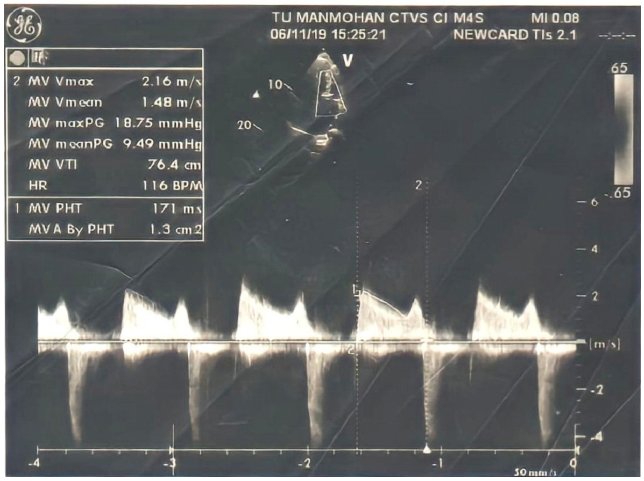


FIGURE 2 Transthoracic Echocardiographic (2D, Doppler) showing Mitral stenosis with mitral valve area 1.3 cm2 and Mitral Valve mean diastolic gradient of 9 mmHg.

Diagnosis of SLE in our patient was established by meeting EULAR/ ACR Revised Euler criteria for SLE.[7] Our patient had ANA titer 1:40 on Hep-2 cells(which is more than 1:80) plus joint involvement (6 points) and Anti-dsDNA antibody(6 points) so total of 12 points and presence of one or more clinical criterion would suffice our patient as a definite case of SLE.

Based on World Heart Federation Guidelines,[8] using TTE, a diagnosis of definite RHD is made without prior history of ARF. Our patient had severe MS with a mean gradient of 9 mmHg (more than 4) and non-rheumatic causes of mitral annular calcification excluded, like ageing[9] and CKD.[10] Our patient was 46 years old with a normal serum creatinine level of 0.6 mg/dL, which excluded the above-mentioned relatively rare causes of non-rheumatic calcification. Globally, RHD remains the cause of 99% of mitral valve stenosis.[11] Additionally, our patient also had moderate mitral regurgitation with a thickened valve.

Both individual diseases are found to have a decrease in regulatory T-cells, which infers the possibility of overlap of immunological pathways causing common manifestations in these diseases,[12,13] but the exact mechanism for this has not been explored till now, which makes it open for further studies in the near future. Our case also supports this, as polyarthralgia can be present in SLE and ARF, which is one of the important criteria for the diagnosis of either disease. One of the case reports also showed the presence of Sydenham's chorea in a patient with SLE,[14] which is one of the components of the Jones criteria for ARF.[15] The commonest heart manifestation of SLE is pericarditis,[16] but mitral valve involvement is also seen, especially sub-valvular changes, as in Libman-Sacks Endocarditis.[17] It corresponds to our result, where the TTE of our patient showed sub-valvular changes in the mitral valve. One study showed SLE caused mitral regurgitation in 74% of patients and mitral stenosis in 3%–4% of patients, which shows MS itself in SLE is rare.[18] One of the pathophysiologies behind valvular involvement in SLE could be citrullination of vimentin, fibronectin, and fibrinogen protein by enzymes produced in chronic inflammation due to SLE, leading to the formation of neo-epitopes[19] making heart valves more susceptible to exaggerated damage by cross-reactivity of antibodies produced against M protein in RHD.[20] It is in contrast with our case, where both mitral stenosis with calcification and mitral regurgitation are present. One of the studies also showed the presence of RHD in 40% of patients with recurrent diastolic heart failure, where they too fulfilled the criteria for diagnosis of SLE without a history of past rheumatic fever.[21] It is comparable to our case, warning us about the possibility of this disastrous consequence.

4 | CONCLUSION

SLE and RHD may present in completely different ways, but clinicians need to know that they can sometimes coexist together. Hence, this coexistence should be taken cautiously and needs to be investigated further.

Thus, we presume that there is a significant association between SLE and RHD, and further studies are needed to further strengthen this linkage for a better understanding of the pathophysiology of these diseases in a more comprehensive way.

AUTHOR CONTRIBUTIONS

Sanjay Dhungana: Conceptualization; project administration; resources; writing – original draft; writing – review and editing. **Abhishek Pandey:** Resources; writing – original draft. **Nijita Aryal:** Supervision; writing – review and editing. **Kshitiz Kayastha:** Writing – review and editing. **Santosh Pandey:** Supervision; writing – review and editing. **Mukunda Thakur:** Supervision.

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The authors claim no conflicts of interest.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable.

CONSENT

Written informed consent form was obtained from the patient to publish this report in accordance with the journal's consent policy.

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