

# Rhupus syndrome and Chiari's network

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## ABSTRACT

A 69-year-old female patient was admitted to our clinic with photosensitivity, symmetric erosive polyarthritis, and cutaneous vasculitis of lower extremities. Rhupus syndrome was diagnosed, and Chiari's network in the right atrium and interatrial septum patent foramen ovale was achieved on transthoracic and transesophageal echocardiography. If it is thought that increased prevalence of antiphospholipid antibodies in patients with rhupus, this congenital remnant is important for the thrombosis risk, cardiac event, and stroke. The association of both diseases may lead to more serious events and cause worse prognosis. Here, our aim is to present a 69-year-old female patient with rhupus syndrome presenting with cutaneous vasculitis and Chiari's network in the right atrium.

**Keywords:** Chiari's network, rheumatoid arthritis, rhupus syndrome, systemic lupus erythematosus

## Introduction

The locomotor system symptoms are observed in systemic lupus erythematosus (SLE) and generally associated with transient, nonerosive, or erosive polyarthritis of proximal interphalangeal, metacarpophalangeal joints, and wrists.<sup>[1,2]</sup> In 1%–3% cases, the joint involvement pattern may be symmetric erosive polyarthritis as in rheumatoid arthritis (RA). The coexistence of SLE and RA called as rhupus syndrome. The clinical features of this syndrome are generally related to RA, also with SLE (mild and lower activity), but the major organ involvement is less frequent in patients with rhupus syndrome.<sup>[3–5]</sup>

Chiari's network is a congenital remnant of the right valve of the sinus venosus.<sup>[6]</sup> The anatomic variation is important due to cardiac complications.<sup>[7]</sup> If it is thought that increased prevalence of antiphospholipid antibodies in patients with rhupus, this congenital remnant will be important for developing formation of thrombosis, cardiac events, and stroke.

We present the case of a 69-year-old female patient with rhupus syndrome and Chiari's network in the right atrium.

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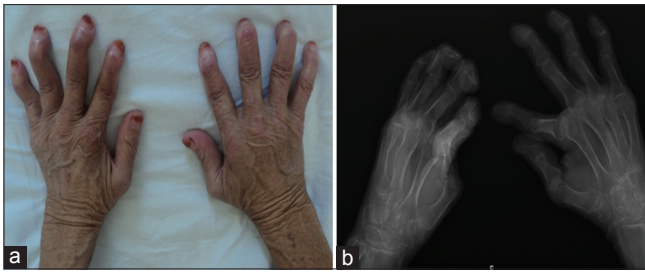
## Case Report

A 69-year-old female patient was admitted to our clinic due to morning stiffness more than an hour and bilateral chronic arthritis. Swan-neck, Boutonnière deformities, and cutaneous vasculitis of lower extremities were observed on physical examination. The blood pressure was 125/65 mmHg, pulse 82 beats/min regularly, and temperature 36.5°C. She had no medical history of taking any drugs. The level of C-reactive protein (CRP) was 18.66 mg/dl and erythrocyte sedimentation rate was 91 mm/h. The patient clinical and laboratory features are shown in Table 1. The patient showed radiological erosion and deformities in the joints [Figure 1a and 1b]. Thoracic computed tomography revealed a mass with a diameter of 51 mm × 28 mm × 32 mm on the right paracardiac area. Chiari's network in the right atrium and interatrial septum patent foramen ovale with normal cardiac motility was achieved on transthoracic and transesophageal echocardiography [Figure 2]. She had 8 points for American College of Rheumatology/European League Against Rheumatism classification criteria for RA and 5 for SLE. She was diagnosed with the rhupus syndrome according to clinical features/laboratory parameters and treated with hydroxychloroquine and prednisolone. Arthritis, vasculitis improved, and the inflammatory markers were decreased to normal values. The patient is still on our follow-up.

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**Figure 1:** (a,b) Erosion and deformities in the joints

**Table 1: Clinical and laboratory features of the case**

Photosensitivity	Positive antinuclear antibody (1/1000 homogeneous)
Vasculitis	Anti-ribonucleoprotein (+)
Morning stiffness	Positive antiphospholipid antibody (anticardiolipin IgG+)
Erosive chronic arthritis	Hypocomplementemia
X-ray changes	Positive rheumatoid factor (156.4 IU/mL)
Lymphopenia	Positive anticyclic citrullinated peptide (200 U/mL)

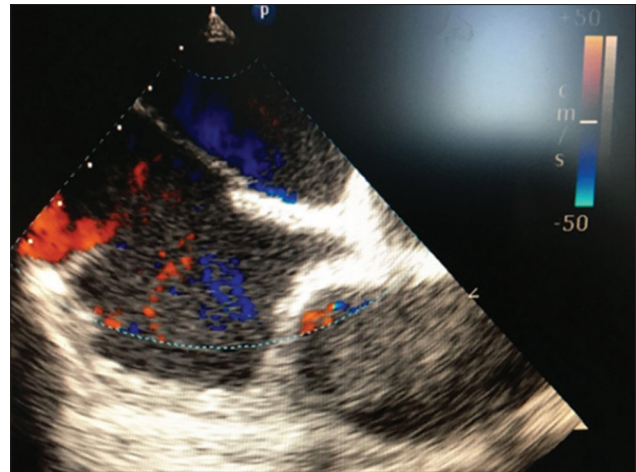
## Discussion

Coexistence of SLE and RA is called as rhus syndrome. The prevalence of the syndrome varies between 0.01% and 2%.<sup>[3,5]</sup> Although it is considered as a variant of lupus arthropathy, the entity is an overlap due to specific autoantibodies such as anti-citrullinated protein antibody (ACPA) for RA and anti-dsDNA, anti-Sm for SLE.<sup>[4,5]</sup>

SLE or RA may be the first presentation (manifested before or simultaneously) of the disease. If the first presentation was SLE, the mean interval time for developing RA was 4.6–16.5 years, in opposite the interval was 4.3–11.<sup>[3-5]</sup> The symptoms of rhus syndrome about RA is associated with erosive arthropathy, pleural, pericardial effusion, skin or hematological disorders, and lower activity for SLE.<sup>[3-5]</sup>

The prognosis is better than SLE and worse than RA.<sup>[3,5]</sup> Furthermore, the autoantibodies are associated with the prognosis. The autoantibodies are suspected in the pathogenesis such as anti-dsDNA for nephritis, anticardiolipin for thrombosis, CRP, anti-RA33, ACPA (citrulline dependence or high levels) for erosive arthritis, and rheumatoid factor (Rf) as the protective factor for nephritis.<sup>[2,8]</sup> The patient had positive antinuclear antibody, antiphospholipid antibodies, Rf, and anti-CCP with hypocomplementemia. We have no information about the first presentation due to the first admission to the hospital. The patient had medical history about erosive polyarthritis since 6 years, without any cutaneous vasculitis or symptoms of SLE.

Pericarditis, heart block, and valvular abnormalities such as dysfunction, atherosclerosis, vegetation, and coronary artery disease are some of the vascular manifestations of SLE. Major organ involvement is less frequent in patients with rhus syndrome.<sup>[3]</sup> In our case, transthoracic and transesophageal echocardiography revealed Chiari's network in



**Figure 2:** Chiari's network in the right atrium and interatrial septum patent foramen ovale

the right atrium. It is a congenital remnant of the right valve of the sinus venosus without any clinical significance and can be found in 2% patients evaluated with transesophageal echocardiography.<sup>[6]</sup> However, it may be associated with valvular endocarditis, embolic events, thrombus formation, and arrhythmias.<sup>[7]</sup> If it is thought that increased prevalence of antiphospholipid antibodies in rhus syndrome, this congenital remnant will become important for developing formation of thrombosis, cardiac events, and stroke. To the best of our knowledge, this is the first reported case of rhus syndrome with Chiari's network.

Consequently, the coexistence of connective tissue diseases is a rare entity such as rhus syndrome. Although Chiari's network is an incidental finding, the rheumatologist should be careful in patients with rhus syndrome and Chiari's network due to complications such as embolic events, thrombus formation, arrhythmias, and worse prognosis.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

## References

1. Ceccarelli F, Perricone C, Cipriano E, Massaro L, Natalucci F, Capalbo G, *et al.* Joint involvement in systemic lupus

- erythematosus: From pathogenesis to clinical assessment. *Semin Arthritis Rheum* 2017;47:53-64.
2. Budhram A, Chu R, Rusta-Sallehy S, Ioannidis G, Denburg JA, Adachi JD, *et al.* Anti-cyclic citrullinated peptide antibody as a marker of erosive arthritis in patients with systemic lupus erythematosus: A systematic review and meta-analysis. *Lupus* 2014;23:1156-63.
  3. Messuti L, Zoli A, Gremese E, Ferraccioli G. Joint Involvement in SLE: The Controversy of RHUPUS. *Int Trends Immun* 2014;4:155-61.
  4. Liu T, Li G, Mu R, Ye H, Li W, Li Z, *et al.* Clinical and laboratory profiles of rhupus syndrome in a Chinese population: A single-centre study of 51 patients. *Lupus* 2014;23:958-63.
  5. Tani C, D'Aniello D, Delle Sedie A, Carli L, Cagnoni M, Possemato N, *et al.* Rhupus syndrome: Assessment of its prevalence and its clinical and instrumental characteristics in a prospective cohort of 103 SLE patients. *Autoimmun Rev* 2013;12:537-41.
  6. Loukas M, Sullivan A, Tubbs RS, Weinhaus AJ, Derderian T, Hanna M, *et al.* Chiari's network: Review of the literature. *Surg Radiol Anat* 2010;32:895-901.
  7. Bae CH, Kwon OC, Lee S, Lee CH, Cho JW. Cystic mass on right atrium of unusual form of Chiari's network: A Case report. *Korean J Thorac Cardiovasc Surg* 2012;45:254-6.
  8. Skare TL, Nisihara R, Barbosa BB, da Luz A, Utiyama S, Picceli V, *et al.* Anti-CCP in systemic lupus erythematosus patients: A cross sectional study in Brazilian patients. *Clin Rheumatol* 2013;32:1065-70.