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Rhupus Syndrom in a Young Male Patient: A Rare Case

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Abstract: Rhupus syndrome refers to the simultaneous presence of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). Rhupus syndrome exhibits a higher prevalence in female and rarely in male, similar to systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). Upon examining the literature, it is evident that the majority of case reports consist of female patients. This case report describes a male patient who diagnosed with rhupus syndrome. **Case Report:** We report the case of a 27-year-old male patient who based on the SLE-SLICC 2012 criteria, the patient met the diagnosis criteria of SLE. Based on the 2010 ACR/EULAR criteria, the patient was diagnosed with rheumatoid arthritis with a total score of 9. The joint criteria showed involvement of more than 10 joints with at least 1 large joint, positive rheumatoid factor results, high CRP and ESR levels, and symptom duration of more than 6 weeks. **Discussion:** Rhupus syndrome is a condition characterized by a symmetric polyarthritis which leads to deformation and erosion. It is preceded by symptoms and signs of SLE and the presence of antibodies that are very specific for diagnosis, such as anti-double stranded DNA, anti-Smith, and anti-cyclic citrullinated peptide. **Conclusion:** In this case, the timely detection of rhupus syndrome is crucial in order to select appropriate treatments, minimize potential complications, and enhance the patient's prognosis, despite its rarity.

Keyword: Rhupus syndrome, systemic lupus erythematosus, rheumatoid arthritis.

INTRODUCTION

Rheumatoid arthritis (RA) is a systemic inflammatory disorder characterized by chronic and advancing inflammation throughout the body. It typically presents with joint stiffness, pain, and swelling, and affects around 1% of the global population. RA is more common in females, with a ratio of 2:1 to 3:1 compared to males. Systemic lupus erythematosus is an autoimmune disorder which can affect many different organs. It is more frequent in females, with a female to male ratio of 9:1 (H.A. Mohammed, 2020). Rhupus syndrome refers to the simultaneous presence of both systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). The patient presents with symmetrical deforming arthritis affecting either both large and small joints, accompanied by signs and symptoms consistent with systemic lupus erythematosus

(SLE) (Özişler, 2018). The combined presentation of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA) was initially documented in 1960, while the name Rhupus syndrome (RhS) was created by Peter Schur in 1971. Rhupus syndrome is an uncommon autoimmune condition that combines characteristics of both rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). It often manifests in sequence, with an estimated prevalence ranging from 0.01% to 2% of patients (Drie, T. et al, 2023).

Women are primarily affected by Rhupus. RA is initially diagnosed in 70% of cases, with subsequent development of SLE. On the contrary, SLE is first observed in 20% of cases, followed by a manifestation of RA characteristics. There have been a small number of reported cases when individuals have experienced symptoms of both systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA) simultaneously. Patients who have rheumatoid arthritis (RA) at a young age or systemic lupus erythematosus (SLE) in old age have a higher likelihood of developing Rhupus. Female patients are particularly vulnerable during the postpartum period and around menopause (Hussain M, et al, 2021). In patients with SLE, the prevalence of rhupus varies widely, ranging from 0.09% to 9.7%. The two most recent and largest reported studies recorded prevalences of 1.3% and 1.4% (Antonini, et al, 2020).

Rhupus is a condition characterized by a symmetric polyarthritis which leads to deformation and erosion. It is preceded by symptoms and signs of SLE (systemic lupus erythematosus) and the presence of antibodies that are very specific for diagnosis, such as anti-double stranded DNA, anti-Smith, and anti-cyclic citrullinated peptide (Carta US, et al, 2017). There are currently no established diagnostic criteria or treatment options specifically for Rhupus syndrome. However, the generally adopted criteria for diagnosing Systemic Lupus Erythematosus (SLE) is Systemic Lupus International Collaborating Clinics (SLICC) 2012, and for Rheumatoid Arthritis (RA) is ACR/EULAR-2010. The diagnosis of Rhupus syndrome relies on certain criteria, including articular erosions, a high level of RA factor, anti CCP, ANA, and anti dsDNA (Lozada-Navarro AC, et al. 2018).

Particularly, the prevalence of rhupus syndrome is higher in female patients, with only a limited number of case studies documenting the disorder in male patients (Chowdhury et al., 2023). This case report describes a male patient who diagnosed with rhupus syndrome.

METHOD

A 27 year old male patient came to the ER at Klungkung Regional Hospital with the main complaint of pain in the joints. Complaints of joint pain felt in the joints of the right and left fingers, knee joints and ankles had been felt since 2 months before the patient was admitted to the hospital and felt increasingly worse since 1 day before entering the hospital. Complaints about joint pain come and go, especially pain every morning when you wake up. Apart from that, the patient also complained that reddish spots appeared on the face, namely on the nose, right and left cheeks and in the area near the ear, which had been felt for 3 months before entering the hospital. The red spots persisted for 2 months and did not disappear, they did not feel itchy or painful. Complaints of weakness for no apparent reason, accompanied by a low grade fever that comes and goes. The patient was said to have no previous history of this kind of illness and a history of other systemic diseases was denied. In the family history, no one complained of complaints like the patient.

Vital signs examination revealed compensatory consciousness, with blood pressure 132/86 mmHg, pulse rate 81 times per minute, respiration rate 18 times per minute. axillary temperature of 36.5ºC, with oxygen saturation of 99% with room air. On physical examination, the face showed a malar rash on the nose, cheeks and around the ears, the eyes did not appear anemic or jaundiced, the oral cavity did not appear to have ulcers, on lung examination there were vesicular breath sounds, no rhonchi or wheezing, regular heart sounds no murmur, on joint examination it appears hyperemic and swollen both the right and left hand of first to fifth

proximal interphalangeal joint, with the range of movement when active is limited due to pain, the left genu region appears swollen, not hyperemic, in the range of movement when active and passive are limited because of pain, swelling appears in the right ankle region, there is no hyperemia, range of movement when active is limited because of pain. Complete blood tests were carried out with hemoglobin results of 12.1 g/dL, leukocytes 3.45 x 10^3/uL, platelets 355 x 10^3/uL, random blood glucose 83 mg/dL, C-reactive protein at 124 mg/dl (normal value 0-8 mg/dl), erythrocyte sedimentation rate (ESR) at 61 mm/h, the immunoserological examination found positive rheumatoid factor with results of 64 IU/mL with interpretation of the results ≥ 8 IU/mL is positive and ≤ 8 IU/ml is negative. Next, an antinuclear antibodies (ANA IF) examination was carried out, resulting in a nucleolar pattern, with a titer of 1:320, with a normal reference value of <1:100.

Based on the SLE-SLICC 2012 criteria, the patient met the diagnostic points, namely the presence of malar rash on the face, arthralgia, leukopenia and positive ANA IF, so the diagnosis of SLE was made. Based on the 2010 ACR/EULAR criteria, the patient was diagnosed with rheumatoid arthritis with a total score of 9. The joint criteria showed involvement of more than 10 joints with at least 1 large joint, positive rheumatoid factor results, high CRP and ESR levels, and symptom duration of more than 6 weeks. The initial treatment given was the analgesic ketorolac 30 mg in drip NaCl 0.9% 100 ml every 8 hours, proton pump inhibitor omeprazole 40 mg intravenously every 12 hours, and methylprednisolone 62.5 mg intravenously every 12 hours. After the patient was hospitalized for 5 days, the patient's complaints improved and he was discharged with therapy of methylprednisolone 4 mg tablet every 12 hours and hydroxychloroquine 200 mg every 24 hours. The patient was then referred to the Rheumatology Division at the central general hospital for further examination and treatment.

RESULT AND DISCUSSION

Rhupus syndrome refers to the simultaneous presence of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). The presence of symmetrical deforming arthritis in both small and big joints, together with accompanying signs and symptoms of systemic lupus erythematosus (SLE), appears clinically. This clinical condition is associated with high specificity autoantibodies, including anti-dsDNA, anti-Sm, and anti-CCP (H.A. Mohammed, 2020). The term "rhupus" was introduced by Peter Schur in 1971 to identify patients who meet the diagnostic criteria for both systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). The occurrence of many connective tissue diseases simultaneously is exceedingly uncommon. Specifically, the occurrence of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA) together is extremely rare, ranging from 0.01% to 2%, in patients with arthritis. Additionally, the occurrence of SLE and RA is less than 2% in individuals with connective tissue illnesses. Rhupus syndrome exhibits a higher prevalence in women, similar to systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). Upon examining the literature, it is evident that the majority of case reports consist of female patients (Özişler, 2018).

Systemic lupus erythematosus is an autoimmune disorder which can affect many different organs. It is more frequent in females, with a female to male ratio of 9:1 (H.A. Mohammed, 2020). Based on the SLE-SLICC 2012 criteria, diagnosis of SLE can be made if 4 of the 17 criteria are met. The criteria consist of clinical and immunological criteria, where there must be at least 1 clinical criterion and 1 immunological criterion. The clinical criteria for this condition include acute cutaneous lupus, chronic cutaneous lupus, oral ulcer, alopecia, arthritis, serositis, renal involvement (proteinuria), neurological involvement (seizures, psychosis), hemolytic anemia, leukopenia (<4.000/mm^3), thrombocytopenia (platelet count less than 100.000/mm^3), immunological involvement such as positive ANA, anti-dsDNA, and

anti-Sm antibodies, presence of anti-phospholipid antibodies, low complement levels (C3, C4, CH50), and a positive direct Coomb's test. The patient had symptoms of rash, arthritis, leukopenia, and ANA positive. Four criteria out of a total of 17 were met.

The presence of rheumatoid factor, which has a high sensitivity and low specificity, is used as the diagnostic criterion for rheumatoid arthritis (RA). However, the presence of positive anti-CCP antibody has a specificity of 98% for diagnosing the condition. Anti-cyclic citrullinated peptide (CCP) antibodies for rheumatoid arthritis (RA) are recognized as a more precise indicator when compared to rheumatoid factor (Akpinar, et al, 2017). The ACR/EULAR 2010 criteria, released in 2010, introduced new categorization criteria. These criteria have a greater sensitivity of 97% but a lower specificity of 55%. The purpose of this new criterion is to facilitate the categorization of rheumatoid arthritis cases during the initial phase, therefore preventing any delays in diagnosis. According to the ACR/EULAR 2010 criteria, a diagnosis of rheumatoid arthritis requires a minimum total score of 6 out of a maximum of 10 (Hidayat, R., et al, 2021). In this case, the patient was diagnosed with rheumatoid arthritis with a total score of 9. The joint criteria showed involvement of more than 10 joints with at least 1 large joint, positive rheumatoid factor results, high CRP and ESR levels, and symptom duration of more than 6 weeks. The patient was referred to the central general hospital for further examination and management.

This case highlights the importance of having a strong sense of suspicion at the initial point of care, as it can help to avoid delays in making a diagnosis and provide valuable learning opportunities. In remote areas of the country where there is no rheumatologist available, the primary care physician provides a crucial role in identifying and either diagnosing or referring cases with rheumatological conditions. Early diagnosis provides a crucial role in improving the prognosis of cases that have problems.

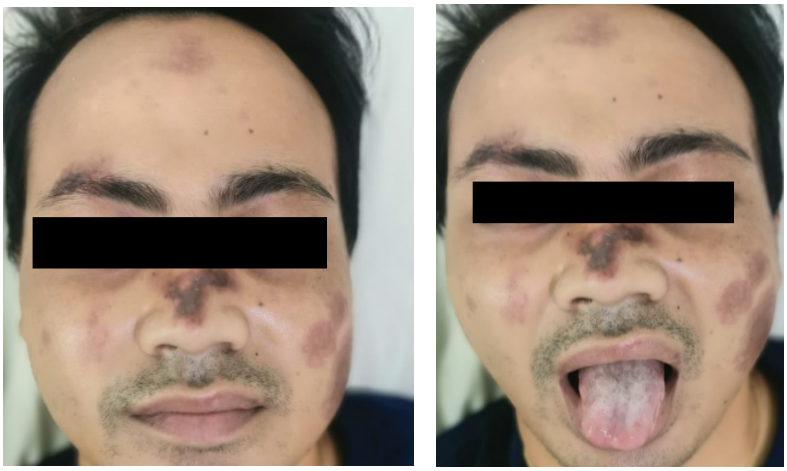

Figure 1. Clinical Manifestation with Malar Rash

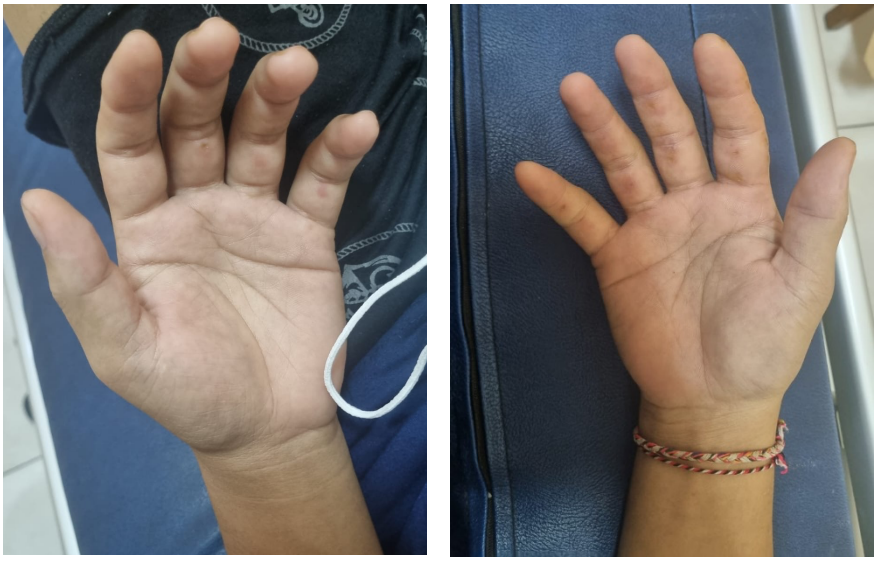

Figure 2. Swelling on Second Digits of Proximal Interphalangeal Joint of Right Hand and Third Digits of Proximal Interphalangeal Joint of Left Hand

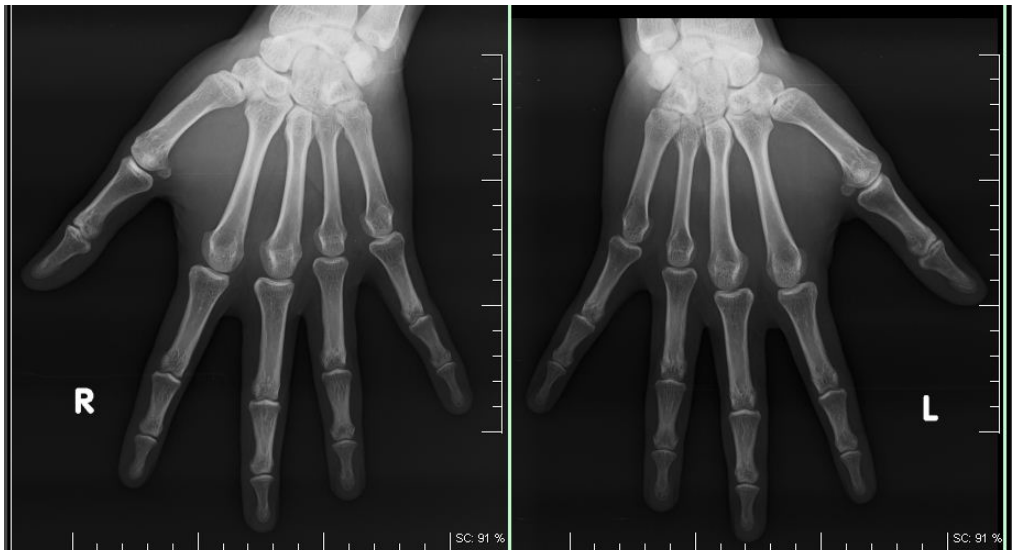

Figure 3. Right and Left Hands X-ray examination showed stage I of Rheumatoid Arthritis without any damage to joints

CONCLUSION

Rhupus syndrome is a rare overlap syndrome that combines features of rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). Therefore, the timely detection of this condition is crucial in order to select appropriate treatments, minimize potential complications, and enhance the patient's prognosis, despite its rarity.

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