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# Systemic Lupus Erythematous and Vitamin D Deficiency in an Adult Sudanese Patient with Sickle Cell Trait

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**Abstract**
systemic lupus erythematous (SLE) is rarely reported in patients with sickle cell trait, here we reported a case who is known sickle cell trait patient presented with skin rash, palpitation, constipation and recurrent frontal throbbing headache with 2 years' history of different joints pain, generalized fatigability, hair loss, feeling hot with normal temperature, the patient was found to have positive antinuclear antibody ANA and low level of vitamin D. The vague symptoms made the suspicious of systemic lupus erythematous so difficult, more attention should be given to such association of autoimmune diseases and sickle cell trait not only sickle cell disease.

**Keywords:** sickle cell trait, systemic lupus erythematous, vitamin D deficiency.

**Abbreviations:**
SCD: sickle cell disease

SCT: sickle cell trait

SLE: systemic lupus erythematous

## Introduction

Sickle Cell Disease SCD encompasses a group of autosomal recessive genetic disorders, including sickle cell anemia (hemoglobin SS), hemoglobin SC, hemoglobin Sb and thalassemia. Sickle cell trait occurs when HbS is inherited from one parent along with normal HbA from the other (e.g., HbSB-thal or HbSC) [1]. SCT patients are typically asymptomatic [2], but problems such as sudden death, vitamin D deficiency and the appearance of SCD-like symptoms may

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occur [3]. Generally, patients with Sickle cell disease present with a wide range of symptoms and signs that overlap with other chronic illnesses like systemic lupus erythematous (SLE). SLE is a chronic autoimmune disease characterized by the production of autoantibodies and the deposition of immune complexes, affecting a wide range of organs [4].

We report a case of a sickle cell trait with newly diagnosed SLE and vitamin D deficiency.

**Case Report**

On March 2020, a 36-year-old female came into Alryan private clinic, Khartoum, Sudan. The patient is a known case of sickle cell trait since childhood after being screened, as her elder brother and sister was diagnosed with sickle cell anemia. Her condition started 2 years ago with generalized fatigability, hair loss, feeling hot with normal temperature, and joint pain. The joints pain involves the elbow, ankle and knee unilateral mainly additive and sometimes migratory, with no swelling except once at the knee without redness or hotness, there was no back pain or body stiffness, the condition aggravated by movement and decreased by simple analgesics. She also noticed a small, painful cervical lymph node with no superficial redness or hotness with no fever or foci of infection, and fine needle aspiration showed signs of inflammation. After that, the lymph nodes disappeared without intervention. At that time, all investigations were normal except for microcytic hypochromic anemia and low vitamin D level (5 ng/ml), for which she received an oral vitamin D and the condition improved but was not completely resolved. Now, the condition worsens with the addition of poor quality and insufficient sleep, loss of social contact, interest and skin rash. The rash on the sun-exposd area, as the skin turns red when she goes out to the sun. In addition, there was a painless oral ulcer and genital ulcer which is sometimes painful. She also complained of shoulder pain when she comp's hair and had never felt weak when she stood up or pain in the hip area.

In the systemic review, there was palpitation, constipation and recurrent frontal throbbing headache that could continue for half the day, moderate in intensity, not interfere with daily activity, associated with nausea but not vomiting, photophobia or photophobia, and there was no obvious trigger, but decreased by paracetamol with metoclopramide or non-steroidal anti-inflammatory drugs and decreased by sleeping in a dark room.

In terms of her past medical history, she had a nasal polyp, gastritis, and recurrent otitis media especially after air fly, and is known to be sensitive to some types of cloth, plastic and some pharmaceutical materials such as salicylic acid powder. She also had a history of recurrent itchy rash on different places in form of redness over the place and there was no history of chronic diseases such as diabetes and hypertension.

There is a strong family history of sickle cell anemia, as one of her brothers had sickle cell anemia and was alive and will. On the other hand, one of her elder sisters died of the disease when she was 13 years old (the status of other family members is not known but no one is symptomatic). There is no known family history of hematological and immunological diseases or a history of recurrent miscarriages and she is not sexually active.

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Current investigations show highly positive antinuclear antibody with strong positivity for SS-A native (60 kda), Ro-52 recombinant, ribosomal P-protein and SS-B in the ANA profile. Vitamin D returned to normal after it was just 5 ng/ml before vitamin D supplementation. Haemoglobin electrophoresis illustrated that HbA was 60.0%, HbA2 was 2.7% and HbS was 37.3%.

The patient showed gradual improvement and all symptoms subsided within a month after she received hydroxychloroquine 200 mg BD and omegas 3.

Diagnosis of SLE was performed by a senior specialist based on the 2019 EULAR/ACR guidelines showed and then followed by both Dr. Ziryab and Dr. Monzer.

**Discussion**

The occurrence of connective tissue diseases, in particular systemic lupus erythematous (SLE), has only been rarely reported in patients with sickle cell disease SCD [5], even though dozens of reported cases are available, but few numbers of case reports published in the association of SLE with sickle cell trait SCT condition [6], that need to be focused on and brought to light more.

SCT patients are typically asymptomatic and spare the serious complications associated with possessing two copies of the mutant allele [2], which would make it easier to diagnose any chronic disease if coexisted, unlike SCD. However, clinical and epidemiological studies have shown that SCT is certainly not an entirely harmless condition [7], and some of these patients can present with complications typical of the SCD phenotype [3], which may be the first differential diagnosis for doctors (symptomatic SCT), delaying the diagnosis of other associated chronic illnesses like SLE that overlap widely with the signs and symptoms of SCD.

In this patient, articular involvement was the main complain, in the association of SLE and SCD, it has been reported that most of the patients were: African, female and middle aged when diagnosis of SLE was made and articular involvement was the most reported complain [8]. In contrast, skin rash is rarely found in the association between SLE and SCD [8], but it has been a major complain by the patient. No data support the similarity of manifestations of SLE when coexisting with SCD and SCT. Moreover, the overlapping clinical features and reporting of ANA positivity in patients with sickle cell disease (SCD) can make diagnosing lupus very challenging for clinicians [8,9].

Some studies suggest the possibility of an association between SCT and the diagnosis of inflammatory and allergic conditions [10] such as skin manifestations in the patient, and a meta-analysis also suggested a higher prevalence of allergic diseases in SLE patients [11]. On the other hand, the patient also suffers from recurrent infections such as otitis media which is probably attributed to the fact that patients with SCD present with a defective activation of the alternate pathway of the complement system [12], which is the reason why there is an increased risk of infection in those patients, but again no data support the occurrence of the same events in patients with SCT.

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Another point of interest was vitamin D deficiency and measuring the level of this vitamin is crucial and not to be missed as it is typical and highly prevalent among those with SLE and SCD [13,14], although no guideline is recommending such practice, but suspicious should be present when such association of diseases exist.

More clinical and epidemiological data are needed to determine the association between SLE and SCT.

**Conclusion**

In this patient, the coexistence of SLE, SCT, and vitamin D deficiency made the diagnosis very challenging, and little information is available regarding the association of these conditions in the literature. More focus should be given, by clinician and researchers, to patients with SCT not only to those with SCD. Additionally, vitamin D levels should be tested in such patients and should not be overlooked.

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