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Systemic Lupus Erythematosus (SLE) with Sickle Cell Disease (SCD)-A Rare Case Series

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ABSTRACT
SLE and SCD are relatively common disorders especially in India and Africa but the combination of connective tissue disease particularly SLE with SCD in a same individual appears to be rare. The actual incidence is not known because most of the published studies are case reports. Only 40 similar cases have been reported in the literature over the past 50 years. Because of the substantial overlap between the clinical features of these disorders, a clinical diagnosis of SLE in patients with SCD is difficult to establish.
Here, we report 2 cases presenting with musculoskeletal and renal complications with fever to the paediatric casualty of MKCG. The complications were initially thought due to SCD with sepsis but when there was no improvement after routine management of SCD, immunological markers were tested which came to be positive. Thus confirming the diagnosis of SLE with SCD.
Keywords: *Connective tissue disease, Systemic lupus erythematosus (SLE), Sickle cell disease (SCD), immunological markers.*

INTRODUCTION

SLE or lupus is chronic, progressive, autoimmune disorder that affects the multiple organ systems with a broad range of clinical and lab manifestations[1]. SCD encompasses agroup of autosomal recessive (AR) genetic disorder that includes sickle cell anemia (hemoglobin SS), hemoglobin SC, and hemoglobin Sb-thalassemia[2]. These patients display a broad spectrum of musculoskeletal, CNS and renal complications that may be associated with either SCD or SLE.[3] Furthermore, approximately 20% of SCD patients have positive ANA antibodies with titers greater than 1/160, making the diagnosis more challenging in clinical practice.[4] The frequency and titers of antibodies in SCD have been reported as relatively higher than in population controls regardless of the presence of autoimmune clinical signs. It has been hypothesized that in SCD, autoantibodies could be induced by a chronic inflammatory state stemming from chronic intravascular and extravascular hemolysis. In an environment of rapid cell turnover, autoantibodies against self-components could be produced. Moreover, environmental stimuli, such as recurrent infections in a permanent inflammatory background, are likely to trigger the production of autoantibodies. Another hypothesis is that the dysfunctional immune status of patients with this disease arising

from functional hyposplenia, complement pathway defects, and impairment of opsonization and phagocytosis could impede clearance of immune complexes.[5] Therefore it has been proposed that when SLE is suspected in a patient with SCD, the best serologic markers appear to be SLE-specific autoantibodies such as anti-dsDNA and anti-Smith. Persistent hypocomplementemia also supports the presence of an immune complex-mediated disease.[6]

CASE REPORT

1ST CASE- A 10 year old male child,born out of consanguineous marriage, was previously diagnosed as a case of Nephrotic syndrome 6 months back, was admitted to the paediatric casualty of MKCG medical college with complains of generalized pain and excruciating pain of the limbs with swelling which was more on the lower limbs without much swelling around eyes and abdomen .The boy had discontinued his medication after taking it for 1 month. Because of the late presentation of Nephrotic Syndrome, all secondary causes was investigated. Following which ANA and Ds DNA was found to be positive. The sickling came positive and HPLC showed sickle cell anaemia. The abnormal swelling of the lower limb was thought to be due to Deep vein thrombosis but on doing doppler, no thrombus was found. A diagnosis of SLE in a patient with SCD was established, with five of the diagnostic criteria of the American College of Rheumatology being met. (Arthritis, serositis, proteinuria, oral ulcer and ANA positive). Fig 1. The final diagnosis was SECONDARY NEPHROTIC SYNDROME DUE TO SLE WITH SCD(SICKLE CELL ANAEMIA).

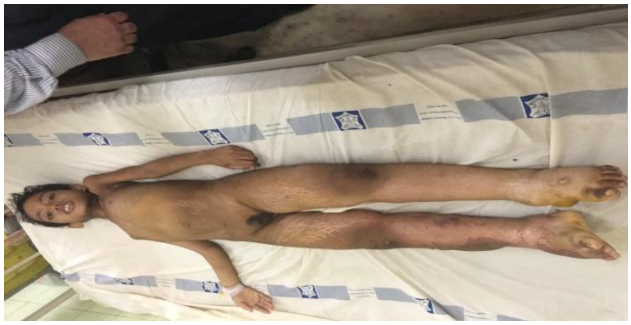

Fig 1 showing swelling of both lower limbs

2ND CASE- A 13 year old female child, born out of consanguineous marriage presented to the paediatric casualty of MKCG medical college with complains of migratory joint pain, fever since last 4 months. With similar history in past but without any history of blood transfusion. A provisional diagnosis of SCD with sepsis was made. On examination hyperpigmentation and rash were found on face with healed ulcers over the lower limbs. Erythematous rashes were found on palm and soles. This pointed towards SLE and on investigating, it was found to be sickle cell heterozygous and immunological markers for SLE like ANA and DsDNA were positive. Five of the diagnostic criteria of the American College of Rheumatology being met. (malar rash, discoid rash, photosensitivity, arthritis, oral ulcer, immunologic disorder). Fig 2 &3. The final diagnosis was SLE WITH SCD (HETEROZYGOUS).

Fig 2 showing malar and discoid rash

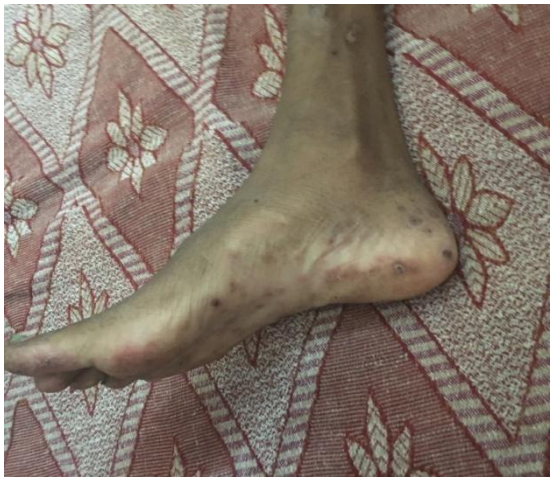

Fig 3 showing rash over sole and leg

DISSCUSSION

Case 1 is a case of SCD complicated by the development of SLE with elements of Nephrotic syndrome. There may be occurance of nephrotic syndrome in SLE but it rarely appears with SCD. Similar case were reported by Francis M.wilso in the year 1964.[7]

Case 2 there is occurance of SLE with sickle cell trait. Similar case were reported by Kabakow and Muchrcke.[8]

Articular involvement is the most frequent lupus related symptom present in 84% followed by serositis (36%), glomerulonephritis class III or IV (11%). Prognosis was favourable in 80%. Cutaneous markers are not frequently mentioned similar to our 1ST case where there was no abnormality of skin. [9]

Toly-Ndour and al. reported that 50% of 88 patients with SCD have positive antinuclear antibodies and 20% with titers greather than 1/200 but only one patient developed rheumatoid arthritis 5 years later and no SLE. In this série, hydroxyurea treated patients have less frequently ANA positive than non-treated patients (p=0.053). Large prospective epidemiological studies are necessary to determine whether the prevalence of immune complex diseases is increased in patients with SCD.[9]

In previous repors by Cherner et al.SCD was diagnosed by 13 years and SLE by 21 years but in our study we have reported early diagnosis of SLE.[10]

Both the cases were treated with prednisolone and showed improvement, the swelling subsided in the first case and the fever and pain subsided in the second case. Both cases were successfully discharged.

CONCLUSION

Physicians should be alerted to the possible development of SLE in patients with SCD not to delay the diagnosis and start appropriate treatment. There is no large prospective epidemiological studies to determine whether there is an increased prevalence of autoimmune disease in patients with SCD. Identifying the etiology of renal abnormality in the setting of co-existing SLE and SCD is important because there are different implementation for morbidity, mortality and therapeutic options.

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