INTRODUCTION

Systemic lupus erythematosus (SLE) is a heterogeneous systemic autoimmune disease. Lupus erythematosus (LE) cells are mature, spherical, homogeneous, and phagocytized neutrophils. In this case report, we describe the observation of LE cells in bone marrow aspirate (BMA) samples from a patient, which raised the suspicion of the presence of SLE, and the cells were analyzed for cytopenia.

Systemic lupus erythematosus is a chronic systemic autoimmune disorder, patients may present with dermatological, hematological, musculoskeletal, pulmonary, and renal symptoms as a result of autoantibody formation directed against self-antigens. The pathological process of lupus starts with a hyperactive immune system. The clinical features of SLE can exhibit a relapsing-remitting course and mimic the course of other diseases, making diagnosis challenging.

LE cells, initially considered to be specific to SLE, have since been observed in other conditions, including rheumatoid arthritis, nephritis, chronic hepatitis, thyroiditis, Sjögren's syndrome, pernicious anemia, ulcerative colitis, red cell aplasia, and mixed connective tissue disease.

CASE REPORT

A 29-year-old female presented with poor appetite, weight loss, and a low-grade fever persisting for 3 months. Upon physical examination, the patient's skin appeared abnormally...
Abdominal examination revealed no organomegaly. There was no axillary, cervical, or inguinal lymphadenopathy. Hematological investigations revealed the following parameters: hemoglobin 89 g/dl, hematocrit 25%, total leukocyte count 3.1 × 10^9 cells/L, and platelet count 252 × 10^9 cells/L. The peripheral blood film showed normochromic; normocytic red cells; leukopenia (neutropenia); no immature cells; and platelets within the normal range. Further investigations were performed, including renal function tests, liver function tests, thyroid function tests, Coombs’ test, chest X-ray, ECG, and an echocardiogram study, which were all normal.

A panel of autoimmune tests was performed, based on the new American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR) criteria, to confirm a diagnosis of SLE in a patient presenting with cytopenia but no other classical signs and symptoms of SLE.

The panel of autoimmune tests demonstrated the following antibody levels: ANA 7.5 IU/ml (positive >1.2), anti-dsDNA antibody >150 U/ml (positive >12 U/ml), antiRo/Sjögren’s syndrome-related antigen A 87.7 U/ml (positive >12), and antiSjögren’s syndrome-related antigen B/La 35.0 U/ml (positive >12). Other autoantibodies, including rheumatoid factor and cyclic citrullinated peptide antibodies, were negative. The patient scored 9 according to the ACR and EULAR SLE criteria (Supplementary Information 1). Next, bone marrow examination was performed to search for the cause of her cytopenia and exclude malignancy or bone marrow failure disorders. She showed no source of infection or any explanation for her cytopenia that had lasted for 3 months. A bone marrow aspirate (BMA) sample from the patient was found to have cellular, normoblastic erythroid hyperplasia with normal myeloid maturation and adequate megakaryopoiesis. Conclusive evidence for a diagnosis of SLE is the presence of unusual LE cells in bone marrow after carrying out all of the necessary investigations mentioned above. Upon further scrutiny, LE cells were observed to be scattered throughout the BMA smear (Figure 1A,B). These results confirmed SLE as the final diagnosis.

3 | DISCUSSION

Various connective tissue diseases are associated with abnormalities in the peripheral blood and bone marrow. Such diseases include SLE, rheumatoid arthritis, nephritis, thyroiditis, pernicious anemia, ulcerative colitis, mixed connective tissue disease, scleroderma, Sjögren’s syndrome, and polymyositis.

Xu et al. detected hematoxylin bodies in BMAs of many pediatric patients with SLE, whereas LE cells were rarely detected. They reported that these findings are a helpful and specific diagnostic indicator that can lead to the detection of SLE when other clinical features are nonspecific.

Cytopenia is common in these diseases due to immune-mediated bone marrow failure and/or excessive destruction of peripheral blood cells. Anemia in SLE may occur due to chronic inflammation, renal insufficiency, immune hemolysis, or, on rare occasions, pure red cell aplasia. Neutropenia occurs in 50% of patients with SLE. Generally, this neutropenia is mild, has little impact on disease, and requires no specific treatment. Any pathogenesis beyond this condition is related to accelerated apoptosis of mature neutrophils. Antineutrophil antibodies have been implicated in this process; however, these can also be present in some patients with SLE without neutropenia. The clinical utility of measuring antineutrophil antibodies in SLE remains questionable. SLE-associated cytopenia may not
present with other classical signs and symptoms of SLE.\(^2\)

In such cases, BMA analysis is performed in addition to or instead of autoantibody analysis to exclude malignancy or bone marrow failure disorders.\(^2\) Although LE cell tests have become overused, they still act as good indicators for planning certain tests for the detection of atypical cases of SLE.\(^9\)

When a patient exhibits a clinical presentation indicative of SLE, clinicians should adhere to the new ACR and EULAR criteria\(^5,6\) to make an accurate diagnosis, and autoantibodies should be tested rather than performing an LE cell assay. However, some patients with SLE might not present with classical symptoms, and many patients present with cytopenia. In these cases, bone marrow biopsies may sometimes be performed in lieu of autoantibody testing to rule out malignancy or bone marrow failure.\(^5\)

Pujani et al.\(^9\) concluded that the unexpected observation of LE cells in BMA films can allow for the diagnosis of unsuspected cases of SLE. In addition, they mentioned two novel insights: First, LE cells can rarely be detected in BMA films prepared immediately without the use of anticoagulants, incubation, or any other manipulation, and that they are an exceptional finding; and second, they reported that accurate cytological observation of BMA samples can provide crucial clues to previously unsuspected conditions.\(^9\)

This report highlights the fact that a simple, immediate, and rapid BMA smear can help in the diagnosis of suspected cases of SLE whenever LE cells are present and may be conclusive for SLE without the need for further tests.

**ACKNOWLEDGEMENTS**

None.

**CONFLICT OF INTEREST**

None.

**AUTHOR CONTRIBUTIONS**

SJM: Conceptualization, Data curation, Formal analysis, Project administration, Resources, Software, Writing- original draft & -review & editing.

**ETHICAL APPROVAL**

None.

**INFORMED CONSENT**

It was obtained to publishing this case.

**DATA AVAILABILITY STATEMENT**

Safaa Jassim Mohammed. (February 1, 2021). Occasionally Detection of Lupus Erythematosus cells in bone marrow sample: A case report. Zenodo. http://doi.org/10.5281/zenodo.4487102

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**SUPPORTING INFORMATION**

Additional supporting information may be found online in the Supporting Information section.

**How to cite this article**: Mohammed SJ. Occasional detection of lupus erythematosus cells in bone marrow samples: A case report. *Clin Case Rep*. 2021;9:e04470. https://doi.org/10.1002/ccr3.4470