International Journal of Research in Medical Sciences
Shaheen SA et al. Int J Res Med Sci. 2023 Sep;11(9):3418-3421
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20232498

Case Report

A rare case report of 32 years old female suffering from lupus panniculitis as an initial sign of systemic lupus erythematosus

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Received: 26 July 2023
Accepted: 10 August 2023

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ABSTRACT

This case report describes a rare case of lupus panniculitis (LEP) in a 32-year-old Egyptian lady who did not have any history of chronic illness. The patient initially presented with leg pain, erythematous nodules, and back ulcers. In addition to proteinuria and increased erythrocyte sedimentation rate (ESR) levels, laboratory tests revealed atypical ANA titers and positive results for anti-DNA, anti-Scl 70, anti-SSa, and anti-SSb antibodies. There was a decline in C3 and C4 levels. A physical examination revealed well-defined ulcers on her back and subcutaneous lumps in both of her legs. The diagnosis of LEP was confirmed by a nodule biopsy. Systemic lupus erythematosus (SLE) was finally determined to be the patient's diagnosis in accordance with the classification standards of the American College of Rheumatology. Methylprednisolone therapy resulted in the resolution of ulcers and nodules, as well as normalization of CBC and C3 levels and a drop in ANA titer. In order to enhance patient outcomes, this example highlights the necessity of treating LEP as a symptom of SLE and the value of early diagnosis and treatment.

Keywords: Lupus panniculitis, SLE

INTRODUCTION

Systemic lupus erythematosus (SLE) is an immune-mediated disease that results in tissue deterioration and inflammation when it affects a person. SLE increases the production of numerous autoantibodies, which causes microvascular inflammation. This medical condition primarily affects women of childbearing age and is caused by a mix of genetic, environmental, and hormonal factors.[1] SLE is characterized by its diverse nature, making its diagnosis and treatment difficult. SLE manifests itself in a variety of ways, ranging from a mild mucocutaneous look to multiple organs and central nervous system (CNS) dysfunction.[2] Photosensitivity, malar rash, hair loss, and discoid lupus are some of the cutaneous signs of SLE. Myalgia, arthralgia, and arthritis are examples of musculoskeletal manifestations.[2]

Furthermore, research has revealed that more than 60 genetic areas are linked to the development of SLE, either directly or indirectly. These genomic areas are also linked to important innate and adaptive immune system mechanisms.[2]

Lupus panniculitis (LEP) is a rare subtype of the various cutaneous manifestations associated with SLE. LEP is also referred to as lupus profundus or subcutaneous LE. Lupus panniculitis is a kind of cutaneous lupus erythematosus that mostly affects the subcutaneous fat, causing indurated plaques or nodules to form. The arms, face, buttocks, chest, and, less frequently, the belly, back, and neck are usually included in the presentation. The lesions in lupus panniculitis may be painful and can exhibit varying degrees of skin involvement, such as erythema, atrophy, or ulceration. Healing of the lesions commonly results in atrophy and scarring.[2]

Here, we discuss a rare case of a 32-year-old Egyptian female who showed up with unusual and atypical SLE symptoms. Notably, this patient's initial manifestation of

her illness was lupus panniculitis. The patient had no history of chronic diseases or autoimmune disorders prior to the current episode. This uncommon presentation demonstrates the complex and sometimes confusing nature of SLE, in which atypical cutaneous signs can precede or develop independently of other systemic symptoms.

CASE REPORT

A 32-year-old Egyptian female with no prior history of any illnesses presented to the clinic with complaints of leg pain, recurrent red nodules on both legs, and a skin ulcer on her back. There was no history of chronic disease or any serious medical conditions in the patient. The physical examination revealed several subcutaneous lumps in both legs showing in Figure 1, as well as several distinct ulcers on her upper back. The patient's temperature was measured at 37°C. There were no other serious symptoms mentioned in addition to the initial complaint.

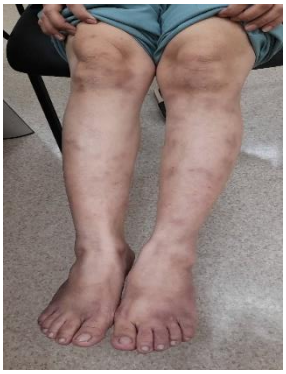

Figure 1: Several subcutaneous lumps in both legs.

During the initial evaluation, immunological tests revealed an abnormal ANA titer of 1/640, as well as positive results for anti-DNA, anti-Scl 70, anti-SSa, and anti-SSb antibodies. The patient also had proteinuria and an erythrocyte sedimentation rate (ESR) of 86 mm. The C-reactive protein (CRP) test, on the other hand, came out negative. Both C3 and C4 levels were found to be low.

A biopsy of one of the nodules was performed to better evaluate the root cause of the patient's complaints. biopsy specimens showed perivascular and cutaneous lymphocytic infiltration, as well as lobular panniculitis with extensive lymphocyte and macrophage infiltration. Adipocyte hyalinization and fibrin thrombosis in the interlobular septa were also observed. Based on these findings, a diagnosis of LEP was made.

To investigate the underlying cause of the patient's condition, the patient was immediately referred to a rheumatology clinic for additional evaluation and treatment. On the basis of the clinical presentation and immunological findings, the patient was diagnosed as systemic lupus erythematosus (SLE) using the American College of Rheumatology's categorization criteria.

The treatment began with an intravenous infusion of 40 mg/d methylprednisolones (equal to 1.5 mg/kg/d prednisone). Over the course of treatment, the dosage was gradually reduced to 24 mg/d oral administration.

The patient's condition improved noticeably after three months of treatment. The ulcers were healed and the nodules had diminished dramatically. The complete blood count (CBC) and C3 levels had returned to normal, and the ANA titer had dropped, according to follow-up tests.

In summary, this case represents a one-of-a-kind presentation of lupus panniculitis as the first symptom of SLE in a 32-year-old female. The rapid diagnosis and treatment with corticosteroids resulted in significant clinical improvement, emphasizing the need for recognizing and treating such unusual signs of SLE as soon as possible. More study and monitoring are needed to better understand the pathophysiology and optimize therapy methods for this rare subtype of lupus erythematosus.

DISCUSSION

LEP was first described by Kaposi in 1883 and was renamed lupus erythematosus profundus by Irgang in 1940. LEP frequently appears as sensitive subcutaneous nodules and plaques with overlaying normal skin, or as erythematous to CCLE-like symptoms (e.g., scaling, follicular plugging, dyspigmentation, telangiectasias, or atrophy). Skin ulceration affects 28% of all LEP patients.[3]

Lupus panniculitis can occur independently or before or after the onset of systemic lupus erythematosus SLE.[4] It has been reported that roughly 70% of patients with lupus panniculitis also have discoid or systemic lupus erythematosus (DLE or SLE), with DLE lesions frequently covering the panniculitis lesions.[5] Lupus panniculitis is often chronic, with periods of remission and exacerbation, and symptoms can last for an average of six years, often resulting in lifelong disfigurement.[5]

The most common histological finding in lupus panniculitis is lymphocytic panniculitis with a lobular predominance pattern.[6] About 20% of individuals may have features similar to DLE, such as lymphocytic infiltration of the upper dermis and basal layer liquefaction.[7] Additionally, as shown in Figure 2, calcification, secondary hyaline degeneration of the basement membrane, blood vessels, and adipose tissue, as well as localized lymphocytic panniculitis with lymphoid nodules harbouring germinal centers in subcutaneous fat or dermis, may also be seen.[4]

The differential diagnosis of lupus panniculitis includes other causes of panniculitis like neoplastic panniculitis (subcutaneous panniculitis-like T-cell lymphoma), inflammatory panniculitis (e.g., erythema nodosum, alpha 1 antitrypsin deficiency, pancreatitis panniculitis, sarcoidosis, and infectious panniculitis (bacterial,

mycobacterial, or fungal). Furthermore, panniculitis caused by other connective tissue diseases such as morphea, dermatomyositis, and overlap syndrome might have clinical and histological similarities to lupus panniculitis.[3,4]

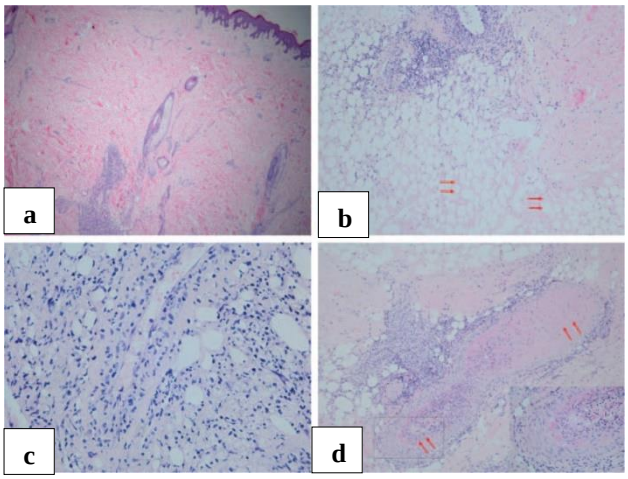

Figure 2: (a) Lymphocytic infiltrations extending from the top dermis to the deep dermis, (b) a description of lymphocytic mixed panniculitis with subcutaneous hyaline necrosis (red arrows), (c) interlobular septal lymphocytic vasculitis, and (d) interlobular septa fibrin thrombosis (red arrows; the region squared in black at the bottom right is magnified).[3,8]

In short, cutaneous lupus erythematosus panniculitis is a distinct manifestation of systemic lupus erythematosus that affects the subcutaneous fat and manifests as distinctive nodules or plaques. It can develop alone or in conjunction with other types of lupus.[9] The disease can be persistent, with individuals frequently experiencing periods of remission and exacerbation. A comprehensive clinicopathologic evaluation is required for an appropriate diagnosis and treatment.

The rarity of LEP as an initial sign of SLE and clinical implications

The case of a 32-year-old girl with lupus panniculitis as the first sign of SLE presented here highlights the rarity of this clinical presentation. While cutaneous signs of SLE are very common, LEP stands out as a distinct type due to its involvement in subcutaneous adipose tissue.[7] The scarcity of such cases adds to the difficulties in diagnosing and recognizing this entity, perhaps leading to delays in effective care.

The clinical symptoms of LEP as a sign of SLE are significant. It emphasizes SLE's complex and unpredictable nature, in which individuals may first appear with distinct cutaneous signs, making early identification difficult.

Healthcare practitioners must be cautious and examine the possibility of underlying systemic involvement in individuals who have cutaneous lesions that do not fit the typical patterns seen in common skin illnesses.

Patients with LEP with a positive ANA should have regular check-ups and be monitored.

Given the likelihood for LEP to precede systemic signs of SLE, individuals with LEP, particularly those with positive anti-nuclear antibodies (ANA), should be evaluated on a regular basis and closely monitored.[10] Regular follow-up visits, thorough physical examinations, and particular laboratory testing are essential for detecting early signs of systemic involvement.[11]

Challenges in diagnosing LEP and preceding systemic manifestations

Diagnosis of LEP is difficult, owing to its rarity and distinctive presentation. Because nodules and plaques are deep-seated, they may mimic other subcutaneous conditions, leading to misdiagnosis or delayed detection of the underlying autoimmune process. Histopathological analysis of a biopsy specimen remains the gold standard for verifying the diagnosis in such circumstances. Even with biopsy, distinguishing LEP from other types of panniculitis can be difficult, necessitating the expertise of dermatopathologists and rheumatologists.[12]

Furthermore, because LEP has the potential to precede systemic signs of SLE, early detection is critical for effective care. Delayed diagnosis may result in systemic disease development, causing damage to important organs such as the kidneys, heart, and central nervous system. Therefore, increased knowledge among healthcare personnel is critical to ensuring quick referral to specialists and prompt diagnosis and management.

The importance of early detection and management for successful outcomes

Early detection and management are critical in improving the prognosis and quality of life of people with LEP-associated SLE. Treatment initiated early in the disease course can avoid or diminish systemic involvement and the risk of irreparable organ damage. Corticosteroids and other immunosuppressive medications are the mainstay of LEP treatment, and rapid initiation can result in cutaneous lesions resolution and disease progression.[13]

CONCLUSION

LEP as an initial manifestation of SLE is uncommon, and its clinical consequences highlight the importance of early detection and care. Healthcare personnel should be aware of this unusual cutaneous manifestation, particularly in individuals with positive ANA and cutaneous features consistent with LEP. Frequent check-ups, attentive monitoring of symptoms, and timely immunology tests are

required to guarantee that these patients have a positive outcome. Early detection and treatment of LEP may be able to avoid or postpone the emergence of systemic signs of SLE, improving the overall prognosis for those affected. More study and education in the medical community are needed to enhance knowledge and management of this rare and challenging SLE manifestation.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Shaheen SA, Barrouq DMS. A rare case report of 32 years old female suffering from lupus panniculitis as an initial sign of systemic lupus erythematosus. Int J Res Med Sci 2023;11:3418-21.