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A Case Study of Systemic Lupus Erythematosus Patient Presenting with Lupus Panniculitis of the Lower Extremities as an Initial Manifestation: A Rare Case

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Abstract. Lupus erythematosus panniculitis is a quite rare entity disease, and to our knowledge occurs in the lower extremity as an initial manifestation of Systemic lupus erythematosus which is very rarely reported. This report presents a case of a 36-year-old female who presented with recurrent subcutaneous nodules on her lower extremities. Further history and physical examination revealed hands and knee joints arthritis with recurrent painless oral ulceration. The blood work performed on the patient revealed positive antinuclear antibody and anti-double strand DNA; hence, the diagnosis of Systemic lupus erythematosus was determined. Skin biopsy was performed and showed lobular panniculitis with heavy infiltration of lymphocyte and plasma cells, and the diagnosis of lupus panniculitis was confirmed. Treatment was started with low doses of systemic steroids and hydroxychloroquine, which resulted in complete clinical remission.

Keywords: Systemic lupus erythematosus, Panniculitis, Chloroquine.

Introduction

Lupus panniculitis which is a variant of erythema nodosum or lupus erythematosus has close resemblance clinically and can only be differentiated histopathologically. This is a rare condition which can occur as a separate entity or rarely along with systemic lupus

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erythematosus. In most cases, it is a disease of women between ages thirty to sixty[1].

Lupus panniculitis or lupus erythematosus profundus (LEP) affects the deep corium of the skin and the subcutaneous tissues are mainly involved. This type of panniculitis differs from other panniculitis by its distribution and clinical changes. Usually, these lesions develop mostly on proximal parts of the upper extremities, trunks, face and head. They rarely appear on the lower extremities[1], which were seen in our case.

This presentation is rare and there are no reports that talk about its incidence or prevalence[2]. Lupus erythematosus profundus (LEP) is an uncommon variant in the clinico-pathological spectrum of lupus erythematosus characterized by chronic inflammation and hyaline necrosis of subcutaneous tissue[3]. Lupus panniculitis occurs in 2% to 5% of systemic lupus erythematosus (SLE) patients. Conversely, 10% to 15% of the patients with this disease as well can have or develop SLE. Lupus panniculitis was seen in 6 out of 228 discoid lupus erythematosus (DLE) patients[4].

Case Report

A 36-year-old Saudi female presented with four months history of recurrent tender subcutaneous nodules on lower extremities primarily on the thighs and upper legs bilaterally, which was initially evaluated and treated by local dermatologist with topical ointments and creams with no significant improvement. These nodules were followed by manifestations of pain in the proximal interphalangeal (PIP) and knee joints bilaterally, and painless recurrent oral ulcerations but no other SLE clinical features.

The physical examination showed multiple tender, rounded firm nodules in the anterior thigh mainly and few in the proximal legs with no superficial skin changes. Joint exam showed few tender PIP joints and mild knee joint effusion bilaterally with joint line tenderness oral mucosa exam showed few ulcers, which according to the patient were painless, rest of the exams were normal.

The blood work revealed positive antinuclear antibodies (ANA) by ELISA and positive anti-(ds-DNA), with normal C3 and C4, anti-smith and anti-phospholipid antibodies were negative. The skin biopsy for the

skin lesions, which was done revealed findings consistent with lupus panniculitis, and based on the clinical and histopathological findings the diagnosis of SLE with lupus panniculitis was confirmed.

She was treated with combination of systemic steroids (prednisolone) in low tapering doses and hydroxychloroquine (Plaquenil) 200 mg twice daily. Within few weeks, the patient showed complete resolution of her clinical features and disappearance of the subcutaneous nodules leaving hyper pigmented skin changes. Complete clinical and serological remission was maintained during 6 months follow-up.

Discussion

Lupus erythematosus panniculitis is a rare and an unusual variant of lupus erythematosus with a chronic recurrent inflammatory process, in which cutaneous infiltrate occurs primarily in a deeper portion of corium giving rise to firm sharply defined nodules of variant sizes lying beneath normal skin. It is often difficult to diagnose as other form of Panniculitis may present similarly.

The patient may have periodic flare-up of panniculitis or have long remissions with no active disease. In most of the cases, there are deep erythematous plaques and nodules, and some ulcers which usually involve the proximal extremities, trunk, breast, face, scalp, buttocks and rarely lower extremities[5,6]. As per our knowledge, there has been only one case reported by Strober[7] of a middle age female presenting with lower extremity lupus panniculitis. Ajubi and Nossent[8] reported two cases presenting with panniculitis elsewhere as first symptom of SLE. Diaz-Jouanen *et al*.[9] reported three out of 270 patients of lupus erythematosus were presented with panniculitis initially. Systemic lupus erythematosus (SLE) should be considered in the differential diagnosis of patients presenting with panniculitis. Diagnosis is confirmed primarily by both clinical and histological findings.

The lesions can spontaneously regress, without ulceration, scarring, and recurrent episodes are common. Moreover, it is a cutaneous process that may be triggered by a wide variety of possible stimuli such as infections, sarcoidosis, rheumatologic diseases, medications, autoimmune disorders such as SLE in this case[10]. This panniculitis affects subcutaneous fat in the skin, usually first evident as an outcropping of erythematous nodules that are highly sensitive to touch[11].

Suspected cases must be confirmed by a series of Serological tests as the ANA test, in addition to specific tests such as the anti-double strand DNA (ds-DNA), anti-smith antibodies (Sm) also confirm the diagnosis of SLE. Levels of certain complement in the blood are also measured to help diagnose and track the disease. The presence of other types of antibodies (anti-phospholipid antibodies) can also aid in diagnosing lupus[12]. Immunofluoresent studies of the biopsy are usually helpful, but it was not done in our patient because of non availability.

In our case, the patient is in fact a case of SLE presenting initially with lower extremity panniculitis, followed with oral ulcers, arthritis of her PIP and knee joints, as well as positive ANA and anti-dsDNA antibodies

Treatment: Antimalaria, like chloroquine and hydroxychloroquine, is the mainstay of treatment for LEP. The chronic and often relapsing course of LEP requires prolonged treatment. Some patients require systemic corticosteroids, usually combined with antimalarials like hydroxychloroquine (Plaquenil) as seen in our case[2]. Furthermore, systemic steroids are a relatively safe therapeutic option if underlying infection has been excluded upon evaluation. Cyclosporin A or thalidomide also have been used to treat LEP[11]. In our case, low doses systemic steroids (Prednisolone) in combination with hydroxychloroquine (Plaquenil) were used to treat the patient. Normally, this drug is used when the patient presents with the milder form of disease such as presence of skin disease and joint pain as seen in this patient.

Conclusion

Lupus panniculitis is a rare disease and difficult to diagnose, which may precede SLE, and the diagnosis of SLE should be considered as an underlying cause of patients presenting with panniculitis. Plaquenil is a safe and effective way of treating this chronic recurrent inflammatory process.

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تقرير حالة نادرة لمريضة بالذئبة الحمراء تظهر في بداية مرضها على شكل التهاب السبلة الشحمية في الأطراف السفلي

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المستخلص. التهاب السبلة الشحمية المصاحب للذئبة الحمراء يعتبر من الحالات النادرة وكما يعتبر ظهوره في الأطراف السفلية كبداية للمرض نادر الحدوث أيضًا. تقرير هذا الحالة يتحدث عن حالة مريضة تبلغ من العمر ٣٦ عامًا تشتكي من عقيدات تحت الجلد في أطرافها السفلية بصفة متكررة. وقد أسفر البحث في تاريخ مرضها وفحصها عن آلام في اليدين والمفاصل وتقرحات متكررة في الفم. نتيجة فحوصات الدم للمريضة كانت موجبة لأضاد النوي والحمض النووي المتكرر وتم تشخيص الحالة بالذئبة الحمراء. كما أخذت عينة من الجلد وتم فحصها ليتم تشخيص التهاب السبلة الشحمية المصاحب للذئبة الحمراء. خضعت المريضة للعلاج بجرعات قليلة من الكورتيزون والهيدروكلوركين مما أعطى نتيجة علاجية إيجابية وأدى إلى خمود المرض اكلينيكيًا.