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Case Study

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CASE REPORT: LUPUS PANNICULITIS – A RARE TYPE OF SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT

Lupus panniculitis is a rare manifestation of Systemic Lupus Erythematous (SLE), characterized by inflammation of the subcutaneous fat. This condition presents diagnostic and therapeutic challenges due to its relapsing nature. We present a case of a 6-year-old female diagnosed with lupus panniculitis. Initial presentation included widespread skin lesions primarily affecting the scalp and axilla. Treatment included steroids, anti-malarials, and immunosuppressants, alongside vitamin supplements. Psychological well-being was also addressed. Medication non-adherence emerged as a significant barrier to effective treatment.

KEYWORDS: Autoimmune diseases, Erythematous lesions, Lupus panniculitis, Lupus profundus, SLE.

INTRODUCTION

Lupus panniculitis, or lupus erythematosus profundus, is a rare form of SLE affecting the subcutaneous fat. It is distinct from other forms of lupus due to its subcutaneous manifestations. While lupus panniculitis occurs in 1-3% of patients with SLE and 10% of those with discoid lupus erythematosus (DLE), it predominantly affects females with a ratio of 2:1. Typically presenting between ages 20-60, this condition is marked by erythematous nodules and ulcerations. Emotional and physical impacts are significant, including severe pain and scarring, which can affect overall quality of life.

CASE REPORT

A 6-year-old female was admitted to the pediatric department on November 23, 2023, for her fourth pulse steroid therapy. The patient had been otherwise well until eight months prior when she developed skin lesions, initially noted on the scalp and axilla. She experienced intermittent fevers over three months, not associated with chills or rigors. Initial diagnosis at a nearby hospital was connective tissue disorder secondary to Sjögren syndrome, based on Anti-RD52 positivity in the ANA profile. Treatment commenced with Hydroxychloroquine, Prednisolone, and vitamin supplements.

Despite initial improvement, the patient experienced a relapse due to medication non-adherence. On readmission to KIMS, lupus panniculitis was confirmed via skin biopsy. The treatment regimen included Methotrexate (15 mg/m²/BSA), Hydroxychloroquine, and pulse steroid therapy. The patient received three rounds of pulse therapy, with the second and third on August 22, 2021, and October 12, 2021, respectively.

At admission, the patient was tachypneic with a saturation of 83% on room air and had multiple healed lesions over the trunk and extremities, with notable 2×2 cm lesions on bilateral knees (Figure 1). She was started on Amoxiclav (50 mg/kg/day) and oxygen therapy. Pulse steroid therapy was paused for 48 hours due to fever spikes. Upon becoming afebrile, she received Methylprednisolone (30 mg/kg/day) for five days.

By day five, her respiratory status improved, and oxygen was discontinued. She was continued on Methotrexate (15 mg/m²) and Hydroxychloroquine (5 mg/kg/day). After a 7-day course of antibiotics, the patient stabilized and was discharged with multivitamins and folic acid supplementation.

DISCUSSION

Lupus panniculitis was first described by Kaposi in 1883. It may be associated with other autoimmune disorders such as Sjögren syndrome and rheumatoid arthritis. Early misdiagnosis as Sjögren syndrome underscores the diagnostic challenge. Although typically seen in adults, our case involved a young child, highlighting the need for vigilance in atypical presentations.

The condition is characterized by relapsing skin lesions, often with watery or bloody discharge and ulceration, leading to scarring and potential psychosocial issues. The histological features, as proposed by Peters and Su, are not universally accepted, and skin biopsies may not always confirm the diagnosis. Positive ANA profiles can support the diagnosis but are not definitive.

Treatment typically involves systemic therapies, as topical treatments are insufficient. Hydroxychloroquine is commonly used, with an onset of action taking up to three months. Alternatives like chloroquine and quinacrine may also be used. Steroid therapy often shows promising results, and in refractory cases, thalidomide, Dapsone, and Rituximab have been employed with varying success. The use of sunscreen is also recommended to prevent further skin damage.

Medication adherence is critical for managing lupus panniculitis, and non-adherence was a significant factor in our patient's relapse. Socio-economic factors may contribute to medication challenges, underscoring the importance of comprehensive support.

List of Tables and Figures Figure 1: Lesions over Hand.

Table: 1 Investigations and Head to toe examination	
Other investigations	**Observations**
RA	Negative
ESR	170 mm/Hr
PSR	Normocytic Hypochromic anemia withneutrophilic leukocytosis with leucoerythroblastic blood picture
Hb	6.9 gm%
PCV	31.2%
Lymphocytes	54.4%
2D Echo	Global Hypokinetic LV Dysfunction (EF: 40%)
Head to toe examination	
Head- Patchy scalp with alopecia, Punched at ulcerated lesions.	
Face- Cracked lips, sunken eyes.	
Abdomen- 2*3 cm oval lesions in the right side of the abdomen.	
Upper limb and lower limb- Multiple circular and oval shaped ulcerated lesions in the elbow and bilateral knees.	
Buttocks- Boggy pant appearance, Multiple punched out lesions over the back.	

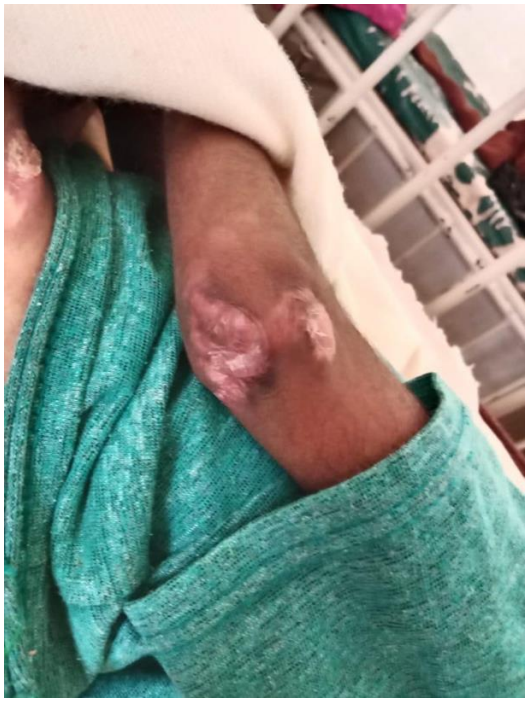

Figure 1: Lesions over Hand.

CONCLUSION

Systemic lupus panniculitis requires careful diagnosis and management. Adherence to treatment is crucial to prevent relapses and improve outcomes. A multidisciplinary approach that includes addressing psychological and socio-economic factors is essential to enhance patient care and quality of life.

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