

## Case report

### Case report on Lupus panniculitis - a rare type of Systemic lupus erythematosus

#### Abstract:

Lupus panniculitis is an infrequent form of Systemic Lupus Erythematosus (SLE). The relapsing nature of skin lesions can make the treatment more challenging. We report a case of a 6-year-old female child with systemic Lupus panniculitis (SLP). The child presented with wounds all over the body initially noted over the scalp and axilla region. The child was managed with Steroids, Anti-malarials and Immunosuppressants including vitamin supplements. Ensuring the psychological wellbeing of the child is also a considered constituent in the care plan. Medication non-adherence of the patient was one of the boundaries of the treatment.

**Keywords:** Autoimmune diseases, Erythematosus lesions, Lupus panniculitis, Lupus profundus, SLE.

#### Introduction

Lupus panniculitis (LP), also known as lupus erythematosus profundus, is a rare type of Systemic Lupus Erythematosus (SLE), which affects the subcutaneous fat. Lupus panniculitis is also known as lupus erythematosus profundus. The diagnosis of lupus panniculitis LP is crucial and needs more attention. The skin lesions must be differentiated from other subcutaneous dermatological conditions. The actual fact is that, 1-3% of patients with SLE and 10% of patients with DLE discoid lupus erythematosus develop lupus panniculitis LP.<sup>1</sup>

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This condition is more frequent in females ~~than in males~~ with a female to male ratio 2:1.<sup>2</sup> The most presenting age group ~~of disease~~ ranges from 20-60.<sup>3</sup> The common manifestations of this autoimmune disease include erythematous nodules and ulcerations. The emotional and physical wellbeing of the patient will be affected due to ~~LPLupus Panniculitis (LP)~~ associated severe pain, atrophy and scarring.

### Case Report

A 6-year-old female child admitted in the pediatric department on 23<sup>rd</sup> November 2021. She was a known case of ~~lupus panniculitis~~LP came for ~~the~~ fourth pulse therapy of steroid. ~~The~~ ~~child~~ was apparently normal 8 months back then ~~she~~ developed wounds all over the body initially noted over the scalp and axilla ~~region~~. ~~Child-She~~ had intermittent fever ~~on and off~~ ~~type for~~ about 3 months ~~but~~ which was not associated with chills and rigors. ~~She was then~~ ~~Then the child was~~ diagnosed with connective tissue disorder secondary to Sjogren syndrome on the basis of Anti RD52 positivity in the anti-nuclear antibody (ANA) profile from nearby hospital. The child was started with ~~Tab. Hydroxychloroquine, Tab. Prednisolone and Vitamin supplementation~~. Following the discharge child was alright for two months then again ~~she~~ developed wounds all over the body due to non-adherence to medications. ~~The child~~she was admitted to KIMS pediatric department ~~was~~ and diagnosed with lupus panniculitis by ~~skin biopsy~~. The child was given ~~with~~ Tab. Methotrexate (15mg/m<sup>2</sup>/BSA), Hydroxychloroquine and ~~was~~ started ~~on~~ ~~pulse steroid- therapy for five days~~. The second and third pulse therapy received by the patient on 22/08/2021 and 12/10/2021 respectively. Other relevant investigations and head to toe examination of the patient are mentioned in table 1.

She was tachypneic at the time of admission with increased work of breathing and a saturation of 83% in room air. She was connected to ~~oxygen prongs~~. She had multiple healed

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1>Include a photo of the slide  
2>Describe the findings in histology

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2.How frequent is the pulse therapy? (e.g. every three weeks?)

**Comment [MMMSA10]:** How many liters?

lesions over the trunk and extremities with 2×2 cm and oval lesions over bilateral knees (figure1 ). Respiratory system showed NVBS bilaterally and coarse crepitations over bilateral subscapular and inframammary area. Injection Amoxiclav (50 mg/kg/day) was started along with oxygen supplement. She had two fever spikes. So, pulse therapy was withheld for 48 hours until she was afebrile. Injection Methyl prednisolone was given 30mg/kg/day 3.3 ml in 100 ml NS over 3 hours for 5 days. On day 5 of antibiotics, her tachypnea reduced and she started maintaining saturation ~~ion~~ room air. Oxygen was tapered and stopped. Tablet Methotrexate (15mg/m<sup>2</sup>) and Tablet Hydroxychloroquine (5mg/kg/day) were advised to be continued. Antibiotics were stopped after 7 days and the child is hemodynamically stable, taking orally well, hence planned ~~to~~ for discharge. On discharge the child was given ~~with~~ ~~Multivitamins~~ multivitamins including Folic acid supplementation.

Comment [MMMSA11]: What does it stand for?

## Discussion

The clinical presentation of systemic lupus panniculitis was first described by Kaposi in 1883.<sup>1</sup> Systemic lupus ~~Panniculitis~~ panniculitis (SLP) may also ~~be~~ associated with other autoimmune conditions like Sjogren syndrome, ~~and~~ rheumatoid arthritis, ~~;~~ T- cell Lymphoma, ~~;~~ traumatic fat necrosis and some other forms of connective tissue disorders. In ~~this the current~~ case, ~~also~~ the earlier diagnosis was Sjogren syndrome secondary to a connective tissue disorder. So, the differential diagnosis is ~~most~~ challenging. Although the most common presenting age group is 20-60 years, here the child was diagnosed with the disease at the age of five. The relapsing type of skin lesions are ~~the~~ characteristics ~~of this~~ disease. Lesions may present with ulcerations preceded by watery or bloody discharge and heal with punched out scars. Sometimes watery or bloody discharges may occur from the lesions followed by ulceration. Healing process may leave punched out scars on the skin. The ~~ulcerating-ulceration~~-scarring cycles do not only affect the ~~natural~~ skin texture but it also impair ~~the~~ mental wellbeing of ~~the~~ patients manifesting as depression and mood swings.

~~mandating, sometimes therapeutic intervention. Depression and mood swings are associated problems in such patients, sometimes needs therapeutic interventions~~ apart from ~~the~~ psychological support. Peters and Su proposed clinical criteria for the histological manifestations of SLP.<sup>4</sup> Although these criteria have not been well accepted, most researchers agreed that SLP has distinctive histological features. Skin ~~Biopsies~~ biopsies are not a completely reliable diagnostic method, ~~however, for the diagnosis of SLP. But~~ an expert dermatologist will be able to distinguish the entities. ~~Although Aa~~ positive ANA profile is one of the digestible diagnostic tests, ~~for the diagnosis of SLP. But~~ the actual role of this parameters is not well established taking into consideration the questionable sensitivity in the diagnosis of lupus panniculitis. Because there is reported lupus panniculitis cases even though the ANA profiling indicated negative results. Other CBC manifestations include leucopenia, anemia, decreased C4 levels and positive rheumatoid factor.

Deep inflammatory process taking place within the subcutaneous adipose layer mandates the prescription of systemic agents as topical preparations are inadequate. are needed to be prescribed in SLP due to the inflammation in subcutaneous adipose layer. Topical preparations may not be good enough to resolve these conditions. Antimalarial drugs were the most commonly recommended drugs which are believed to bring positive response in SLP. Hydroxychloroquine is widely used at a dose of < 6.5mg/kg/day based on ideal body weight.<sup>5</sup> Antimalarials needs up to three months to show its action. The use of chloroquine is also noticed in some cases but hydroxychloroquine is more preferred over chloroquine due to its improved-favourable safety profiles especially in case of retinal toxicity.<sup>5</sup> Some studies demonstrated the beneficial effect of quinacrine in combination with other antimalarials.<sup>6</sup> Steroid therapy is also one of the promising therapies among others. Several studies suggested the successful regression of lesions by the using steroids. Administration of thalidomide has been proved to be effective if the patient is failed to respond with-to

antimalarials.<sup>7-9</sup> But the entire therapy should be monitored carefully due to the hazardous ~~Side-side~~ effects of the drug. The oral use of Dapsone is also verified in the treatment of SLP at ~~the a~~ dose of 25- 75 mg daily.<sup>10</sup> McArdle et al reported ~~the a successful~~ use of Rituximab for the treatment of refractory SLP.<sup>11</sup> ~~There the patient showed a significant clinical response to the treatment with better skin improvement.~~ Some of the ~~other immunomodulatory immunosuppressive~~ agents like azathioprine have also been tried as an adjuvant therapy with antimalarials and steroids.<sup>12,13</sup> Use of sunscreen in SLP patients should be encouraged as ~~they~~ ~~it helps~~ to prevent ~~the~~ further skin damage ~~to some extent~~. All therapeutic drugs ~~for used in~~ SLP are ~~Off~~off-label in the United States.<sup>5</sup> The unavailability of validated clinical results makes controlled and systemic studies more laborious. In this patient, ~~nonadherence to the prescribed medication non-adherence~~ was found to be one of the ~~primary e~~contributing factors for reoccurrence and morbidity. The socio-economic factors of the patient were unfavorable so it could be the probable reason for medication non-adherence.

### **Conclusion**

Systemic lupus panniculitis needs more attention during diagnosis and selection of therapeutic regimen. Appropriate clinical care can significantly reduce the morbidity and mortality rate among such patients. Complete medication adherence needs to be encouraged in patients to achieve maximum therapeutic outcome and improvement in quality of life.

### **COMPETING INTERESTS DISCLAIMER:**

**Authors have declared that no competing interests exist. The products used for this research are commonly and predominantly use products in our area of research and**

country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

UNDER PEER REVIEW

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**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 with neutrophilic leukocytosis with leucoerythroblastic blood picture
Hb	6.9 gm%
PCV	31.2%
Lymphocytes	54.4%
2D Echo	Global Hypokinetic LV Dysfunction ( EF: 40% )
<b>Head to toe examination</b>	
<b>Head</b> - Patchy scalp with alopecia, Punched <del>at</del> -out ulcerated lesions.	
<b>Face</b> - Cracked lips, sunken eyes.	
<b>Abdomen</b> - 2*3 cm oval lesions <del>in</del> -on the right side of the abdomen.	
<b>Upper limb and lower limb</b> - Multiple circular and oval shaped ulcerated lesions in the elbow and bilateral knees.	
<b>Buttocks</b> - Boggy pant appearance, Multiple punched out lesions over the back.	

Figure 1: Lesions over Hand

