

CASE REPORT

Lupus Panniculitis in Children: A Rare Finding

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Correspondence to: Dr. Aijaz Ahmed, Email: aijaztalani19@gmail.com, ORCID: [0000-0002-8648-8064](https://orcid.org/0000-0002-8648-8064)**ABSTRACT**

Lupus panniculitis (LP) is a rare condition that primarily affects adults and is even more uncommon in the pediatric population. This rarity, combined with variable clinical presentations and limited diagnostic approaches in children, often leads to its misdiagnosis or being overlooked. Recently, a case involving a six year's old boy demonstrated the importance of recognizing early skin manifestations and providing timely treatment. The child was presented with painful swelling in the right hand and erythematous plaques on the face and trunk. Although the autoimmune workup yielded weakly positive results, a skin biopsy was performed, which confirmed the diagnosis of LP. This case report serves to emphasize the significance of identifying early skin manifestations and the advancements in diagnostic methods that aid in accurate diagnosis.

Keywords: Child, Lupus Erythematosus, Panniculitis.

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INTRODUCTION

Lupus panniculitis (LP) is a rare condition, with an uncommon clinical variant of cutaneous lupus erythematosus. It accounts for about 2-3% of lupus patients with the rare association of systemic lupus erythematosus, subcutaneous nodules, or plaques that characterize the cutaneous lesion of LP.^{1,4} LP is also known as lupus profundus or subcutaneous lupus erythematosus, the disease was first described by Kaposi in 1869, as a small subset of all the cutaneous LE, it usually occurs in 3rd and 4th decade but children can also develop LP.^{1,2} It is characterized histologically by the skin and subcutaneous adipose tissue with lymphocytic infiltrates in the lower dermis and adipose tissue with features of chronic periadnexal and perivascular inflammation deep in the dermis, which extends into subcutaneous fat.² This present case, one of the very few diagnosed cases in children from Pakistan signifies the importance of correlating the skin and histological findings in making the diagnosis of this little-known disease.

CASE REPORT

A 6 year's old male child was admitted to the department of pediatric medicine, National Institute of Child Health with a complaint of painful swelling over the right hand with erythema and erythematous plaque

for the last 20 days. The patients' mother also notified the sudden onset of right-hand swelling and erythema that was associated with pain and fever. The right thumb was more swollen, and the patient was unable to move fingers and wrist joint due to pain; the patient also had erythematous plaques over the cheeks, neck, and anterior and posterior trunk region. After a few days of admission, the patient developed erythematous small annular purpura over both legs extending up to the hip region. The patient had no history of photosensitivity with no oral ulcers, hair loss, livedo reticularis, husky voice, diarrhea, flu, runny nose, medicine and steroid use, no history of skin thickening, apart from right-hand interphalangeal joint, wrist and elbows joint, no other joint pain was there.

On Skin examination there were multiple well-demarcated erythematous plaques, with scattered adherent crust, plaques on the face starting left maxillary bone up to left lower mandible of around 4*6 cm with no sclerosis seen. Few erythematous plaques with central clearing over the trunk. Right-hand thumb and fingers were more edematous and erythematous (Figure 1). It was extending up to the elbow joint, with restricted painful movement at the right wrist and elbow joint there was a positive temperature gradient (overlying skin was warm). There were multiple purpuras over both legs extending up to the hip region. (Figure 2) The patient was of average height and normal build with vitals and subtitles in the normal range

except clubbing was noted as grade 4. (Figure 3)
The patient was investigated extensively with tests including complete blood count (CBC), blood sugar taken at random (RBS), liver and kidney function tests (KFT) (LFT), thyroid tests, coagulation testing, chest X-rays, antinuclear antibody (ANA), anti-smooth muscle was weakly positive. Moreover, for confirmation of diagnosis, skin biopsy was performed. The specimen for the patients' anti-double stranded DNA (ANTI dsDNA) was 12, which was negative, but the lupus anticoagulant antibodies, and anti-double stranded deoxyribonucleic acid tests. Moreover, echocardiography, ultrasound of the abdomen, and biopsy of the skin was also performed.

Serum ANA and ASMA both were positive with fine cytoplasmic SPE, Extractable Nuclear Antigen Antibodies (ENA) profile shows SCL-70 positive (6.71). A rheumatologist was taken on board who advised to

start tablet deltacortril, hydroxychloroquine (HCQ) with Histamine Type-2 (H2) receptor blockers. Direct Coombs and C3 and C4 levels were further investigated. Direct coombs were positive with low C4 levels (0.36). biopsy was taken from the left side of the trunk which showed lymphocytic infiltration in the lower dermis while adipose tissue reactive lymphocytes showed positivity for CD3 (PAN t). CD4 expression was greater than CD8. These features were consistent with LP. After the biopsy report, a second opinion was taken from the rheumatologist. The consultation suggested the continuation of tablet methotrexate and tablet rapicort for two weeks and then follow up after two weeks with CBC, Serum Glutamic Pyruvic Transaminase (SGPT), urine D/R. On the follow-up visit, the patients' condition clinically improved. The range of movements at the wrist joint improved and swelling subsided (Figure 4).

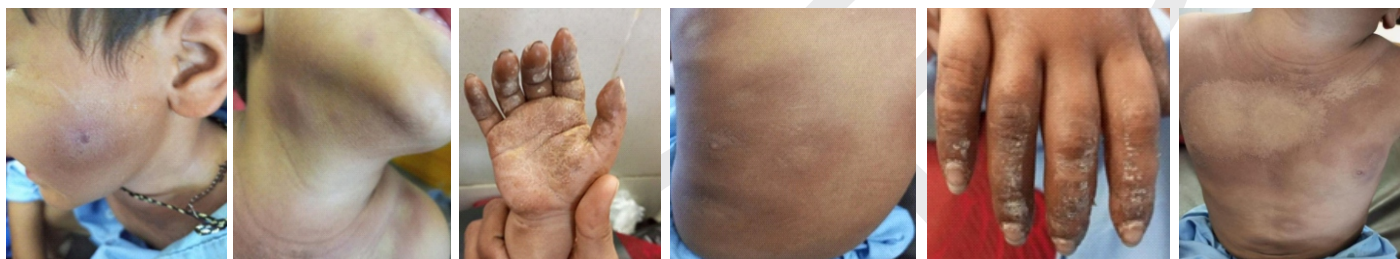


Figure 1: Few erythematous plaques with central clearing over the trunk. Right-hand thumb and fingers were more edematous and erythematous. It was extending up to the elbow joint, with restricted painful movement at the right wrist and elbow joint there was a positive temperature gradient (overlying skin was warm)



Figure 2: Multiple purpuras over both legs extending up to the hip region



Figure 3: Grade 4 clubbing



Figure 4: Range of movements at the wrist joint improved and swelling subsided

DISCUSSION

LP is a rare form of lupus with typical involvement of the dermis and the tissues beneath. This disease has a primary predisposition for middle aged women who are generally more inclined to be affected by autoimmune nature of diseases, but a few cases have also been observed in the pediatric population.⁵⁻⁷

LP can appear alone or in association with discoid or systemic lupus. Patients with systemic lupus have a two to seven percent chance of progression to LP; however, some patients with lupus erythematosus lesions develop systemic manifestations rendering abnormal test results.⁸

As the disease has very similar presentation and histology to connective tissue disease, it is of utmost importance that patients suspected of LP should be thoroughly investigated with investigations including complete comprehensive history, thorough physical examination as well as detailed serologic marker tests to make a conclusive diagnosis and rule out other differential diagnosis. Multiple nodules with indurations or plaques with predilection for trunk and proximal extremities are typical for LP.⁹

The progression of LP is associated with the innate immune system. Adipocytes were found to express toll-like receptors (TLR) 1, TLR2, TLR3, TLR4, and TLR6, while the stromovascular fraction of adipose tissue expressed TLR5, TLR7, TLR8, TLR9, and TLR10. This autoimmune condition involves the recognition of self-DNA and self-RNA, facilitated by the RNA and DNA pattern recognition abilities of TLR7 and TLR9, respectively.^{10,11} Following that, the adaptive immune system was triggered, leading to the production of autoantibodies against nucleic acids and nucleic acid-bound proteins. A prior investigation demonstrated the use of hydroxychloroquine as a treatment for LP due to its capacity to inhibit intracellular TLR in laboratory experiments.¹² Methotrexate, cyclophosphamide, and cyclosporine A are additionally employed in cases of LP that recur or exhibit a more aggressive nature in patients who do not respond to other treatment options. Moreover, thalidomide was also reported as an alternative therapy. However, its use requires strict monitoring of the patient due to the potential occurrence of neuropathies, which are observed in approximately 70% of cases and are deemed irreversible.¹³

The current case report has reported LP associated with systemic lupus erythematosus in a pediatric male patient. This is very significant as it highlights the fact that pediatricians should be aware of etiologies of

dermatologic manifestations such as erythematous swellings and plaques in children. These can be early manifestations of pediatric systemic lupus erythematosus. Monitoring of serologic tests like ANA and anti DSDNA along with skin biopsy are recommended to ensure early recognition and treatment of the disease. All these steps will ensure an improved quality of life for the patients.

CONCLUSION

The current case report has reported LP associated with systemic lupus erythematosus in a pediatric male patient. This is very significant as it highlights the fact that pediatricians should be aware of etiologies of dermatologic manifestations such as erythematous swellings and plaques in children. These can be early manifestations of pediatric systemic lupus erythematosus. Monitoring of serologic tests like ANA and anti DSDNA along with skin biopsy are recommended to ensure early recognition and treatment of the disease. All these steps will ensure an improved quality of life for the patients.

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