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 the skin lesions can make the treatment more challenging. We report the case of 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 considering constituent in the care plan. Medication non-adherence of the patient was one of the limits of the treatment.

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

Introduction

Lupus panniculitis is a rare type of Systemic Lupus Erythematosus (SLE), which affects the subcutaneous fat. It is also known as lupus erythematosus profundus. The diagnosis of lupus panniculitis 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 develop lupus panniculitis.[1] This condition is more frequent in females than in males with a female to male ratio of 2:1.[2] The most common age

at presenting this group of disease ranges from 20-60.[3] The common manifestations of this autoimmune disease include erythematous nodules and ulcerations. The emotional and physical wellbeing of the patient are severly affected due to Lupus Panniculitis (LP)-associated severe pain, atrophy and scaring.

Case Report

A 6-year-old female child was admitted in the pediatric department on 23rd November 2021. She was a known case of lupus panniculitis and came for the fourth pulse of steroid therapy. The child was apparently normal 8 months back and then developed wounds all over the body initially noted over the scalp and axilla region. The child had intermittent fever on and off type about 3 months, which was not associated with chills and rigors. 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 a nearby hospital. The child was started with hydroxychloroquine, prednisolone tabs. and vitamin supplementation. Following the discharge, the child was alright for two months then again developed wounds all over the body due to non-adherence to the medication. The child was admitted to KIMS pediatric department and diagnosed with lupus panniculitis by skin biopsy. The child was given with methotrexate tab. (15mg/m^2/BSA), hydroxychloroquine and started pulse steroid therapy for five days. The second and third pulse therapy was 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 an oxygen saturation of 83% in room air. She was connected to oxygen prongs. She had multiple healed lesions over the trunk and extremities with 2×2 cm and oval lesions over bilateral knees

(figure1). The respiratory system showed NVBS bilaterally and coarse crepitations over both subscapular and inframammary areas. Injection of amoxiclav (50 mg/kg/day) was started along with oxygen supplementation. She had two fever spikes. So, pulse therapy was withheld for 48 hours until she was afebrile. Methyl prednisolone injection 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 in room air. Oxygen was tapered and stopped. Methotrexate tablet (15mg/m^2) and Hydroxychloroquine tablet (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 be discharged. On discharge the child was given multivitamins and folic acid supplementation.

Discussion

The clinical presentation of systemic lupus panniculitis was first described by Kaposi in 1883.[1] Systemic lupus panniculitis (SLP) may also be associated with other autoimmune conditions like Sjogren syndrome, rheumatoid arthritis, T- cell lymphoma, traumatic fat necrosis and some other forms of convective tissue disorders. In this case, also the earlier diagnosis was Sjogren syndrome secondary to connective tissue disorder. So, the differential diagnosis is most challenging. Although the most common presenting age group is 20-60, here a child was diagnosed with the disease at the age of five. The relapsing type of skin lesions is the characteristic of this disease. Sometimes watery or bloody discharge may occur from the lesions followed by ulceration. The healing process may leave punched out scars on the skin. The ulcerating scarring cycles not only affect the natural skin texture but also the mental wellbeing of the patient. Depression and mood swings are associated problems in such patients, which sometimes need therapeutic interventions apart from psychological support. Peters and Su proposed clinical criteria for the histological manifestations of SLP.[4] Although these criteria have not been well accepted, most researchers agreed that SLP has distinctive

histological features. Skin biopsies are not a completely reliable method for the diagnosis of SLP. But an expert dermatologist will be able to distinguish the entities. A positive ANA profile is one of the mandatory tests for the diagnosis of SLP. However, the actual role of these parameters is not well established in the diagnosis of lupus panniculitis because there are 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. Systemic agents are needed to be prescribed in SLP due to the inflammation in the 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.[5] Antimalarials need up to three months to show their action. The use of chloroquine is also noticed in some cases but hydroxychloroquine is more preferred over chloroquine due to its improved safety profile especially concerning retinal toxicity.[5] Some studies demonstrated a beneficial effect of quinacrine in combination with other antimalarials.[6] Steroid therapy is also one of the promising therapies among others. Several studies suggested the successful administration of the steroids. Administration of thalidomide has been proved to be effective if the patient has failed to respond to antimalarials.[7-9] But the entire therapy should be monitored carefully due to the hazardous side effects of the drug. The oral use of dapsone is also verified in the treatment of SLP at a dose of 25 - 75 mg daily.[10] McArdle et al reported the use of rituximab for the treatment of refractory SLP.[11] Their patient showed a significant clinical response to the treatment with considerable skin improvement. Some of the immunosuppressive agents like azathioprine have also been tried as an adjuvant therapy with antimalarials and steroids.[12,13] Use of sunscreen in SLP patients should be encouraged as they help to prevent further skin damage to some extent. All therapeutic drugs for SLP are off-

label in the United States.[5] The unavailability of validated clinical results makes controlled and systemic studies more laborious. In this patient, medication non adherence was found to be one of the prime 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.

References

1. Bednarek A, Bartoszak L, Samborski W. Case report on a patient with lupus panniculitis. Postepy Dermatol Alergol [Internet]. 2015;32(1):59–62.
2. Abu Bakar Tauseef,[1] *Muhammad Sohaib Asghar,[1] Narmin Khan,[1] Maryam Zafar,[1] Uzma Rasheed,[1] Munir Hussain Siddiqui,[3] Umar Farooq,[4] Marium Tauseef[5]Lupus panniculitis in association with anti-phospholipid antibody syndrome on a background of systemic lupus erythematosus: A case report and literature review. EMJ Allergy Immunol. 2020;Vol 37 Issue 4
3. Zhang R, Dang X, Shuai L, He Q, He X, Yi Z. Lupus erythematosus panniculitis in a 10-year-old female child with severe systemic lupus erythematosus: A case report. Medicine (Baltimore) [Internet]. 2018;97(3):e9571.
4. Dhaou B. Ben Dhaou et al. Journal of Dermatology & Dermatologic Surgery. 2017;21:110–2.
5. Braunstein I, Werth VP. Update on management of connective tissue panniculitides: Panniculitis. Dermatol Ther [Internet]. 2012;25(2):173–82.
6. Chung HS, Hann SK. Lupus panniculitis treated by a combination therapy of hydroxychloroquine and quinacrine. J Dermatol [Internet]. 1997;24(9):569–72.
7. Cortés-Hernández J, Torres-Salido M, Castro-Marrero J, Vilardell-Tarres M, Ordi-Ros J. Thalidomide in the treatment of refractory cutaneous lupus erythematosus: prognostic factors of clinical outcome: Thalidomide in refractory CLE. Br J Dermatol [Internet]. 2012;166(3):616–23.

8. Ordi-Ros J, Cortés F, Cucurull E, Mauri M, Buján S, Vilardell M. Thalidomide in the treatment of cutaneous lupus refractory to conventional therapy. J Rheumatol. 2000;27(6):1429–33.

9. Thomson KF, Goodfield MJ. Low-dose thalidomide is an effective second-line treatment in cutaneous lupus erythematosus. J Dermatolog Treat [Internet]. 2001;12(3):145–7.

10. Ujiie H, Shimizu T, Ito M, Arita K, Shimizu H. Lupus erythematosus profundus successfully treated with dapsone: review of the literature. Arch Dermatol [Internet]. 2006;142(3):399–401.

11. Mcardle A, Joshua &., Baker F. A case of "refractory" lupus erythematosus profundus responsive to rituximab Adrian McArdle & Joshua F. Clinical Rheumatology. 2009;28:745–6.

12. Díaz-Jouanen E, DeHoratius RJ, Alarcón-Segovia D, Messner RP. Systemic lupus erythematosus presenting as panniculitis (lupus profundus). Ann Intern Med [Internet]. 1975;82(3):376–9.

13. Nousari HC, Kimyai-Asadi A, Provost TT. Generalized lupus erythematosus profundus in a patient with genetic partial deficiency of C4. J Am Acad Dermatol [Internet]. 1999;41(2):362–4.

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%)
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