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. The relapsing nature of skin lesions can make the treatment more challenging. We report a case of 6-year-old female child with systemic lupus panniculitis. The child presented with wounds all over the body initially notedover 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: Autoimmunediseases,Erythematosus lesions, Lupus panniculitis, Lupus profundus, SLE.

Introduction

Lupus panniculitis (LP), also known as lupus erythematous profundus, is a rare type ofsystemic lupus erythematosus(SLE), which affects the subcutaneous fat. The diagnosis of 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 discoid lupus erythematousdevelop LP.[1] This condition is more frequent in females with a female to male ratio 2:1.[2] The most presenting age groupranges from 20-60.[3] The common manifestations of this autoimmune disease include erythematosus

modulus and ulcerations. The emotional and physical wellbeing of the patient will be affected due to LPassociated severe pain, atrophy and scaring.

Case Report

A 6-year-old female child admitted in the pediatric department on 23rd November 2021. She was a known case of LP came for the fourth pulse therapy of steroid. Thechild was apparently normal 8 months back then she developed wounds all over the body initially noted over the scalp and axilla. Shehad intermittent feverfor about 3 monthswhich was not associated with chills and rigors. She was then 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(5mg/kg/day),Tab.Prednisolone(1mg/kg/day) 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. She was admitted to KIMS pediatric department was and diagnosed with lupus panniculitis by skin biopsy. The child was givenTab. Methotrexate(15mg/m^2/BSA), Hydroxychloroquine(5mg/kg/day) and was started on pulse steroid(30mg/kg/day) therapy for five days. Parents were advised to repeat the pulse steroid therapy every 3-4 weeks. 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(2 lpm). She had multiple healed lesions over the trunk and extremities with 2×2 cm and oval lesions over bilateral knees (figure1). Respiratory system showed normal vesicular breath sound 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 n room air. Oxygen was tapered and stopped. Tablet Methotrexate (15mg/m^2) 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 fordischarge. On discharge the child was given multivitamins including folic acid supplementation.

Discussion

The clinical presentation of systemic lupus panniculitis was first described by Kaposi in 1883.[1] Systemic lupuspanniculitis (SLP) may alsoassociate with other autoimmune conditions like Sjogren syndrome andrheumatoid arthritis;T- cell Lymphoma;traumatic fat necrosis and some other forms of convective tissue disorders. In the currentcase, the earlier diagnosis was Sjogren syndrome secondary to a connective tissue disorder. So, the differential diagnosis is 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 arecharacteristics. Lesions may present with ulcerations proceeded by watery or bloody discharge and heal with punched out scars. Theulceration-scaring cycles do not only affect the skin texture but it alsoimpairsthe mental wellbeing of patients manifesting as depression and mood swings, mandating, sometimes therapeutic intervention Apart from the 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. Skinbiopsies are not a completely reliable diagnosticmethod; however, an expert dermatologist will be able to distinguish the

entities. Although a positive ANA profile is one of the digestible diagnostic tests,the actual role of this parameters is not well established taking into consideration the questionable sensitivity. 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.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 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 itsfavorablesafety profiles especially in case of retinal toxicity.[5] Some studies demonstrated the 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 regression of lesions by the using steroids. Administration of thalidomide has been proved to be effective if the patient failed to respond toantimalarials.[7-9] But the entire therapy should be monitored carefully due to the hazardous sideeffects of the drug. The oral use of Dapsone is also verified in the treatment of SLP at adose of 25- 75 mg daily.[10] McArdle et al reporteda successful use of Rituximab for the treatment of refractory SLP.[11]Some other immunomodulatory 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 ithelps to prevent further skin damage. All therapeutic drugs used inSLP areoff-label in the United States.[5] The unavailability of validated clinical results makes controlled and systemic studies more laborious. In this patient, nonadherence to the prescribed medicationwas found to be one of the primarycontributing factors for reoccurrence and morbidity. The socio-

economic factors of the patient were unfavorable so it could be the probable reason for medication nonadherence.

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.

Conflict of interest

The authors attest to the absence of a conflict of interest.

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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 left ventricular Dysfunction (EF: 40%)
Head to toe examination	
Head- Patchy scalp with alopecia, Punched outulcerated lesions.	
Face- Cracked lips, sunken eyes.	
Abdomen- 2*3 cm oval lesions on 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