Lupus panniculitis – an unusual but distinct clinical entity of systemic lupus erythematosus (SLE)

S A Sathananthan[1], K N L Pathiraja[2], C N Gunasekera[3]

Sri Lanka Journal of Dermatology, 2016, **18**, 50-52

Introduction

Lupus panniculitis or lupus profundus, is a rare presentation of SLE (systemic lupus erythematosus)/ DLE (discoid lupus erythematosus). Lupus panniculitis occurs more frequently as a separate disease than in association with SLE or DLE. It is characterized by chronic inflammation and hyaline necrosis of subcutaneous tissue. The lesions are most frequent on the face and proximal limbs. We report a classical presentation of LP presenting as multiple subcutaneous nodules resistant to long term steroid and Hydroxychloroquine (HCQ) leading to residuallipoatrophy. Our literature survey revealed that rituximab is an exciting treatment option for treatment resistant lupus panniculitis.

Case report

A 31 year old unmarried female presented with recurrent multiple tender subcutaneous nodules in the proximal limbs, trunk and face for 5 years. Nodules have never ulcerated and not produced discharge. Individual nodules healed over 2-3 months with atrophy and pigmentation of overlying skin. She developed 2-3 exacerbations per year and was rarely free of lesions. She had arthralgia for 4 years and recent onset of Raynaud's phenomenon. No other skin lesions or photosensitivity. There was no history of bleeding manifestations or trauma preceding the lesions or history of neuropsychiatric manifestations. Family history was negative.

O/E- Multiple tender subcutaneous nodules on the cheeks, inner aspect of the upper arms and thighs with bluish pigmentation on the overlying skin. Multiple linear atrophic areas appeared as grooves in the both upper limbs and thighs. She also has left hemi facial lipoatrophy. No malar/discoid rash or palatal ulcers. In addition she hadhepatosplenomegaly and lymphadenopathy. She was pale but not icteric.

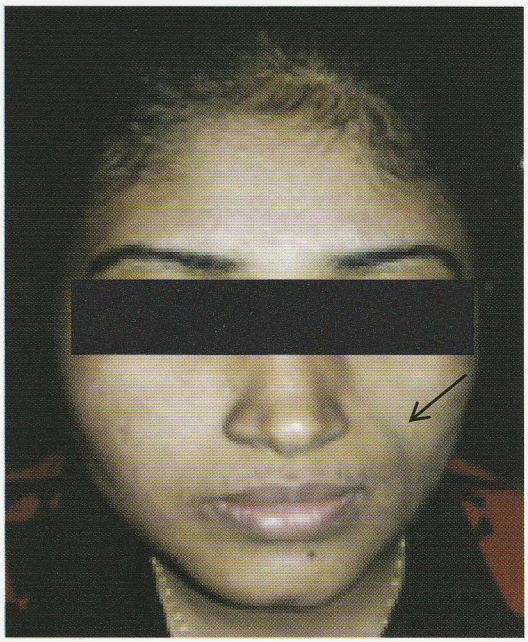

Figure 1. *Left hemifacial lipoatrophy.*

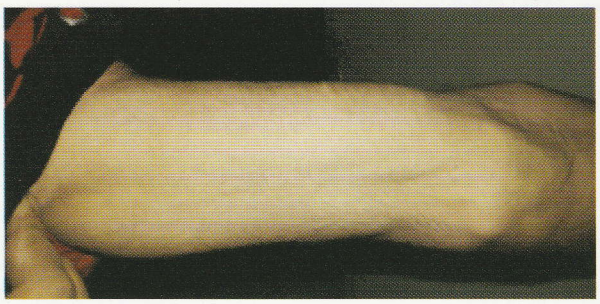

Figure 2. *Multiple lipoatrophy (grooves) in left arm.*

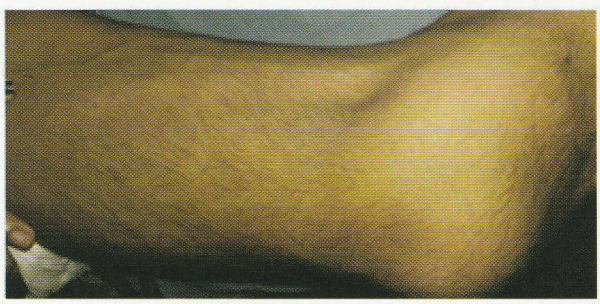

Figure 3. *Lipoatrophy in the right thigh.*

[1,2] *Senior Registrar,* [3]*Consultant Dermatologist, National Hospital of SriLanka, Colombo.*

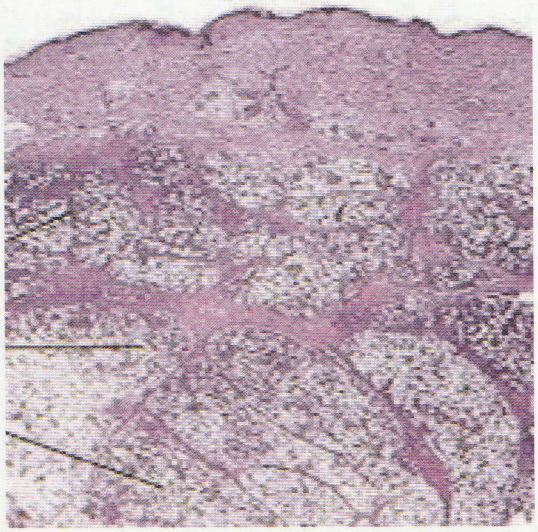

Figure 4. *Lobular panniculitis (×10).*

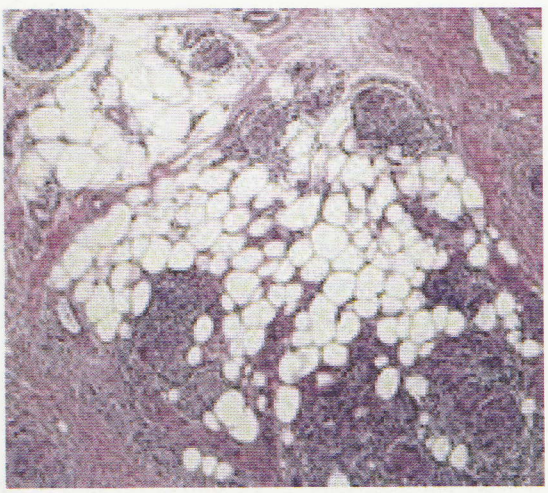

Figure 5. *The dermis shows perivascular and periappendageal inflammatory infiltrate of lymphocytes.*

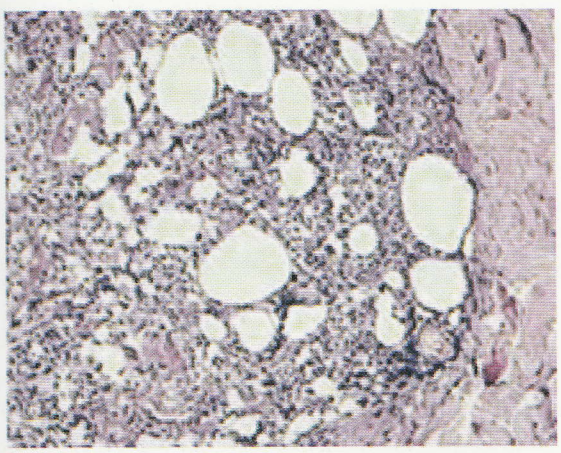

Figure 6. *Subcutaneous tissue shows lobular panniculitis with dense infiltrates of lymphocytes, histiocytes, macrophages, and focal hyalinization.*

Investigations:

WBC/DC -3.9, N-56%, L-36%, M-6%, E-2%, Hb-10 5.g/dl, PLT- 124, ESR-25mm/hr LFT-normal initially but markedly deranged with time (AST>600, ALT>800IU/L), RFT-normal, ANA/DsDNA - negative, Anti Ro/Anti La-negative, U1RNP antibody-negative Complements (C3 and C4) – normal, PT/INR and APTT – normal, Serum ferritin-1166 ng/ml (8-140), Lupus anticoagulant – normal Bone marrow – pancytopenia could be due to autoimmune process.

Skin biopsy – Epidermis shows mild hyperkeratosis with follicular plug formation and basal degeneration. Deep dermis and subcutaneous fat shows predominant lobular panniculitis with a heavy lymphocytic infiltration with histiocytes. Marked perivascular lymphocytic infiltrations are noticed. No vasculitis. Conclusion – The features are more in favour of lupus panniculitis.

We could not perform immunofluorescent studies because of non-availability.

Treatment

She was diagnosed as having lupus panniculitis and treated with oral prednisolone 1mg/kg and HCQ 200mg/bd for more than 3 years with poor response. Azathioprine was stopped after 6 months because of her deranged liver functions (AST>800IU/l,ALT> 1000IU/l). Liver biopsy revealed drug induced hepatitis and liver enzymes improved 4 months after stopping drug. MMF (mycophenolatemofitil) was recently introduced and stopped again due to raised liver enzymes (>5 times) in 4 weeks time. IV dexamethasone pulse was started monthly and prednisolone was tailed off. She developed multiple tender nodules (5-6) while on dexamethasone pulse. We plan to add cyclosporine as a steroid sparing agent because of her uncontrolled disease activity causing permanent disfigurement.

Discussion

The first observations of subcutaneous nodules in SLE were made by Kaposi in 1883. The diagnosis of lupus panniculitis is challenging where involvement of the subcutis is the only manifestation of disease. 1-3% of patients suffering from SLE and up to 10% of those suffering from DLE develop lupus panniculitis. The inflammatory reaction in lupus panniculitis takes place primarily in the deep dermis and the subcutis leading to deep indurated nodules or sharply defined plaques. The overlying skin usually appears normal but there may be erythema, atrophy, ulceration or poikilodermatous or hyperkeratotic changes[1, 2, 3, 4].

As the serological abnormalities are rare, the diagnosis of lupus panniculitis is confirmed primarily by both clinical and histologic findings. In more than 50% the histologic findings closely mimic DLE, in rest the changes are confined to the subcutis only as lobular panniculitis. Hyaline necrosis is a hallmark of lupus panniculitis[8]. Lupus band test is a useful test for the diagnosis. The test is carried out upon non-lesional skin biopsy and the positive result, which indicates the deposits of IgG, IgM and C^3 antibodies, is obtained in 36-70% of cases[5].

Lupus panniculitis is a chronic relapsing condition in which painful lesions may heal with significant disfigurement and lipoatrophy. Hence early diagnosis can prevent permanent disfigurement and the ensuing psychological consequences. Treatment options include intralesional and systemic steroids, antimalarials, mycophenolatemofetil, dapsone, cyclosporine, and thalidomide. The management of this entity can be difficult because of the lack of response to conventional treatments[7]. Surgical correction of lipoatrophy should only be done after complete cessation of activity of the disease. Autologous fat transfer or micro lipoinjection would be the best choice for the treatment of the lipoatrophy[12].

Two patients of a recent report presented with lupus panniculitis refractory to several therapies and demonstrated a remarkable improvement to the infusion of the anti-CD20 monoclonal antibody, rituximab at a dosage of 375 mg/m2/week. After the first infusion, painful lesions had resolved without the appearance of new lesions. Rituximab may be an effective treatment for patients with lupus panniculitis when other therapies are ineffective. To date, there is only one case report of lupus panniculitis treated with rituximab in the literature.

Some authors believe that lupus panniculitis represents a spectrum of subcuticular T-cell lymphoid dyscrasia because of the overlap among lupus panniculitis, atypical lymphocytic lobular panniculitis, and SPTCL (subcutaneous panniculitis like Tcell lymphoma). However, at present the relationship among these entities is not completely clear. As lupus panniculitis overlaps with SPTCL, further investigation and long term follow-up is warranted if lupus panniculitis remains refractory to treatment or systemic symptoms arise because future subcutaneous lymphoma cannot be ruled out[10, 11].

References

1. Tuffanelli DL. Lupus erythematosuspanniculitis (profundus): clinical and immunologic studies. *Arch Dermatol* 1971; **103**: 231-42.
2. Black MM, Cunliffe WJ. Inflammatory disorders of subcutaneous fat. In: Champion RH, Burton JL, Burns DA *et al*, editors. Textbook of Dermatology. London: Blackwell Science; 1998. P. 2420-1.
3. Peters MS, Su WP. Eosinophils in lupus panniculitis and morpheaprofunda. *J Cutan Pathol* 1991; **18**: 189-92.
4. Arnold HL Jr. Lupus erythematosusprofundus (Kaposi-Irgang): Historical review and report of a case. *Arch Derm Syphilol* 1948; **57**: 196-203.
5. Martens PB, Moder KG, Ahmed I. Lupus panniculitis: clinical perspectives from a case series. *J Rheumatol* 1999; **26**: 68-72.
6. Izumi AK, Takiguchi P. Lupus erythematosus-panniculitis. *Arch Dermatol* 1983; **119**: 61-4.
7. Requena L, Sanchez Yus E. Panniculitis Part II. Mostly lobular panniculitis. *J Am Acad Dermatol* 2001; **45**: 325-61; quiz 362-4.
8. Chung HS, Hann SK. Lupus panniculitis treated by a combination therapy of hydroxychloroquine and quinacrine. *J Dermatol* 1997; **24**: 569-72.
9. Peters MS, Su WP. Lupus erythematosuspanniculitis. *Med Clin North Am* 1989; **73**: 1113-26.
10. Aggarwal K, Jain VK, Dayal S. Lupusery thematosus-profundus. *Indian J Dermatol Venereol Leprol* 2002; **68**: 352-3.
11. Winkleman RK. Panniculitis in connective tissue disease. *Arch Dermatol* 1983; **119**: 336-44.
12. Rao TN, Ahmed K, Venkatachalam K. Lupusery themat osusprofundus. *Indian J Dermatol Venereol Leprol* 2010; **76**: 448.