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Case Report

Discoid Lupus Erythematous with Systemic Manifestation: Case Report

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Abstract:

Lupus erythematosus (LE) is a chronic autoimmune disorder with multisystem involvement with varying spectrum of symptoms in between localized multiple dermatological manifestations cutaneous LE (CLE) on one end of the spectrum and severe systemic LE (SLE) on the other end. Aetiology is multifactorial and polygenic. Discoid lupus erythematosus (DLE) is the most common form of chronic cutaneous lupus erythematosus, it is disfiguring skin disease with a significant impact on the patient's everyday life. DLE patients display well-defined skin lesions, often in sun-exposed areas. The disease often has a chronic and relapsing course. It is important to confirm a CLE diagnosis with histopathological examination by biopsy sample. (1)

Classic DLE lesions begin as red-purple macules, papules or small plaques and rapidly develop a hyperkeratotic surface. Most patients with untreated classic DLE lesions suffer indolent progression to large areas of cutaneous dystrophy and scarring alopecia that can be psychosocially devastating.(2)
Here in this case, presentation began with skin manifestation with progression towards systemic involvement patient was later labelled as Systemic Lupus Erythematosus with Discoid Lupus Erythematosus

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Case Report

A 36-year-old female with a congenital port wine stain on left half of her face presented to Medicine OPD with chief complaints of joint pain of bilateral lower limbs with swelling and tenderness and intermittent fever since last 6 months associated with multiple skin lesions and facial oedema. Six months back patient developed an intermittent fever episodes and malaise associated with pruritus for which patient took symptomatic treatment. Later she noticed rashes over both cheeks associated with oral ulcers inside mouth Figure 4. Which developed within 5-6 days causing decreased appetite due to burning sensations while eating. The rash started as a flat mucosal lesion in mouth to raised red-purple lesions approximately each measuring of pea-sized lesions later forming fluid filled lesions which later ruptured with clear fluid, it progressed to the chest, back and below the knee Figure 2. With this complains patient was taking regular treatment from dermatologist with on and off oral steroid but was not relieved of her symptoms. Two months later patient developed an event of loss of appetite, sleep, and irritability with increased suspiciousness for which she started taking psychiatric treatment later patient was on tablet clonazepam 0.25 mg HS.

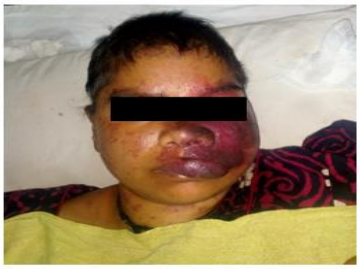


Figure 1. *Port wine stain over left half of face*

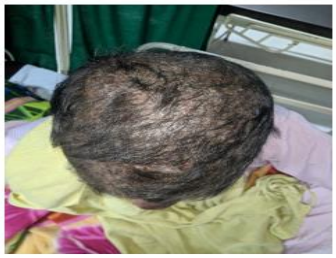


Figure 2. *Scarring Alopecia*

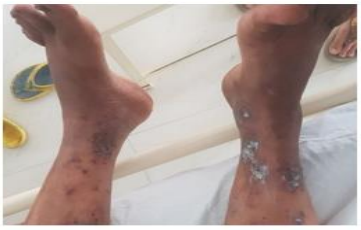


Figure 3. *Well defined ulcerative lesions over legs*

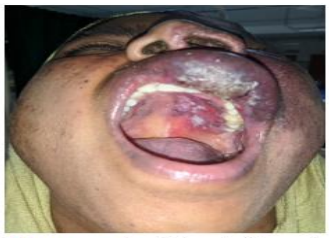


Figure 4. *Facial oedema with oral ulcerative lesions*

Patient present to our medicine OPD with increased rash, facial oedema and bilateral lower limb swelling associated with joint pain and intermittent fever and multiple skin lesions all over body **Figure 3.** & alopecia. **Figure 2.**

Past medical history revealed congenital haemangioma with port wine stain over left half of face **Figure 1.**, history of blood transfusion during pregnancy, history of haemorrhoids.

On examination patient is fairly nourished, afebrile with bilateral pedal oedema below the level of ankle and left facial oedema. Head and neck examination, revealed discrete erosive lesions over the cheek, nose and scalp, measuring approximately 0.5 X 1 cm in diameter. Scalp lesions causing scarring alopecia should be noted which illustrates erythematous, disc-like, scaly plaques showing signs of healing accompanied by scarring and hypopigmentation. Posterior Neck reveals scarring with erythema and hyperpigmentation Neck Lymph nodes are not palpable. Intraoral examination revealed small multiple white lesions which are non-painful along mucosa and hard palate

Examination of the trunk revealed multiple, well-defined, round erosive lesions measuring approximately 1 X 2 cm in dimension. Below Knee patient had bilateral round well-defined ulcerative lesions largest one measuring 2x3 cm. Based on history and clinical examination a provisional diagnosis of Discoid Lupus Erythematosus (DLE) with systemic manifestation was made. Differential diagnosis of Systemic Lupus Erythematosus (SLE) and other vasculitis disorders was considered.

Investigations:

Complete Blood Count suggestive of Bi-cytopenia with Haemoglobin 12.2 gram%, with Total leukocytic count 2090 cells/mm^3 and platelet count reduced to 60000 cells/mm^3. ANA (Anti-Nuclear Antibody) was Positive, Electrocardiogram revealed long Qtc with Left axis Deviation with Left Bundle Branch Block (LBBB), 2decho was suggestive of Ebstein's Anomaly. Urine examination revealed protein=+1 trace, no urinary red cells, 24-hour urine protein 1734 mg / day. Sr. creatinine 15.58 mg/dl, protein / creatinine 0.99 suggestive of low-grade proteinuria, Serum AST 292 U/L, Serum ALP 140 U/L, Serum Albumin 3.0 mg/dl, blood culture and urine culture had insignificant growth, RA factor Negative, HbA1c 5.7%

Histopathological Examination:

The biopsy shows extremely atrophic epidermis with lamellated hyperkeratosis. Papillary dermis shows sclerosis with melanin incontinence. Focal vacuolar dermatitis with colloid bodies is seen. Dermo-epidermal junction shows smudging with pyknotic neutrophils. Basement membrane is thickened and prominent **Figure 5.** There is also thickening of collagen bundles seen in upper two-third of reticular dermis. Follicular infundibulum is dilated and

plugged with parakeratotic stratum corneum. There is mild perifollicular infiltrate of lymphocytes seen which is suggestive of Sclerotic and atrophic variant of DLE

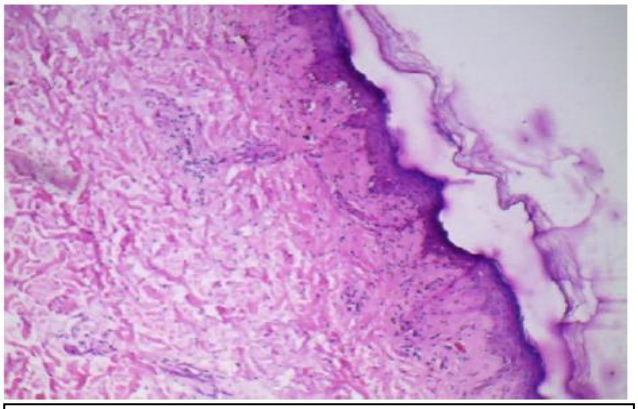
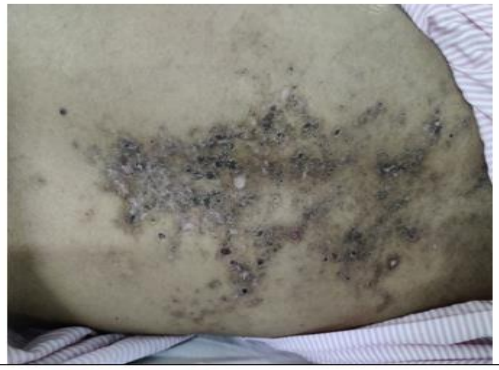


Figure 5. *Histopathological Examination slide of skin biopsy*



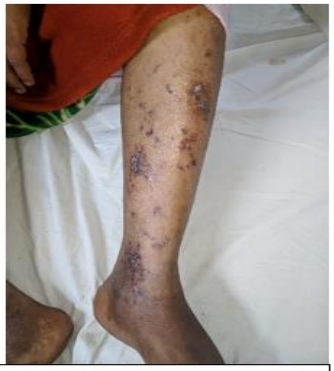


Figure 6. **Figure 7.** **Figure 8.**

Images suggestive of improvement of lesions after pulse steroid therapy with injection methylprednisolone 1gm intravenous once a day for three days In figure 6 is after pulse therapy reduced facial swelling , lesions in figure 7,8 shows scab formation with improvement in ulcerative lesions.

Based on clinical findings, laboratory and histopathological examination. Patient was diagnosed as Discoid Lupus Erythematosus with Systemic Lupus Erythematosus with Ebstein's Anomaly and patient was mainly started on injectable steroid with injectable methylprednisolone 1 gm intravenous once a day for initial 3 days as a 'pulse therapy' later shifted to oral steroid with oral azathioprine 100mg once a day and was given multivitamins, antibiotics, mupirocin ointment, sunscreen lotion, intralesional steroid therapy with injection triamcinolone **Figure 9.** Etc. With this therapy lesions started improving with scab formation **Figure 7 & 8** & oedema over face significantly reduced **Figure 6.**

Figure 9. *Intralesional triamcinolone given 1mg/per injection/per 1 site*

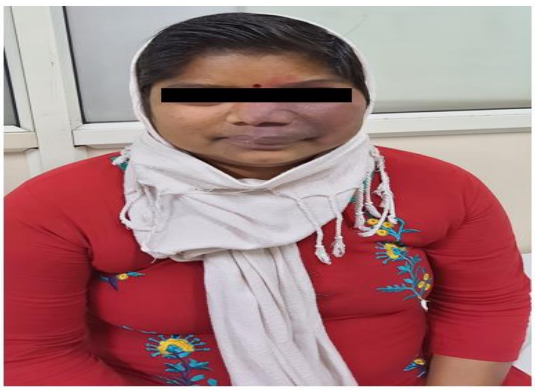


Figure 10. *OPD follow up after 3 months from discharge*

Discussion

DLE is skin disorder with minimal systemic involvement. DLE begins as erythematous, oedematous, scaling papules that spread centrifugally and coalesce into plaques, the size may vary from a few millimetres to a few centimetres. Scales produces a carpet-tack appearance after lifting and reveals dilated pilosebaceous orifices occupied by horny plugs. The healing of lesion begins in the centre which producing atrophy and scarring, telangiectasia, and pigmentary changes. Scarring alopecia is a significant finding(3).

Mucosal involvement is seen in about 24% of the DLE patients. Lesions appearing as chronic plaques or lichen planus-like oral lesions, ulcerations, cheilitis and plaque-like palatal lesions are seen.

The pathophysiology of DLE is poorly understood. It has been suggested that a heat shock protein is induced in the keratinocyte following ultraviolet (UV) light exposure or stress, and this protein may act as a target for gamma (delta) T-cell-mediated epidermal cell cytotoxicity.(3)

Patients may manifest any symptom of SLE. Therefore, the history should include an assessment for symptoms of pleuritis, pericarditis, neurologic involvement, and renal involvement. It may present with constitutional symptoms such as fever, malaise, multiple joint pain, myalgia, myopathy etc. to systemic manifestation of various system in which cardiopulmonary involvement with pleuritis, pericarditis, myocarditis, endocarditis, pleural effusion etc.

Neuropsychological manifestation like headaches, personality changes, seizures, cranial and peripheral neuropathies, cerebellar dysfunction etc. Renal manifestations include glomerulonephritis, proteinuria, haematuria, urinary casts, hypertension etc. Haematological manifestations like anaemia, leukocytopenia, thrombocytopenia, lymphopenia, lymphadenopathy, vasculitis etc. Gastrointestinal manifestations like impaired bowel motility , bleeding or perforation , pancreatitis etc. are seen(4)

The treatment of DLE includes avoiding exacerbating factors and suppression of lesions. Those who are sensitive to sunlight should apply UVA/UVB-15 protective sunscreen daily and covering the body parts while being outdoors. Topical steroid and/or Intralesional cortisone injections is the first line of treatment for localized cutaneous and mucosal lesions. Systemic corticosteroids, Calcineurin inhibitors, pimecrolimus cream or tacrolimus ointment, aminoquinoline antimalarial, dapsone or imiquimod 5% may also be used along with topical antifungal therapy. Topical hydroquinone is used for the treatment of hyperpigmented scars. More recently, biological therapies with agents like etanercept and tumour necrosis factor have demonstrated an overall decline in the disease activity. Efalizumab, a monoclonal antibody and a T-cell modulator has also shown a good response in patients with DLE. Patient should follow-up every six monthly as DLE is pre-cancerous condition, for the early detection of SLE and to minimize scarring.

In patients of SLE with mild features of fever, arthralgia, myalgia can be managed with NSAIDs. Hydroxychloroquine (HCQ) can be added if required as skin lesions and arthritis also respond to HCQ. In patient with severe life-threatening symptoms systemic steroid should be used. Acutely Ill and patient with Lupus Nephritis Pulses of Methylprednisolone (1gm/ day) should be given for 3 days.

Conclusion

SLE may begin with skin manifestation of discoid lupus erythematosus with progression to systemic involvement patient in the course of disease. Here we have case who presented with skin manifestation of DLE and upon further investigation found to have systemic manifestation of SLE.

Recent clinical trials

New disease-modifying conventional and biologic agents used alone, in combination or sequentially, have improved rates of achieving treatment goals, including limiting of glucocorticoid use. More specifically, studies have shown that MMF or enteric-coated mycophenolate sodium is effective like azathioprine in patients with general lupus or Lupus Nephritis. Calcineurin inhibitors added to standard-of-care induction therapy for Lupus Nephritis (so called

'multitarget' therapy) may increase complete renal remission rates and maintain remission. The first regimen tested included tacrolimus in combination with Mycophenolate mofetil and glucocorticoids, as both induction and maintenance therapy(5). Rituximab is anti cd 20 monoclonal antibody tried in patients of SLE. Belimumab, a monoclonal human antibody that inactivates B lymphocyte stimulator can also be used for treatment of patients with active disease, who are refractory to standard treatment. (6)

References

1. Grönhagen C, Nyberg F. Cutaneous lupus erythematosus: An update. Indian Dermatol Online J. 2014;5(1):7.
2. Sreejan C, Gopakumar R, Babu S, Roopashri R. Discoid lupus erythematosus: A case report with review of the literature. J Indian Acad Oral Med Radiol. 2009;21(1):37.
3. Discoid lupus erythematosus: A case report with review of the literature Sreejan C K, Gopakumar R, Babu S, Roopashri R K - J Indian Acad Oral Med Radiol 2009;(1);37
4. Maidhof W, Hilas O. Lupus: An overview of the disease and management options. P T 2012 Apr;37(4):240–9.
5. Fanouriakis A, Tziolos N, Bertsias G, Boumpas DT. Update in the diagnosis and management of systemic lupus erythematosus. Ann Rheum Dis. 2021;80(1):14–25.
6. Trial Watch BLYS-targeted antibody shows promise in Phase III SLE trial. *Nat. Rev. Drug Discov.* 2009; 8:688